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DATABASE ESTABLISHMENT AND AN INVESTIGATION OF QUALITY OF LIFE, SELF-ESTEEM, AND SELF-CONCEPT OF PATIENTS WITH DEVELOPMENTAL DENTAL ANOMALIES

A thesis submitted to the University of Dublin in fulfillment of the requirements

For

The Degree of Doctor of Philosophy by

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March 2012

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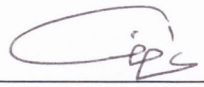
Signature: 

Date: 29.03.2012

Summary

In this thesis a cohort of patients attending the Dublin Dental University Hospital (DDUH) affected by developmental dental disorders (DDDs) such as hypodontia, amelogenesis imperfecta (AI) and dentinogenesis imperfecta (DI) were investigated.

Purpose: The aims of this project were to establish an Irish National Database of patients with (DDDs) and to investigate the impact of these disorders on the quality of life (QoL) of affected individuals and their families.

Database establishment and analysis: A web-based database was established for patients with (DDDs) and is hosted in the (DDUH). The database contains data of 401 patients affected by hypodontia, (AI), and (DI). The database was developed in collaboration with all disciplines involved in providing patient care in the Special Dental Needs Clinic in the (DDUH). It stores all relevant clinical and biographical information for this patient population in order to identify the prevalence, clinical presentation and geographical distribution of these developmental dental disorders. The information can also be used to help allocate services and outline the resources needed to optimise the management of patients with these conditions. The database allows compilation and searching of data in multiple ways. It was specifically designed to facilitate searching patterns of hypodontia with multiple fields searched simultaneously. Hypodontia was the most common (DDDs) referred to the Special Dental Needs clinic since its inception in 2001. The majority of referrals came from the Health Service Executive and were from in and around the Dublin area. The pattern of tooth agenesis of Irish patients appears to be similar to that previously reported in the literature.

Impact of (DDDs) on (QoL) and Self-Esteem of affected individuals and their families: Given the emphasis society places on facial aesthetics, health care providers have come to question what effects (DDDs) of teeth have on a person's psychosocial health. It is essential these providers understand how dental health and appearance fit into the larger picture of overall health and well-being. No Irish data on the psychosocial influence of severe (DDDs) was previously available. This project provided information regarding the importance of teeth and oral aesthetics in psychosocial development in affected children and adult populations.

Impact of hypodontia and (AI) on the (QoL) of affected adult patient population

Materials and Methods: Adult patients (18 years or older) from the Special Dental Needs Database with clinical and radiological diagnoses of moderate to severe

hypodontia and (AI) were contacted. Those who agreed to participate were included and age and gender matched with control patients attending the Dublin Dental University Hospital for routine dental care. A postal package of two well-established questionnaires (Oral Health Related Quality of Life [OHIP-49] and Rosenberg Self Esteem Scale), information leaflet, instructions, and prepaid envelopes was sent to all potential participants. A blinded researcher collated all data and the paired data were analysed with a statistical significance level of $P \leq 0.05$.

Results and Conclusions: These (DDDs) had a marked negative impact on the (QoL) of this patient population under most quality of life domains. The psychosocial impact of severe hypodontia on the (QoL) of affected patients is greater than that experienced by patients affected with moderate hypodontia, and is similar to the impact of (AI). However, Self esteem was not equally affected and was not statistically different between the test and control groups.

Impact of hypodontia and (AI) on the (QoL) of affected children and their families

Materials and Methods: Parents/Guardians of young patients (under 18 years of age) from the Special Dental Needs Database with clinical and radiological diagnoses of moderate to severe hypodontia and (AI) were contacted to give their consents and their child's assent for participation. Those who agreed (parent and child) to participate were included and then age and gender matched with control patients attending the Dublin Dental University Hospital for routine dental care. A postal package of three established questionnaires (Parental Perception Questionnaire (PPQ), Family Impact Scale (FIS), and Piers Harris Self Concept Scale), information leaflet, instructions, and prepaid envelopes was sent to all potential participants. Parents were advised to complete two questionnaires (PPQ and FIS); whereas, children were asked to complete the Piers Harris Self Concept Scale, A blinded researcher collated all data and the paired data were analysed with a statistical significance level of $P \leq 0.05$.

Results and Conclusions: (DDDs) can have detrimental effects on a young patient's (QoL) as perceived by their parents. However, as far as the children were concerned only the behavioural adjustment domain of the Piers Harris Self-Concept Scale was significant for hypodontia patients, and the intellectual and school status domain for children with (AI). (DDDs) did not appear to have a major impact on family members of the affected child.

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Definitions

Definitions

Allele

An allele is one of two or more versions of a gene. An individual inherits two alleles for each gene, one from each parent. If the two alleles are the same, the individual is homozygous for that gene. If the alleles are different, the individual is heterozygous. Though the term "allele" was originally used to describe variation among genes, it now also refers to variation among non-coding DNA sequences (Health, 1997).

Autosomal Dominant

Autosomal dominance is a pattern of inheritance characteristic of some genetic diseases. "Autosomal" means that the gene in question is located on one of the numbered, or non-sex, chromosomes. "Dominant" means that a single copy of the disease-associated mutation is enough to cause the disease. Huntington's disease (which is a neurodegenerative disease inherited within families characterized by congnitive and movement problems) is a common example of an autosomal dominant genetic disorder (Health, 1997).

Autosomal Recessive

It is a pattern of inheritance of some genetic diseases in which two copies of the mutation are needed to cause the disease (Health, 1997).

Confounding factor

A confounding factor in a study is a variable, which is related to one or more of the variables defined in a study. A confounding factor may mask an actual association or falsely demonstrate an apparent association between the study variables where no real association between them exists. If confounding factors are not measured and considered, bias may result in the conclusion of the study (Board, 2010).

Dizygot twins

Fraternal, or dizygotic, twins result from the fertilization of two separate eggs during the same pregnancy. They share half of their genes, just like any other siblings. Fraternal twins can be of the same or different sexes (Health, 1997).

Epistatic genes

Epistasis is a circumstance where the expression of one gene is affected by the expression of one or more independently inherited genes. For example, if the expression of gene #2 depends on the expression of gene #1, but if gene #1 becomes inactive, then the expression of gene #2 will not occur. In this example, gene #1 is said to be epistatic to gene #2 (Health, 1997).

Frameshift mutation

A frameshift mutation is a type of mutation involving the insertion or deletion of a nucleotide in which the number of deleted base pairs is not divisible by three. "Divisible by three" is important because the cell reads a gene in groups of three bases. Each group of three bases corresponds to one of 20 different amino acids used to build a protein. If a mutation disrupts this reading frame, then the entire DNA sequence following the mutation will be read incorrectly (Health, 1997).

Gene

The gene is the basic physical unit of inheritance. Genes are passed from parents to offspring and contain the information needed to specify traits. Genes are arranged, one after another, on structures called chromosomes. A chromosome contains a single, long DNA molecule, only a portion of which corresponds to a single gene. Humans have approximately 23,000 genes arranged on their chromosomes (Health, 1997).

Genetic penetrance

Penetrance is the characteristic phenotypic effect of a genotype. If the phenotype is always expressed in the presence of the genotype, the genotype is completely penetrant. If it is not always expressed, it is incompletely penetrant (Health, 1989).

Genotype

A genotype is an individual's collection of genes. The term also can refer to the two alleles inherited for a particular gene. The genotype is expressed when the information encoded in the genes' DNA is used to make protein and RNA molecules. The expression of the genotype contributes to the individual's observable traits, called the phenotype (Health, 1997).

Heterozygous

Heterozygous refers to having inherited different forms of a particular gene from each parent. A heterozygous genotype stands in contrast to a homozygous genotype, where an individual inherits identical forms of a particular gene from each parent (Health, 1989).

Homeobox gene

A homeobox is a DNA sequence found within genes that are involved in the regulation of patterns of development (morphogenesis) in animals, fungi and plants (McGinnis et al., 1984).

Homeodomain

The homeodomain fold is a protein structural domain that binds DNA or RNA and is thus commonly found in transcription factors. The fold consists of a 60-amino acid helix-turn-helix structure, in which three alpha helices are connected by short loop regions. Many homeodomains induce cellular differentiation by initiating the cascades of co-regulated genes required to produce individual tissues and organs (Gehring, 1993).

Homozygous

Homozygous is a genetic condition where an individual inherits the same alleles for a particular gene from both parents (Health, 1989).

Internal Consistency

Internal consistency estimates reliability by grouping questions in a questionnaire that measure the same concept. A reliable questionnaire should produce the same or very similar results, on two or more administrations to the same respondents, provided that, there is good reason to believe that the health status of the patients has not changed. It is measured using Cronbach's alpha statistics or the Kuder Richardson (KR-20). The closer the correlation to one, the higher the reliability estimate of the instrument (Colosi, 1997; Jenkinson and McGee, 1998c).

Likert Scale

It is a response scale in which respondents select from a range of options, which are placed on a continuum, such as Never, Rarely, Sometimes, Often, Very Often. A number of related questions tapping the aspects of the same phenomenon will, ideally,

have the same response categories and can be summed. When responding to a Likert questionnaire item, respondents specify their level of agreement to a statement (Jenkinson and McGee, 1998c). The scale is named after its inventor, psychologist Rensis Likert (Likert, 1932).

Locus

A locus is the specific physical location of a gene or other DNA sequence on a chromosome, like a genetic street address (NHGRI, 1999).

Missense mutation

A missense mutation is when the change of a single base pair causes the substitution of a different amino acid in the resulting protein. This amino acid substitution may have no effect, or it may render the protein non-functional (Health, 1997).

Monozygot twins

Identical twins are also known as monozygotic twins. They result from the fertilization of a single egg that splits in two. Identical twins share all of their genes and are always of the same sex.

Nonsense mutation

A nonsense mutation is the substitution of a single base pair that leads to the appearance of a stop codon where previously there was a codon specifying an amino acid. The presence of this premature stop codon results in the production of a shortened, and likely nonfunctional, protein (Health, 1997).

Phenotype:

A phenotype is an individual's observable traits, such as height, eye color, and blood type. The genetic contribution to the phenotype is called the genotype. Some traits are largely determined by the genotype, while other traits are largely determined by environmental factors (Health, 1997).

Probands

A proband is an individual being studied or reported on. A proband is usually the first affected individual in a family who brings a genetic disorder to the attention of the medical community (Health, 1997).

Test- retest reliability

Reliability is the consistency of the measurement, or the degree to which an instrument measures the same way each time it is used under the same condition with the same subjects. In short, it is the repeatability of a measurement. A measure is considered reliable if a person's score on the same test given twice is similar. The two occasions need to be far enough apart so that previous responses cannot be remembered but close enough in time so that change in in the true score is minimal (Colosi, 1997; Jenkinson and McGee, 1998c).

Test Validity

Validity is the extent to which an instrument measures the desired underlying concept (Jenkinson and McGee, 1998c)

For example, say one is studying the effect of strict attendance policies on class participation. In this case, it was observed that class participation did increase after the policy was established. Each type of validity would highlight a different aspect of the relationship between the treatment (strict attendance policy) and the observed outcome (increased class participation) (Colosi, 1997).

Types of Validity:

There are four types of validity:

1. Conclusion validity: It asks is there a relationship between the program and the observed outcome? Or, in the previous example, is there a connection between the attendance policy and the increased participation observed?

2. Internal Validity: It asks if there is a relationship between the program and the outcome observed and is it a causal relationship? For example, did the attendance policy cause class participation to increase?

3. Construct validity: This determines if there is a relationship between how the concepts were operationalised in a study to the actual causal relationship being observed? Or in the previous example, did the treatment (attendance policy) reflect the construct of attendance, and did the measured outcome—increased class participation—reflect the construct of participation? Overall, one tries to generalise the conceptualised treatment and outcomes to broader constructs of the same concepts.

4. External validity: It refers to the ability to generalise the results of any study to other settings. In the previous example, is it possible to generalise the results to other classrooms? (Colosi, 1997).

X-linked: It refers to a gene on the X chromosome (Health, 1989).

X-linked dominant: it describes a dominant trait or disorder caused by a mutation in a gene on the X chromosome. The phenotype is expressed in heterozygous females as well as in hemizygous males (having only one X chromosome); affected males tend to have a more severe phenotype than affected females (Health, 1989).

X-linked recessive: A mode of inheritance in which a mutation in a gene on the X chromosome causes the phenotype to be expressed in males who are hemizygous for the gene mutation (i.e., they have only one X chromosome) and in females who are homozygous for the gene mutation (i.e., they have a copy of the gene mutation on each of their two X chromosomes). Carrier females who have only one copy of the mutation do not usually express the phenotype, although differences in X-chromosome inactivation can lead to varying degrees of clinical expression in carrier females (Health,1989)

Chapter 1

Literature Review

Chapter 1: Literature Review

1.1 HYPODONTIA

1.1.1 Definitions

Hypodontia is the term used to describe the developmental absence of one or more primary or secondary teeth, excluding third molars. It is the most common developmental dental anomaly and can be challenging to manage clinically (Larmour et al., 2005).

1.1.2 Classifications

At birth, all primary teeth and first permanent molars crypts are visible radiographically. Calcifications of crowns of the permanent teeth, except the third molars, start at three years of age and are normally completed by six years of age (Zhu et al., 1996). Clinical examination at 3 to 4 years is suitable for diagnosis of congenitally missing primary teeth and at 12-14 year old for permanent teeth (Bailleul-Forestier et al., 2008b). However, Wisth et al. reported that the prevalence of missing teeth is higher when examined at the age of seven years of age compared with nine years of age (Wisth et al., 1974a). At seven years of age, 7.1% of the children had missing teeth, while two years later only 6.6 percent of the same sample was diagnosed with hypodontia.

Hypodontia commonly refers to the condition where there is absence of only one or a few teeth only. Oligodontia is the term usually used to describe six or more missing teeth, and anodontia is the complete absence of teeth.

Hypodontia can also be classified according to the severity of the condition as:

Mild: one to three teeth developmentally missing.
Moderate: four to six teeth developmentally missing.
Severe: more than six teeth developmentally missing.

Hypodontia can affect both the primary and permanent dentitions, and there is strong correlation between hypodontia in primary and permanent dentitions. Children with hypodontia in the primary dentition nearly always present with hypodontia of the permanent successors (Bailleul-Forestier et al., 2008b). Hypodontia is rare in the primary dentition with a prevalence of less than 1% in Caucasians. When it does occur

in the primary dentition, it most commonly involves the mandibular incisors (Jorgenson, 1980).

1.1.3 Aetiology

Hypodontia has been regarded as a multifactorial condition with both genetic and environmental influences playing a role (Parkin et al., 2009). However, investigators offer different opinions regarding the relative importance of each factor (Dhanrajani, 2002; Larmour et al., 2005; Parkin et al., 2009). If genetic factors are the main influence and specific genes can be isolated, then it may be possible to screen for hypodontia and manipulate these genes in the future in order to "develop" teeth (Sharpe and Young, 2005).

1.1.3.1 Genetic Factors

Family and twin studies

A strong genetic influence in hypodontia has been established by studies completed on families and twins (Brook, 1984; Svinhufvud et al., 1988; Stimson et al., 1997; Liu et al., 1998). Some disagreement exists about whether a single gene defect is involved or if it is due to the interaction of a number of genes, i.e., a polygenetic trait. In a large family study, it was proposed that in the majority of cases, hypodontia had a polygenetic inheritance mode (Brook, 1984). The risk of any relatives having hypodontia depended upon a combination of numerous genetic and environmental factors, each with a small effect. Genetic influences were confirmed with a much higher prevalence rate amongst relatives of the probands when compared to the general population, and the proportion of relatives affected varied with the severity of the condition in the probands.

Another study proposed that hypodontia is caused by a single gene defect and follows an autosomal dominant pattern of inheritance (Burzynski and Escobar, 1983). However, at a clinical level, it is known that there is variable expression of the trait, suggesting variable gene penetration. This suggests a polygenic mode of inheritance with epistatic genes and environmental factors interfering with the phenotypic expression of the hypodontia gene (Parkin et al., 2009).

Monozygotic twin studies

The classical approach for separating the effects of genetic and environmental influences involves comparing identical (monozygous) twins. Differences between monozygous twin pairs should reflect environmental factors, whereas differences between dizygous twin pairs are due to both genetic and environmental factors. The majority of such studies confirmed a variable expression of hypodontia in monozygotic twin pairs, suggesting an environmental influence (Gravely and Johnson, 1969; Nik-Hussein and Salcedo, 1987; Lapter et al., 1998). However, further studies have not determined monozygosity absolutely. In order to confirm or refute monozygosity in such studies, it has been proposed that fingerprinting technique, such as that outlined by Jeffreys et al (Jeffreys et al., 1985), be employed (Kindelan et al., 1998).

Molecular genetics

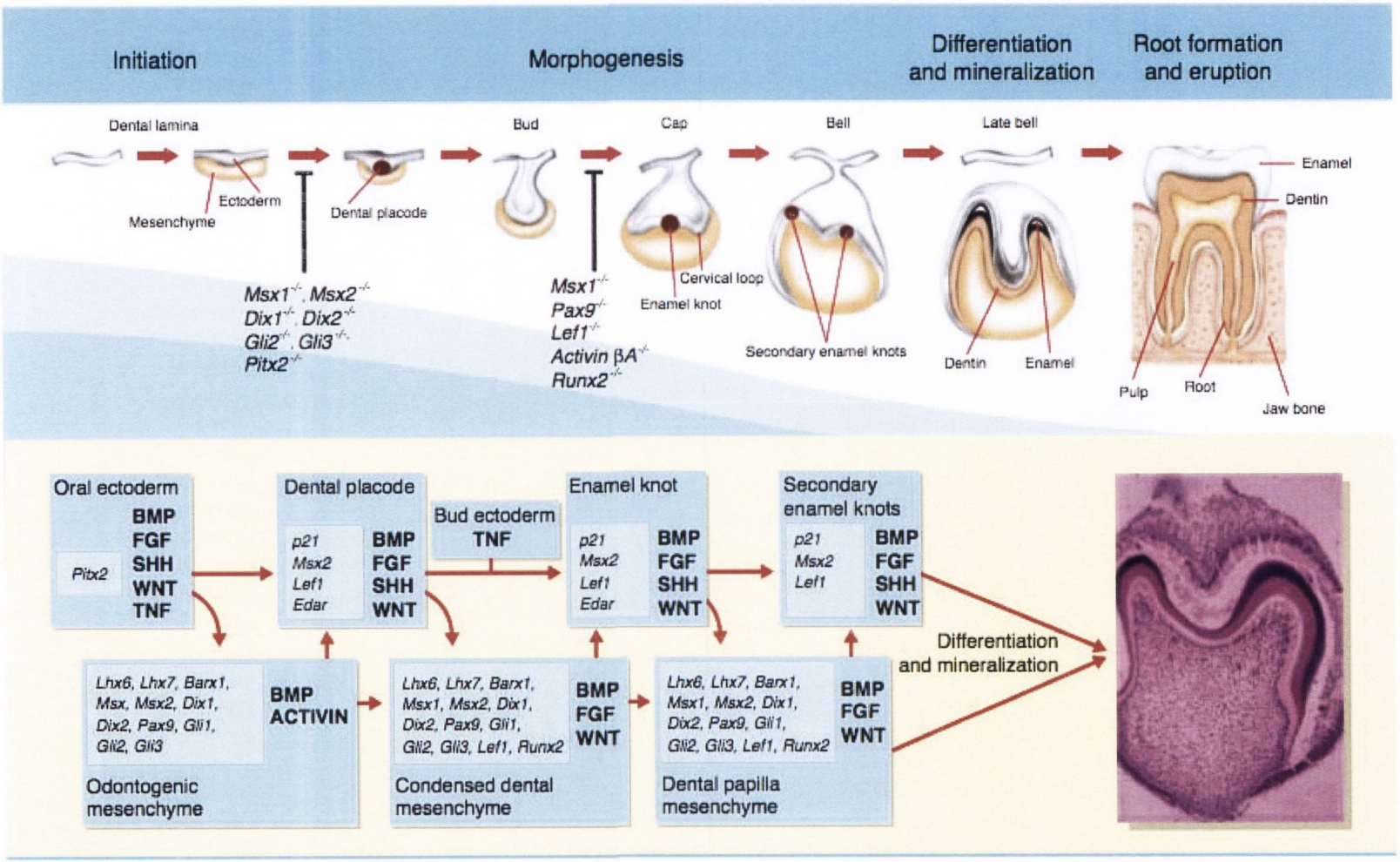
Advances in molecular genetics and the Human Genome Project (NHGRI, 1999) have provided substantial insight regarding the identification of genes involved in the pathogenesis of human diseases. These include dental diseases affecting enamel and dentine formation and anomalies in tooth number. Genetic diseases affecting tooth structure have been classified by the affected tissue (enamel versus dentine), specificity (syndromic versus non-syndromic) and pattern of inheritance: autosomal dominant (AD), autosomal recessive (AR), or X-linked recessive (XLR) (Bailleul-Forestier et al., 2008b).

In a human embryo, deciduous and permanent teeth develope from the oral ectoderm and the underlying neural mesenchymal cells, which have migrated from the cranial neural crest to the facial process. During the 6^{th} week of human development, a line of oral epithelial cells thicken to form the dental lamina, which develops several buds, that penetrate the underlying mesenchyme. The epithelial compartment of each bud invaginates to form a cupule-shaped structure, the enamel organ, with an inner part, the enamel reticulum. The enamel organ covers a densified mesenchymal tissue, the dental papilla. Mesenchymal cells of the dental papilla, which are adjacent to the enamel organ, differentiate into odontoblasts and start to secrete organic dentine matrix that serves as a scaffold for the deposition of hydroxyapatite crystals (Fleischmannova et al., 2008). The central part of the papilla forms the dental pulp. Epithelial cells adjacent to the differentiating odontoblasts differentiate into ameloblasts, which secrete enamel, the

outer cover of teeth. Mineralisation begins at the tips of the cusps and proceeds towards the cervical area. Root development follows crown formation. Reciprocal interactions of the epithelial and mesenchymal tissues regulate dental development (Thesleff, 2003; Fleischmannova et al., 2008). The mesenchyme surrounding the enamel organ and the growing roots form a fibrous structure, the periodontal ligament which ties the tooth to the alveolar bone (Diekwisch, 2002). While this process leads to the differentiation of deciduous teeth, the pedicle that connects the growing enamel organ to the surface epithelium produce an expansion that follows the same differentiation process, and lead to the formation of permanent teeth. A distal extension of the dental lamina provides the tooth germs for permanent molars. Simultaneously, the outer part of the dental lamina degenerates, separating the tooth from the gingival epithelium.

Biomineralisation begins during weeks 14-18 of pregnancy, and the crowns of all 20 primary teeth are partly mineralised at birth. All permanent tooth crowns except the third molars have begun their mineralisation by 6 years of age. Studies of odontogenesis, mostly using mouse teeth as models, have indicated that the position, number, size, and shape of different teeth are under genetic control (Sharpe, 2001). Tooth development is initiated by signals from the epithelial dental lamina to the mesenchyme (Mitsiadis et al., 1998). Thereafter, the mesenchyme regulates epithelial morphogenesis. Regulation of development is mediated by complex interactions between the epithelium and mesenchyme (Figure 1-1). The centre of the epithelial bulk, the enamel knot, functions as an important signalling centre regulating tooth shape (Lisi et al., 2003). During tooth development, the epithelium and mesenchyme interact through different families of signalling molecules and their receptors. These comprise the transforming growth factor β (TGFβ), bone morphogenetic proteins (BMP), fibroblast growth factor (FGF), epidermal growth factor (EGF), hedgehog (Hh) and wingless (Wnt) families (Sarkar et al., 2000).

Figure 1-1: Signalling and families of signalling molecules in tooth development (adapted from Thesleff, 2003).



Disruption of any one of the odontogenic signalling pathways can affect tooth development and may contribute to developmental dental disorders. Defects can affect not only the number of teeth (missing teeth, extra teeth) but also the form (shape, size) and mineralisation of resultant teeth (Fleischmannova et al., 2008).

Recent advances in the fields of human genetics and molecular biology have provided a greater understanding of tooth development. Embryogenesis of the craniofacial structures, including teeth, is genetically determined through neural crest cell migration. The migration of neural crest cells and their specification to form different types of teeth is now thought to be under the control of a family of genes known as the homeobox genes (Cobourne, 1999). They can be regarded as the master genes of craniofacial development, controlling patterning/ specification, induction, and all epithelial/mesenchymal interactions. Those of particular interest in tooth development are the muscle-specific homeobox genes Msx1 and Msx2. The proteins encoded by these homeobox genes are known as transcription factors, and can switch other genes on or off, thereby controlling gene expression. This hierarchy of gene interactions can be thought of as a coordinated cascade. The effects of the master or homeobox genes on other genes in this cascade of the interactions can be referred to as down-streaming effects.

Oligodontia with mutations in MSX1 (4p16.1)

Mutations in the homeobox gene MSX1 lead to specific hypo/oligodontia pattern. Second premolars and third molars are the most commonly affected teeth ("Online Mendilian Inheritance in Man 106600" OMIM106600) (McKusick, 1995), and are often associated with cleft lip and/or cleft palate (Modesto et al., 2006). First maxillary premolars, first mandibular molars, upper lateral incisors and lower central incisor may be absent. The primary dentition is usually normal (Vastardis et al., 1996; Lidral and Reising, 2002; Kim et al., 2006a; Fleischmannova et al., 2008).

Oligodontia with mutations in PAX 9 (14q12-q13)

Mutations in the transcription factor gene PAX9 lead to absence of most permanent molars with or without hypodontia in the primary dentition (OMIM 604625) (Stockton et al., 2000; Fleischmannova et al., 2008). Some individuals also have missing maxillary and/or mandibular premolars as well as mandibular central incisors. Smaller teeth observed in affected individuals suggested that PAX9 is involved not only in the

positioning and development of certain teeth but also in the morphogenesis of the entire dentition (Goldenberg et al., 2000). There appear to be genotype-phenotype correlations in PAX9 mutations: missense mutations give a milder phenotype than nonsense and frameshift mutants (Bailleul-Forestier et al., 2008b).

Both Pax9 and Msx1 are homeobox transcription factors expressed in the dental mesenchyme during early morphogenesis under the control of Fibroblast Growth Factor (FGF)/ Bone Morphogenetic Protein (BMP) signalling. Epithelial Bmp4 induces Msx1 expression in the underlying mesenchyme. Epithelial FGF8 expressed at the sites of future tooth formation induced Pax9 expression in the adjacent mesenchyme (Tucker et al., 1998).

Studies using knockout mice have revealed that Pax9 is necessary for the dental mesenchyme to condense around the tooth bud epithelium. Mesenchymal expression of odontogenic molecules, such as Msx1, lymphoid enhancer binding factor 1 (Lef1), and Bmp4, are down-regulated in the absence of Pax9, and tooth development is arrested at the bud stage (Peters et al., 1998). The interplay between Pax9 and its downstream target, Msx1, is thought to be necessary for the expression of Bmp4 in the mesenchyme, an essential prerequisite for the induction of primary enamel knots. Bmp4 expression is dramatically decreased in Msx1 knockout and tooth bud mesenchyme of molars fail to develop beyond the bud stage (Satokata and Maas, 1994). Ectopic Bmp4 expression partially rescues the Msx1 knockout tooth phenotype (Zhao et al., 2000). Mice deficient in either Pax9 or Msx1 bear similar defects to humans with mutations in these genes. Msx1 knockout mice show defective development of oro-facial structures and nails, including a highly penetrant arrest of tooth development at the bud stage and orofacial clefts. These findings are typical of hypodontia and Witkop syndrome (Satokata and Maas, 1994; Jumlongras et al., 2001). Pax9-deficient mice die shortly after birth: they present with cleft palate, have multiple skeletal defects, the thymus and parathyroid glands are absent, and tooth development is again arrested at the bud stage (Peters et al., 1998). Unlike hypodontia in humans, the tooth germ arrest in the Msx1 and Pax9 knockout mice affects all teeth. Dental defects only are observed in hypomorphic mice (a mice with a mutation that causes a partial loss of gene function i.e. a reduction in gene expression, but not a complete loss of gene function), where the expression of Pax9 is reduced. A lower Pax9 gene dosage results in phenotypic variability,

preferentially missing different types of teeth, which is typical of human hypodontia (Kist et al., 2005).

Oligodontia with mutations in AXIN 2 (17q23-24)

Mutations in AXIN2 gene cause tooth agenesis and colorectal cancer (OMIM 608615). The patients who carry the mutation lack 8-27 permanent teeth. Penetrance of colorectal cancer is very high. Defects in the deciduous dentition were observed in only one patient (Lammi et al., 2004). No specific tooth agenesis patterns have been identified.

Oligodontia with locus mapped to chromosome (10q11.2)

He-Zhao deficiency has been characterized in a large Chinese kindred with a distinct form of permanent tooth agenesis (OMIM 61096) (Liu et al., 2001). The Oligodontia was transmitted in an autosomal dominant fashion with incomplete penetrance. The affected members in the kindred had normal primary dentitions, followed by the absence of most permanent teeth with the exception of first and/or second permanent molars and maxillary central incisors. The gene locus was mapped to chromosome 10q11.2. KROX-26/ZNF22 expressed in human tooth development, is a potential candidate gene because it is thought that it may play a potential function in the molecular regulation of tooth formation in humans (Gao et al., 2003).

1.1.3.2 Environmental Factors

There has been little published research about the specific environmental influences involved in hypodontia. Major environmental factors that have been reported include infection, e.g., rubella (Gullikson, 1975), drugs, such as thalidomide, irradiation and chemotherapeutic agents (Nasman et al., 1997). The higher prevalence of hypodontia in cleft lip and palate patients has been widely reported with the maxillary lateral incisor in the cleft site being the most commonly missing tooth. This can be explained by a localised disturbance of the dental lamina in the cleft area (Ranta, 1986; Laatikainen and Ranta, 1994).

1.1.3.3 Other Factors

The different prevalence of hypodontia at different tooth sites may be associated with developmental timing. It is the later developing teeth in each morphological class which are most frequently absent, e.g., the lateral incisor, second premolar, and third molars. There is more evidence from animal and human studies that if the teeth which develop

early in a given segment are large, those developing later are smaller or absent (Sofaer et al., 1971; Sofaer, 1975; Brook, 1984).

Developmental absence of teeth is a consequence of:

- Physical obstruction or disruption of the dental lamina
- Space limitation
- Functional abnormalities of the dental epithelium
- Failure of initiation of the underlying mesenchyme (Nunn et al., 2003)

Hypodontia may arise as a familial condition, with a high proportion of affected individuals occurring in families with a previous history of the condition (Brook, 1984). The nature of the inheritance is complex and not well understood, but it is thought to be related to more than one gene (Bailit, 1975; Thesleff, 2000). Several reports suggested that an autosomal dominant gene with variable penetrance might be responsible (Tal, 1981; Phillip and Caurdy, 1985; Ruprecht et al., 1985). Hypodontia may also arise in individuals with no family history of the disorder.

Hypodontia is an important condition in which both aesthetics and function can be detrimentally affected. Children with moderate to severe hypodontia, where four or more teeth are developmentally absent tend to have delayed tooth development, reduced mesiodistal crown diameter, and abnormal morphology and position of the teeth (Rune and Sarnas, 1974).

A number of systemic conditions, such as hypohidrotic ectodermal dysplasia, Down syndrome, chondroectodermal dysplasia and Van der Woude syndrome have hypodontia as a feature. The developmental disruption due to the presence of a cleft lip and palate involving the alveolus may also result in an absence of teeth in that region, notably the maxillary lateral incisors (Winter and Geddes, 1967; Gorlin and Cohen, 2001; Lam et al., 2010).

1.1.4 Prevalence

Studies assessing hypodontia vary widely in their reports of prevalence in the permanent dentition, as demonstrated in Table 1-1. The majority of these studies report prevalence rates varying from 2.6% in Saudi Arabia (Salama and Abdel-Megid, 1994) to 11.3% in Ireland (O'Dowling and McNamara, 1990). Studies in the United Kingdom suggest a prevalence rate of 4% to 4.5% (Rose, 1966; Brook, 1974). Most of these

studies were based on clinical observation, which were not as reliable as radiographic studies or patients accessed through orthodontic clinics rather than subjects chosen at random from the population (al-Emran, 1990). The results presented by the investigators indicate that differences exist between various populations.

A problem with population studies is the age variation of the populations studied. This may account for some of the differences in hypodontia prevalence, as younger children may still have teeth to calcify, whereas in older patients teeth may have been extracted. Some of these problems would be avoided by carrying out prospective population studies rather than retrospective radiographic studies (Larmour et al., 2005).

Table 1-1: Prevalence of hypodontia

Country	**Author**	**Population type**	**Age range (yr)**	**Number of patients**	**Prevalence (%)**	**Most frequently absent tooth**
Norway	(Aasheim and Ogaard, 1993)	School children	7.8-10.4	1953	6.5%	Mandibular second premolar
Saudi Arabia	(al-Emran, 1990)	School children	13.5-14.5	500	4%	Mandibular second premolar
Sweden	(Bergstrom, 1977)	School children	8-9	2589	7.4%	Mandibular second premolar
England	(Brook, 1974)	Nursery and school children	3-5 & 11-14	958 & 1183	4.4%	Mandibular second premolar
Hong Kong	(Davis, 1987)	School children	12	1093	6.9%	Mandibular incisor
Iceland	(Johannsdottir et al., 1997)	School children	6	396	5.0%	Mandibular second premolar
Denmark	(Locht, 1980)	School children	9-10	704	7.7	Mandibular second premolar

Australia	(Lynham, 1990)	Australian defence force recruits	16-26	662	6.3%	Maxillary lateral incisor
United States	(Muller et al., 1970)	School students	11-15	14940	3.5%	Mandibular second premolar
Malaysia	(Nik-Hussein, 1989)	Children attending a dental hospital	6-15	1583	2.8%	Maxillary lateral Incisor
Ireland	(O'Dowling and McNamara, 1990)	Orthodontic patients	7-17	3056	11.3%	Mandibular second premolar
Denmark	(Rolling, 1980)	School children	9-10	3325	7.8%	Maxillary second premolar
England	(Rose, 1966)	Orthodontic patients	7-14	6000	4.3%	Mandibular second premolar
Saudi Arabia	(Salama and Abdel-Megid, 1994)	Children attending a dental hospital	5-10	1300	2.6%	Mandibular second premolar
Brazil	(Gomes et al., 2010)	Orthodontic patients	10-15	1049	6.3%	Maxillary lateral incisor

1.1.5 Tooth Predilection

The mandibular second premolar is the most affected tooth, followed by the maxillary lateral incisor and second maxillary premolar. Unilateral occurrence of dental agenesis is more common than bilateral occurrence. However, bilateral agenesis of maxillary lateral incisors is more common than unilateral agenesis. The overall prevalence of agenesis in the maxilla is comparable with that in the mandible, but a marked difference was found between both jaws regarding tooth type showing a tooth predilection of lateral incisors and second premolars in the maxilla as opposed to second premolars in the mandible. When third molars were included more third molars were missing in the maxilla than mandible (Mok and Ho, 1996; Bartzela et al., 2010; Hashem et al., 2010).

Agenesis of the third molars is the most commonly reported ranging from 24% (Rozkovcova et al., 2004) to 28.5% (Mok and Ho, 1996). However, agenesis of this tooth is not included in hypodontia studies.

1.1.6 Severity of Hypodontia

Population studies of hypodontia prevalence suggest that the majority of patients (80%+) will present with one or two congenitally missing teeth (Muller et al., 1970; Bergstrom, 1977). Less than 10% will present with four more missing teeth and less than 1% will present with six or more absent teeth. Anodontia or congenital lack of all teeth without associated abnormalities is extremely rare. Some case reports of anodontia have suggested that anodontia of permanent teeth is a homozygote state of the gene responsible for pegged or missing maxillary incisors (Witkop, 1987; Hoo, 2000; Gadda et al., 2010). The more severe hypodontia cases also have an increased risk of being associated with a syndromic condition (Hobkirk and Brook, 1980).
It is not within the scope of this thesis to discuss the associated syndromes and systemic conditions associated with hypodontia.

1.1.7 Associated Dental Anomalies

1.1.7.1 Microdontia

The association between hypodontia and microdontia is well established (Brook, 1984; Baccetti, 1998). A common example is a congenital absence of a maxillary lateral incisor, which is frequently associated with a diminutive contralateral incisor. In 1974

Brook reported that hypodontia and microdontia are more prevalent in females (5.7% and 3.1%) as compared to (2.4% and 1.9%) males, whereas supernumeraries and macrodontia are more common in males (2.4% and 1.2%) as compared to (1.7% and 0.9%) females. It was suggested that this implicated a common genetic linkage between the different dental anomalies (Brook, 1974). In a follow-on study the same group found that combined hypodontia/microdontia was more prevalent in females (8.8%) as compared to males (5%), whereas the combined supernumeraries/macrodontia was more prevalent in males (3.6%) than females (2.6%) (Brook, 1984).

1.1.7.2 Impaction of Permanent Canines

The association between hypodontia and impaction of the maxillary canine has also been reported (Brin et al., 1986; Mossey et al., 1994; Peck et al., 1996). More specifically this involves the absence or reduced size of the maxillary lateral incisor. It is thought that the maxillary canine is guided in eruption by the lateral incisor root. If the lateral incisor root is absent or diminutive, eruptive guidance is lost and the canine can impact in a palatal position. Peck *et al* in a study of two samples of nonsyndromic subjects possessing either maxillary canine-first premolar transposition or palatal displacement of the maxillary canine reported that absence of the lateral incisor increased the prevalence of maxillary canine impaction by a factor of thirteen with 26% of lateral incisor hypodontia cases experiencing impaction (Peck et al., 1996).

In a clinical and radiographical assessment of 2440 adolescent Israeli school children (aged 14 to 18) investigating dental anomalies of the maxillary lateral incisors and its relation to the position of maxillary canine it was reported that up to 42% of cases had palatally placed (but not necessarily impacted) canines (Brin et al., 1986).

1.1.7.3 Maxillary Canine/First Premolar Transposition

It has been reported that patients with a canine/first premolar transposition experience hypodontia in 49% of cases (Peck et al., 1993). It was suggested that hypodontia and transposition may have a common genetic linkage.

1.1.7.4 Taurodontism

Taurodontism literally means "bull-like", and is manifested by teeth (most commonly mandibular first permanent molars) having an enlarged and elongated pulp chamber. It has been reported that it is more common in individuals with hypodontia, with a

reported prevalence of 35% in a sample of 66 patients as compared with 7.5% of a control group without hypodontia (Seow and Lai, 1989). However, the study did not relate the findings to the severity of hypodontia.

1.1.8 Associated Skeletal Defects

1.1.8.1 Anteroposterior Skeletal Pattern

There have been a wide range of results reported from cephalometric studies which assessed the skeletal pattern of hypodontia patients. Some studies reported predominately healthy class I skeletal patterns (Roald et al., 1982; Dermaut et al., 1986; Yuksel and Ucem, 1997). Other studies suggested that patients with hypodontia have, on average, smaller and more retrognathic maxillae, i.e. they tended to have class III skeletal patterns (Wisth et al., 1974b; Sarnas and Rune, 1983; Woodworth et al., 1985; Chung et al., 2000). This tendency becomes more significant as the severity of the hypodontia problem worsens, especially when more than one type of tooth is missing (Chung et al., 2000).

1.1.8.2 Vertical Skeletal Pattern

Cephalometric studies report that hypodontia patients have a tendency to have a reduced anterior lower face height (Woodworth et al., 1985; Ogaard and Krogstad, 1995). This also becomes significant with increasing severity of hypodontia (Chung et al., 2000).

1.1.9 Dental Management of Hypodontia

The dental management of hypodontia depends on the severity (the number and location of missing teeth), the dental complexity of the case (the presence of associated dental and/or skeletal anomalies) and also the presence of associated systemic conditions/syndromes i.e., ectodermal dysplasia. In most cases it requires an interdisciplinary approach to provide care for this population. In 2010 a classification for patients with oligodontia (six or more missing teeth) which could act as an aid in treatment planning was proposed (Singer et al., 2010). The author used panoramic radiographical records of 70 patients with oligodontia to categorise the extent of the disability and treatment modality. Patients were classified into three types depending on the number of missing primary and permanent teeth, as well as in relation to their

prosthodontic requirements.

The author established a set of guidelines which he then used to classify patients according to the complexity of the case and its prosthodontic needs (Table 1-2).

1.1.9.1 Type1

The dental arches are often intact and well developed due to the presence of permanent teeth, and when absent, the retention of their primary predecessors (missing teeth range from 6-13). There is always bilateral molar support and amble alveolar bone with few areas of localized bone deficiency. Edentulous spans are one to two teeth wide. The number of sites that require prosthodontic replacement range from zero to seven and depend on the number of congenitally missing teeth.

1.1.9.2 Type 2

These cases are characterised by large numbers of missing permanent teeth (8-22) in localised areas and the retention of variable numbers of primary teeth. There may be a lack of unilateral or bilateral posterior molar support and in some cases the absence of permanent teeth distal to the canine in one or more quadrants. Supra-eruption of opposing teeth into these spans may be observed. There is ample alveolar bone where teeth are present. However, significant areas of localised, and in some cases generalised, bone deficiency are associated with the large edentulous spans.

1.1.9.3 Type 3

There is almost a complete absence of permanent and primary teeth. There is always an absence of bilateral support and a lack of permanent teeth distal to the canines in all quadrants. Consequently, there is a significant deficit in alveolar bone. The basal bone in the maxilla is sparse and represents a significant technical challenge to prosthetic rehabilitation.

The clinical application and suggested benefits of this classification are yet to be established and validated. In addition this classification was proposed to provide guidance for treatment planning and economic elements in the treatment of patients with hypodontia but no conclusions were possible from the data presented.

Table: 1-2: Morphologic features and oligodontia type (adapted from Singer et al, 2010).

NA: not applicable; V: Varied; A: applicable.

		Type1	Type 2	Type 3
Molar status				
	No bilateral molar support	NA	V	A
	No posterior teeth in one arch	NA	A	A
Alveolar bone volume				
	Ample alveolar bone	A	A	NA
	Ample alveolar bone with localised deficiency	A	A	NA
	Significant generalised alveolar bone deficiency	NA	A	A
Orthodontic requirements				
	No orthodontic therapy indicated	NA	V	A
	Limited objective orthodontics	A	V	NA
	Complex orthodontics	A	V	NA
	Adjunctive implant-assisted anchorage	A	V	NA
	Implant-supported arch-wire	NA	V	NA
Residual spaces				
	Edentulous spans limited to 1-2 teeth wide	A	V	NA
	At least 1 span greater than 2 teeth wide	NA	A	A
Prosthodontic requirements				
	Postorthodontic interim prosthodontics	A	A	NA
	Simple prosthodontics or implant prosthesis	A	NA	NA
	Complex prosthodontics	NA	A	A

1.1.10 The Interdisciplinary Management of Hypodontia

The developmental absence of teeth can seriously disable a young person both physically and emotionally, especially during the turbulent years of adolescence. Early management is indicated, yet there are reports of patients with hypodontia being referred late with all that that implies- treatment disrupting the examination years and reluctance to wear appliances because of the impact on the young person's social life (Hobkirk et al., 1994).

There is much to be gained from the interdisciplinary management of young people who have hypodontia. Many patients are looked after by multidisciplinary teams—and each clinician does their treatment in isolation. True interdisciplinary working involves the close working of committed team where each member contributes their expertise to achieve an optimum outcome for the patient and their family (Hobkirk et al., 1994; Bergendal et al., 1996; Nunn et al., 2003).

The two main dental disciplines that must be involved are orthodontics and restorative dentistry. However, depending on the severity of hypodontia, or the presence of an underlying systemic disturbance or skeletal nature of malocclusion, consultation with a paediatric dentist, paediatric physician, oral surgeon, general dental practitioner, and psychiatrist have been advocated (Nunn et al., 2003). The aims of management are to provide the patient with well-aligned arches and the overall result should be aesthetically acceptable to the patient, as well as being occlusally and periodontally stable. The interdisciplinary team for management of hypodontia patient should include:

- A paediatric dentist.
- A restorative dentist.
- An orthodontist.
- An oral and maxillofacial surgeon and a periodontist.
- A special care dentist.
- A general dental practitioner.

1.1.10.1 The Role of Paediatric Dentist

The key factor for patients with hypodontia is early referral. This is particularly helpful in the long term planning of patients' care. There are reports of patients with

hypodontia being referred to hypodontia clinics as late as 12 years of age suggestive of either a poor demand from patients/parents or lack of awareness from primary care providers (Hobkirk et al., 1994). This late diagnosis and referral of hypodontia cases may also be caused by the retention of primary teeth so that hypodontia is not identified early in life by patient or parents.

The role of paediatric dentist can be summarised in the following points:

1. Behaviour management in a challenged young person
2. Prevention of caries
3. Management of infra-occlusion of primary teeth
4. Intermediate restoration of diminutive teeth with composite resin additions
5. Endodontic care
6. Removable and fixed prosthesis

1.1.10.2 The Role of the Restorative Dentist

The need to await eruption and root formation of permanent teeth frequently delays restorative intervention. Short clinical crowns of recently erupted permanent teeth and, in particular, retained primary teeth, can compromise the retention of both adhesive and conventional bridgework due to reduced surface area available for bonding (Sano et al., 1994; Wyatt, 2007).

With the interdisciplinary management of hypodontia the areas of primary concern to the restorative dentist include:

- Early consideration of the likely final restoration.
- Maintenance of appearance and function.

Treatment options for hypodontia patients include the following:

1. Direct bonding techniques
2. Removable Prostheses
3. Resin bonded fixed partial dentures (RBFPD)
4. Conventional fixed partial dentures
5. Implant retained/ supported prosthesis

1.1.10.3 The Role of Orthodontist

The orthodontist plays an important role in the interdisciplinary management of patients with hypodontia in order to achieve an optimum outcome. The orthodontic management of hypodontia patients includes:

1. Space management
    a. Space Closure
    b. Space opening and re-distribution
2. Up-righting and aligning teeth
3. Management of an increased vertical and horizontal overlap
4. Retention and stability

1.1.10.4 The Role of the Periodontist and the Oral and Maxillofacial Surgeon

1. Aid orthodontics
2. Supercede orthodontics
3. Aid prosthodontics

1.1.10.5 The Role of Special Care Dentist

Dougall and Fiske stated that special care dentist seeks to address the oral health needs of people with a range of primary conditions, which may result in their oral health being compromised directly through the condition itself, or indirectly through medication or poor access to care and that even when specialist care is required, it is often the patient management rather than the dentistry that is complex (Dougall and Fiske, 2008).

A special care dentist might be required if the person had an impairment or disability that meant care could not adequately be provided by a general dental practitioner. Faulks and Hennequin described three groups of people who require special care dentist (Faulks and Hennequin, 2006), this included:

1. People who experience disability due to impairment of oral function and/or structure and who are limited in their activity and/or participation directly by their oral status.

2. People who have a condition that has direct or indirect repercussions on their oral health.

3. People who are disabled by their social, environmental or cultural context, which reflects on their oral health.

1.1.10.6 The Role of the General Dental Practitioner

Many patients with moderate or severe hypodontia will need to wear prosthesis from an early age. This has the potential to increase the risk of caries, periodontal disease and denture stomatitis (Hobkirk et al., 1995). These patients need to be instructed about meticulous oral and denture hygiene, the use of topical fluoride, and regular professional checkups (Thind et al., 2005). The role of the general dental practitioner would include:

- Provisional diagnosis and maintenance.
- Initial referral.
- Treatment, often with some restorative care.
- Maintenance following completion of treatment.

1.2 AMELOGENESIS IMPERFECTA

Enamel is a unique tissue characterised by a low protein and high mineral content. Ameloblasts play two roles during amelogenesis. They secrete the four major enamel matrix proteins and proteases: amelogenin (80-90%), ameloblastin (5-10%), enamelin (3-5%), and enamelysin (1%) (Goldberg et al., 1995). Later, they contribute to the maturation of the enamel, which is accompanied by a loss of organic matrix and an increase in mineralisation. Enamel is made up of strictly organised, oriented and tightly packed crystallites (Robinson and Kirkham, 1986). These crystallites are extremely long relative to their thickness. They generally extend from the underlying dentine toward the surface of the tooth and are organised into bundles, called prisms (Robinson et al., 1997). As a result of its peculiar properties, enamel mirrors and records, during its development, the variations in metabolic status of the individual. Hence enamel anomalies may reflect both environmental and systemic disturbances (Goodman and Armelagos, 1985). Diagnosis of an enamel defect requires a detailed record of the medical and clinical history of the patient, the potential exposure to environmental factors and the family history. Defects in genes encoding enamel proteins generally cause enamel malformations without affecting other parts of the body, although several

genetic syndromes are associated with enamel defects (Bailleul-Forestier et al., 2008b). It is not the scope of this thesis to discuss syndromic amelogenesis imperfecta.

1.2.1 Definitions

Amelogenesis imperfecta (AI) is a genetically and clinically heterogeneous group of inheritable disorders primarily affecting enamel formation. The phenotype varies from localised enamel deficiencies (hypoplasia) to complete absence of enamel (aplasia).

AI represents a group of conditions, which affect the structure and clinical appearance of enamel of all or nearly all the teeth in a more or less equal manner and which may be associated with morphological or biochemical changes elsewhere in the body (Aldred et al., 2003a).

The AI can be autosomal dominant, autosomal recessive, or X-linked. One large epidemiological study of 51 Swedish AI families found that approximately 6% of cases to be X-linked, 63% autosomal dominant (AD), and 12% autosomal recessive (AR) (Backman and Holmgren, 1988). The remaining 19% of AI cases were sporadic with neither family history nor discernable mode of transmission evident. The distribution of AI types is also known to vary in different populations. Autosomal recessive AI was the most prevalent AI type reported in a study evaluating 70,000 Israeli school children (Chosack et al., 1979). Diagnosis involves exclusion of extrinsic environmental or other factors, establishment of a likely inheritance pattern, recognition of phenotype and correlation with the dates of tooth formation to exclude a chronological developmental disturbance (Crawford et al., 2007).

1.2.2 Epidemiology

Reports vary widely depending upon the gene pool. Values of 1:14000 in the USA (Witkop and Sauk, 1976) to 1:700 in Sweden (Backman and Holm, 1986) have been reported (Crawford et al., 2007).

1.2.3 Clinical and Radiographic Characteristics of AI

Enamel findings in AI are highly variable, ranging from deficient enamel formation (hypoplastic AI) to defects in the mineral and protein content (hypomineralised and hypomature AI) (Witkop, 1988). In the latter forms, the enamel demonstrates more or

less severe discoloration, from opaque white enamel to yellowish or brown appearance (Table 1-3). The dentine itself can be exposed after the loss of enamel cap.

Table: 1-3: Clinical and radiographical characteristics of amelogenesis imperfecta, (adapted from Bailleul-Forestier, 2008).

	AI type	**Clinical aspect**	**Radiographic aspect**
Hypoplastic (type 1)	Hypoplastic, generalised (type IG)	Thin, patchy, rough aspect of the enamel layer Reduced contact between the teeth	Hypoplasia of the enamel layer, more pronounced in the coronal two-thirds of the crown. Reduced thickness of enamel
	Hypoplastic localised (type I A-F)	Similar aspect, but limited part of the crown	Hypoplasia may be difficult to observe
Hypomineralised	Hypocalcified (type III)	After eruption, the enamel persists only on the cervical part	Normal thickness of the enamel before tooth eruption Enamel radiolucency similar to dentine
	Hypomature (type II)	Chalky appearance, with orange, brown or white colour	Normal thickness of the enamel Enamel radiolucency slightly greater than dentine

1.2.4 Classification of Amelogenesis Imperfecta

Classifications of AI have evolved since the original division into hypoplastic and hypocalcified types in 1945 (Weinmann et al., 1945) (Table1-4). Some have been exclusively based on the phenotype (appearance) while others have used the phenotype as the primary discriminate and the mode of inheritance as a secondary factor in diagnosis.

The most commonly used classification was proposed in 1988 by Witkop (Witkop, 1988), and revised by Nusier in 2004 (Nusier et al., 2004) (Table 1-4). Although this system allows some apparent ordering of AI, practical application of the Witkop system is hampered by the important intrafamilial variability of the AI phenotype and the lack of close correlation between the AI type, and the causative molecular defect (Backman, 1988; Hart et al., 2002a; Crawford et al., 2007).

Based on enamel appearance and hypothesised developmental defects AI can be classified into hypoplastic (secretory defect), and hypocalcified (mineralisation defect), and hypomaturation (protein processing and crystallite maturation defect) forms (Witkop, 1988).

It would be ultimately useful to classify conditions by specific gene and subsequent biochemistry and molecular mechanisms (Aldred and Crawford, 1995; Hart et al., 2002b). Combining AI and their associated phenotypes with the mode of inheritance may be more clinically relevant and is critical for patient counselling and investigative approaches directed at identifying the molecular basis of these conditions (Wright, 2006).

Table 1-4: Classification systems applied to amelogenesis imperfecta (adapted from Crawford et al, 2007 with modification).

Weinmann et al, 1945	Two types based solely on phenotype: hypoplastic and hypocalcified (Weinmann et al., 1945).
Darling, 1956	Five phenotypes based on clinical, microradiographical and histopathological findings (Darling, 1956). Hypoplastic: • Group 1 Generalised pitting • Group 2 Vertical grooves (now known to be X-linked AI) • Group 3 Generalised hypoplasia Hypocalcified: • Type 4A Chalky, yellow, brown enamel • Type 4B Marked enamel discolouration and softness with post-eruptive loss of enamel • Type 5 Generalised or localised discolouration and chipping of enamel
Witkop, 1957	This classification was based primarily on phenotype. Five types have been described (Witkop, 1957): • Hypoplastic • Hypocalcification • Hypomaturation • Pigmented hypomaturation

	• Local hypoplasia Mode of inheritance has since been incorporated as further means of delineating cases.
Schulze, 1970	This classification was based on phenotype and mode of inheritance (Schulze et al., 1999).
Witkop and Rao, 1971 Witkop and Rao, 1971 (continued)	This classification was based on phenotype and mode of inheritance. Three broad categories: hypoplastic, hypocalcified, hypomaturation (Witkop, 1971). 1. Hypoplastic a. Autosomal dominant smooth hypoplastic with eruption defects and resorption of teeth. b. Autosomal dominant rough hypoplastic c. Autosomal dominant pitted hypoplastic d. Autosomal dominant local hypoplastic e. X-linked dominant rough hypoplastic 2. Hypocalcified A. Autosomal dominant hypocalcified 3. Hypomaturation B. X-linked recessive hypomaturation C. Autosomal recessive pigmented hypomaturation

	D. Autosomal dominant snow-capped teeth E. White hypomature spots.
Winter and Brook, 1975 Winter and Brook, 1975 (continued)	This classification was based primarily on phenotype. Four main categories: hypoplasia, hypocalcification, hypomaturation and hypomaturation-hypoplasia with taurodontism. Mode of inheritance was proposed as a secondary means of sub-classification (Winter and Brook, 1975). a. Hypoplasia • Type 1. Autosomal dominant thin and smooth hypoplasia with eruption defects and resorption of teeth • Type II. Autosomal dominant thin and rough hypoplasia • Type III. Autosomal dominant randomly pitted hypoplasia • Type IV. Autosomal dominant localised hypoplasia • Type V. X-linked dominant rough hypoplasia b. Hypocalcification • Autosomal dominant hypocalcification c. Hypomaturation • Type 1. X-linked recessive hypomaturation • Type II. Autosomal recessive pigmented hypomaturation • Type III. Snow-capped teeth d. Hypomaturation-hypoplasia with taurodontism

	• Type I. Autosomal dominant smooth hypomaturation with occasional hypoplastic pits and taurodontism • Type II. Autosomal dominant smooth hypomaturation with thin hypoplasia and taurodontism
Witkop and Sauk, 1976	This classification was based on phenotype and mode of inheritance, similar to classification of Witkop and Rao (1971) (Witkop and Sauk, 1976).
Sundell and Koch, 1985	This classification was based solely on phenotype (Sundell and Koch, 1985).
Witkop, 1988 revised by Nusier et al 2004 (in italics)	Four major categories based primarily on phenotype (hypoplastic, hypomaturation, hypocalcified, hypomaturation-hypoplastic with taurodontism) subdivided into 15 subtypes and secondarily by mode of inheritance (Witkop, 1988; Nusier et al., 2004). **Type I Hypoplastic:** • IA—hypoplastic, pitted, AD (autosomal dominant) • IB—hypoplastic, local, AD • IC—hypoplastic, local, AR (autosomal Recessive) • *IC—local hypoplastic, AR* • ID—hypoplastic, smooth, AD • IE—hypoplastic, smooth, X-linked dominant • IF—hypoplastic, rough, AD

	• IG—enamel agenesis, AR • *IG—generalised thin hypoplastic, AR* **Type II Hypomaturation:** • IIA—hypomaturation, pigmented, AR • *IIA—Pigmented hypomaturation, AR* • IIB—hypomaturation, X-linked recessive • IIC—hypomaturation, snow-capped teeth, X-linked • IID—hypomaturation, snow-capped teeth, AD? **Type III Hypocalcified** • IIIA, AD • IIIB, AR • *IIIB—hypocalcified, AR* **Type IV Hypomaturation-hypoplastic with taurodontism** • IVA—Hypomaturation-hypoplastic with taurodontism, AD • IVB—Hypoplastic-hypomaturation with Taurodontism, AD
Aldred and Crawford, 1995	This classification was based on (Aldred and Crawford, 1995): • Molecular defect (when known)

	• Biochemical findings results (when known) • Mode of inheritance • Phenotype
Hart et al. 2002	Proposed a molecular defect sub classification of the AMELX conditions (Hart et al., 2002b) 1.1 Genomic DNA sequence 1.2 cDNA sequence 1.3 Amino acid sequence 1.4 Nucleotide and amino-acid sequences 1.5 AMELX mutations described to date
Aldred et al., 2003	This classification was based on (Aldred et al., 2003b): • Mode of inheritance • Phenotype— Clinical and Radiographical • Molecular defect (when known) • Biochemical result (when known)
Wright JT, 2006	Based on mode of inheritance and molecular genetics, three types of AI were described (Wright, 2006): **X-LINKED AMELOGENESIS IMPERFECTA** • *AMELX* mutations that are diverse and include Deletion, Missense, and Nonsense Mutations

	• *AMELX* Major Deletions and Mutations in the Signal Peptide Coding Region • *AMELX* Mutations in the N Terminus Coding Region • *AMELX* Mutations in the C Terminus Coding Region **AUTOSOMAL DOMINANT AMELOGENESIS IMPERFECTA** • *ENAM* Mutations: Genotype and Phenotype • Autosomal Dominant Hypocalcified AI • Autosomal Dominant Hypoplastic Hypomaturation AI With Taurodontism **AUTOSOMAL RECESSIVE AMELOGENESIS IMPERFECTA** • Enamelysin (*MMP20*) Mutations: Genotype and Phenotype

Although AI is considered here as a non-syndromic defect, many other dental anomalies may be associated with the enamel defects; such as pulp calcification, taurodontism, delayed eruption and sometimes, gingival overgrowth (Collins et al., 1999). Ravassipour and co-workers investigated 54 AI affected and 34 unaffected family members from 18 different kindred using cephalographs and found that forty two percent of AI affected individuals and 12% of unaffected family members had dental or skeletal open bite malocclusion. He also found that skeletal open bite malocclusion was variably expressed in AI affected individuals and that enamel phenotype severity did not necessarily correspond with the presence or severity of open bite malocclussion. He further concluded that the formation of maxillary and mandibular bones may be abnormal, leading frequently to open-bite malocclusion and less commonly to prognathism (Ravassipour et al., 2005).

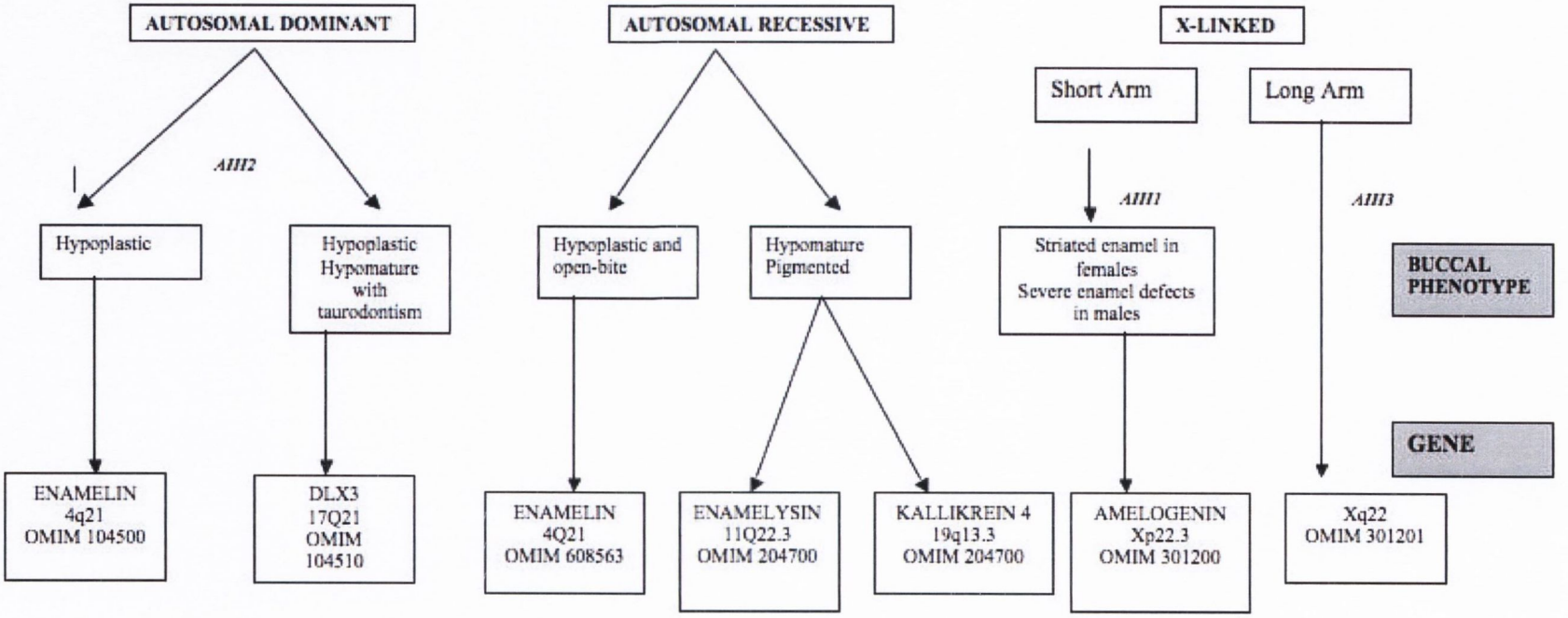
1.2.5 Aetiology

Most AI are inherited in a dominant manner, with major intrafamilial variability in expressivity (Kim et al., 2006b). The most commonly affected gene is enamelin [ENAM (4q21) AIH2-OMIM 104500] (Dong et al., 2000). Enamelin mutations are preferentially associated with hypoplastic AI (Rajpar et al., 2001). Eight enamelin mutations have been identified to date (two of these are however associated with a recessive transmission mode of inheritance). In a family with AI type hypoplastic-hypomaturation with taurodontism and autosomal inheritance (AIHHT) a mutation was reported within the homeodomain of DLX3 (17q21) (Dong et al., 2005). Previous studies have shown a DLX3 mutation outside the homeodomain associated with tricho-dento-osseous syndrome (TDO), suggesting that TDO and some forms of AIHHT are allelic (Wright et al., 1997). Figure 1-2 shows a diagnostic tree of non-syndromic amelogenesis imperfecta.

Several other candidate genes proposed on the basis of their expression in ameloblasts were considered: tuftelin (TUFT1) (1q21) (Deutsch et al., 1994), ameloblastin (AMBN) (4q21) (MacDougall et al., 1997), amelotin (AMTN) (4q13.3) (Iwasaki et al., 2005). Moreover AI has been mapped to 8q24.3 in a large pedigree, segregating an autosomal dominant form (Mendoza et al., 2007). In several surveys, all known loci have been excluded (Hart et al., 2003; Kim et al., 2006b; Santos et al., 2007) indicating that not all candidate genes have been identified to date.

The genetic analysis, inheritance pattern of AI and its associated syndromes are beyond the scope of this thesis.

Figure 1-2: Diagnostic tree of non-syndromic amelogenesis imperfecta (adapted from Bailleul-Forstier et al, 2008).



Diagnostic tree of non-syndromic amelogenesis imperfecta. Major clinical variations could be observed considering the important variability of the AI phenotype, even in a single patient or in a same family. Candidate genes: ameloblastin (4q21), tuftelin (1q21) amelotin (4q13.3) or genes related with syndromic disease (MSX2) are not included in the clinical diagnostic tree.

1.2.6.1 Clinical

The family history, pedigree plotting, clinical observation and meticulous recording form the backbone of diagnosis in this, as in any potentially inherited condition. Extra-oral radiographs may reveal the presence of unerupted and sometimes spontaneously resorbing teeth. Intra-oral radiographs will reveal the relative contrast between enamel and dentine in cases where mineralisation may have been affected. The same films in conjunction with clinical observation will provide information on the degree of any enamel hypoplasia (Ng and Messer, 2009).

1.2.6.2 Genetic Diagnosis

Laboratory genetic diagnosis is presently only a research tool and has not been deployed as a routine clinical diagnostic tool yet. This is particularly difficult because of ethical as well as financial issues.

1.2.6.3 Differential Diagnosis

Extrinsic disorders of tooth formation, chronological disorders of tooth formation and localised disorders of tooth formation should be considered in the differential diagnosis. The teeth form a record of dysplasia (abnormal growth) during different life stages. Dental enamel does not "heal" other than at the microscopic level in the context of early demineralisation caused by caries (Crawford et al., 2007). As a result, many disturbances of metabolism, which may have resolved, as well as genetic conditions, will be reflected in the appearance of the teeth. Four simple questions have been proposed to facilitate a differential diagnosis (Barron et al., 2008):

(1) Does any one else in the family have a similar presentation?

(2) Are all of the teeth affected in a similar manner?

(3) Is there a chronological distribution to the clinical appearance?

(4) Is there anything in the past medical history which might have caused sufficient metabolic disturbance to affect enamel formation?

The answers to these four questions form the foundation for the differential diagnosis.

The commonest differential diagnosis is dental fluorosis (Sapir and Shapira, 2007). The variability of this condition, from mild white "flecking" of the enamel to profoundly

dense white colouration with random, disfiguring areas of staining and hypoplasia, requires careful questioning to distinguish from AI.

Fluorosis may present with areas of horizontal white banding corresponding to periods of more intense fluoride intake and may show the first or second premolars to be spared (chronological distribution). In the latter case, the history will often reveal excessive fluoride intake either in terms of a habit such as eating tooth paste in childhood, or related to a local water supply (Sapir and Shapira, 2007).

A similar distribution of findings—chronological enamel hypoplasia—can arise from one of many causes during the time of tooth formation. Ranging from gastrointestinal upset of a prolonged nature, such as coeliac disease (a diagnosis which may not be confirmed until later in life) and anti-leukaemic therapy. Causes such as these may be identified from the patient history and the chronological distribution of the markings seen. However, there are cases where no identifiable cause can be elucidated and the diagnosis of AI remains tentative in apparently sporadic cases of enamel defects. It is anticipated that molecular genetic tools will allow more precise diagnosis in the future (Crawford et al., 2007).

1.2.6.4 Genetic Counselling

There is a growing acceptance that a classification of (inherited) enamel defects based primarily or exclusively on phenotype (appearance) is problematic. For this reason, the mode of inheritance and underlying genomic changes are important discriminators. This is particularly so in relation to genetic counselling of affected individuals and their families. These conditions are often embarrassing, distressing and may lead to social exclusion. Sensitive interview and early supportive intervention are essential (Crawford et al., 2007).

1.2.7 Treatment

The supportive clinical care needed by these individuals is substantial both in terms of clinical and emotional demands. Patients have been known to cover their teeth with pieces of paper, chewing gum or other materials in order to mimic an "ordinary" appearance with at least one individual steeling money to fund his dental care (Aldred et al., 2003a).

The treatment is, as ever, based on the principles of prevention before intervention. However, in these patients' cases, intervention will likely be earlier and more radical than for other dental conditions (Crawford et al., 2007).

1.2.7.1 Dental Management of Affected Children and Adolescents

Common clinical problems include poor aesthetics, tooth sensitivity, and extensive tooth wear. AI management in childhood and adolescence is, therefore, aimed at improving aesthetics, reducing sensitivity, correcting or maintaining vertical dimension and restoring masticatory function (Ng and Messer, 2009). AI can have marked psychosocial effects on young patients with more severe phenotypes (hypocalcified phenotypes tend to present with more severe post-eruptive breakdowns, wear and tooth sensitivity than hypoplastic phenotypes) (Coffield et al., 2005). Consequently, early diagnosis, followed by appropriate preventive and restorative care performed over several treatment phases is essential to optimise the final outcome (Seow, 1993; Bouvier et al., 1996; Ayers et al., 2004; Coffield et al., 2005). The initial phase of treatment should begin soon after diagnosis in the primary or mixed dentition and is followed by a transitional phase which provides the patient with a functional and aesthetic permanent dentition prior to the permanent phase in adulthood (Bouvier et al., 1996). Early multidisciplinary management maximises the treatment options for the permanent dentition (Ayers et al., 2004).

Casein and sensitivity prevention

As the quantitative and/or qualitative enamel defect results in an inherently elevated risk of caries and tooth wear, aggressive preventive protocols optimizing the use of topical fluorides, Casein PhosphoPeptide-Amorphous Calcium Phosphate (CPP-ACP) products, and dietary modification should be implemented early. Generalised sensitivity must be managed, as it may affect the oral health-related quality of life and/or interfere with effective tooth-brushing (Coffield et al., 2005). Yengopal investigated the current literature in a systematic review with meta-analysis to investigate the effects of CPP-ACP on caries and concluded that CPP-ACP had short-term remineralization effect on demineralised enamel (Yengopal and Mickenautsch, 2009). Topical fluoride and CPP-ACP products applied to dysmineralised enamel may aid remineralisation, reducing tooth sensitivity to thermal stimuli and reducing caries risk (Bader, 2010). The possible benefits of CPP-ACP in amelogenesis imperfecta have yet to be elucidated.

Maintaining good oral hygiene is important, as poor gingival health complicates restorative management and can contribute to future periodontal disease. Oral hygiene measures may be complicated by vertical grooves involving the crown in X-Linked AI females, which may extend sub-gingivally in young children. Rough hypoplastic or dysmineralised enamel at gingival margins may also enhance plaque accumulation and calculus formation (Seow, 1993).

Bonding with adhesive restorations

Bonding to AI enamel is challenging because of uncertain acid-etch patterns and bond strengths. Etch patterns of five AI variants were studied including: (1) pitted hypoplastic; (2) smooth hypoplastic; (3) X-linked hypoplastic (male); (4) X-linked hypoplastic (female); and (5) hypoplastic enamel. The three classical etch patterns (Silverstone et al., 1975) were found in the pitted hypoplastic (type I etch pattern, where etching of prism core is predominant), X-linked hypoplastic (female; type II etch pattern, where etching of prism peripheries is predominant), and hypocalcified (type 1-III etch patterns, [type III, where no prism structure is evident]) samples, suggesting bonding may be possible in these forms of AI (Seow and Amaratunge, 1998). Typical etch patterns were absent in the smooth hypoplastic sample, possibly due to abnormalities in prism structure, insufficient etchant concentration or etching time (Seow and Amaratunge, 1998). The X-linked hypoplastic (male) sample showed a patchy loss of surface structure without evidence of etch patterns (Seow and Amaratunge, 1998).

In vitro, enamel, and dentine shear bond strengths for primary teeth from a 12 year old subject with hypocalcified AI were significantly lower than that for the primary teeth from children with no apparent developmental defects of enamel (Saroglu et al., 2006). Pretreating hypocalcified enamel with 5% sodium hypochlorite produced more favourable patterns by removing the acid insoluble proteins surrounding crystals and increased enamel shear bond strengths significantly (Venezie et al., 1994; Saroglu et al., 2006). Very little clinical evidence exists, however, to support the use of such preparative procedures.

Intra-coronal restorations

Direct restorative material, such as glass ionomer cements, resin-modified glass ionomer cements, compomers, resin composites, and stainless steel crowns (SSCs) or

indirect resin composites or cast onlays and crowns, may be used to improve aesthetics and/or teeth with posteruptive breakdown due to caries or wear. Glass ionomer cements, compomers, and resin composites may be successful as interim restorations, but ongoing fracture of hypocalcified enamel may result in defective margins. Intra-oral dental radiographs can provide information regarding enamel thickness and the extent of mineralisation of erupted teeth and anticipate the condition of enamel of unerupted teeth (Collins et al., 1999).

Although bonding to hypoplastic enamel is feasible, thus reducing tooth sensitivity in local hypoplastic and smooth hypoplastic phenotypes, sufficient enamel must be available for bonding or full coverage restorations may be required (Collins et al., 1999). In patients affected by dysmineralisation phenotypes where posteruptive breakdown has resulted in enamel loss and subsequent dentine exposure, using dentine bonding agents or glass ionomer cements prior to resin composite placement may enhance bonding of resin composite veneers or anterior strip crowns (Seow, 1993). Full coverage restorations are required commonly for posterior teeth, particularly in hypocalcification phenotypes due to extensive wear and the need to prevent further posteruptive breakdown (Seow, 1993).

Full coverage restorations

In the primary and young permanent dentition, full coverage restorations are perhaps the most effective method to manage tooth sensitivity and poor aesthetics. In young children, stainless steel crowns (SSCs) are often the restoration of choice to restore severely broken down primary and permanent molars, maintain vertical dimension, and manage tooth sensitivity (Kwok-Tung and King, 2006; de Souza-e-Silva et al., 2010). Older patients may receive cast onlays or full crowns. When SSCs and cast restorations were used to treat permanent first molars affected by AI or severe enamel defects, no significant differences in longevity or quality of the restorations were found over 24 months (Zagdwon et al., 2003). Stainless steel crowns are simple and quick to place, particularly when managing young children under general anaesthesia. However, they require more tooth preparation, committing teeth to full coverage restorations at 18 to 20 years of age for periodontal reasons. Use of separating elastics prior to tooth preparation allows SSCs to be cemented without proximal tooth reduction (Seow, 1993). Cast onlays fabricated from base metal or gold alloys are more conservative as

they may be fabricated without prior tooth preparation (Harley and Ibbetson, 1993; Hunter and Stone, 1997).

Onlays are frequently placed in supraocclusion (up to 3 mm at the permanent first molars), yet 7-18 year olds re-equilibrated their occlusion within 3 months after cementation (Harley and Ibbetson, 1993). Onlays, however, are costly and technique sensitive. They require a minimum of two chairside appointments and rely heavily on child cooperation, which may be compromised by sensitivity to thermal stimuli during impression making and cementation. Cast onlays are, therefore, used primarily to provide occlusal coverage in less severely affected molars in older cooperative patients (Sengun and Ozer, 2002).

Aesthetic improvements

Several clinical reports advocated different treatment modalities which included an interim aesthetic improvement of discoloured or hypoplastic primary and permanent teeth which could be achieved by bonding direct or indirect resin composite restorations including strip crowns (Seow, 1993; Sengun and Ozer, 2002; Vitkov et al., 2006). To reduce the need for mechanical tooth preparation, discoloured permanent teeth may benefit from microabrasion and/or tooth whitening prior to resin composite veneers or crowns (Ashkenazi and Sarnat, 2000; Ng and Manton, 2007). Heavily discoloured teeth may require layering techniques in resin placement and opaquers to improve the aesthetic outcome (Vitkov et al., 2006). These treatment approaches may help improve aesthetic and function for amelogenesis imperfecta patients; however, their long-term outcomes still require further prospective investigation.

1.3 Quality of life of patients suffering from Hypodontia and Amelogenesis Imperfecta

1.3.1 Dental genetic disorders

The disorders, which are the primary focus of this thesis, are hypodontia and amelogenesis imperfecta.

1.3.1.1 Hypodontia

Hypodontia is a failure of development of teeth. It varies in severity from single tooth absence to complete lack of any teeth (anodontia). Sporadic forms, as well as X-linked, autosomal-dominant and autosomal-recessive forms, may occur (Bailleul-Forestier et al., 2008b). In some cases there may be associated ectodermal abnormalities, best illustrated in cases of X-linked hypohidrotic ectodermal dysplasia (Bailleul-Forestier et al., 2008a).

1.3.1.2 Amelogenesis Imperfecta

Amelogenesis imperfecta (AI) is a disfiguring condition of the dental enamel, which can be inherited as autosomal dominant (63%) autosomal-recessive (12%) or X-linked autosomal (6%) forms (Backman, 1988; Nusier et al., 2004). Sporadic cases are also encountered and it is anticipated that ultimately the molecular basis of such cases will be elucidated (Wright, 2006). The enamel may be reduced in quantity (hypoplasia) or of poor quality, appearing yellow/brown as a result of a mineralisation defect (variously described as hypomineralisation, hypocalcification, or dysmineralisation). AI can occur alone or in a combination of variety of syndromes (Bailleul-Forestier et al., 2008a).

1.3.2 Background

1.3.2.1 Public Awareness of Developmental Dental Anomalies

Many individuals and families with genetic disorders are aware of the significant advances in molecular genetics (Aldred et al., 2003a). The ethical issues regarding pre-natal diagnosis and other options, including gene therapy for life-threatening disorders maintain a high profile in public debate. The issues of pre-natal testing, or the options afforded by *in vitro* fertilisation and selection as well as gene therapy in dental genetic disorders, have not been considered. Whilst some might hold the view that "it's only teeth", for some of these families this single issue may be the reason for deciding

whether or not to have children (Aldred et al., 2003a). A better informed public is now asking what reproductive options are available to them. This family pressure and the increasing body of knowledge allows the possibility—and demands the facility—for families to be given the best possible information on which to base their decisions (Aldred et al., 2003a).

1.3.2.2 Effects on Families

Developmental dental disorders are not associated with significant mortality "defined as a fatal outcome or death"; however, there is a high degree of morbidity "defined as the rate of disease or proportion of diseased persons in a given locality or nation" and that the effects on individuals and their families should not be under-estimated.

The diagnosis of developmental dental disorders such as hypodontia and amelogenesis imperfecta can have a number of implications for the affected individuals and their family. This is particularly the case for a diagnosis of severe hypodontia or anodontia (the total absence of teeth). This diagnosis can carry a number of dental, medical, psychosocial, functional, educational and financial implications (Bergendal et al., 1998). Patients and immediate family members should be counselled about such implications when a diagnosis of developmental dental disorder is made to enable the patient and his/her family to prepare for these issues and minimise their future impact.

1.3.2.3 Psychosocial Implications

Given the current focus on aesthetics, it is reasonable to speculate that deviations from "ideal" or "normal" dental and orofacial aesthetics could be detrimental to a person's psychosocial well-being (Locker, 1989b; Klages et al., 2004). Studies have documented the effects of malocclusion and motivations for orthodontic treatment and orthognathic surgery (Helm et al., 1985; Phillips et al., 1997; Phillips et al., 1998). In a follow up study Helm and co-workers investigated 977 Danish adolescents who were recorded as having malocclusion in a national survey of schoolchildren without access to organised orthodontic care (original sample size was 1,252). Fifteen years later, a questionnaire containing questions about body image and specific questions concerning self perception and social implications of dental appearance was sent to this patient population. Only 10% of this patient population had orthodontic treatment (which were removed from the study), and the remaining 758 subjects were investigated and

compared to 152 patients used as controls with no malocclusion. The authors concluded that some traits of malocclusion such as extremely increased horizontal or vertical overlap were identified as a potential threat to one's body image and self-concept in both adolescence and adulthood, with dissatisfaction with dental appearance and schoolmate's teasing as important factors (Helm et al., 1985).

Many patients who opt to have orthognathic surgery have primarily a self-image motivation for treatment, as reported by Phillips and co-workers who investigated the motives for treatment of 135 patients whose orthodontic conditions required orthognathic surgery. He found that 16% of the patients had primarily a self-image motivation, whereas 4% had an oral function motivation. He also found that 6% of the total population had a strong dual self-image/oral function motivation, 5% reported dual oral function/TMJ motivation, and 6% reported self-image/TMJ motivation. Only 2% of the total population reported strong motives for all dimensions. The remaining 63% reported no strong motive dimensions. (Phillips et al., 1997). A second study of 194 patients who required orthognathic surgery investigated the level of psychological distress in this patient population during their initial visit using questionnaires and found that over 15% of patients reported experiencing a level of psychological distress concerning their skeletal disharmony that warranted intervention (Phillips et al., 1998). No control group was used but scores were compared to the scale's norm scores. It would have also been interesting to see if surgical intervention had reduced the level of psychological stress after treatment.

Shaw investigated the social and psychological influences of dental and facial appearance in 840 school children and 840 adults using a modified black and white photographs of an attractive boy and girl and an unattractive boy and girl in which the face was standardised and only the dentofacial arrangements have been modified. Each photograph was viewed by forty two children and forty two adults to evaluate their social attractiveness on a visual analogue scale. He concluded that normal dental appearance has an important influence on people's perception of friendliness, social class, popularity and intelligence of an individual (Shaw, 1981). In a second study of similar design, using 800 young school children to evaluate the modified photographs, Shaw concluded that the most salient feature was the protrusion of the upper teeth, and that a unilateral cleft consistently attracted low ratings. He also concluded that the

background facial attractiveness appears to be more influential than the individual's dental condition (Shaw et al., 1985).

A further problem with facial and dental deformity is that in social interaction it is invariably impossible to hide or disguise it, because in normal interaction the eyes attend the face (Macgregor, 1970).

Bullying within school children is a common problem. The prevalence of bullying and victimisation in elementary schools (grade 1-5) varies from 11.3% in sample of 5813 students in Finland (Kumpulainen et al., 1998) to a high of 49.8% in a nationwide survey of 8249 students in Ireland (O'Moore and Kirkham, 2001). The difference in prevalence could be related to methodogical differences between the two studies in grouping subjects' responses. In the former study anchor statements were ("Is bullied occasionally" and "Is Bullied certainly"), whereas in the second study the anchor statements were ("Once or twice", "sometimes", "about once a week", and "several times a week").

In a meta analysis review of cross sectional studies of available literature on bullying, it was concluded that chronic bullying can lead to depression, loneliness, anxiety, low self-esteem and underachievement at school (Hawker and Boulton, 2000). This was further validated and related to dental appearance and the effects of malocclusion (DiBiase and Sandler, 2001).

In school children, deviations in dental appearance have been found to be a target for teasing, although the group of children to whom this applied was also found to be at a higher risk of harassment generally. The greater the deviation of the dental appearance, the greater the implication to the child, and comments about teeth appeared to be more hurtful than those about other features (Shaw et al., 1980; Akram et al., 2011).

Developmental dental disorders can impact significantly on dental aesthetics. Factors, which contribute to a decrease in dental aesthetics, include:

- The severity of the condition.
- Abnormalities in tooth shape and colour i.e. microdontia, pitted enamel, and discoloured teeth.
- Abnormal tooth position.

Features such as spacing, as in hypodontia, or discolouration, as in amelogenesis imperfecta, may become a focus for bullies and parents may insist upon treatment on this basis. However, research in this area is lacking.

1.3.2.4 Functional Implications

A paucity of research has been carried out on the functional implications of hypodontia. One study suggested that masticatory difficulty is seldom a problem with severe hypodontia (Hobkirk et al., 1994) although this was solely based on a questionnaire. In a systematic literature review of amelogenesis imperfecta it was reported that dental hypersensitivity was the symptom which severely influenced the patients' daily lives and was subsequently reported in twenty clinical case reports (Poulsen et al., 2008).

1.3.2.5 Educational Implications

The dental management of developmental dental disorders can place a considerable commitment on the family with frequent appointments required for treatment and reviews over childhood and adolescence. Orthodontic treatment, which can last up to three years, can be during an educationally critical period of childhood and if appointments are spaced every six weeks may result in significant time away from school. A child may suffer from other medical problems that require appointments with other hospital specialists, which may complicate matters further. In one study the dental pain experience and its impact on four hundred and nine Saudi Arabian school children and their families was investigated (Stewart et al., 2002). Students were randomly selected and asked about their dental pain experience over the last four weeks. Those who reported having pain during these four weeks completed an interview questionnaire related to the severity, duration and impact of pain, and were clinically examined to determine the cause. He found that approximately 67% of the total sample reported a previous history of dental pain, and 40% had pain within the last month prior to the interview. Among those reporting dental pain within the last month 82% of the pain was due to pathological reasons. Dental pain prevented students from eating (60%), playing (42%), sleeping (43%) and going to school (27%). Also, 15.6% of parents/guardians had to leave their work place to take children to the dentist. He concluded that dental pain clearly affected the quality of the children's lives and had an impact on families and the society as a whole.

It is important for clinicians to be empathetic about parental concerns in such cases. In order to reduce the impact of dental treatment on education, the following advice should be considered (Gill et al., 2008):

- Parents should be advised to have a school diary when making appointments so that these can be placed during holiday periods whenever possible.
- Children undergoing treatment during examination years should be prioritised to appointment slots after school hours.
- If appointments must be scheduled during school times, care should be taken that the same subjects are not missed on each occasion and less educationally demanding subjects are interrupted instead, such as physical education.

1.3.2.6 Financial Implications

A diagnosis of developmental dental disorders can have financial implications for the affected individual and his/her family. It has been reported in the literature that a patient affected by amelogenesis imperfecta stole money to fix his teeth after years of teasing and failed relationships (Aldred et al., 2003a).

In Ireland the Health Service Executive (HSE) funds dental treatment for young patients in Primary Care and Orthodontics, but funding for patients with special dental needs was not organised and depended on the region where the patient was resident. In 2002 a Special Dental Needs clinic was established in the Dublin Dental University Hospital. This tertiary referral centre is dedicated to the treatment of patients with developmental dental disorders and associated conditions.

In the United Kingdom the National Health Service funds dental care for children and implant-related treatment for young adults with six or more missing teeth. Some secondary and tertiary care is available for patients affected by these conditions from the Community Dental Service in childhood and by the dental teaching hospitals throughout adult life. However, there is no clear policy on this issue and there are insufficient resources in the system to undertake the continuing care of all such individuals. The General Dental Service (National Health Service) do not include developmental dental defects as a reason for exemption from charges (Aldred et al., 2003a).

Australian families report substantial costs of up to $A 40, 000 (≈ $US 37, 000 and ≈ € 27,600) for full mouth dental rehabilitation (tooth restoration), none of which is covered under the Australian Federal Government Medicare Scheme and little of which can be recovered through private health insurance. Although Australian states and territories provided free or subsidised dental treatment for children, the complexity of treatment needs, limited funding, and the fact that much of this treatment is required in late adolescence, means that most children are unable to benefit from any such provisions (Aldred et al., 2003a).

In Canada dental care has largely been dealt with as a market mechanism, meaning that it is mainly privately financed and delivered. It is delivered by dentists and other dental workers in private practices, and financed through out-of pocket expenditures and/or employment-based health insurance. Canada has also dealt with dental care in terms of welfare-based provision, meaning that it targets public support for individuals considered in social need (e.g., social assistance recipients, low-income children, disabled individuals). Delivery to such groups can either be direct and/or indirect through part-funding. Finally, Canada has dealt with dental care in terms of service-based provision, meaning that it finances selected dental services delivered in hospitals that are considered a medical need (e.g., surgical-dental services for those with craniofacial anomalies) (Carlos, 2009).

Another financial implication to be considered is the time parents have to take off work to accompany their children for treatment (particularly self-employed parents) and travel costs (Gill et al., 2008).

Quality of life in the present context depends on the inherent severity of a genetic disorder and also on the financial and social resources available for a person to cope with that abnormality. Inherited dental disorders can be considered to fall into two categories: those which have minor-to-moderate impact on an individual's life, and those which have a serious impact on their life. Some shocking extracts from correspondences with families of patients affected by amelogenesis imperfecta and dentinogenesis imperfecta ranging from being upset about their child's dental condition, to not wanting to have children in the future, or even reports of children wanting to die because of their dental conditions have been reported (Aldred et al., 2003a).

1.3.3 Definitions

1.3.3.1 Definition of Health

A state of complete physical, mental and social well-being and not merely the absence of disease or infirmity (WHO, 1946).

1.3.3.2 Definition of Quality of Life (QoL)

Quality of life is an amorphous concept that has a usage across many disciplines—geography, literature, philosophy, health care economics, advertising, health promotion and the medical and social sciences (e.g. sociology and psychology). It is a vague multidimensional concept and theoretically incorporates all aspects of an individual's life (Bowling, 1995a).

Quality of life is defined by the World Health Organisation Quality of Life Group as "an individual's perception of their position in life in the context of the culture and value systems in which they live and in relation to their goals, expectations, standards and concerns. It is a broad ranging concept affected in a complex way by the person's physical health, psychological state, level of independence, social relationships, and their relationships to salient features of their environment" (WHOQOL, 1993b).

1.3.3.3 Definition of Health Related Quality of Life (HRQoL)

This is a patient-based concept which focuses more on the impact of a perceived health status on the ability to live a fulfilling life (Bullinger et al., 1993). It is defined as "optimum levels of mental, physical, role (e.g. work, parent, carer, etc.) and social functioning, including relationships, and perceptions of health, fitness, life satisfaction and well-being". It should also include some assessment of the patient's level of satisfaction with treatment, outcome and health status and with future prospects. It is distinct from quality of life as a whole, which would also include adequacy of housing, income and perceptions of the immediate environment (Bowling, 1995a).

1.3.3.4 Definition of Oral Health and Oral Health Quality of Life

"When talking about oral health, our focus is not on the oral cavity itself but on the individual and the way in which oral disorders, disease and condition threaten health, well-being and quality of life" (Locker, 1997). Kressin broadly defined oral health quality of life as "a broad conception of health, encompassing the traditional definition

of health, as well as an individual's subjective impact of oral health on well being and functioning in every-day life" (Kressin, 1997). Later oral health quality of life was defined simply as "the extent to which oral disorders affect functioning and psychosocial wellbeing" (Locker et al., 2000). Oral health quality of life was also defined as "the symptoms and functional and psychosocial impacts that emanate from oral disease and disorders" (Locker et al., 2002b). More recently Locker and Allen defined subjective oral health as "the functional and psychosocial impacts of oral diseases and conditions as perceived by the individual" (Locker and Allen, 2007). They also defined oral health quality of life as "the impact of oral disorders on aspects of everyday life that are important to patients and persons, with those impacts being of sufficient magnitude, whether in terms of severity, frequency or duration, to affect an individual's perception of their life overall" (Locker and Allen, 2007). Locker and Allen further explained that measures of subjective health status offer a description of a person's current status while measures of health related quality of life also offer a subjective evaluation of that status (Locker and Allen, 2007).

1.3.3.5 Oral Health Related Quality of Life (OHRQoL)

To date the term "oral health-related quality of life" has no strict definition. However, an example of a practical definition is one provided by the United States Surgeon General's report on oral health which defines OHRQoL as a multidimensional construct that reflects (among other things) people's comfort when eating, sleeping, and engaging in social interaction; their self-esteem; and their satisfaction with respect to their oral health (Satcher, 2000). If HRQoL is an individual's assessment of how the following factors affect his/her well being: experience of pain/discomfort, physical function, psychology and social function, it is reasonable that when those considerations center on oro-facial concerns, oral health-related quality of life is assessed (Inglehart and Bagramian, 2002).

Locker and Allen demonstrated that some of the definitions of oral health, oral health quality of life and indeed oral health related quality of life suggest that health related quality of life equates health, while others imply that it is something more than health, encompassing additional and broader dimensions of human experience (Locker and Allen, 2007). Leplege and Hunt explained that the shift in nomenclature from "oral health status" to "oral health related quality of life" was based on the assumption that

since measures address aspects of functioning that are compromised by oral diseases and disorders of various kinds, they must necessarily indicate how these diseases and disorders affect the quality of life (Leplege and Hunt, 1997). They further explained that this was not the case as the "implications of measuring health status are quite different from those of measuring quality of life" (Leplege and Hunt, 1997). Carr and Higginson found that patients with chronic conditions reported their quality of life as good, in spite of severe physical conditions. This clearly shows that health and health related quality of life are both conceptually and empirically distinct (Carr and Higginson, 2001).

1.3.4 The International Classification of Impairments, Disabilities and Handicaps (ICIDH)

Based on the work of Wood in 1980 (Wood, 1980a; Wood, 1980b) the WHO endorsed the ICIDH as a conceptual framework for disability with a glossary of terms such as impairment, disability and handicap. These three principal components operate independently with impairment addressing impact on the body; disability to impact on the person; and handicap to impact on the person interacting with the environment.

In recent times there has been a surge of interest in quality of life research. Contemporary assessment of health status should routinely involve looking into patient's experiences and perceptions. Many factors may have an impact on quality of life and on health that are not directly related e.g. finance, and environment so it is important to distinguish between QoL and HRQoL.

Locker adapted the ICIDH framework to portray the functional and psychosocial impact of chronic oral disease and dysfunction as a progression from impairment to handicap, through intermediary steps of functional limitation and disability (Figure 1-3 below) (Locker, 1988).

1.3.4.1 Impairment

Impairment is defined as any anatomical loss, structural abnormality or disturbance of physical or psychological process, either present at birth or arising out of disease or injury. Measures of impairment give an indication of the nature and extent of

anatomical loss or structural abnormality associated with disease, injury or birth defects (Locker, 1988; Locker, 1989b).

1.3.4.2 Functional limitation

Functional limitation completes the set of concepts necessary for measurement of disease and its biophysical consequences. Functional limitations are defined as restrictions in the functions customarily expected of the body or its component organs or systems (Locker, 1988; Locker, 1989b).

1.3.4.3 Discomfort

It is a self-reported physical and psychological distress, including pain and other feeling states not directly observable (Locker, 1988; Locker, 1989b).

1.3.4.4 Disability

It is defined as any limitation in or lack of ability to perform the activities of daily living. While early measures of disability focussed upon a narrow range of such activities, most frequently mobility, body movement and self-care. Current approaches encompass the distinct dimensions of physical, psychological and social well-being (Locker, 1988; Locker, 1989b).

Physical well-being may be affected in two ways (Locker, 1988)

- Activity restrictions, which consist of acute or chronic limitations in such areas of life as ambulation, mobility, self-care, eating, sleep and rest.
- Limitations in usual social role, which involves problems in work, household management and recreation.

Psychological well-being may be affected in two ways (Locker, 1988)

- The disease may give rise to changes in emotional and affective states, all of which involve distress. Anxiety, depression, uncertainty, emotional fatigue, loss of volition and feelings of hopelessness are common complaints among those with chronic conditions.
- Cognitive functioning may also be affected and result in confusion, forgetfulness, and inability to make decisions with problems in thought and concentration.

Social well-being may be affected in three ways (Locker, 1988)

- Social integration, which reflects the extent of an individual's participation in community life and is assessed on the basis of membership of formal and informal organisations.
- Social contacts, which reflects the frequency of interactions with friends and relatives.
- Social and emotional intimacy, which is assessed in terms of the presence of one or more individuals who provide psychosocial and instrumental support.

1.3.4.5 Handicap

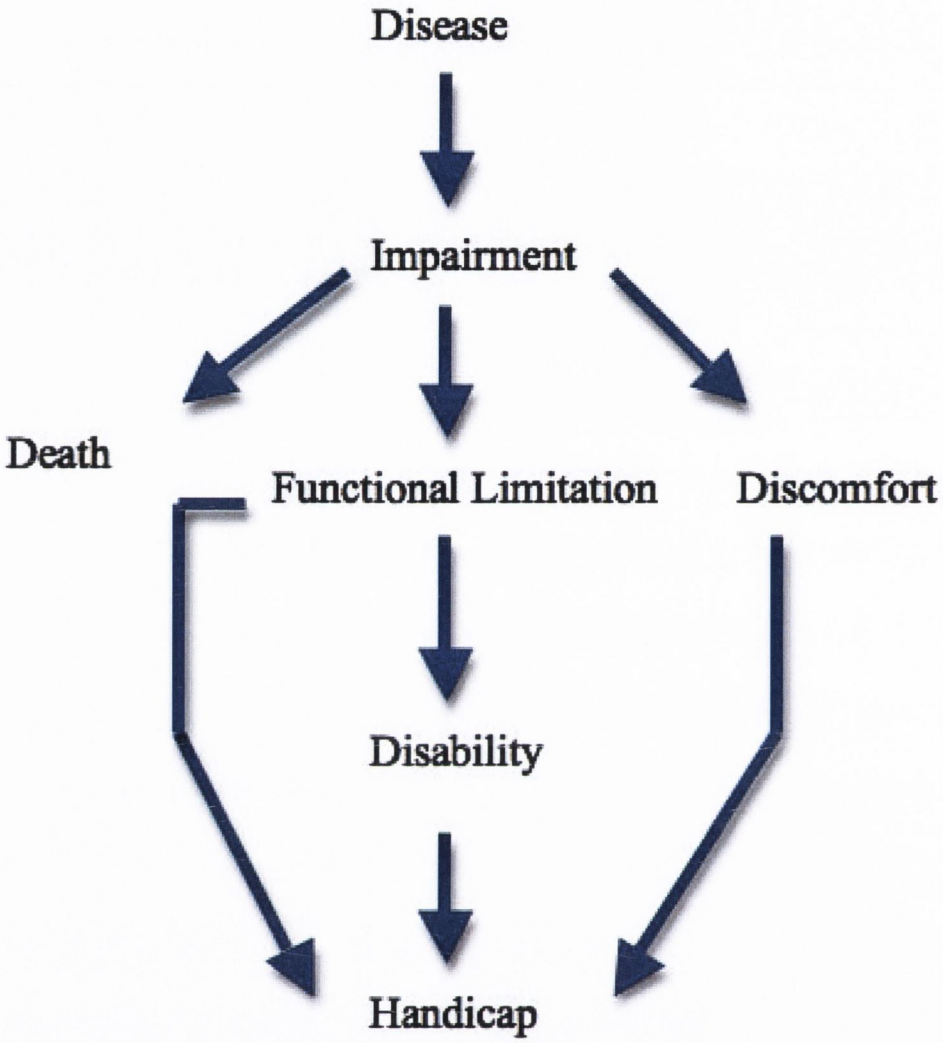
Handicap is defined as the disadvantage experienced by impaired and disabled people because they do not or cannot conform to the expectations of society or the social groups to which they belong. Disease and disability can have a negative effect on marriage and family formation, employment prospects, income, self-image and self-esteem, loneliness and social isolation. Some of these effects have been reported with respect to dental and oral disease. These diseases, either directly or because of their relationship to physical attractiveness, have been shown to affect the social acceptability and school performance of children (Shaw et al., 1980; Broder et al., 1994; Hunt et al., 2005). In a study of 196 cleft lip/palate patients it was found that as a group patients with clefts marry at a significantly lower rate when compared to unaffected siblings and controls. They also found that people with facial clefts are more likely to marry later in life, have less children or childless marriages (Peter and Chinsky, 1974). Similar findings were also reported by Broder and co-workers (Broder et al., 1994).

The conceptual model adapted by Locker can be summarised in the following section:

Handicap may come from impairment such as arthritis. This condition involves discomfort that causes a functional limitation and may lead to disability and handicap. Impaired dental function such as found in hypodontia and amelogenesis imperfecta may not cause disability or severe functional limitation but there still may be a social handicap. These developmental dental anomalies may lead to tooth loss, and if progressive, to an edentulous state. Dentures may not be well tolerated or may be poorly fitting which cause difficulty eating and this is described as a functional limitation. This results in a disability because people find it harder to eat. As many

patients may not be able to eat the food of their choice and are embarrassed by the eating difficulty they may have eating certain foods, they may eat in private, which is social handicap. Poor dental appearance may also be a social handicap, interfering with social interaction (Locker, 1988).

Figure 1-3: Conceptual Model (WHO) adapted from Locker (1988).



1.3.5 Recent changes in Perception

In the previous section, the International Classification of Impairments, Disabilities, and Handicaps adopted by the WHO was discussed and was based on "Social Role Theory" portraying disablement as a negative and socially unacceptable consequence of impairment. It has been the major conceptual influence on the construction of psychometric tools for dentistry (WHO, 2001). However, the current views of chronic disease are refocused on the influence on coping strategies used by people to prevent or limit disability and handicap called the "Positive Health Theory". Consequently, the WHO adopted the International Classification of Functioning, Disability and Health (ICF) as an alternative description of health and health-related states based on an existential view of the body, the person and society (MacEntee, 2006).

1.3.5.1 Social Role Theory (Utilitarian Model)

This theory presumes that everyone has a predetermined identity and set of biopsychosocial needs that demand conformity to the established modes of society (Hodge, 1990). It can be expressed as "functionalism", and it portrays each person as a functioning unit of society, and disease as a disturbance to social function and productivity (MacEntee, 2006).

1.3.5.2 Positive Health Theory (Existential Model)

This endorses the social environment as a dominant influence on personal values, behaviours, and interpretations of disability (Hodge, 1990). It emphasises that positive outlooks on health are associated usually with a social role determined by personal preferences rather than authoritarian principles.

1.3.6 The International Classification of Functioning, Disability and Health (ICF)

The International Classification of Functioning, Disability and Health (WHO, 2001) provides a standard language and framework for the description of health and health-related states. It is a classification of health and health-related domains. These domains describe changes in body function and structure, what a person with a health condition can do in a standard environment (their level of capacity), as well as what they actually do in their usual environment (their level of performance).

These domains are classified from body, individual and societal perspectives by means of two lists:

(1) A list of body functions and structure, and (2) A list of domains of activities and participation.

In the ICF classification, the term *functioning* refers to all body functions, activities and participation, while the term *disability* is an umbrella term for impairments, activity limitations and participation restrictions. ICF also lists environmental factors that interact with all these components.

The ICF classification document runs to 228 pages and is published by the World Health Organisation. It places its stress on health and functioning, rather than on disability, and shifts the focus from emphasising people's disabilities to their level of health. By shifting the focus from cause to impact, it places all health conditions on an equal footing allowing them to be compared using a common metric—the ruler of health and disability (WHO, 2001).

1.3.6.1 The Need for an ICF

Diagnosis alone does not predict service needs, length of hospitalisation, level of care or functional outcomes. Nor is the presence of a disease or disorder an accurate predictor of receipt of disability benefits. This means that using medical classification of diagnosis alone will not provide the information needed for health planning and management purposes as it lacks data about the level of functioning and disability (WHO, 2001). ICF allows the collection of these vital data in a consistent and internationally comparable manner.

1.3.6.2 The Model of the ICF

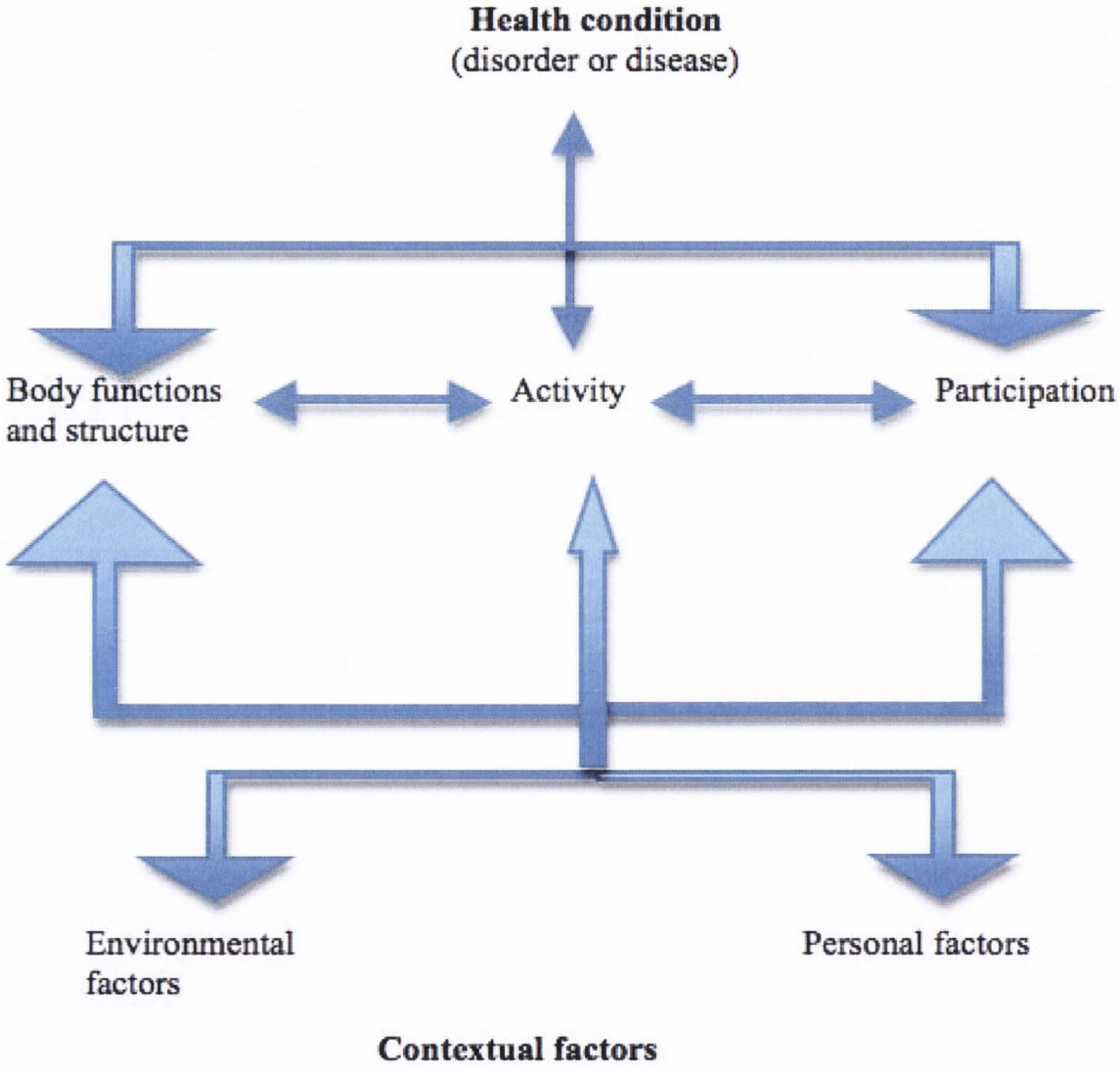
Two major conceptual models of disability have been proposed.

- The Medical Model, which views disability as a feature of the person, directly caused by disease, trauma, or other health condition, which requires medical care provided in the form of individual treatment by professionals. Disability, in this model, calls for medical or other treatment or intervention, to "correct" the problem with the individual.

- The Social Model, which identifies disability as a socially-created problem and not at all an attribute of the individual. Disability here demands a political response, since the problem is created by an unaccommodating physical environment brought about by attitudes about other features of the social environment.

On their own, neither model is adequate, although both are partially valid. The ICF adopts a biopsychosocial model based on integrating the medical and social models (WHO,2001).

Figure 1-4: The interaction between components of the International Classification of Functioning, Disability and Health (adapted from WHO, 2001).



Concepts of Functioning and Disability

As indicated in the previous figure, the ICF disability and functioning are viewed as outcomes of interactions between health conditions (disease, disorders and injuries) and contextual factors.

Among the contextual factors are external environmental factors, e.g. social attitudes, architectural characteristics, legal and social structures, as well as climate and terrain, in addition to internal personal factors, which include gender, age, coping styles, social background, education, profession, past and current experience, overall behaviour pattern, character and other factors that influence how disability is experienced by the individual (WHO, 2001).

The diagram also identifies the three levels of human functioning classified by the ICF:

- Functioning at the level of body or body part
- Functioning at the level of the whole person
- Functioning at the level of the whole person in a social context

Disability therefore, involves dysfunctioning at one or more of these levels: impairments, activity limitations and participation restrictions. The ICF classification defines these components as follows (WHO, 2001):

- *Body functions* are physiological functions of body systems (including psychological functions)
- *Body structures* are anatomical parts of the body such as organs, limbs and their components
- *Impairments* are problems in body function or structure such as a significant deviation or loss
- *Activity* is the execution of a task or action by an individual
- *Participation* is the involvement in a life situation
- *Activity Limitations* are difficulties an individual may have in executing activities
- *Participation restrictions* are problems an individual may experience in involvement in life situations

- *Environmental Factors* make up the physical, social and attitudinal environment in which people live and conduct their lives

1.3.7 Application of ICF in Dentistry

Despite the fact that ICF has been developed in 2001 and that it represented a huge departure from portraying disablement as a negative and socially unacceptable consequence of impairment to refocusing on the influence on coping strategies used by people to prevent or limit disability and handicap, its applications to dentistry have been very modest. Examples of these applications include developing new models of oral health (MacEntee, 2006), defining the population requiring special care dentistry (Faulks and Hennequin, 2006), and an attempt to develop the ICF into a brief head and neck cancer patient questionnaire (Rogers et al., 2010).

1.3.7.1 A new model of oral health

Application of the ICF to the information on the significance of the oral cavity offers a conceptual and an empirical foundation for a new interactive model illustrating relationships between the principal components of oral health. *A spherical representation of the relationships offers three interacting layers. The outer contextual or environmental layer consists of interacting personal and social factors and skills. The middle functional and structural layer contains the potential for impairments or deviations in structure that may or may not restrict participation or limit activities, while the inner layer contains the three domains of oral health that are particularly significant to people. Finally, the multidirectional arrows around and between the layers indicate the dynamic and fluctuating relationships both within and between the layers* (MacEntee, 2006).

Potential applications of the new psychosocial model

The developer of the model (MacEntee, 2006) stated some applications for the model in dentistry which can be summarised in the following points:

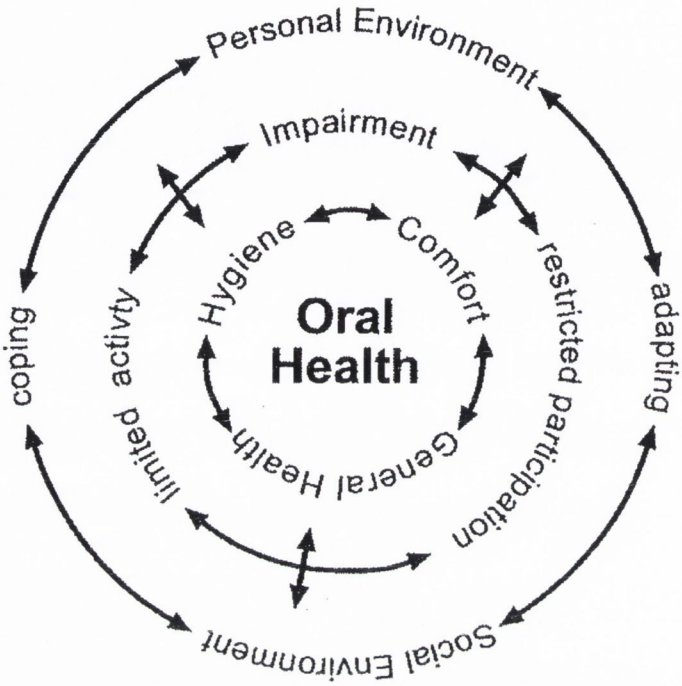
- It can be used to explore and explain dynamic relationships between major components or domains of oral health in lives of different populations. An example for this, is the acceptability of the impairment caused by missing maxillary anterior teeth in a football player because it may contribute positively to

his professional environment, while the same impairment may be considered environmentally as a severe handicap for a professional saxophonist.

- It can be used as a conceptual foundation for building and assessing new psychometric scales relevant to situations where there is a potential for positive as well as negative impact on oral health related quality of life. Patient's acceptance of the "shortened dental arch" in contrast to more sophisticated prosthodontic procedures highlights the functional adaptability of patients, and the need for a model that can accommodate both positive and negative responses to change. This may direct attention away from treatments that are particularly invasive and expensive when there is a possibility that patients can cope well with less expensive methods such as conventional dentures and implant overdentures.

The concepts embodied in the new ICF classification justify the revision of existing conceptual frameworks and models to provide a broader and more optimistic model of the external and internal factors influencing oral health (WHO, 2001).

Figure 1-5: New Model of Oral Health (Adapted from MacEntee, 2006).



1.3.7.2 ICF Derived Definition of Populations Requiring Special Care Dentistry

Faulks and Hennequin used the ICF classification to define populations requiring special care dentistry (Faulks and Hennequin, 2006). They argued that defining patient populations is essential in order to gather epidemiological data to describe the current health status and project the economic, medical and social consequences of disability and disease. They further explained that public health policy concerning health care and prevention can not be planned or implemented without the knowledge of the numbers and needs of the population concerned (Faulks and Hennequin, 2006).

Based on the fact that ICF provides a single classification and a common language for the description of functional ability in three dimensions: body function and structure, individual activity, and participation in society and that these dimensions are modified by environmental and contextual elements and personal factors; Faulks and Hennequin gathered and analysed all ICF items relating directly to the oral sphere, items having an influence on oral function and health, and all environmental factors affecting oral status. The analysis of all these items was then used to identify three groups with respect to oral health:

- First group: it was made up of persons who experience of disability due to impairment of oral function and/or structure and who were limited in their activity or participation directly by their oral status i.e. a child with cleft lip and palate (impairment of oral structure leading to impaired facial aesthetics and speech quality).
- Second group: consisted of those persons who had a given condition that had direct or indirect repercussions on their oral health i.e. a child with Down syndrome (the underlying oral impairment is aggravated by the limited possibility for self-maintenance of oral hygiene and access to care).
- Third group: it was made up of persons who were disabled by their social, environmental or cultural context, which reflected on their oral health i.e. a child with nursing caries (there is no underlying impairment of function, but limitation in activity and participation due to dental pain).

Based on the above groups Faulks and Hennequin defined populations in need of special care dentistry as " those with disability or activity restriction that directly or indirectly affects their oral health, within the personal and environmental context of the individual". Those patients were unable to express need or access care, for reasons of reduced communication, fear, inability to cooperate, professional prejudice, inappropriate service provision, amongst other factors, represented the population in need of special care dentistry (Faulks and Hennequin, 2006).

1.3.7.3 Developing an ICF Based Questionnaire for Head and Neck Cancer Patients

The ICF is a clinician-rated evaluation. This poses potential practical disadvantages in routine practice because assessment have to be undertaken as face to face evaluations, requiring extra time and resources in a busy outpatient setting (Guyatt et al., 1986). Clinician-rated scores might not correspond to patient perceptions as they do not reflect patients' own feelings about specific domains (Gill and Feinstein, 1994; Guyatt and Cook, 1994).

From the comprehensive ICF classification, specific Core Sets have been developed (Cieza et al., 2004b). The goal of the ICF Core Sets is to select disease-specific sets of categories that can serve as minimal standards for the assessment and documentation of functioning and health in clinical studies. ICF Core Sets have been developed for 16 health conditions including heart disease (Cieza et al., 2004b; Becker et al., 2009), obstructive pulmonary disease (Stucki et al., 2004b), stroke (Geyh et al., 2004), diabetes mellitus (Ruof et al., 2004), obesity (Stucki et al., 2004a), rheumatoid arthritis (Stucki et al., 2004c), depression (Cieza et al., 2004a), breast cancer (Brach et al., 2004) and head and neck cancer (Tschiesner et al., 2009).

In a very innovative attempt Rogers and co-workers developed ICF Core Set as a patient self-completed questionnaire "Brief Core Set Questionnaire for Head and Neck Cancer Patients (BCSQ-H&N)" which consisted of ninteen ICF items. Their rationale was that a simple questionnaire is easier to apply as a standardised outcome measure than a clinician-rated scale (Rogers et al., 2010). They, however, acknowledged some difficulties such as the fact that ICF is written in a very specified scientific language and it was necessary to re-word some domains and the scoring system into a language more easily understood by the patients. Secondly, for certain domains, it was felt that a

more detailed level of questioning in greater depth about problems with structure and functioning was appropriate and would have been easier for patients to understand (Rogers et al., 2010).

Rogers and co-workers identified some key findings related to ICF Core Set for head and neck patients and suggested that while (BCSQ-H&N) is the first of its type and that it had the potential for international acceptance, considerable development was required. Some of the key findings related to the differences between the patient self-completed responses using University of Washington v.4 Quality of Life Measure (UW-QoLv4) and the clinician-rated scoring using (BCSQ-H&N). They further explained that the language of the ICF was very specific and that it was likely patients may have interpreted some statements differently. Another finding was that the questionnaire did not address certain domains, which were considered very important by many patients through the discussions at interviews. These domains are however included in the Comprehensive ICF Core Set but not in the Brief (Rogers et al., 2010).

1.3.8 How is quality of life measured?

There are two broad approaches which have been employed in health related quality of life research (Bowling, 1995a):

1.3.8.1 The functionalist approach

The functionalist approach is the most widely used approach. It measures the traits based on pre-conceived assumptions of health related quality of life and their relevance to all individuals (Ware, 1995). This approach employs scales that attempt to cover all pertinent domains of life affected by health using self administered or interview-assisted questionnaires. Summing up responses to individual items can generate a quantitative score for overall health related quality of life or its subdomains to be utilised as outcome measures. In an overview of some of the principle factors to be considered in choosing or developing a quality of life measure Aaronson suggested that employing batteries of questions can bias responses and over-estimate the impact of health status on life quality, and that a mixture of quantitative and qualitative strategies may be most fruitful (Aaronson, 1988). A study of quality of life in twenty patients undergoing hip replacement questioned the content validity of many health related quality of life measures by stating that items that are included in the scales are

questionably pertinent and meaningful to all individuals because most of these instruments assume that health and illness alone determine the QoL of patients, whereas several respondents in his study did not include health when asked to nominate the five most important aspects of their lives (O'Boyle et al., 1992). Further more, the relevance or importance of particular behaviours or events does not necessarily remain static for a given individual with the passage of time or over the course of an illness (O'Boyle et al., 1992).

1.3.8.2 The hermeneutic approach

This was developed to assess quality of life from the individual's perspective (McGee et al., 1991; O'Boyle et al., 1993; Hickey et al., 1996; Akram et al., 2011) and aimed to measure the meaning and significance of health and illness to the individual's quality of life (O'Boyle et al., 1992). It is an interview-based instrument derived from a decision analysis technique known as judgment analysis "which is a process in which a variety of life areas nominated as important by individual patients and the differences in relative weights attached to these areas are identified" (O'Boyle et al., 1992). It externalises judgment policy by using statistical methods to derive algebraic models of the judgment process. Its goal is to describe, quantitatively, the relations between someone's judgment and the information, or "cues", used to make that judgment (Hammond et al., 1975). It employs more qualitative type research using open-ended interviews in comparison to pre-coded closed questions. It investigates how an individual constructs subjective phenomena such as quality of life without a preconceived notion of their dimensions or domains (Stewart, 1988; Akram et al., 2011; Meaney et al., 2011).

1.3.8.3 Oral Health and Qualify of Life Measures

McGrath et al investigated the perceived relationship between oral health and quality of life using open-ended questions *" In what way does the condition of your teeth, gums, mouth or false teeth reduce your quality of life (negative effects)?"* and *"In what way does the condition of your teeth gums, mouth or false teeth add to your quality of life (positive effects)?"* in 1778 adults aged 16 years or older (McGrath et al., 2000). The results indicated that 75 percent of respondents felt that oral health status has either enhanced or reduced their quality of life. Sixty one percent (1,065) reported their oral health status enhanced their QoL in some way while 51 percent (902) claimed that their

oral health status reduced their QoL in one way or another (McGrath et al., 2000). Cohen and Jago recognised the need to develop patient-based measures of oral health status when they first indicated the lack of data relating to psycho-social impact of oral health problems (Cohen and Jago, 1976).

Fundamentally, there are three categories of OHRQoL measures as indicated by Slade (Slade, 2002). These are (1) social indicators, (2) global self-rating and (3) multiple item questionnaires of OHRQoL. Social indicators are used to assess the effect of oral conditions at the community level (Reisine, 1984; Locker, 1989a). Typically, large population surveys are carried out to express the burden of oral diseases on the whole population by means of social indicators such as days of restricted activities, work loss and school absence due to oral conditions (Reisine, 1984). While social indicators are meaningful to policy makers, they have limitations in assessing OHRQoL. For example, using work loss to measure the impact of oral diseases is not an appropriate indicator for those who are not working (Slade, 2002). This approach only indicates the trends in uptake of health care services and provides little information on an individual level (Allen, 2003).

Global self-rating of OHRQoL which are also known as single-item ratings, refer to asking an individual a general question about their oral health. Response options to this global question can be in categorical or visual analogue scale (VAS) format. For example, a global questions asking: "how do you rate your oral health today?" can have categorical responses ranging from "Excellent" to "Poor" or VAS responses on a 100 mm scale (Slade, 2002).

Some limitations of this approach include that VAS only gives an indication of the current status, but does not give any insight as to WHY a problem is happening or being reported. Global scales are relatively crude, and there is some concern about construct validity (Wewers and Lowe, 1990).

Multiple items questionnaires are the most widely used method to assess OHRQoL and they can be classified into generic instrument that measure oral health over-all versus specific measure which are specialised to measure specific oral health dimensions such as dental anxiety (McNeil and Rainwater, 1998), head and neck cancer (Terrell et al., 1997), or dentofacial deformity (Cunningham et al., 2000) or to assess specific

populations such as children (Jokovic et al., 2002). The advantages of using questionnaires are that they are relatively inexpensive, are familiar and acceptable to most people (Mays and Pope, 2000) and can be used in further studies in other populations to compare results and at different stages in treatment to assess response to treatment (Akram et al., 2011). There are some disadvantages to using self-report questionnaires, the most frequent being that they can be intrinsically manipulative. This draw back can be overcome in the early stage by using qualitative research methods to generate the items for the questionnaire (Mays and Pope, 2000; Akram et al., 2011; Meaney et al., 2011)

1.3.8.4 Generic Measures

Generic instruments are designed to measure across a wide range of conditions. This allows comparisons between different conditions to assess their impact on quality of life but will not be sufficiently sensitive to determine the impact of more subtle oral conditions (Locker and Allen, 2007). Single indices have advantages and disadvantages over profile measures of outcomes. The first advantage is that they appear to summarise a great deal of information in a simple single number. The second advantage, which remains controversial, is that they can be used in cost-benefit analysis, which is undertaken by health economists and attempts to assess the benefits of treatment, taking into account the cost of intervention (Jenkinson and McGee, 1998b). The disadvantages include that some single index measures are derived from simply summing up scores from a profile, which makes them too complex or that they are derived from other psychometric techniques which do not provide social valuations and consequently are inappropriate for use in cost-benefit analysis (Jenkinson and McGee, 1998b). The discriminant validity and responsiveness to change properties of the generic measures may be poor (Corson et al., 1999; Allen et al., 2001).

1.3.8.5 Specific Measures

Specific questionnaires are directed towards a particular disease or setting. They have an advantage over generic measures in that they are more likely to detect subtle changes in specific conditions, thus having better responsiveness (Locker and Allen, 2007). These measures also contain statements and domains, which are only relevant to the clinical condition under investigation thus reducing patient burden and increasing questionnaire acceptability (Allen, 2003; Akram et al., 2011). The more specific the

measure is, the more likely it is to pick up on subtle changes but it will be unable to make broader statements across groups (Corson et al., 1999; Locker and Allen, 2007), and that interpretation of specific measures is often difficult as it is not possible to "norm" against a general population sample (Jenkinson and McGee, 1998b).

A suggestion was made to use a disease specific measure and a generic measure. The rationale is to have a measure with core quality of life statements, and disease specific statements to improve responsiveness (Bowling, 1995a).

1.3.9 Methods of Developing Oral Specific Measures

Various methods have been used to develop oral specific measures (Allen, 2003) including assigning an over-all score (which is ordinal or interval in nature) to indicate the extent of a range of functional and psycho-social consequences. This approach was used in the General Oral Health Assessment Index (GOHAI)(Atchison and Dolan, 1990). GOHAI contains 12 statements (e.g. "how often did you feel uncomfortable eating in front of people because of problems with your teeth or dentures") with a Likert response format (i.e. 0 = Never, 1 = Seldom, 2 = Sometimes, 3 =Often, 4 = Very often, 5 = Always). Response codes are summed across the 12 statements to give a 0-60 overall score. The same approach was employed in the Social Impacts of Dental Disease (Cushing et al., 1986) and the Subjective Oral Health Status Indicators (Locker and Miller, 1994).

A second approach was to evaluate the patients' perceptions salience of events, as demonstrated in the Dental Impact Profile (Strauss and Hunt, 1993). This measure contains 25 statements using the format "do you think your teeth or dentures have a good (positive) effect, a bad (negative) effect or no effect on your eating". The 25 statements are divided into 4-subscales (eating, health/well being, social relations, romance), and an overall profile score is calculated as the proportion of positive or negative responses among all items answered.

A third approach has been to place functional disorders and their social consequences in a hierarchy of outcomes. This approach differs from an index in that a respondent can indicate whether a problem is entirely internal (such as some difficulty chewing), or whether this in turn causes interpersonal or social consequences (such as avoiding the company of others). The hierarchy of outcomes is based on the WHO classification of

Impairments, Disabilities and Handicaps (Wood, 1980a), and the theoretical framework for measuring oral health developed by Locker (Locker, 1988; Locker, 1989a). Using this method, a "profile" of social impacts can be described. Examples of some measures which were developed using this approach include: The Oral Health Impact Profile [OHIP-49] (Slade and Spencer, 1994), the Dental Impact on Daily Living [DIDL] (Leao and Sheiham, 1996), and the Oral Impacts on Daily Living [OIDP] (Adulyanon et al., 1996). Each of these measures attempts to measure both the frequency and severity of oral problems on functional and psychosocial well being.

More recently, a forth approach based on the WHO International Classification of Functioning, Disability and Health (ICF) using the ICF Core Set for head and neck cancer patients was used to develop a patient self completed questionnaire as a standardised outcome measure which has the potential for international acceptance (Rogers et al., 2010). However, this measure is specific to head and neck cancer patients and it needs to be further investigated and validated.

OHRQoL instruments vary widely in terms of the number of questions (items), and format of questions and responses. Table 1-5 shows the thirteen most common OHRQoL instruments that have been proposed and tested to assess their psychometric properties such as reliability, validity and responsiveness (Al Shamrany, 2006).

Table 1-5: The thirteen most commonly used OHRQoL questionnaires (adapted from Al Shamarany, 2006 with modifications).

Measure (questionnaire) **Author (developer)**	**Dimensions measured**	**No. of questions**	**Example of question**	**Response format**
Sociodental Scale (Cushing et al., 1986)	Chewing, talking, smiling, laughing, pain, appearance	14	Are there any types of foods you have difficulties chewing?	Yes/no
RAND Dental Health Index (Dolan et al., 1991)	Pain, worry, conversation	3	How much pain have your gums and teeth caused you?	4 categories: "not at all" to "a great deal"
General Oral Health Assessment Index (Atchison and Dolan, 1990)	Chewing, eating, social contacts, appearance, pain, worry, self-consciousness	12	How often did you limit the kinds or amounts of food you eat because of problems with your teeth or dentures?	6 categories: "always to "never"
Dental Impact Profile (Strauss and Hunt, 1993)	Appearance, eating, speech, confidence, happiness	25	Do you think your teeth or dentures have a good effect (positive), a bad effect (negative, or no effect on feeling comfortable?	3 categories: good effect, bad effect, no effect

Oral Health Impact Profile (Slade and Spencer, 1994)	Function, pain, physical disability, psychological disability, social disability, handicap	49	Have you had difficulties chewing foods because of problems with you teeth or dentures?	5 categories: "very often" to "never"
Subjective Oral Health Status Indicators (Locker and Miller, 1994)	Chewing, speaking, symptoms, eating, communication, social relations	42	During the last year, how often have (dental problems) caused you to have difficulty sleeping?	Various, depending on question format
Oral Health Quality of Life Inventory (Cornell et al., 1997)	Oral health, nutrition, self-related oral health, overall quality of life	56	Two-part questions: (A) How important is it for you to speak clearly? (B) How happy are you with your ability to speak clearly?	Part A: 4 categories ("not at all important" to "very important"), Part B: 4 categories ("unhappy" to "happy")
Dental Impact on Daily Living (Leao and Sheiham, 1996)	Comfort, appearance, pain, daily activities, eating	36	How satisfied have you been, on the whole, with your teeth in the last 3 months?	Various, depending on question format
Oral Health-Related Quality of Life (Kressin et al., 1996)	Daily activities, social activities, conversation	3	Have problems with your teeth or gums affected your daily activities such as work or hobbies?	6 categories: "all of the time" to "none of the time"

Oral Impacts on Daily Performances (Adulyanon et al., 1996)	Performance in eating, speaking, oral hygiene, sleeping, appearance, emotion	9	Four-part questions: (A) In the past 6 months, have [dental problems] caused you any difficulty in eating and enjoying foods? (B) Have you had this difficulty on a regular/periodic basis or for a period/spell? (C) During the last 6 months, how often have you had this difficulty? (D) Using a scale from 0 to 5, which number reflects what impact the difficulty in eating and enjoying food had on your daily life?	Various, depending on question format
Child Perception Questionnaire (CPQ11-14) (Jokovic et al., 2002)	Four health domains: symptoms, functional limitations, emotional well-being, and social well-being. In addition there are two global rating questions	36	Questions asked about the frequency of events in the previous three months. The total score of the CPQ11-14 ranges from 0-144.	"Never=0"; "Once/twice=1"; "Sometimes=2"; "Often=3"; and "Every day/almost every day=4"
Child Oral Impact on Daily Performance (Child-OIDP) (Gherunpong et al., 2004)	Difficulties carrying out eight daily activities namely eating, speaking,	8	Two-part questions: (A) In the past 3 months, have [dental problems] caused you any difficulty in eating and enjoying	Never; once or twice a month; once or twice a week; every day/ nearly every

	cleaning mouth, sleeping, smiling, school work, emotion and social contact for 11-12 years old.		foods? Scored on 0-3. (B) Identify the oral condition that caused the specific impacts by answering for each reported item (1) yes or (0) no to a list of oral conditions. Two scores can be obtained: Additive score (range 0-24) and Simple count score (range 0-8) by summing the dichotomised frequency items of (0) not affected and (1) affected.	day.
Early Childhood Oral Health Impact Scale (ECOHIS) (Pahel et al., 2007).	Impact on child and family. Ages 3-5 years old. To be answered by parents/ caregivers. Six domains: Child symptoms, Child function, Child psychology, Child self-image/social interaction, parent distress, and family function domains.	13	How often has your child had pain in the teeth, mouth or jaws? How often have you or another family member……..because of your child's dental problems or treatment? (A) been upset, (B) felt guilty, (C) taken time off from work	Never; Hardly ever; Occasionally; Often; Very often; Don't know. Scores for the child section range from 0-36. Whereas, those for the family section range from 0-16. Higher response indicate greater impact and poorer OHRQoL.

1.3.10 Criteria for evaluating health related quality of life measures

In a critical appraisal of the face validity of patient based outcome measurements in clinical trails Gill and Feinstein identified a number of criteria that must be met if a measure is to be used to assess "quality of life" (Gill and Feinstein, 1994). A second set of criteria was also published in the same issue by Guyatt and Cook and was more properly characterised as "health related quality of life" (Guyatt and Cook, 1994). A third set of criteria, which are similar to the ones outlined by Guyatt and Cook, was developed by Locker and Allen (Locker and Allen, 2007). These criteria are summaries in table 1-6.

The criteria identified mainly focus on two broad and overlapping questions, which need to be considered when evaluating a measure in terms of the underlying construct being assessed:

- Is the measure patient or person-centred?
- Does the measure incorporate aspects of daily life that are important to patients or persons, which may be compromised by disorders of various kinds?

Table 1-6: Criteria for evaluating health-related quality of life measures (adopted from Gill and Feinstein; Guyatt and Cook; and, Locker and Allen).

Criteria of Gill and Feinstein (Gill and Feinstein, 1994).
• Did the investigators conceptually identify what they meant by quality of life? • Did they state the domains they wanted to measure as components of quality of life? • Did the investigators give reasons for choosing the instruments they used? • Did the investigators aggregate the results from multiple items, domains or instruments into a single composite score? • Were patients asked to give their own global rating for quality of life? • Was overall quality of life distinguished from health-related quality of life? • Were patients invited to supplement the items listed in the instruments offered by the investigators? • If so, were these supplemental items incorporated into the final rating? • Were patients asked to indicate which items were personally important to them? • If so, were these importance ratings incorporated into the final rating?
Criteria of Guyatt and Cook (Guyatt and Cook, 1994).
• Do the authors show that aspects of patients' lives they have measured are important to the patients? • If not, have previous studies demonstrated their importance? • Do the investigators examine aspects of patients' lives that clinical experience indicates patients value? • Are there aspects of HRQL that are important to patients that have been omitted? • Were individual patients asked to directly place a value on their lives?
Criteria of Locker and Allen (Locker and Allen, 2007).
• Is the stated aim to measure health-related quality of life or quality of life and is this explicit? If so, are these constructs defined and their constituent domains identified? • If not, is an alternative construct measured by the instrument specified and defined and its constituent domains identified? • Do the investigators specify the contexts in which the measure is to be used? Was it developed to be used with groups (as in surveys or clinical trials) or individuals (as in clinical practice)? • Were the items comprising the questionnaire derived from qualitative interviews with those who will be completing the questionnaire? • Is there evidence that the aspects of life the items address are important to those who will be completing the questionnaire? • Does the questionnaire contain global ratings of health-related quality of life or quality of life? • How was the measure validated? Was it tested against oral health indicators or were broader indicators that may capture aspects of quality of life used?

Fitzpatrick and co-workers outlined the requirements for judging the appropriateness of out-come measures in clinical trails in the following points (Fitzpatrick et al., 1998):

- Is the content of the measure appropriate to the questions, which the clinical trial is intended to address?
- Does the instrument produce reliable results (i.e. that are reproducible and internally consistent)?
- Is the measure valid (i.e. does the measure detect what it claims to measure)?
- Is the measure responsive (i.e. does the measure detect changes over time that matter to patients)?
- How precise are the scores on the measure (i.e. how able is the measure to detect differences in health)?
- How interpretable are the scores on the measure?
- Is the measure acceptable to patients (e.g. is it too long or potentially distressing)?
- How feasible is the measure for the study (i.e. is the instrument easy to administer and process)?

1.3.10.1 Patient Vs. Expert centred

There seems to be a consensus that measures addressing overall or components of quality of life should reflect the perspectives of patients and/or the lay public (Frisch et al., 1992; Leplege and Hunt, 1997). Measures that meet this criterion are patient or person-centred rather than expert-centred (Leplege and Hunt, 1997).

Gill and Feinstein in a review of the quality of life measures concluded that the patient's opinion is the only one that matters when it comes to rating quality of life (Gill and Feinstein, 1994). The authors explained their views by stating that quality of life is a uniquely personal perception, denoting the way that individual patients feel about their health status and/or non-medical aspects of their lives, and that most measurements of quality of life in the medical literature seemed to aim at the wrong targets. If the patient reported that their quality of life is excellent then the authors feel that it must be because the patient reports this fact (Gill and Feinstein, 1994). Many measures that claim to assess health related quality of life or quality of life have been

criticised because they reflect the values and concerns of physicians, social scientists or other experts rather than patients or persons and what they consider to be relevant (Bowling, 1995b; Leplege and Hunt, 1997; Carr and Higginson, 2001). Leplege and Hunt stated that patients are often asked to complete questionnaires that do not reflect their concerns (Leplege and Hunt, 1997).

Guyatt and Co-workers stated that the main mechanism by which a patient or lay person's perspectives can be accommodated is through the use of qualitative interviews. According to Guyatt the items comprising questionnaires that claim to capture quality of life issues must be derived from in-depth interviews with those who will ultimately be expected to complete the questionnaire (Guyatt et al., 1986; Juniper et al., 1996). Consequently, one way of evaluating OH-QoL measures is the way in which the items comprising the measure where developed. Were they derived from qualitative interviews with the target population? If not, and since items may legitimately be obtained from other sources such as literature reviews or clinical opinion (Guyatt et al., 1986), is there any evidence from these other sources or elsewhere that the items do fully address the concerns of those who will be expected to complete the questionnaire? That is, did the investigators achieve content coverage, a component of content validity that is as important as criterion validity but most often neglected? (Locker and Allen, 2007).

1.3.10.2 Establishing item importance

Gill and Feinstein state that 'The need to incorporate patient's values and preferences is what distinguishes quality of life from all other measures of health'. In other words, do the items comprising the questionnaire refer to aspects of daily life that are important to the target population (Gill and Feinstein, 1994)?

Recently, qualitative research investigated the issues of importance for patients affected with developmental dental disorders using focus groups of patients with hypodontia (Akram et al., 2011; Meaney et al., 2011). The two studies interviewed patients affected with hypodontia in small focus groups to identify any emerging themes until saturation (when no new themes were emerging from the participant's responses) was achieved. The similarities and/or dissimilarities between the themes of the two studies are noteworthy. In the first study, the themes included: treatment, appearance, activities,

and other people's reaction (Akram et al., 2011). Whereas those of the second study included: transition from childhood to adulthood, waiting time for treatment, and importance of aesthetics (Meaney et al., 2011).

Locker and Allen outlined the limitations of qualitative research and stated that although qualitative studies are a first step in eliciting the views of patients and persons regarding what is important, they are insufficient in and of themselves (Locker and Allen, 2007). The reason for this is that while qualitative interviews can reveal what is salient to the interviewees, the interview may reveal a range of experiences some of which are relatively more and some relatively less important. This is clearly manifested in the differences between the themes, which emerged from the two qualitative studies quoted earlier despite achieving saturation (Akram et al., 2011; Meaney et al., 2011). Locker and Allen further explained that, if the questionnaire developed from qualitative research is to be used in clinical trials or other studies where the unit of analysis is the group, some way has to be found of identifying which experiences are of most importance on average to the group who will be participating in the study (Locker and Allen, 2007). Guyatt and Co-workers proposed an item impact study as a way of identifying importance. Items derived from qualitative interviews are given to a group of patients or persons who indicate if they experience the problem described by the item and, if so, how much bother or distress it causes. Item impact scores are calculated by multiplying the prevalence of the problem by its mean bother rating and items ranked according to these scores and the top ranking items selected for the final questionnaire (Guyatt et al., 1986). The limitation of this approach is that a high prevalence and low impact item may be more highly scored than a low prevalence high impact item, so that items of importance to a minority of patients or people may not make it into the final questionnaire. Consequently, the item-impact approach produces a group-centred rather than a patient-centred questionnaire (Prutkin and Feinstein, 2002; Locker and Allen, 2007). While group centred-questionnaires may be appropriate for clinical trials, they concede that they may not be appropriate for clinical practice. When clinicians are making treatment decisions they should consider an individual patient's values since these may differ from or be masked by the aggregated values of those participating in an item impact study (Locker and Allen, 2007). This points to the fact that 'quality of life – even when health-related – has different components, significance and meaning

that are unique to the individual' (Prutkin and Feinstein, 2002), and that the main 'challenge to measuring quality of life lies in its uniqueness to individuals' (Carr and Higginson, 2001). Gill and Feinstein proposed solutions for improving the measurement of health-related quality of life. These included: rate frequency, severity and importance; allowing respondents to add supplemental items, and using global ratings which are summary variables which reflect the differing values and preferences of a group of individuals (Gill and Feinstein, 1994).

This means that a questionnaire must have a complex structure – questions that document experiences; importance ratings that indicate the value an individual attaches to those experiences; open-ended questions to elicit experiences not covered by the questionnaire, and global ratings of quality of life and health-related quality of life. If these requirements are not met a measure is not addressing health-related quality of life; rather it is a measure of health status (Locker and Allen, 2007).

Carr and co-workers proposed the use of individualised measures (Carr and Higginson, 2001). These measures allow respondents to select issues and concerns that are affected by health rather than providing a standardised predetermined list (Fitzpatrick et al., 1998). However, they acknowledge that the completion of such questionnaires is time consuming and difficult. Locker and Allen argue that the aggregation analysis and interpretation of the data are challenging, particularly with respect to group comparisons and change over time. This raises the interesting question of whether or not we can legitimately refer to the quality of life of a group and, if so, how quality of life might be measured at the group level (Locker and Allen, 2007)?

1.3.11 What is Being Measured by OHRQoL Measures?

In dentistry there has been a proliferation of instruments and scales seeking to assess "oral health related quality of life" and/or "quality of life" of patients with various oral conditions. This however has led to a certain lassitude and a failure to continue to address the conceptual and methodological issues involved in measuring perceptions of oral disorders at the individual and population level (Locker and Allen, 2007). Most available quality of life measures have been investigated with special focus on their content, scoring methods, and technical properties rather than with the more fundamental question of what a measure actually measures and the value on which it is

based (DeJong, 1997).

There is clearly a conceptual and methodological issue in defining the attribute to be measured and how measures of that attribute were developed. For instance, the construct validity of most available measures has been investigated but scores derived from these measures, which discriminate between different populations or show associations with global ratings of oral health or show changes in the expected directions after treatment import very little about the underlying construct being addressed (Locker and Allen, 2007).

Fitzpatrick and co-workers believe that the use of the term quality of life is unhelpful since it 'misleadingly suggests an abstract or philosophical set of judgments or issues relating to life in the broadest sense of factors outside the person, such as living standards, political or physical environments'. Moreover: 'Because, rightly or wrongly, hardly any of the vast array of so-called quality of life measures used in health settings address matters beyond the health-related, we avoid using this terminology as much as possible' (Fitzpatrick et al., 1998). They suggested the use of 'patient-based outcome measures' rather than 'quality of life measures' as a collective term for instruments that assess perceptions of health, its consequences and the benefits derived from therapeutic interventions. Other researchers suggest that the notion of quality of life, even when "health-related", should be abandoned altogether and replaced with the more 'easily handled and rigorously defined notion of subjective health status' (Leplege and Hunt, 1997). They further explain that measures of subjective health are valuable in their own right since the outcomes being sought by most of those seeking health care are a reduction in symptoms and improvements in function, irrespective of their impact on overall life quality. This is consistent with their view that the proper role of health care is the improvement of health status; that is, health care is concerned with the removal of potential barriers to the quality of life rather than with the quality of life itself (Leplege and Hunt, 1997).

Locker and Allen reported that for those who feel that quality of life is a legitimate concern of health professionals and the health care system, the problem of how it can be measured remains (Locker and Allen, 2007). They further outlined the two main options used, which include the use of individualised measures or the relatively

cumbersome approach suggested by Gill and Feinstein, which includes the use of global ratings, rating severity and importance, and allowing supplemental items that patients can add to denote important factors that may not have been included in the instrument (Gill and Feinstein, 1994). Locker and Allen favored the option suggested by Prudkin and Feinstein in which they acknowledge that an individual's health status can have a broader impact with respect to how they perceive themselves and their lives and allows this impact to be measured more simply (Prutkin and Feinstein, 2002). This entails the use of subjective health status measures along with global ratings of quality of life and health-related quality of life; that is ratings of the extent to which quality of life is compromised by symptoms or functional and psychosocial problems. These ratings incorporate individuals' own beliefs, values and concerns and the relative importance of different life domains. Consequently, they 'allow adequate expression of the way in which individual patients determine their own quality of life' (Prutkin and Feinstein, 2002). Locker and Allen concluded that "providing that these measures and scales are patient/person centred and that they consist of items concerning symptoms or aspects of functioning that patients or persons consider to be important", the last option solves the problem of what existing measures actually measure, provide one way in which data on a unique, highly individualized phenomenon can be grouped for analysis, and provide a way of ascertaining the meaning of scores derived from instruments such as the OHIP, which, they believe, have no intrinsic meaning (Locker and Allen, 2007).

1.3.12 Defining and Measuring Oral Health-Related Quality of Life

Locker and Allen reviewed five of the most commonly used OHQoL/OHRQoL measures and explained that they all address the functional and psychosocial outcomes of oral disorders and can be readily used in population surveys or clinical trials and that they document the frequency of impacts emanating from oral diseases that clinicians, investigators or research subjects have suggested are important (Locker and Allen, 2007). However, based on the work of Leplege and Hunt who stated that 'patients are replying to questions designed by experts is not sufficient to claim that scores calculated from these responses reflect the patient's viewpoint' (Leplege and Hunt, 1997) Locker and Allen questioned the ability of these measures to adequately capture the views and concerns of those completing the questionnaire. They also posed the question what do these measures actually measure? Do they assess subjective oral

health, OH-QoL or quality of life? They concluded that none of the measures reviewed fully comply with the 'patient / person-centred', 'importance' and 'validation' criteria for the development of measures which can be deemed to assess OH-QoL and that in order to answer the first question, more qualitative work with sufficiently large samples to ensure that the views of people and patients are being properly represented by existing measures is needed. They further explained that in order to answer the second question, more precise definition of the main constructs that measures are intended to address is required (Locker and Allen, 2007).

1.3.13 Which Oral Health Quality of Life Measure Should be Used?

In the oral health context, the question of which measure to use has been the subject of intense research effort in recent years (Gill and Feinstein, 1994; Guyatt and Cook, 1994; Jenkinson and McGee, 1998a; Locker and Allen, 2007). Currently, both generic and disease specific measures of health status are used. These measures are developed using scientific and statistical methods. Questions are not chosen randomly but along lines of an existing model or from reference groups of those with experience in the field of interest. This can and should include patients, their relatives and healthcare professionals (Corson et al., 1999). Qualitative research methods, based on informal interviews, have been recently used to identify issues of importance for patients with developmental dental disorders such as hypodontia (Akram et al., 2011; Meaney et al., 2011). This qualitative research is aimed at identifying emerging key themes to be utilised in developing QoL questionnaires for patients with developmental dental disorders (Akram et al., 2011).

While all of the measures appear to have been well validated and based on sound theoretical frameworks, some practical issues remain. For instance, which of these measures should be used? Determining which measure to use is unclear, as no substantive work to compare the relative performance of the various measures across a range of clinical situations has been published. This would be useful to clinicians hoping to use these measures to assess outcomes of clinical procedures, as short measures such as GOHAI are much easier to use than sophisticated measures such as OHIP (Allen, 2003). Short versions of health status measures have an inherent appeal in clinical situations, but it is a well known psychometric property that the sensitivity of a measure diminishes as statements are removed (Streiner and Norman, 1989; Awad et

al., 2008). Some efforts have been made to shorten existing measures while retaining such important psychometric properties as reliability and precision. The methods used include internal reliability analysis, factor analysis and regression analysis to identify items that had the strongest associations with the original long versions of the measures (Ware and Sherbourne, 1992; Slade, 1997). The short version of OHIP contains 14 items derived from the 49 – item OHIP, and appears to have good validity and reliability properties (Slade, 1997; Allen and Locker, 2002). In addition, a subset of OHIP items derived using the item impact method has been developed for use as an outcome measure of tooth replacement procedures (Allen and Locker, 2002). The responsiveness of this subset of OHIP items seems to be better than the OHIP-14, and an argument can be made for using an item impact to derive a subset of items for use in specific clinical trial contexts (Allen and Locker, 2002). Measures, which use weights to allow the severity of an impact to be described, are likely to be better outcome measures. Scoring methods based on ordinal scores are prone to produce skewed results, particularly when the range of possible responses is narrow. However, the contribution of weights to the performance of health status measures has been questioned (Allen, 2003). Streiner and Norman suggested that weighting of statements does not improve the performance of a measure consisting of more than forty items (Streiner and Norman, 1989). This concurred with the findings of Locker and co-workers who reported that weighting the Surgical Orthodontic Outcome Questionnaire (SOOQ) (Berka, 2004) did not substantially improve its performance (Locker et al., 2007). Allen and Locker found that the discriminant, predictive and concurrent validity of OHIP was only moderately improved by weights (Allen and Locker, 1997). This finding was consistent with that of Leao and Sheiham for the DIDL questionnaire (Leao and Sheiham, 1996). However, weights increase the complexity of use and interpretation of health status measures. This may act as an impediment to the use of these measures in clinical settings. The issues which need to be resolved are (1) whether the methods for developing weights are appropriate; (2) if the ranges of weights wide enough to discriminate; and (3) whether weights are of any benefit at all (Allen, 2003).

As assessment of change is a fundamental requirement of all longitudinal study designs, the issue of ability of health status measures to quantify change is of interest. Locker described four ways of measuring change, namely: 1) comparison of "before" and

"after" measurements; 2) change scores, calculated by subtracting the baseline score from the follow-up score; 3) global transition judgments, and; 4) global transition scales (Locker, 1998). All of these methods may be used, but none are universally accepted. Comparing baseline and follow up measurements is straightforward, but positive and negative changes may cancel each other out, thus giving the impression of no change. Change scores, also known as raw gain scores, are difficult to accept because intrinsically they have no meaning. It is, therefore, not possible to describe a change score in either a positive or negative direction as clinically meaningful.

A global transition judgment is a patient's overall assessment of how their health status has changed over the study period in question. Changes in health status as measured by a health status measure can be compared with the global transition judgment. Researchers have found that changes in global judgments varied over time and were consistent with self-reported health status indicators and GOHAI scores (Dolan et al., 1998; Locker, 1998). This would suggest that discriminant validity properties of global transition judgments may be at least as effective as multi-item oral health quality of life measures. However, it is unlikely that a comprehensive picture of responsiveness to change can be gained from using global measures alone (Allen, 2003). Global transition scales are derived from a series of global transition statements applied to different dimensions of health. The scale scores are calculated by summing the response codes, and monitoring changes over the time period of the study. The use of this method has not been widely reported in the literature (Allen, 2003).

Slade has illustrated the complexity of measuring change in quality of life (Slade, 1998) . In a longitudinal observational study, he used OHIP to measure oral health related quality of life at baseline and two-year follow-up visits. He reported that both improvement and deterioration in oral health related quality of life can occur simultaneously. Using three risk predictors (tooth loss, problem related dental visits, financial hardship) to assess effects of various methods of measuring change, high risk and low risk groups were compared. High risk groups had both higher rates of deterioration and improvement in quality of life than low risk groups. The example used to explain this phenomenon was that loss of teeth may increase chewing difficulty, but decrease pain. Tooth loss may, therefore, improve quality of life for some individuals, while decreasing it for others.

1.3.12 Quality of Life and the Child

Since paediatric oral disorders such as hypodontia and amelogenesis imperfecta are numerous and are likely to have negative effects on the quality of life, there is a need for measures that document oral health outcomes in child populations.

Developments of health status measures in children have to take into account the developmental and methodological issues that relate to children. A child starts to develop abstract thinking and self concept about six years of age according to Hetherington and they also begin to compare themselves with others in terms of physical and personality traits (Hetherington et al., 1999). At eight years of age children develop a sense of time and their recall period lengthens. By 11-12 years of age children have an understanding of their emotions and have developed a concept of self that encompasses popularity. Similarly, the content of daily activities, understanding of feeling states, perceptions of relationships, and communication skills evolve with children's age (French and Christie, 1996; Theunissen et al., 1998). In adolescence, the peer group becomes increasingly important and there is a preoccupation with the views of others. These stages of development ideally require separate methods of assessment (Hetherington et al., 1999). Until recently, Children's health related quality of life was measured using parents as informants. This was based on concerns that children's reports of their health and QoL would not meet accepted psychometric standards of validity and reliability because of limitations in their cognitive capacities and communication skills (Theunissen et al., 1998; le Coq et al., 2000b). However, a number of recently developed instruments have demonstrated that with appropriate questionnaire techniques, it is possible to obtain valid and reliable information from children concerning their HRQoL (Christie et al., 1993; Landgraf et al., 1996; Jokovic et al., 2002).

One issue with respect to measuring the HRQoL of children, which remains a concern is that of parent versus child reports and that whether parents could be used as a proxy for their children (Glaser et al., 1997; Whiteman and Green, 1997; Parsons et al., 1999; le Coq et al., 2000b; Zhang et al., 2007a). Some studies indicate high agreement for some health domains (Canning et al., 1992; Glaser et al., 1997; Jokovic et al., 2003a), others have found low concordance between parent and the Child's self assessment (Theunissen et al., 1998; Parsons et al., 1999; le Coq et al., 2000b; Zhang et al., 2007a).

In a review of the existing literature of the oral health quality of life in children, Barbosa and Gaviao explained that during early childhood and for children with severe orofacial disorders, parent-child relationship is somewhat closer so that parents are more familiar with their activities and feelings. This however changes for older children as they get more time with their peers and away from parental supervision and tend to share their experiences with parents to a lesser extent (Barbosa and Gaviao, 2008b).

1.3.15 Future Developments of QoL Measures

It is of paramount importance that the international research community if the field of QoL or HRQoL must agree on a strategy which facilitates comparison of data. Locker and Allen stated that it would be helpful if national norms were established for more frequently used measures and that cross-cultural relevance of the consequences of dental disorders must be considered (Locker and Allen, 2007). Allison and co-workers explored this issue and reported that the nature and magnitude of impacts could vary between populations with different cultural backgrounds (Allison et al., 1999). Models of health are becoming more sophisticated, and it remains to be seen if the conceptual underpinning of existing oral health status measures is now sufficiently robust, or whether new measures, based on more recent models, should be developed (Allen, 2003). This means that the measures currently used in oral health research may need revision and further development. MacEntee suggests that the language, definitions and theoretical model contained within the International Classification of Functioning, Disability and Health (ICF) may provide a useful departure for the further development of patient-based oral health measures (MacEntee, 2006). Locker and Allen point out to the fact that the (ICF) definitions and the concepts that comprise the model are wholly concerned with health and functioning and that there is no reference to issues, such as health-related quality of life or quality of life (Locker and Allen, 2007). These concerns were also stated by Rogers and co-workers who identified some key issues in relation to using the ICF model as a quality of life tool (Rogers et al., 2010).

1.3.16 Oral Health Impact Profile

The social impact of dental disease is measured using the Oral Health Impact Profile (OHIP), which was developed in Australia from an initial set of 535 statements which were obtained from open-ended interviews of 64 dental patients (Patient group) from

private practices, dental hospital primary care and dental hospital prosthodontic clinics (Slade and Spencer, 1994). These patients were not a random sample but selected because of the likely range of oral diseases with consequent social impact. These items were then reduced to a set of 46 unique statements based on their form and content and ability to represent one of six domains derived from the conceptual model. Since statements about handicap were rare, three additional statements representing the concept of handicap were taken from an existing generic health status measure (Hunt et al., 1986). It consisted of 49 items that were weighted according to their severity and based on the opinion of 328 people from community groups, dental practitioners and students who became known as the judging panel. Consequently, the weights may not reflect the severity and/or importance of the events described by the items as perceived by individual dental patients or dental patients as group (Carr and Higginson, 2001). They represented a broad range of opinions and whilst it is possible that such a large group is unlikely to be able to reach a definite consensus, the OHIP-49 has performed well in tests of reliability and validity and OHIP-49 questionnaire has been translated to many languages (Slade and Spencer, 1994; Pires et al., 2006; Awad et al., 2008; John et al., 2008).

The OHIP-49 provides a comprehensive measure of self-reported dysfunction, discomfort and disability attributed to oral conditions (Slade and Spencer, 1994). It provides information about the "burden of illness" within populations and the potential of measuring effectiveness of health services in reducing the burden of illness. Responses to the questions were made on a Likert scale indicating if the problem had been experienced "very often" (code=4), "fairly often" (code=3), "sometimes" (code=2), "hardly ever" (code=1) or "never" (Code=0) with total scores ranging from (0 to 196). Higher scores indicate higher levels of dysfunction, discomfort, and disability attributable to oral conditions.

The questionnaire is subdivided into seven domains to measure:

1. Functional limitation (nine questions)
2. Physical pain (nine questions)
3. Psychological discomfort (five questions)
4. Physical disability (nine questions)

5. Psychological disability (six questions)
6. Social disability (five questions)
7. Handicap (six questions)

The 49 questions constitute a lengthy questionnaire, and average time for administration by an interviewer is 17 minutes. This is the reason that it is mainly used as a self-complete questionnaire (Slade and Spencer, 1994). The questionnaire was later shortened to 14 items (OHIP-14) to make completion easier (Slade, 1997). It has proven validity and reliability and has the potential to be used on an individual basis. However, when the numbers of items of the OHIP-49 questionnaire were reduced, whilst its responsiveness could be maintained there was a decrease in reliability, especially within the Psychological and Social disabilities and Handicap domains for the (35 to 65-year group). Its construct validity was also reduced in Physical pain, Psychological and Social Disabilities domains for the (35 to 65-year group), as well as on Physical pain, Psychological discomfort, Physical and Psychological disabilities in the 65 year and older group. This occurred primarily when reducing one to two items per domain (Awad et al., 2008).

It is worth mentioning that during the OHIP 49 development process the range of statements and judgments about importance could have been influenced by the non-random method of patient selection and the number of persons involved. It is also worth mentioning that the OHIP-49 scale was developed for assessing social impact in groups or populations and that the author did not attach "clinical significance" to individual subscale scores, nor summed standardised scores. Only recently John and co-workers defined the Minimum Important difference (MID) for the OHIP-49 as six OHIP units lower (better, 95% confidence interval) (John et al., 2009). This is defined as "the smallest difference in score in the domain of interest which patients perceive as beneficial and which would mandate, in the absence of troublesome side effects and excessive cost, a change in the patient's management" (Jaeschke et al., 1989). This allows the clinical as well as the statistical significance of differences between groups to be better appreciated. This novel approach however remains to be validated.

A major advantage of this measure is that the statements were derived from a representative patient group, and were not conceived by dental research workers

(patient-centred). This increases the possibility of the measure "tapping into" social consequences of oral disorders considered important by patients (improving its content validity), and is considered to be the most sophisticated measure of oral health (Slade and Spencer, 1994; Locker, 1998). However, the item reduction process was expert-centred, designed to select items according to their fit with a conceptual framework rather than on the basis of their importance to the patient from whom they were derived and that the process of retaining or eliminating some statements was not described (Locker and Allen, 2007). An additional advantage of using this questionnaire is that it covers a wide range of social aspects including items that have low prevalence and high impact and high prevalence and low impact. This makes comparison between groups (test and control) feasible (group-centred questionnaire). While group-centred questionnaires may be appropriate for clinical trails, they have a limitation for use in clinical practices where individual patients are being considered (Locker and Allen, 2007).

1.3.17 Measuring Child Oral Health Related Quality of Life (COHRQoL)

In a systematic review of the existing published literature from January 1985 to March 2007, Barbosa identified 89 articles related to child perception of OHRQoL. Only 13 articles satisfied the inclusion criteria and hence were included in the review. Nine articles used the CPQ instrument and four studies used the child-oral impacts on daily performances (Child-OIDP) (Barbosa and Gaviao, 2008a).

The development of a tool to assess the OHRQoL of children must function on many levels. Jokovic et al stated that given the cognitive and developmental differences at different ages, a single standardised self-report health status measure for children of all age groups could not be developed, and that an age-specific questionnaires for 6-7, 8-10, and 11-14 years-olds are required as these groups are homogeneous in terms of roles, emotional, cognitive abilities and language development (Jokovic et al., 2002). Jokovic and co-workers also described and validated the Child Perceptions Questionnaires (CPQ 8-10) and (CPQ 11-14) which has been developed to measure the OHRQoL of children aged eight to ten years in one questionnaire and eleven to fourteen years in a separate questionnaire (Jokovic et al., 2002). A sample of the

questionnaires used is shown in Appendix I. The 3 components used to measure COHRQoL are:

- Two age specific Child Perception Questionnaires: 8-10, and 11-14
- Parental-Caregiver Perception Questionnaire (PPQ)
- Family Impact Scale (FIS)

All components or items in the questionnaire (CPQ 11-14) are based on two previously reported models (Guyatt et al., 1986; Juniper et al., 1996).

The development of this set of questionnaires marked a change in instruments to measure health related quality of life in children. Previously the parents completed the questionnaires by proxy in order to avoid any methodological difficulties in measuring QoL in children. For example, the impact of compromised health status on income, employment and sexuality will not apply to young children. Also there is a concern that children may not provide accurate or reliable reports of their own health status when they respond to a questionnaire. This could simply be due to factors such as limited understanding of the questions asked or limited understanding of their disease or health in general. Therefore, child's cognitive and emotional development affects the overall reliability of self-reported health outcomes (Connolly and Johnson, 1999).

A study on 228 Dutch children investigated the reproducibility, construct validity and responsiveness of self-report and parent-report using a condition specific quality of life questionnaires [How Are You (HAY) for 8-12] (le Coq et al., 2000a; le Coq et al., 2000c), which has two sections (generic and condition specific) for patients with asthma and their parents compared to healthy children and their parents as controls. The results indicated that in discriminative studies child and parental reports could be substituted on a group-level. He concluded that parents are valuable sources of information regarding their children but their knowledge is limited (le Coq et al., 2000b). With this in mind, the COHRQoL questionnaire has been developed, using an age-specific questionnaire for the child as the chief source of data, augmented by parental information (Jokovic et al., 2002). However, later research has shown that parents do not always have an insight to their children's quality of life status even with the use of these questionnaires. However, the CPQ questionnaires have been reported to be the most widely used (Jokovic et al., 2003b; Zhang et al., 2007a; Barbosa and

Gaviao, 2008a). Barbosa stated that when evaluating child HRQoL, children's understanding of oral health and well-being is affected by a number of variables which include: age, age-related experiences, gender, race, education, culture, experiences related to oral conditions, opportunities for treatment, childhood period of changes, back-translating questionnaire and self-perceived treatment need (Barbosa and Gaviao, 2008a).

1.3.17.1 Child Perception Questionnaires (CPQ 8-10)

Participants in the development process of the child perception questionnaires (CPQ 8-10) were selected from a convenient sample of two groups of children; one with dental decay and the second targeted group were children with clefts of the lip and palate between the ages of 8-10. The items of the questionnaire were selected from literature review, data related to child psychology, and from a focus group of parents and teachers. Items were selected using item impact study (Jokovic et al., 2004). This item selection process does not meet the "patient/person centred" criteria for evaluating a quality of life measure set by Gill and Feinstein and other workers who stated that most available quality of life measures are social scientists or experts-centred and do not address what patients/persons consider to be relevant (Gill and Feinstein, 1994; Guyatt and Cook, 1994; Locker and Allen, 2007). The final questionnaire consists of 25 questions organised into four health domains [oral symptoms (N=5), functional limitations (N=5), emotional well-being (N=5), and social well-being (N=10)]. The questions ask about the frequency of events in the previous 4 weeks in relation to the child's oral/orofacial condition. The response options are: "never=0", "once/twice=1", "sometimes=2", "often=3", and "everyday/almost everyday=4". It also has contains two global ratings of the child's oral health and extent to which the oral/orofacial condition affected his/her overall well-being. Global ratings were worded as "when you think about your teeth or mouth, would you say that they are….." and "How much do your teeth or mouth bother you in everyday life?". A 4-point response format, ranging from "very good=0" to "poor=3" and from "not at all=0" to "a lot=3", respectively (Jokovic et al., 2004). The total score of the CPQ 8-10 ranges from 0-100. The instrument showed good construct validity; however, there are conflicting reports describing its discriminantive validity in relation to different dental conditions (Barbosa et al., 2009; Wogelius et al., 2009; Barbosa et al., 2011; del Carmen Aguilar-Diaz and Irigoyen-

Camacho, 2011). The instrument also showed substantial internal consistency reliability and that test re-test reliability was also acceptable (Jokovic et al., 2004).

1.3.17.2 Child Perception Questionnaire (CPQ11-14)

Development and testing

The sample group for questionnaire development and testing were children aged 11-14 years with dental diseases (primarily caries), orthodontic disorders, and orofacial conditions (primarily cleft lip and/or palate) recruited from patients attending the Craniofacial Unit, Hospital for Sick Children, Toronto and the Paediatric Orthodontic Clinics, Faculty of Dentistry, University of Ontario. The inclusion criteria were:

- Presence of impact from oral and orofacial conditions
- Restorative or orthodontic treatment not yet commenced or in early stages
- Absence of cognitive impairment or other diagnosis
- Fluency in English
- Parental and child consent

Items for the questionnaire were generated in a similar way to those for the CPQ 8-10 using literature review, experts/parents, and item impact study; hence the instrument shares the same limitations for being not patient/person centred and that low prevalence-high impact items may not make it to the final questionnaire. However, it meets the criteria of covering aspects of daily life that patients value (Gill and Feinstein, 1994; Guyatt and Cook, 1994; Locker and Allen, 2007).

Initially 46 items were chosen by reviewing existing oral health and child health measures in four health domains: symptoms, functional limitations, emotional well-being, and social well-being.

The item impact study identified those items that arise most frequently and had the greatest importance to the target population. The items were tested on a group of 83 children from the three groups described. Each child was asked about each item and asked to rate how important it was on a four point Likert scale of 0 ("does not bother me at all") to 4 ("bothers me very much"). An impact score was calculated by multiplying the percentage of children with positive responses by the item's means importance rating (as rated by the children on the four point Likert scale) (Jokovic et al., 2002). They were then ranked within the four health domains according to the

impact scores. Those above the median in each ranking were selected for the final questionnaire. The final questionnaire had 36 items (Jokovic et al., 2002).

In the CPQ11-14, questions asked about the frequency of events in the previous three months. The response options were: "Never=0"; "Once/twice=1"; "Sometimes=2"; "Often=3"; and "Every day/almost every day=4". The questionnaire also included two global ratings questions of the child's oral health and the extent to which the condition affected his/her overall well-being were also. The questions were worded as follows: "Would you say that the health of your teeth, lips, jaws and mouth is...?" and "How much does the condition of your teeth, lips, jaws or mouth affect your life overall?" These global ratings had a five-point response format ranging from "Excellent= 0" to "Poor= 5" for oral health, and from "Not at all=0" to "Very much=5" for well-being. Scores for the questionnaire are calculated as total/overall scores by summing the response codes for the questionnaire items or scores for each health domain. The total score of the CPQ11-14 ranges from 0-144. The construct validity and discriminative properties were satisfactory and internal consistency reliability was acceptable to good (Jokovic et al., 2002; Agou et al., 2008; Barbosa et al., 2011; Massarente et al., 2011). However, its evaluative properties as an outcome measure remains to be validated.

1.3.17.3 Parental-Caregiver Perception Questionnaire (PPQ)

Jokovic and co-workers developed a measure for parental perceptions as a component of Child Oral Health Related Quality of Life Questionnaire called the Parental-Caregiver Perception Questionnaire (PPQ) (Jokovic et al., 2003b). Parental knowledge may not be all-inclusive so it is to be considered as complementary and not a replacement for the child's report (le Coq et al., 2000b).

The questionnaire was used to measure the parental perception of their child's OHRQoL and was not intended as a proxy measure where parental opinion was a substitute for that of the child. Parents/caregivers are intimately involved in the daily life, health and healthcare of their children. It was reasonable to assume that the outcome of treatment for any condition was likely to be influenced by parental perceptions of the needs of the child in addition to the actual needs of the child. The procedures used to develop the questionnaires were based on the process described previously (Guyatt et al., 1986) which included:

- A literature review
- Expert opinion from a panel of relevant health care professionals and parents of affected children
- An item impact method used to select the items for the questionnaire

The same four health domains of the CPQ which included: oral symptoms (six questions), functional limitations (seven questions), emotional well-being (eight questions), and social well-being (ten questions) were used, and internal consistency reports demonstrated a Cronbach's alpha of 0.94 and its test re-test reliability was good (ICC =0.85) with good construct validity

(Jokovic et al., 2003a; Jokovic et al., 2003b; Zhang et al., 2007b; Agou et al., 2008; Do and Spencer, 2008; Malden et al., 2008; Berger et al., 2009; Goursand et al., 2009).

The questionnaire consists of thirty-one questions worded as "During the last 3 months, how often has your child (had/been) "mouth sore" or "upset"? The response options were: "never=0", "once or twice=1", "sometimes=2", "often=3", "every day or almost every day=4". These questions are scored separately on a Likert (Likert, 1932) response format with a total score ranging from 0 to 124. It also has two Global Rating Questions to evaluate the overall well being of the child " How would you rate the health of your child's teeth, lips, jaws and mouth?" and "How much is your child's overall wellbeing affected by the condition of his/her teeth, lips, jaws or mouth". These global rating questions had a five-point response format from "Excellent" to "Poor" for oral health and from "Not at all" to "Very much" for well-being (Jokovic et al., 2003b).

As the parental knowledge of their children could be limited, particularly with respect to activities or relationships that exist outside the home and with respect to internal feeling states, a "Don't know" response was also allowed. This was deemed essential in studies in which participants report their perceptions of the health or qualify of life of another individual. The author further explains that, while the use of "Don't Know" option was not an issue in longitudinal studies where the aim is to assess within-subject change, it is clearly an issue in cross-sectional studies, where differences between groups are being explored. The following three options were suggested to deal with "Don't Know" responses (Jokovic et al., 2003b):

1. The exclusion of cases with "don't know" responses, but this leads to the loss of valuable data, and may compromise studies with small sample size.
2. Dropping items from the questionnaire that have a high proportion of "don't know" responses. This approach has been shown to have detrimental effects on the criterion validity of the scale (Marshman et al., 2007).
3. Replacing all "don't know" response by (0). This option was shown not to have any effects on the internal consistency of the PPQ (Marshman et al., 2007; Goursand et al., 2009).
4. The fourth is using the adjusted score by giving all "don't know" responses the value (0). The PPQ scores are then calculated by summing the response codes to all the 31 items and dividing this sum by the number of items for which a valid response was obtained. This adjusted score represents the mean item score for those items that were answered. Using this option showed good discriminant and construct validity (Jokovic et al., 2003b; Marshman et al., 2007).

Jokovic and co-workers investigated the agreement between mothers and children aged 11-14 years in rating child oral related quality of life using CPQ11-14 and the PPQ. These two questionnaires are analogous and have 31 items in common. The relationship between the scores of the two questionnaires was investigated using (1) comparisons of scores, (2) correlation analysis for scores and global ratings, (3) correlation analysis for individual questions, and (4) scatter plots of the scores of parent-child pairs. The comparison analysis assess agreement at the level of the group, while the correlation analysis assess agreement at the level of the individual pairs (Jokovic et al., 2003a). Jokovic and co-workers concluded that, at the group level, agreement between mothers and children was good. However, at the individual levels and for the emotional and social well being the agreement was only moderate (Jokovic et al., 2003a). This is explained by the fact that these domains address events that mothers do not have insight into as 11-14 year old children are intensely peer-oriented, and the peer-group dynamics is central in their lives (Bee, 1998; Hetherington et al., 1999).

1.3.17.4 Family Impact Scale (FIS)

The last component of the COHRQoL questionnaire is the family impact scale (Locker et al., 2002a). The family has an important role in child health: a sick child is likely to impact on the rest of the family, healthcare intervention often addresses both parental

and child concerns and finally, a parental report will be affected by the extent to which the family is affected by the child's condition. The FIS was developed as a generic measure for any dental or oro-facial condition. It evaluates the impact of dental/orofacial conditions of a child on parents and other family members as reported by parents.

The item impact study model, which was described previously, was used to develop the Family Impact Scale (Guyatt et al., 1986; Juniper et al., 1996), and hence the scale shares the same limitations of CPQ8-10, CPQ11-14, and PPQ as items were developed by reviewing the literature and were not patient/person centred, and that some low prevalence high impact items may have been excluded from the final questionnaire. However, it has the advantage that it covers aspects of daily life that are considered important by patients/parents. An additional advantage is that the measure has a wide range of high prevalence items, which reduces floor effects and promotes sensitivity to change (Locker and Allen, 2002). It was shown to be sensitive to variations in family impact and almost 75% of parents/caregivers reported that the condition had affected the family "sometimes or often/everyday" in the last three months. The measure was good at discriminating between the three groups affected by dental and orofacial conditions and had good construct validity, and hence was used in this research to discriminate between the test and control groups. The internal consistent reliability reported a Cronbach's alpha of 0.83 and was reproducible (ICC =0.80) for those who reported their child's condition as stable (Locker et al., 2002a; Jokovic et al., 2003b; Agou et al., 2008; Malden et al., 2008; Barbosa Tde and Gaviao, 2009; Berger et al., 2009; Goursand et al., 2009).

The questionnaire consists of fourteen questions scored on a Likert response format (Likert, 1932), which asked about the frequency of events in the previous 3 months " During the past 3 months, how often.... (has there been disagreement or conflict in your family)...because of your child's teeth, lips, mouth or jaws"; the response options with respect to frequency were as follows: "never=0"; "once or twice=1"; "sometimes=2"; often=3"; everyday or almost every day=4". A don't know response was also allowed. Parents/caregivers were also asked to give overall or global assessments of the child's oral health and the extent to which the oral or oro-facial condition in question affected the child's overall well-being. These global ratings had a 5-point response format from

"Excellent" to "Poor" for oral health and from " Not at all" to "very much" for well being. The total scores for the Family Impact Scale range from 0 to 56 (Jokovic et al., 2003b). The responses of parents are scored under four domains, which include: parental/family activity (five questions), parental emotions (four questions), family conflict (four questions), and financial burden (one question).

1.3.18 Self Esteem

1.3.18.1 Definition

In 1965 Rosenberg defined self-esteem as "a stable sense of personal worth or worthiness" (Rosenberg, 1965a). In 1969 Branden established a more comprehensive definition of self esteem, which is "the disposition to experience oneself as being competent to cope with the basic challenges of life and of being worthy of happiness. It is confidence in the efficacy of our mind, in our ability to think. By extension, it is confidence in our ability to learn, make appropriate choices and decisions, and respond effectively to change. It is also the experience that success, achievement, fulfillment and happiness are right and natural for us" (Branden, 1969).

1.3.18.2 The Three Types of Self-Esteem

Global self-esteem (Trait Self-Esteem)

Sometimes self-esteem is used to refer to a personality variable that represents the way people generally feel about themselves. Researchers call this form of self-esteem "global self-esteem" or "trait self-esteem", as it is relatively enduring across time and situations. Depictions of global self-esteem range widely. Some researchers take a cognitive approach, and assume that global self-esteem is a decision people make about their worth as a person (Crocker and Wolfe, 2001; Crocker and Park, 2004), whilst others emphasise emotional processes and define global self-esteem as a feeling of affection for oneself that is not derived from rational and judgmental processes (Brown and Mankowski, 1993). However defined, global self-esteem has been shown to be stable throughout adulthood, with a probable genetic component (Kernis, 2006).

Feelings of Self-worth (State Self-Esteem)

This is what people mean when they talk about experiences that "threaten self-esteem" or "boost self-esteem". A person might say his/her self-esteem was sky-high after getting a big promotion or a person might say his/her self-esteem plummeted after a

divorce. These self-evaluative emotional reactions of feeling proud or pleased with ourselves (on the positive side), or humiliated and ashamed of ourselves (on the negative side) are referred to as feelings of self-worth. Global self-esteem persists while feelings of self worth are temporary (Kernis, 2006).

Self-Evaluations (Domain Specific Self-Esteem)

This refers to the way people evaluate their various abilities and attributes. For example, a person who doubts his ability in school may be said to have "low academic" self esteem and a person who thinks she is good at sports may be said to have "high athletic" self esteem. These self-evaluations or self-appraisals refer to the way people evaluate or appraise their physical attributes, abilities, and personality characteristics. Following on from this people can have different levels of self-esteem in different areas. One person could have high athletic self-esteem but low artistic self-esteem, while another person could have low mathematic self-esteem but high social self-esteem (Kernis, 2006).

1.3.19 Self Concept

The esoteric description and inter-changeability of terminology has led to concepts such as self image and self esteem being used synonymously (Hughes, 1984) and the expansion of a plethora of ill-defined and confounding labels such as self worth, self belief, self concept, self awareness and self regard being proposed (McGuire, 1994).

Many definitions of self-esteem and self-concepts have been proposed, yet there remains noticeably little agreement in nomenclature. However, evident themes within the literature suggest the following notions with regards to self: (1) the global over-arching view of self may be regarded as "self-concept"; (2) the evaluative aspect relates to worth and "self-esteem"; (3) the descriptive facets referring to characteristic available to an individual in defining self may be understood as "self image"; and (4) a notion of competency in terms of how effective a person considers they will be in undertaking a task has been referred to as "self-efficacy" (Butler and Gasson, 2005).

1.3.19.1 Self-Esteem/Self-Concept Measures

The range and variety of self concept/self esteem scales is vast. Blascovich suggested that at least 200 measures of self esteem have been developed, yet most measures tend to be short lived and of debatable quality (Blascovich and Tomaka, 1991).

A PsychINFO database search for the last twenty years, using key word matches for self concept, self image, self worth, self esteem, self competence, self confidence, self efficacy and self construing retrieved 1426 articles and collated information only from 1226 articles after eliminating unrelated articles (Butler and Gasson, 2005). It was reported that the Piers-Harris Self Concept Scale was the most frequently reported measure (433 times), followed by Rosenberg Self Esteem Scale (263 times), with only four scales reported on fifty or more occasions.

1.3.20 The Piers-Harris Children's Self-Concept Scale

The Piers-Harris Children's Self-Concept Scale was originally developed in the early 1960s to provide a brief, self-reporting instrument for the assessment of self-concept in children and adolescents. As defined by the scale authors, self-concept is "a relatively stable set of self attitudes reflecting both description and evaluation of one's own behaviour and attributes". Self concept is regarded as synonymous with self esteem, and although originally conceived as unidimensional, psychometric analysis indicated that the scale is multi-factorial, with up to six clusters/domains (Piers and Herzberg, 2007). The scale originally consisted of 25 positively and 35 negatively phrased items presented as first person declarative statements (e.g. "I have a nice hair"; "I am dumb about most things") where children respond to each item with either a "yes" or "no" answer.

The Piers-Harris 2 is a 60-item self-report questionnaire, subtitled *The Way I Feel About Myself.* It is designed for administration to children who are at least 7 years old and have at least a second-grade reading ability. The measure can be used with adolescents up to 18 years of age. The Piers-Harris 2 items are statements that express how people may feel about themselves. Respondents are asked to indicate whether each statement applies to them by choosing a "yes" or "no" answers.

The Piers-Harris 2 includes the same Self-Concept and Validity scales as the original Piers-Harris. The Self-Concept scales comprise the Piers-Harris 2 Total (TOT) score,

which is a general measure of the respondent's overall self-concept, and the six domain scales, which assess specific components of self-concept. The domain scales include (1) Behaviour Adjustment (BEH), (2) Intellectual and School Status (INT), (3) Physical Appearance (PHY), (4) Freedom from Anxiety (FRE), (5) Popularity (POP), and (6) Happiness and Satisfaction (HAP). The Self-Concept scales are scored so that a higher score indicates a more positive self-evaluation in the domain being measured. The Piers-Harris 2 Validity scales include the Inconsistent Responding (INC) index, which is designed to identify random response patterns, and the Response Bias (RES) index, which measures a child's tendency to respond yes or no irrespective of item content (Piers, 1969; Piers, 1984; Piers, 1996; Piers and Herzberg, 2002; Piers and Herzberg, 2007).

1.3.20.1 Principles of Use

The Piers-Harris 2 is appropriate for use in any research, educational, or clinical setting that requires efficient quantitative assessment of children's reported self-concept (Butler and Gasson, 2005). As with many self-reported measures, users should bear in mind that the intent of the Piers-Harris 2 is readily apparent to most children and adolescents. For this reason, the responses may be subjected to conscious and unconscious distortions, usually in the direction of greater social desirability as some questions may elicit answers that are socially unacceptable or undesirable. Answers reporting behaviours or attitudes that conform to the norms are deemed socially desirable, and those that report deviations from the norms are considered socially undesirable and hence this may lead to social desirability bias.

Although the scale is a useful instrument, it cannot by itself provide a comprehensive evaluation of a child's self-concept (Piers and Herzberg, 2007). Such an evaluation is a complex task requiring clinical sensitivity and familiarity with the applicable research literature. In making clinical judgments concerning Piers-Harris 2 results, users should integrate other sources of data, which may include clinical interviews with the child and other informants, prior history, school records, classroom observations, and results from other psychological tests. The full details of scoring, calculating the validity of scores, plotting the profile, and interpreting results are all available from the Piers-Harris 2 developers (Piers and Herzberg, 2007).

1.3.20.2 Total Score (TOT)

TOT is the number of items endorsed in the direction of positive self-concept, and thus has a raw-score of 0 to 60. TOT is a measure of general self-concept: Higher scores (≥ 60T) indicate favourable self-concept (i.e. high degree of self-esteem), whereas lower scores (≤ 39T) are associated with lower self-concept. When the TOT score is low, it may reflect deficits in specific domains of self evaluation, or a more generalised deficiency in self-concept. TOT is not calculated by summing up the raw scores from the six domain scales, because some items appear on more than one scale, and the TOT raw score is not an equivalent to the sum of the domain scale raw scores (Piers and Herzberg, 2007).

1.3.20.3 Behavioural Adjustment (BEH)

The 10-item BEH scale measures admission or denial of problematic behaviours. Item content ranges from specific behaviours (e.g., Item 45, "I get into a lot of fights") to more general statements concerning problem situations in the home or at school (e.g. Item 14, "I cause trouble to my family"). The item most strongly correlated with the BEH score is Item 19, "I do many bad things". Scores can range from (≥ 56T to ≤ 39T).

1.3.20.4 Intellectual and School Status (INT)

The INT scale consists of 16 items that reflect a child's assessment of his or her abilities with respect to intellectual and academic tasks. The items also cover general satisfaction with school and future expectations about achievement. Some items address how a child's intellectual capabilities affect key relationships (e.g. Item 16, "I am an important member of my family"; Item 26, "My friends like my ideas"). The item most strongly correlated with the INT score is Item 5, "I am smart". Scores can range from (≥ 56T to ≤ 39T).

1.3.20.5 Physical Appearance and attribute (PHY)

The 11-item PHY scale measures a young person's appraisal of his or her physical appearance, as well as attributes such as leadership and the ability to express ideas. The item most strongly correlated with the PHY score is Item 44, "I am good-looking". Scores can range from (≥ 56T to ≤ 39T).

1.3.20.6 Freedom From Anxiety (FRE)

The FRE scale consists of 14 items that reflect anxiety. Individual items tap a variety of specific emotions, including worry, nervousness, shyness, sadness, fear, and general feeling of being left out of things. The item most strongly correlated with FRE score are Item 29, "I worry a lot" and Item 32, "I feel left out of things". Scores can range from ($\geq$ 56T to $\leq$ 39T).

1.3.20.7 Popularity (POP)

The 12-item POP scale represents a child's evaluation of his or her social functioning. The item content covers perceived popularity, ability to make friends, and feelings of inclusion in activities such as games and sports. The item most strongly correlated with the POP score is Item 41, "I have many friends". Scores can range from ($\geq$ 56T to $\leq$ 39T).

1.3.20.8 Happiness and Satisfaction (HAP)

The HAP scale consists of 10 items that reflect feelings of happiness and satisfaction with life. Most items on the HAP scale are general and broadly phrased; only one item refers to a specific personal attribute (Item 49, "I have a pleasant face"). The item most strongly correlated with the HAP score is Item 40, "I am unhappy". Scores can range from ($\geq$ 56T to $\leq$ 39T).

1.3.21 Rosenberg Self Esteem Scale (SES)

The Rosenberg self-esteem scale was developed by Rosenberg and has been used in as much as one quarter of all the research that exists on self esteem (Rosenberg, 1965a; Tafarodi and Swann, 1995). The original sample for developing the scale was a group of 5024 high school juniors and seniors from 10 randomly selected New York State high schools.

The SES is the standard against which new measures are evaluated. Its ease of administration, scoring, reliability, validity, and brevity underlie the recommendations for its use as a straightforward measure of positive or negative feelings about ones self. The Rosenberg SES, however, is not without possible complications. For example, the items may be susceptible to socially desirable responding as previously discussed (Robinson et al., 1991).

This scale was originally designed to measure adolescents' global feelings of self-worth or self acceptance. The 10 items that make up the Rosenberg Self-Esteem Scale (SES) were designed to optimise ease of administration, economy of time, unidimensionality (set of items forming an instrument and all measure just one trait) (Hattie, 1985), and validity. Self-Esteem Scale items require the respondent to report their feelings directly. The SES is constructed of five positively and five negatively phrased items composed exclusively of first person evaluative statements about the self (the five negatively scored items are reversed before the total score of the scale is calculated) (Rosenberg, 1965a). It is typically scored using a four point response format (strongly agree, agree, disagree, strongly disagree) designated with numerical values of (0, 1, 2, 3) or in some studies (1, 2, 3, 4) resulting in a scale range of 0 to 30 or 1 to 40 with higher scores representing higher self esteem. Some authors, however, have adopted more familiar Likert-style formats employing 5 or 7 point scales resulting in broader ranges of SES scores.

1.3.22 Administration Methods

Guyatt and co-workers described the modes of administration of HRQoL measures (Guyatt et al., 1986).

- An interview administration will ensure that all questions are completed and that any misunderstandings are clarified. This approach is expensive, time consuming and respondents may be unwilling to acknowledge problems.
- A telephone-administered questionnaire may be more effective because it is cheaper and also allows any ambiguities to be resolved.
- A self-administered method requires few resources but many may not return it or leave it incomplete.

Surrogate responders, defined as "individuals who provide information regarding a specific issue in place of the individual involved in the study" can also be used and they have the advantage that it may reduce stress for the target group but they may not reflect the patient's position accurately as detailed in studies that examined the use of parents as proxies when examining hypodontia patients (Jokovic et al., 2003a; Zhang et al., 2007a).

1.3.23 Limitations of Psychometric Measures (Questionnaires and/or Scales)

In a review of the available literature for the psychometric scales/questionnaires used to measure personality trait such as self esteem and self concept (Butler and Gasson, 2005) the limitations of these psychometric measures were summarised in the following points:

1. Scales tend to be normed on geographically limited samples, with potential problems in generalisation.
2. Most scales/questionnaires are generally "author generated", and constructed in a way suggesting the author's direct involvement.
3. Scale and questionnaires are usually on the functionalist approach and are often closed ended and do not allow individuals to provide their own descriptions of their self-perception or quality of life (O'Boyle et al., 1993).
4. The "language of anchors" varies across scales measuring the same item making comparison between scales difficult (e.g., "Strongly agree", "Strongly disagree", "not at all", "very much so", "yes", "no").
5. Current scales are predominantly of Western origin taking little account of "cultural philosophies". However, most psychometric scales have been translated and validated in different languages.
6. The length of time to complete a scale/ questionnaire may arise as a salient issue especially for young children. Selecting scales with a greater number of items fosters more thorough sampling from the domain of possible descriptions, producing a more heterogeneous and representative instrument with increased validity and generality. On the other hand comprehensiveness and reliability may be reduced with brevity (Awad et al., 2008).
7. Multi-word descriptions may involve complex, multifaceted evaluations that can be conceptually difficult for young children.
8. Repetition of items of a scale/questionnaire can be a frustrating experience for respondents, especially young subjects. Piers-Harris has 15 very similar paired items to provide an estimate of "inconsistent responding" (Piers and Herzberg, 2007).

9. Response bias is a major issue as scales/questionnaires remain vulnerable to social desirability. Subjects tend to respond in a way that is socially appropriate rather than how the individual feel or perceive his/herself or their quality of life (Tourangeau and Yan, 2007).

This literature review covered the areas of developmental dental anomalies and also the measures currently employed to investigate oral health related quality of life in people with orofacial conditions in addition to measures of self-esteem and self-concept.

Chapter 2

Development of an Irish National Special Dental Needs Database

Chapter 2: Development of an Irish National Special Dental Needs Database

2.1 Background

Since the establishment of the Special Dental Needs Clinic in the Dublin Dental University Hospital in 2001, consistent patient referrals have been received by this tertiary care clinic.

Patients are referred for comprehensive treatment planning specific to each individual case, and frequently treatment is also completed on site.

Due to the high cost and complexity of treatment of this patient population and the limited availability of funding for treatment in the Dublin Dental University Hospital, a subgroup of patients with special dental needs was identified in the Dental Hospital.

This cohort of patients has specific dental conditions many details of which could not be captured and recorded in the main database of the Dental Hospital. Additionally, the ability to search specific fields and identify patterns was not achievable with the main database. Hence the decision was made to develop a purpose specific database appropriate for this patient population to collate and to delineate better this group.

2.3 Purpose of database

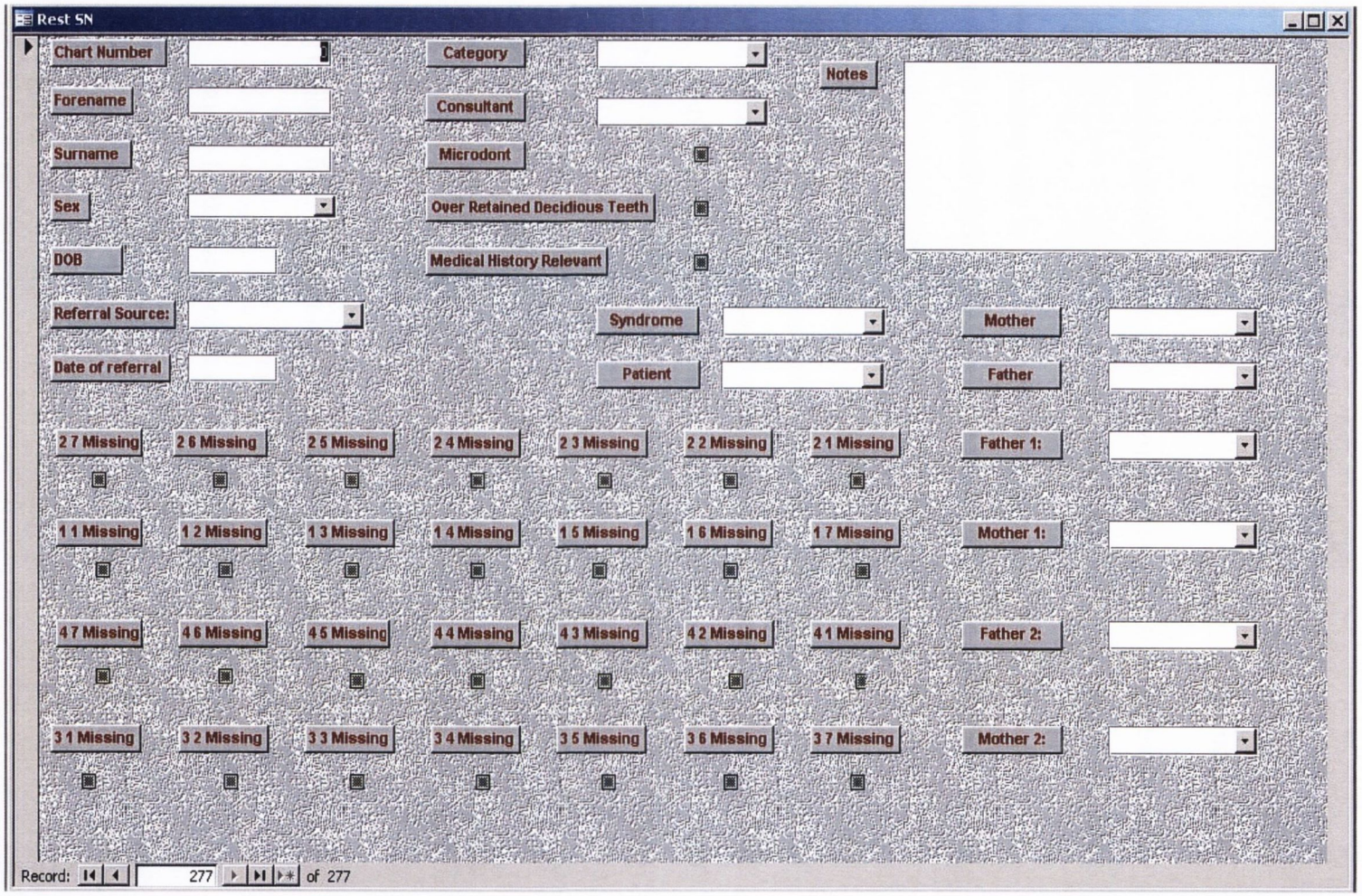
- To record relevant information specific to special dental needs patients not captured in the current electronic dental or paper recording systems.
- To facilitate retrieval of information efficiently and effectively.
- To store patient information safely.
- To permit interrogation of patients' biographic and clinical data to examine service levels and regional information.
- To facilitate collation of clinical records for this patient cohort.

2. 4 Initial projects

2.4.1 Microsoft Access® database

An initial version of the database was designed using proprietary products such as Microsoft Access® for Windows®. An Access® template was designed with the relevant clinical details (Figure 2-1).

Figure 2-1: Microsoft Access initial database template



Relevant patient data were entered into the Microsoft Access® database from individual patient clinical notes and radiographs. Soon after compiling the database, it was clear that Microsoft Access® was not flexible enough to satisfy the requirements of the purpose specific database required. The main drawbacks of the Microsoft Access® database were as follows:

- Screen Cluttering

  The Microsoft Access® database was designed on one template with all relevant information placed on the same template. This led to cluttering of fields on screen and as a consequence it was very easy to omit fields, or enter information incorrectly.

- Lack of important information

  By the nature of a dental database, twenty-eight fields (the number of teeth excluding third molars) were necessary to describe dental patterns alone. This limited the number of extra fields available for other relevant information, restricting the amount of data that can be recorded.

- Limited search ability

  In order to search specific patterns (i.e. number and pattern of developmentally missing teeth), at least 28 fields had to be searched for each individual query. The option of examination of the database across all fields was not possible.

- No edit/save facility

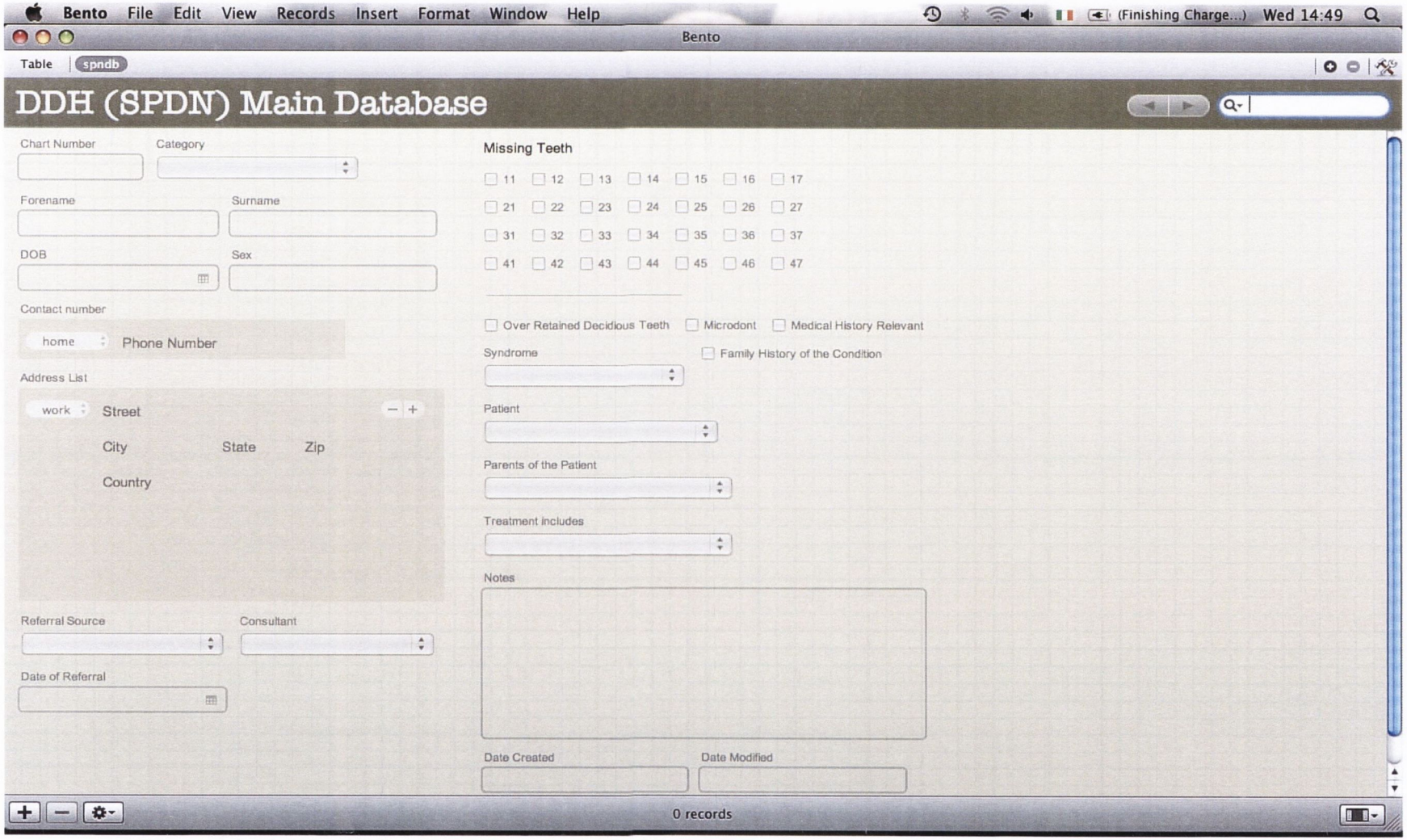
  In the initial version of Access® database, there was no facility to save or edit data as required. Records were easy to accidently modify or delete, and there was no traceability to identify when the record was created or last modified.

2.4.2 Bento® database

A more functional proprietary database was designed using Bento® software for Apple Mac® [Version 1.0V2 (392), FileMaker, inc. USA 2007-2008] in order to overcome identified shortcomings of Microsoft Access®. The database template was designed to contain all relevant clinical and biographical details and have an integral searching ability. All patient data were mapped and imported from Microsoft Access® to Bento® database. All further data, which could not be entered into the Microsoft Access®

database were gathered from patient records and entered into the Bento® database (Figure 2-2).

Figure 2-2: Bento Template of Special Dental Needs Database (SPNSDB).



As the database grew from 277 to 400 entries and new records were continuously being added, some limitations became evident which included:

- Screen Cluttering

  Even though the Bento® database allowed the addition of more fields (as compared to Microsoft Access® database i.e. biographical fields) this was still insufficient as new fields were required. The addition of these new fields led to screen cluttering and an ergonomic screen arrangement could not be formatted.

- Lack of additional important data

  On assessing the Bento® database, it was evident that it lacked important clinical data, which were important from a diagnosis and future research perspective. Examples of this include the detailed records of over-retained primary teeth and family history of the condition, which would facilitate future genetic research to investigate these conditions.

- Limited searching ability

  Despite the fact Bento® had a more powerful searching ability (maximum of ten filters at a time), it did not actually satisfy the searching requirements of the database, which required a searching mechanism of a minimum of twenty-eight filters at a time.

- Security issues

  One major drawback of the Bento® database is the fact that it was installed on a password protected Macbook® which was stored securely. This has the associated risk of theft of the Macbook® with patient data.

- Creation of double records

  Due to the fact that the SPNSDB is stored separately from the hospital's main database "Salud®" (tth Salud Dental Suite, Copyright 1998-2006, Two-Ten Health, Version 1.12.2, Revision 21), this would inevitably lead to the creation of duplicate records. This means that, if patient's data were modified on the hospital's database, these changes will not be simultaneously modified on the SPNSDB. There was also no facility to trace back changes. Consequently the SPNSDB would be out of date and of limited use after a short period of time.

2.5 Rationale for the development of the new web based database system

The SPNSDB has very specific software requirements. Initial attempts at project implementation were completed using proprietary software products, such as Microsoft Access® for Windows® and Bento® for Apple Mac®. The shortcomings of both products became apparent shortly after the first prototypes were developed: both products were not sufficiently flexible to accommodate all the requirements of SPNSDB. Also, development of a product exclusively for Microsoft Windows® or Apple Mac OS® platform would limit any future potential usage to users of these specific operating systems. Any attempts to modify an initial prototype into a developed system would face the challenges of supporting multiple users and operating systems and consequently would likely result in the need for a complete code rewrite of the proprietary product.

As a result, a new development approach had to be adopted. To accommodate the SPNSDB requirements open source web-based technologies were employed. The use of these agile technologies allowed easy adaptation of the system and streamlining of its development, so that the demanding SPNSDB software requirements could be met. During its development the project was significantly further enhanced and expanded beyond the original scope.

A set of requirements and guidelines was established to design a new database. These included:

- The requirement to contain all relevant clinical and biographical patient information.
- Security: The database would have to be password protected and accessible only to authorised persons.
- Automated identification of new patients: The database should identify all new patients who are registered in the Dublin Dental University Hospital and tagged as Special Dental Needs Patients.
- Updatable: The facility must exist to update database automatically on a scheduled basis by integrating with the hospital's main database.

- User friendly: A simple method should be devised to enter and extract information easily.

- Single records: The database should integrate with the Dublin Dental University Hospital's electronic records to avoid creating duplicate records.

- Editing: The database should have the facility to edit and save data in a controlled manner to avoid modifying/deleting data accidentally.

- Access to database: the option should exist to provide different levels of access to multiple users under an overall operation control.

2.5.1 Software Requirements:

The SPNSDB had to provide the following capabilities:

1. To hold and display the list of special dental needs (SPNS) patients.
2. To allow integration of patient electronic dental records via various parameters, including biographical information and specific dental information.
3. To facilitate the automated updates of new patients added to the database. In addition the facility was required to allow patients to be screened to ensure that they meet the inclusion criteria prior to joining the database. This ensures that information was independently checked for a second time to ensure greater accuracy of database.
4. To integrate and synchronise biographical patient data from the Dublin Dental University Hospital's "Salud®" electronic patient database.

2.5.2 Software Stack

After the unsuccessful use of proprietary software systems, a new development approach was chosen, which relied on open web-based standards. These included:

- Apache web server, version 2 (Apache, 2004b).
- Hypertext PreProcessor Scripting Language (PHP), version 5 (PHP, 2004).
- MySQL database, version 5 beta (MySQL, 2005).
- Drupal website development framework, version 5.0 beta 1 (Drupal, 2006).

The use of these products eliminated dependence on proprietary software products and associated licenses while providing full functionality for a new database.

2.5.2.1 Apache Web Server, Version 2

The Apache HTTP Server Project is a collaborative software development effort aimed at creating a robust, commercial-grade, and freely available source code implementation of a Hyper Text Transfer Protocol (HTTP) web server. The project is jointly managed by a group of volunteers located around the world, using the Internet to communicate, plan and develop the server and its related documentation. The apache web server project is part of the Apache Software Foundation. In addition, hundreds of users have contributed ideas, codes, and documentation to the project (Apache, 2004a).

A study conducted by NetCraft (an internet services company based in Bath, England, which provides Internet research data) reported that as of March 2010, Apache accounts for 54.55% of total public web servers on the Internet (Netcraft, 2010).

2.5.2.2 Hypertext Preprocessor PHP (Version 5)

PHP is a widely used general-purpose scripting language that is particularly suited for Web development and can be embedded into Hyper Text Mark Up Language (HTML). It allows website developers to write dynamically generated sites and pages quickly and efficiently (PHP, 2004). PHP is the language used to develop Drupal Web Development frameworks.

2.5.2.3 MySQL database, version 5 beta

MySQL is a relational database management system, which provides a backend for storing, searching and retrieving data. MySQL is characterised as a free, fast, and reliable open source relational database. However, it does lack some sophistication and facilities but provides a feasible alternative to commercial databases management systems, such as Oracle®, Microsoft's SQL and IBM's DB2 (MySQL, 2005).

2.5.2.4 Drupal website development framework (Version 5.0 beta 1)

Drupal is a content management system (CMS) written in PHP language. It provides a solid framework for designing and developing websites of arbitrary complexity via a combination of Drupal Core and 3rd party modules (plugins). Drupal can run on any computing platform that supports a web server capable of running PHP code (Apache)

and a database (MySQL) to store its content and configuration data. Developing websites in Drupal requires minimal programming experience.

2.5.3 Database structure

The database structure of SPNSDB was represented as a table that contained a list of patients, their biographical details, dental records and other comments. However, in reality the MySQL database used to develop the SPNSDB contained many additional tables (about 60). This is attributed to Drupal's internal and 3rd party module implementation.

It must be noted that the database structure has been specifically designed to allow personal record integration from Salud database – a database that contains personal and clinical records of patients attending the Dublin Dental University Hospital.

The advantages of integrating the SPNSDB to the hospital's main database "Salud" are:

- Automatic updates to patients biographical details.
- Addition of new patients is facilitated minimizing administration time and increasing efficiency.

The data was split into 6 categories:

- General Information
- Category and Medical History
- Referral Information
- Missing and Over-retained primary Teeth
- Hyperdontia
- Family History

More detailed descriptions of each field are as follow:

2.5.3.1 General Information

Human-readable Name	Machine Name	Type	Purpose
Patient's Status	field_patient_status	Integer	It sorts patients according to their tagging status.
Chart Number	field_patient_chart_number	Text	
First Name	field_patient_first_name	Text	
Middle Name	field_patient_middle_name	Text	
Last Name	field_patient_last_name	Text	
Gender	field_patient_sex	Text	
D.O.B.	field_patient_dob	Date	Date of Birth
Address 1	field_patient_address1	Text	Salud requires 3 fields for address, so it had to be mapped in our database in the same way to facilitate synchronisation.
Address 2	field_patient_address2	Text	
Address 3	field_patient_address3	Text	
City	field_patient_city	Text	
Country	field_patient_county	Text	
Post Code	field_patient_zip	Text	
Area Code	field_patient_area_code	Text	
Telephone	field_patient_landline	Text	
Telephone 2	field_patient_mobile1	Text	
Telephone 3	field_patient_mobile2	Text	
Email	field_patient_email	Text	

2.5.3.2 Category and Medical History

Human-readable Name	Machine Name	Type	Purpose
Category	field_patient_category	Text	
Amelogenesis Imperfecta	field_patient_category_amelogenesis	Text	
Dentinogenesis Imperfecta	field_patient_category_dentinogenesis	Text	

Hypodontia	field_patient_category_hypodontia	Text	
Consultant	field_patient_consultant	Text	
Other consultant	field_patient_consultant_other	Text	
Relevant Medical History	field_patient_medical_history	Text	Y/N tick boxes
Relevant Medical History Notes	field_patient_medical_history	Text	
Associated Syndrome	field_patient_associated_syndrome	Text	Y/N tick boxes
Associated Syndrome Description	field_patient_associated_syndrome	Text	

2.5.3.3 Referral Information

Human-readable Name	Machine Name	Type	Purpose
Referral Source	field_patient_referral	Text	
Health Service Executive branch	field_patient_referral_details	Text	
Referring Dentist Name	field_patient_referring_dentist	Text	
Referral Date	field_patient_referral_date	Date	
Referring Dentist Address	field_patient_referring_dentist_0	Text	
Referring Dentist Number	field_patient_referring_dentist_1	Text	
Referring Dentist Email	field_patient_referring_dentist_2	Text	

2.5.3.4 Missing and Over-retained Teeth

Human-readable Name	Machine Name	Type	Purpose
Over-retained Teeth	field_patient_overretained_teeth	Text	Y/N tick boxes
patient_tooth_55	field_patient_tooth_55	Integer	
patient_tooth_54	field_patient_tooth_54	Integer	
patient_tooth_53	field_patient_tooth_53	Integer	
patient_tooth_52	field_patient_tooth_52	Integer	
patient_tooth_51	field_patient_tooth_51	Integer	
patient_tooth_61	field_patient_tooth_61	Integer	

patient_tooth_62	field_patient_tooth_62	Integer		
patient_tooth_63	field_patient_tooth_63	Integer		
patient_tooth_64	field_patient_tooth_64	Integer		
patient_tooth_65	field_patient_tooth_65	Integer		
patient_tooth_85	field_patient_tooth_85	Integer		
patient_tooth_84	field_patient_tooth_84	Integer		
patient_tooth_83	field_patient_tooth_83	Integer		
patient_tooth_82	field_patient_tooth_82	Integer		
patient_tooth_81	field_patient_tooth_81	Integer		
patient_tooth_71	field_patient_tooth_71	Integer		
patient_tooth_72	field_patient_tooth_72	Integer		
patient_tooth_73	field_patient_tooth_73	Integer		
patient_tooth_74	field_patient_tooth_74	Integer		
patient_tooth_75	field_patient_tooth_75	Integer		
Missing Teeth	field_patient_missing_teeth	Text	Y/N tick boxes	
patient_tooth_17	field_patient_tooth_17	Integer		
patient_tooth_16	field_patient_tooth_16	Integer		
patient_tooth_15	field_patient_tooth_15	Integer		
patient_tooth_14	field_patient_tooth_14	Integer		
patient_tooth_13	field_patient_tooth_13	Integer		
patient_tooth_12	field_patient_tooth_12	Integer		
patient_tooth_11	field_patient_tooth_11	Integer		
patient_tooth_21	field_patient_tooth_21	Integer		
patient_tooth_22	field_patient_tooth_22	Integer		
patient_tooth_23	field_patient_tooth_23	Integer		
patient_tooth_24	field_patient_tooth_24	Integer		
patient_tooth_25	field_patient_tooth_25	Integer		
patient_tooth_26	field_patient_tooth_26	Integer		
patient_tooth_27	field_patient_tooth_27	Integer		

patient_tooth_47	field_patient_tooth_47	Integer
patient_tooth_46	field_patient_tooth_46	Integer
patient_tooth_45	field_patient_tooth_45	Integer
patient_tooth_44	field_patient_tooth_44	Integer
patient_tooth_43	field_patient_tooth_43	Integer
patient_tooth_42	field_patient_tooth_42	Integer
patient_tooth_41	field_patient_tooth_41	Integer
patient_tooth_31	field_patient_tooth_31	Integer
patient_tooth_32	field_patient_tooth_32	Integer
patient_tooth_33	field_patient_tooth_33	Integer
patient_tooth_34	field_patient_tooth_34	Integer
patient_tooth_35	field_patient_tooth_35	Integer
patient_tooth_36	field_patient_tooth_36	Integer
patient_tooth_37	field_patient_tooth_37	Integer
Microdontia	field_patient_microdontia	Text

2.5.3.5 Hyperdontia

Human-readable Name	**Machine Name**	**Type**	**Purpose**
Hyperdontic Teeth	field_patient_hyperdontic_teeth	Text	Y/N tick boxes
17	field_patient_hyperdontic_17	Integer	
16	field_patient_hyperdontic_16	Integer	
15	field_patient_hyperdontic_15	Integer	
14	field_patient_hyperdontic_14	Integer	
13	field_patient_hyperdontic_13	Integer	
12	field_patient_hyperdontic_12	Integer	
11	field_patient_hyperdontic_11	Integer	
21	field_patient_hyperdontic_21	Integer	
22	field_patient_hyperdontic_22	Integer	
23	field_patient_hyperdontic_23	Integer	

24	field_patient_hyperdontic_24	Integer
25	field_patient_hyperdontic_25	Integer
26	field_patient_hyperdontic_26	Integer
27	field_patient_hyperdontic_27	Integer
47	field_patient_hyperdontic_47	Integer
46	field_patient_hyperdontic_46	Integer
45	field_patient_hyperdontic_45	Integer
44	field_patient_hyperdontic_44	Integer
43	field_patient_hyperdontic_43	Integer
42	field_patient_hyperdontic_42	Integer
41	field_patient_hyperdontic_41	Integer
31	field_patient_hyperdontic_31	Integer
32	field_patient_hyperdontic_32	Integer
33	field_patient_hyperdontic_33	Integer
34	field_patient_hyperdontic_34	Integer
35	field_patient_hyperdontic_35	Integer
36	field_patient_hyperdontic_36	Integer
37	field_patient_hyperdontic_37	Integer

2.5.3.6 Family History

Human-readable Name	Machine Name	Type	Purpose
Family History of the Condition	field_patient_family_history_of	Text	Drop down list selection
Family History	field_patient_family_history_co	Text	Drop down list selection
Patient	field_patient_family_history__0	Text	Drop down list selection
Parents of the Patient	field_patient_family_history__1	Text	Drop down list selection
Grandparents of the patient (Paternal)	field_patient_family_history__2	Text	Drop down list selection

Grandparents of the patient (Maternal)	field_patient_family_history__3	Text	Drop down list selection
Treatment Included	field_patient_treatment	Text	Multiple choice selection
Number of Implants	field_patient_implants	Integer	
Additional Notes	field_patient_additional_notes	Text	Text Area

These fields were designed to fully detail information necessary to work with SPNS patients.

2.5.4 System integration with Salud

System integration with Salud was facilitated with the help of the DDUH Information Service Staff. Particularly designed fields of patient records in Salud® were exposed to SPNSDB in read-only mode. The view was limited only to personal records of patients who have had been previously identified as SPNS.

The synchronisation was performed in "pull" mode, which means that SPNSDB initiated synchronisation and pulled information from Salud®, not Salud® pushing its updates onto SPNSDB.

The advantages of "Pull" mode over "Push" mode include:

1. Convenience: The Salud database was an already established database and changing its configuration to push updates to SPNSDB would have been more difficult and may have had knock-on effects on other applications.
2. Simplicity: It is much easier to design the SPNSDB at its initial stages to query and pull out specific information fields from Salud. The Salud database contains the electronic records of 236,317 patients (Accessed: 18/05/2010), so the alteration of the newly designed SPNSDB was less cumbersome.
3. Cost: Changing Salud to push data to SPNSDB would require additional expertise and support from the providing company, which would add to costs.

The SPNSDB currently updates itself by synchronising with Salud every morning at 07:00. Such updates allow two events:

- Updating all patients' biographical details. Many patients in the database are young adults and it is anticipated that their names and addresses may change. Any updates to patients' electronic record on Salud by Central Medical Record Area staff are automatically synchronised to SPNSDB.
- Insertion of new records into SPNSDB whenever any patient in Salud is marked with a SPNS flag. Such records are placed into a "Review Queue" for specialists to review these patients and to include them into SPNSDB if appropriate. The tagging of a patient with the SPNS flag occurs at the time of first referral and is indicated on the referral letter by the appropriate consultant, so that the patient can be set up on Salud as SPNS. Sometimes when the patient is first seen for an assessment his/her diagnosis does not satisfy the criteria for inclusion into the Special Dental Needs scheme. Hence the flag SPNS is then removed and they are relabeled appropriately. If in error this re-tagging does not happen, then these patients are easily identifiable from the "Review Queue", when their clinical notes are carefully examined before entering them into the SPNSDB.

The synchronisation was facilitated using Open Data Base Connectivity (ODBC) technology. Open Database Connectivity (ODBC) is "Microsoft's strategic interface for accessing data in a heterogeneous environment of relational and non- relational database management systems" (Microsoft, 2007). ODBC provides an open, vendor-neutral way of accessing data stored in a variety of proprietary personal computers, minicomputers, and mainframe databases. ODBC provides a universal data access interface. With ODBC, application developers can allow an application to concurrently access, view, and modify data from multiple, diverse databases (Microsoft, 2007).

This synchronisation was established to perform lookup and updates. The following fields are exposed by Salud and synchronised with SPNSDB:

- First name
- Middle name
- Last name
- Address (fields 1,2,3)

- Home city
- Postcode
- Country
- Telephone 1 (combined from area code + telephone fields in Salud)
- Telephone 2
- Telephone 3
- Email
- Gender
- Date of Birth
- Referring Dentist (combined from GDP title, first name, and last name in Salud)
- Referring Dentist's Address (combined from Salud's Pract name, address 1,2,3)
- Referring Dentist's Phone number

It is important to note that the last three fields (referring dentist's information) are only completed when previously empty in SPNSDB. This is to prevent the overwriting of information already entered manually to SPNSDB before synchronisation, which were not present in the main Database "Salud". These fields do not need updating once entered, as the referring dentist is always the first dentist who referred the patient to the Dublin Dental University Hospital.

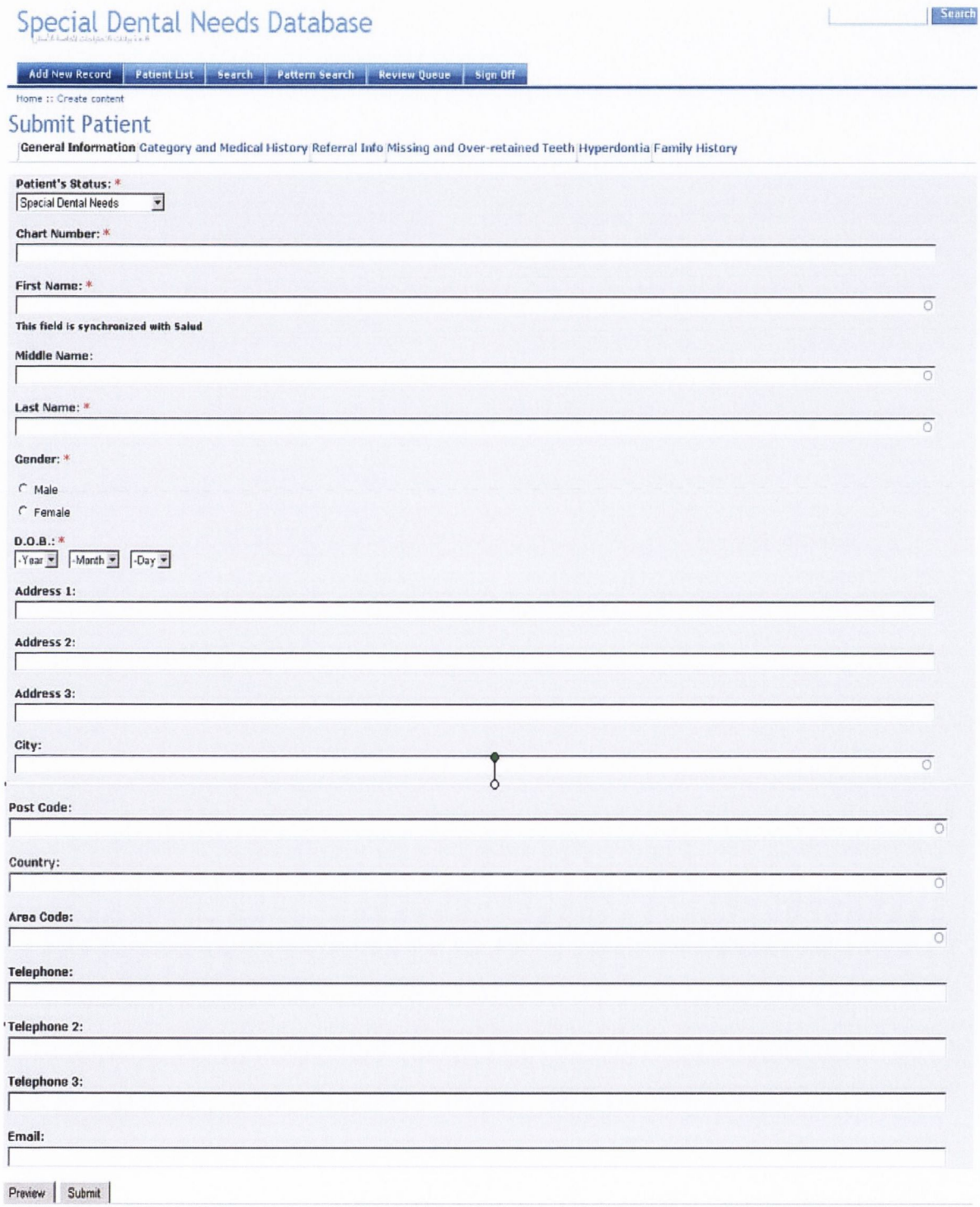
2.5.5 User Interface Design

The SPNSDB interface is based on Drupal's Zen Classic theme. Zen theme allows easy customisation and provides simple and uncluttered interfaces. The choice of development technology partially predetermined the interface look – a typical browser page (Drupal, 2006). Drupal provides a choice of themes that can be customised to accommodate specific requirements of a particular project. The theme was modified to accommodate specific display and ordering requirements of teeth, as well as the large number of choices required during any given search.

2.5.6 General information

All patients' general information is obtained from Salud once the patient is tagged as SPNS, and the SPNSDB synchronises with Salud at 0700 each day.

Figure 2-3: This figure shows the general information tab of SPNSDB

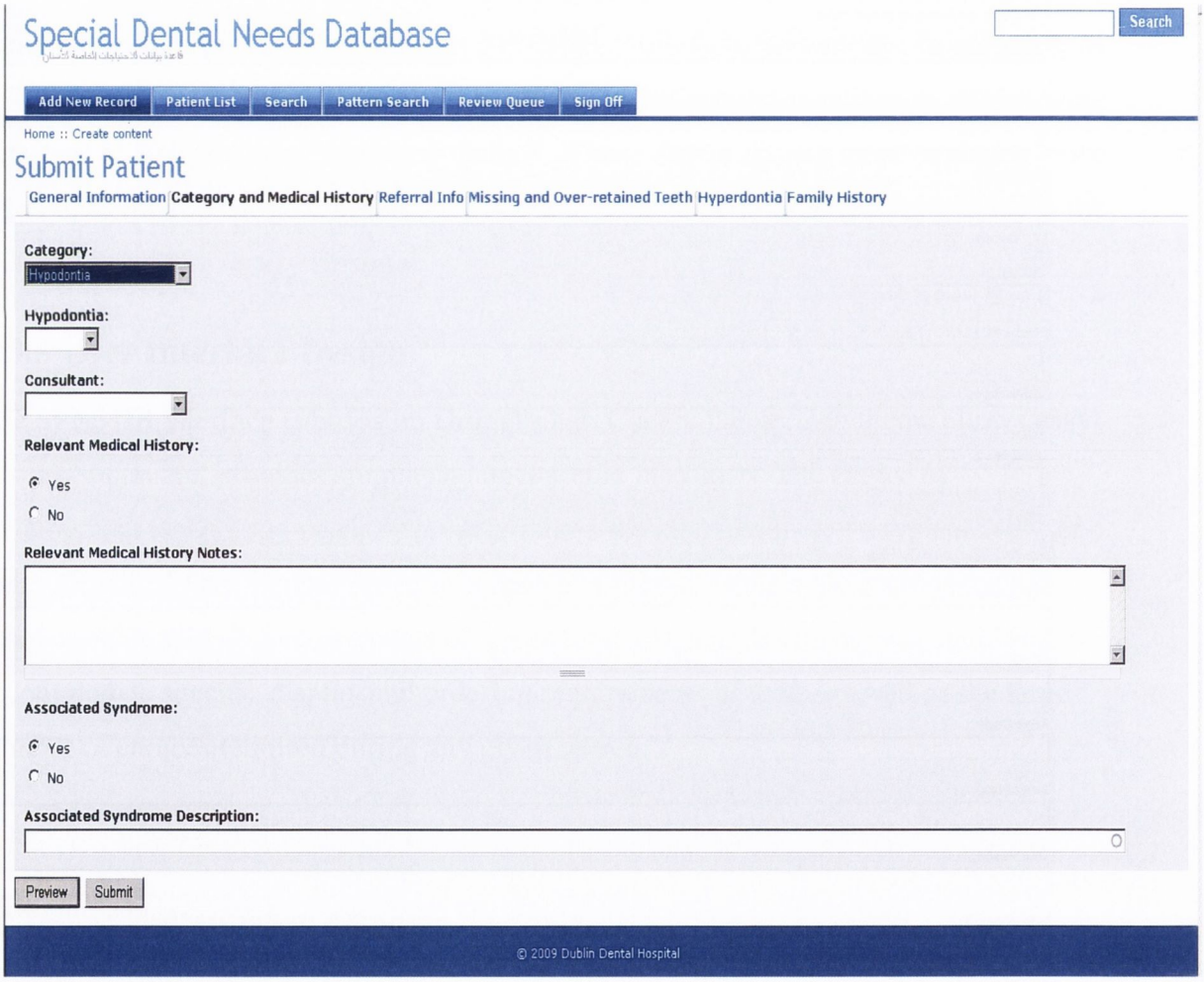


2.5.7 Category and Medical History

In the category and medical history tab the patient is classified according to his/her developmental dental condition. Once the appropriate category is selected the information in this tab changes to allow entering of information related to the specific condition. In this example the condition selected is hypodontia. This opens a second drop down list as to the type of hypodontia (mild/moderate/severe). There is a drop down list for the name of the consultant providing care for the patient.

There is also a tick box for the relevant medical history and any associated syndromes. This opens a free text window for the related information.

Figure 2-4: Category and medical history tab



2.5.8 Referral information

This tab contains all the referral information, which are synchronised from Salud. If the referral was from the Health Service Executive, a second drop down list appears to specify the exact geographical region of the HSE. The referring dentists name is designed to recall names entered to facilitate the procedure.

Figure 2-5: Referral Information tab

2.5.9 Missing and Over-retained Teeth

In this tab all the over-retained and developmentally missing teeth are recorded. If the small box beside over-retained or missing teeth is ticked, the four dental quadrants for both the primary and the permanent dentitions appear to facilitate selection of the appropriate over-retained and/or missing teeth. For ease of use the information on over-retained and missing teeth is included on one tab. Entering the information this way facilitates pattern identification, as detailed later in this chapter.

Figure 2-6: Missing and Over-Retained tab

2.5.10 Hyperdontic teeth

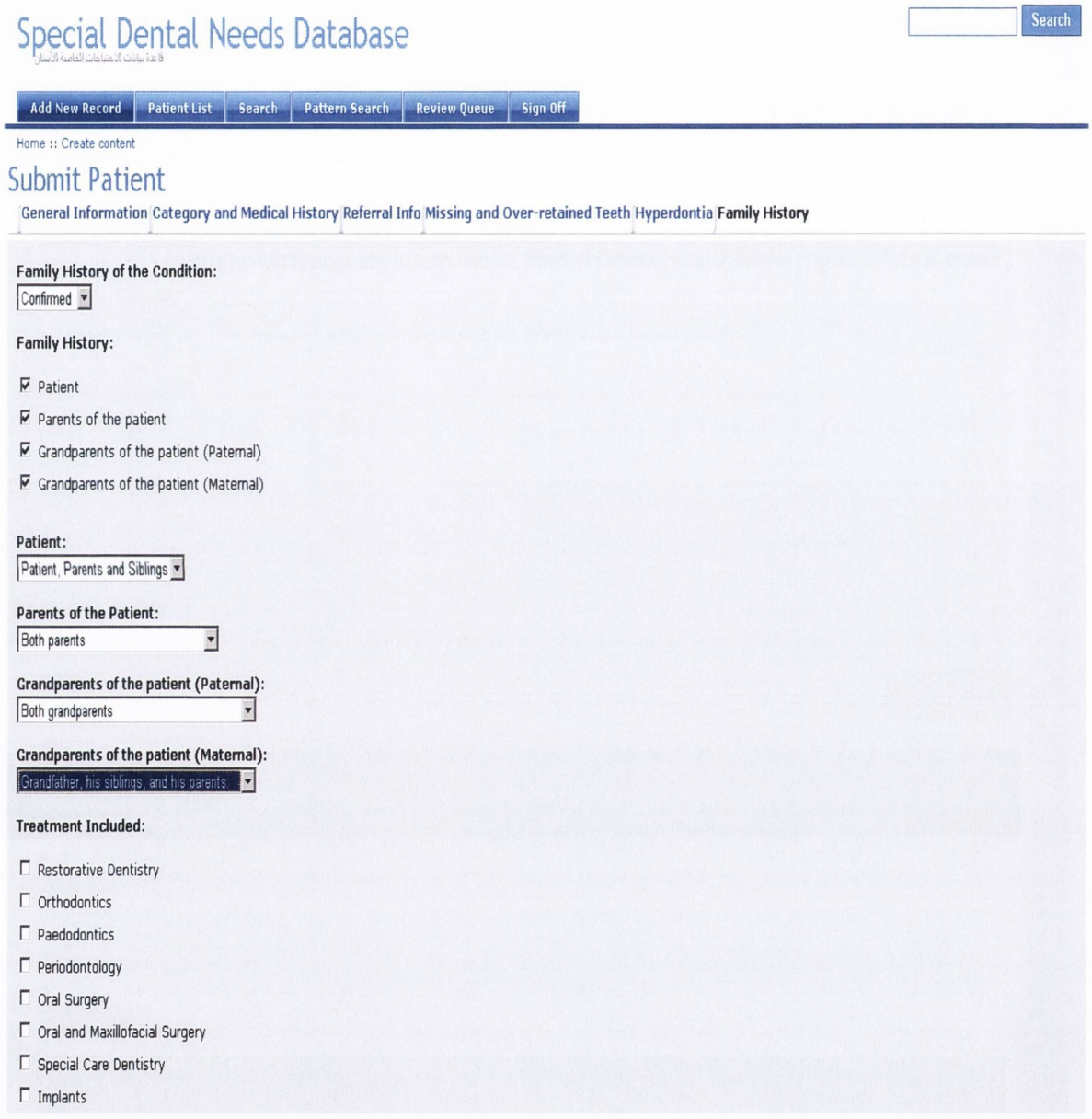
This tab contains the dental information of patients with hyperdontia. This is usually associated with rare medical syndromes such as cleidocranial dysostosis. This was designed to be in a separate tab to avoid confusion with over-retained or missing teeth.

Figure 2-7: Hyperdontic teeth tab

2.5.11 Family history of the condition

The family history tab contains the hereditary information of the developmental dental condition if present. It shows the inheritance pattern for three generations with drop down lists to investigate the pedigree of the condition.

Figure 2-8: Family history tab

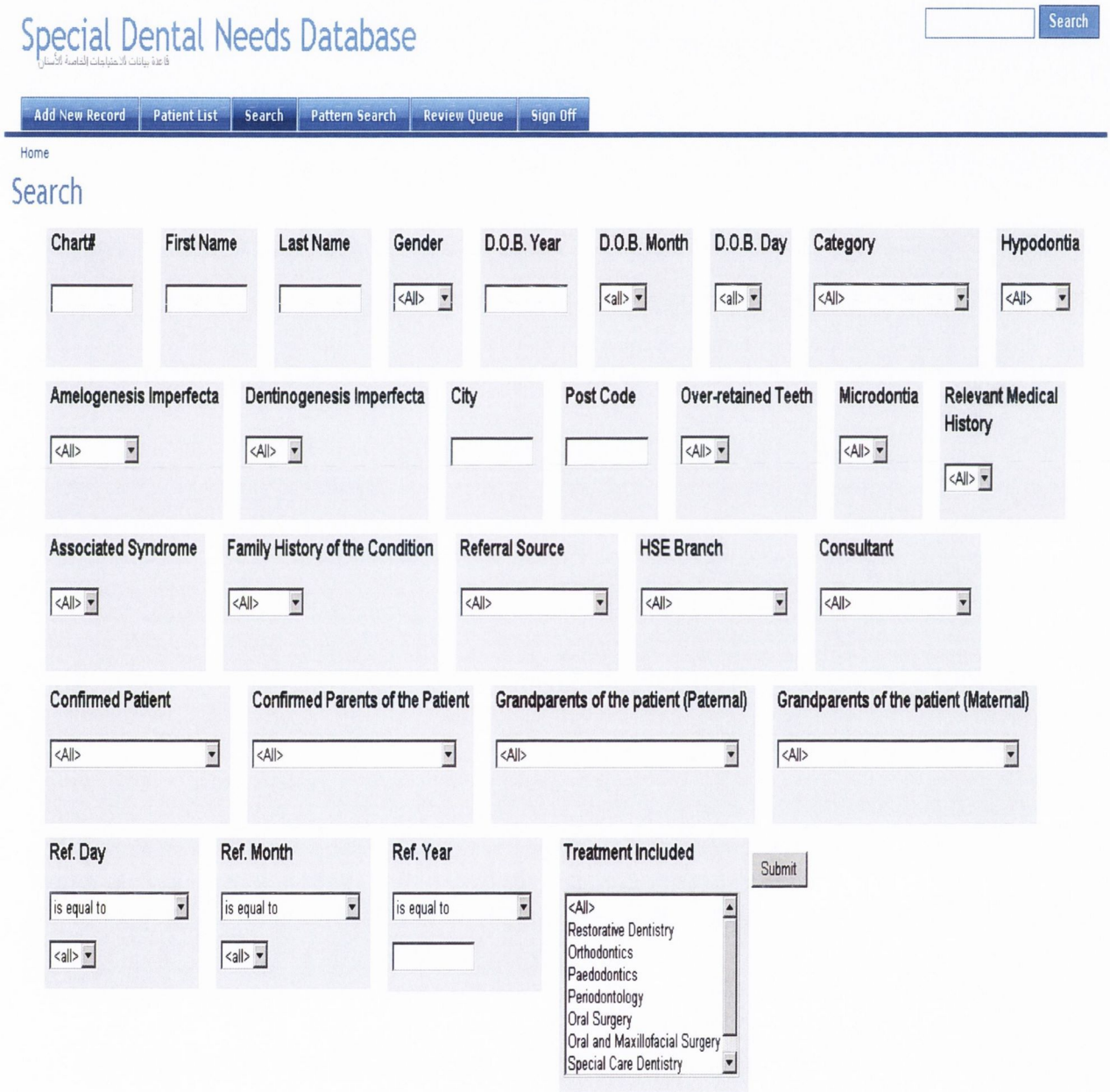


2.5.12 Database Search Engine

2.5.12.1 General Search

The General "Search" tab has 29 filters, which can either be searched individually or collectively. The search engine of the general search tab is so flexible that all 29 filters can be searched simultaneously.

Figure 2-9: General search tab



2.5.12.2 Pattern search

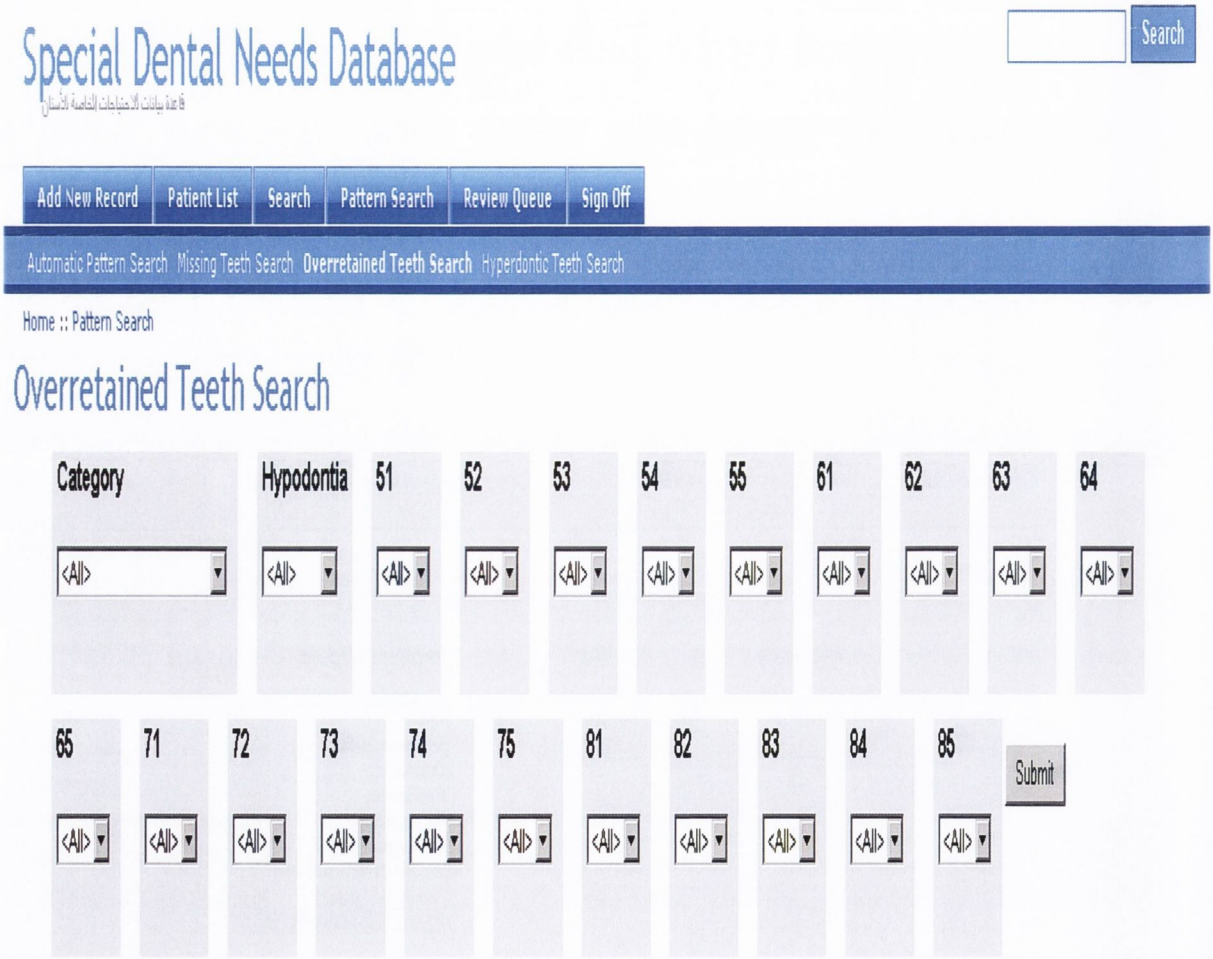
Over-retained teeth

The Over-Retained "Search" tab allows the search for patterns of over-retained teeth. Every primary tooth can have one of three options (All, 0, and 1).

- "All": means that the search engine ignores the tooth so that it can be present or absent.
- "0": the tooth is not over-retained.
- "1": the tooth is over-retained.

All 22 filters shown here can be searched simultaneously or individually in order to identify patterns of over-retained teeth.

Figure 2-10: Over-retained teeth pattern search tab



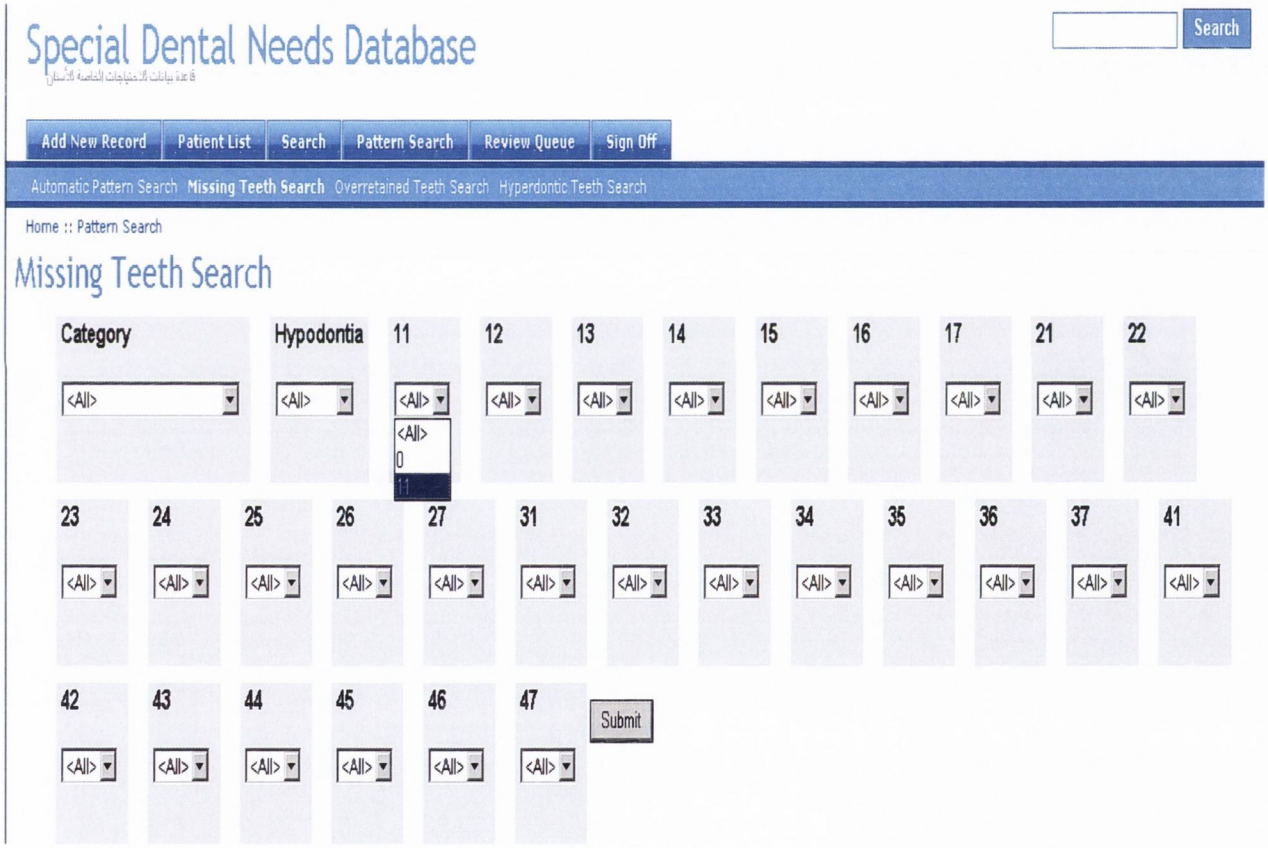
Missing teeth

The Missing Teeth "Search" tab allows searching for a clinical pattern of missing teeth. Every permanent tooth can have one of three options (All, 0, and 1).

- "All": means that the search engine ignores the tooth so that it can be present or absent.
- "0": the tooth is present.
- "1": the tooth is missing.

All 30 filters shown here can be searched simultaneously or individually in order to identify clinical patterns of missing teeth in any combinations.

Figure 2-11: Missing teeth pattern search



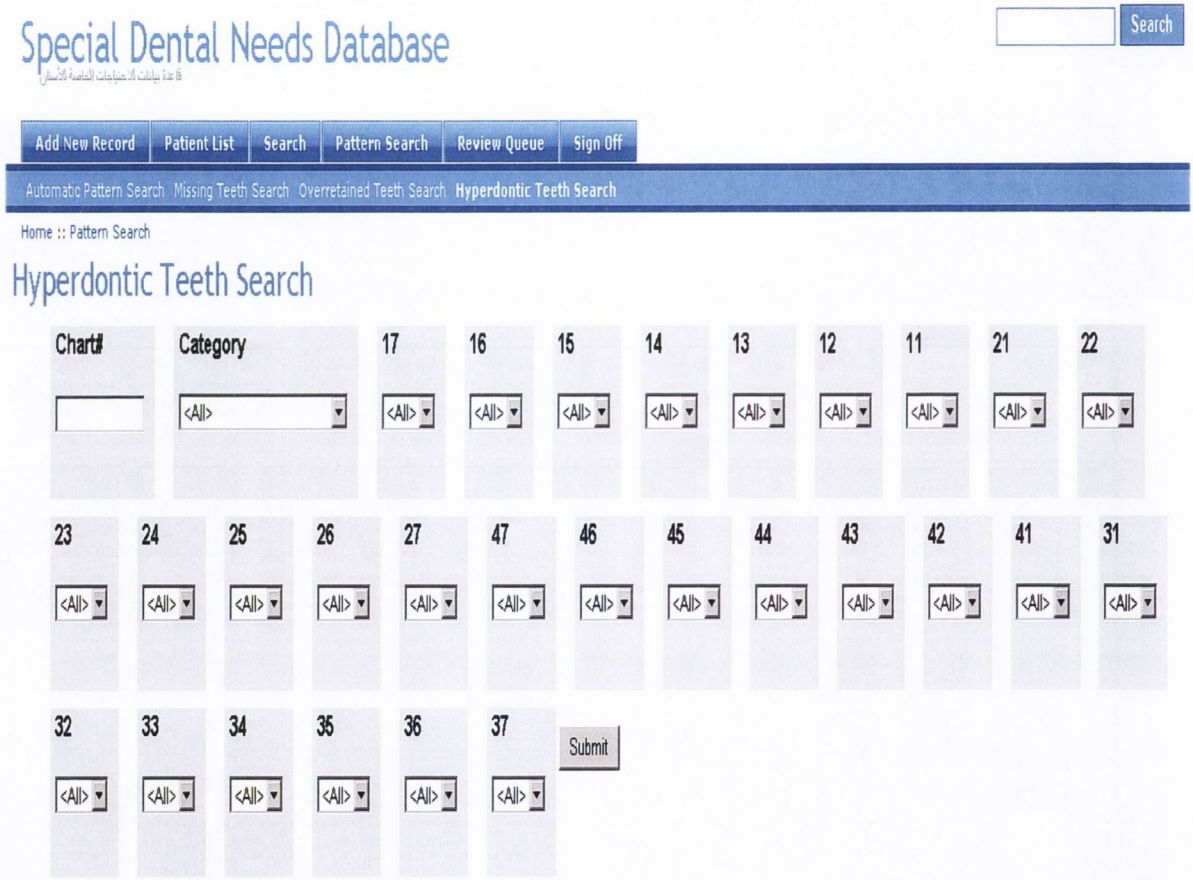
Hyperdontic teeth

The Hyperdontic Teeth "Search" tab allows the search for a clinical pattern of hyperdontic teeth. Every permanent hyperdontic tooth can have one of three options (All, 0, and 1).

- "All": means that the search engine ignores the tooth so that it can be present or absent.
- "0": the tooth is not hyperdontic.
- "1": the tooth is hyperdontic.

All 31 filters shown here can be searched simultaneously or individually in order to identify clinical patterns of hyperdontic teeth.

Figure 2-12: Hyperdontic teeth pattern search



2.5.13 Automatic pattern detection

The Automatic Pattern Detection search engine identifies similar clinical patterns and reports them by grouping patients with similar clinical presentation.

Anecdotally, some patterns of tooth agenesis appear to be more frequent than others. While most hypodontia research is directed toward mapping patients by their genotype and then identifying their clinical presentation (phenotype) (Brook, 1984; Svinhufvud et al., 1988; Stimson et al., 1997; Liu et al., 1998); there is a current research trend to group patients by their phenotype in order to facilitate better understanding and clinical management (van Wijk and Tan, 2006; Creton et al., 2007). With 32 possible teeth, the number of different patterns equals 4,294,967,296 (2^{32}), or more than 4 billion combinations, which would be inefficient to work with (van Wijk, 2010). During the database development process an automatic pattern detection search tool, which groups patients by the pattern of missing teeth, was incorporated into the SPDNS database. It automatically identified patterns of tooth agenesis and groups patients accordingly.

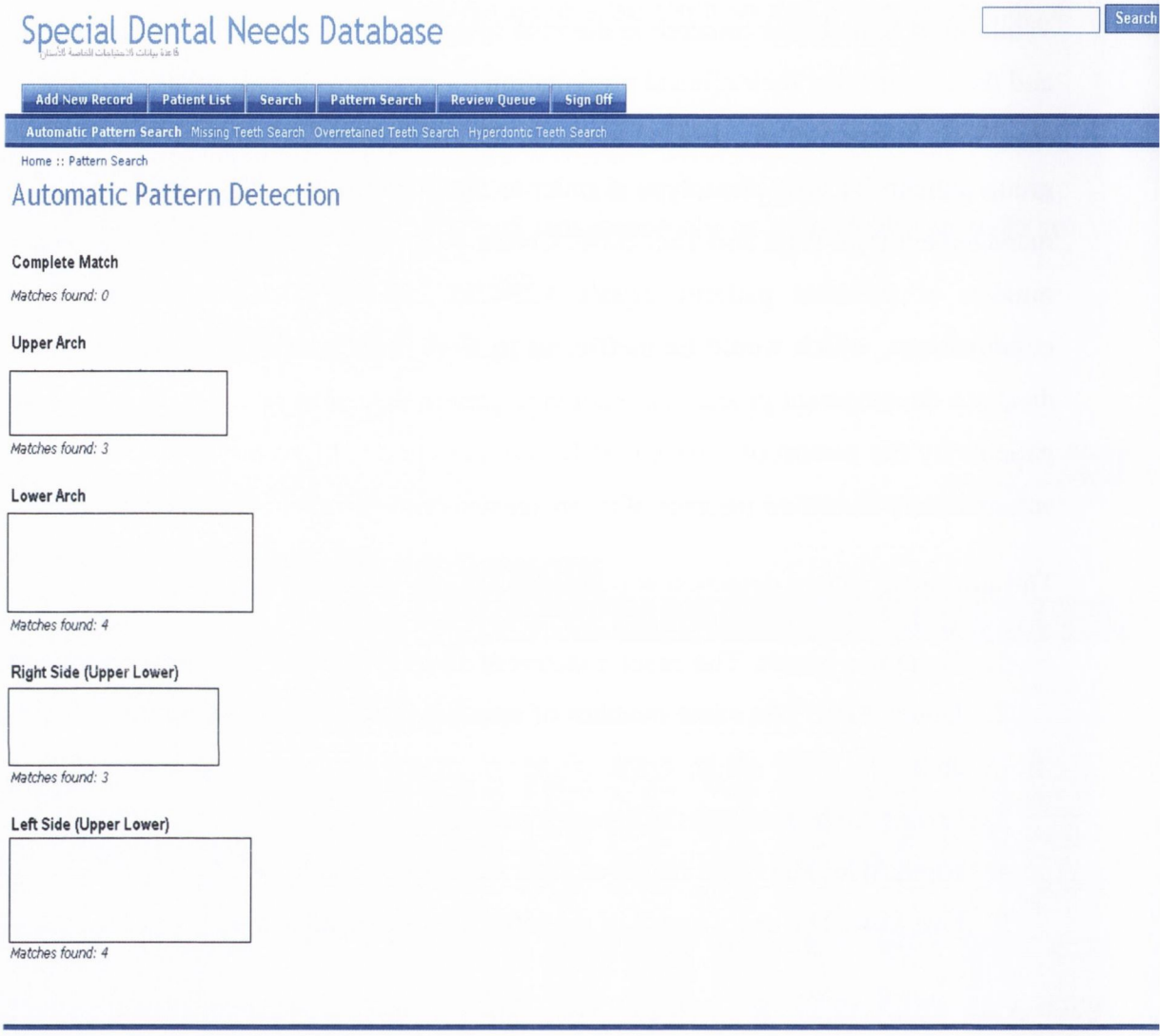
The automatic pattern detection was divided to:

1. Complete Match: The exact matches of all missing teeth were grouped together.
2. Upper Arch: The exact matches of maxillary arch only ignoring the mandibular arch.
3. Lower Arch: The exact matches of mandibular arch.
4. Right Side: The exact match of right side (upper and lower).
5. Left Side: The exact match of the left side (upper and lower).

When an exact tooth agenesis pattern is identified i.e. (<u>432/234</u>) the number of patients with the specific pattern can be increased by broadening the search to include the specific pattern (<u>432/234</u>) in addition to other teeth. The status of other teeth could be present, absent or ignored. This is achieved by using the Missing Teeth Pattern Search engine described earlier.

In this figure patients' names were concealed and the number of matches are shown below each match group.

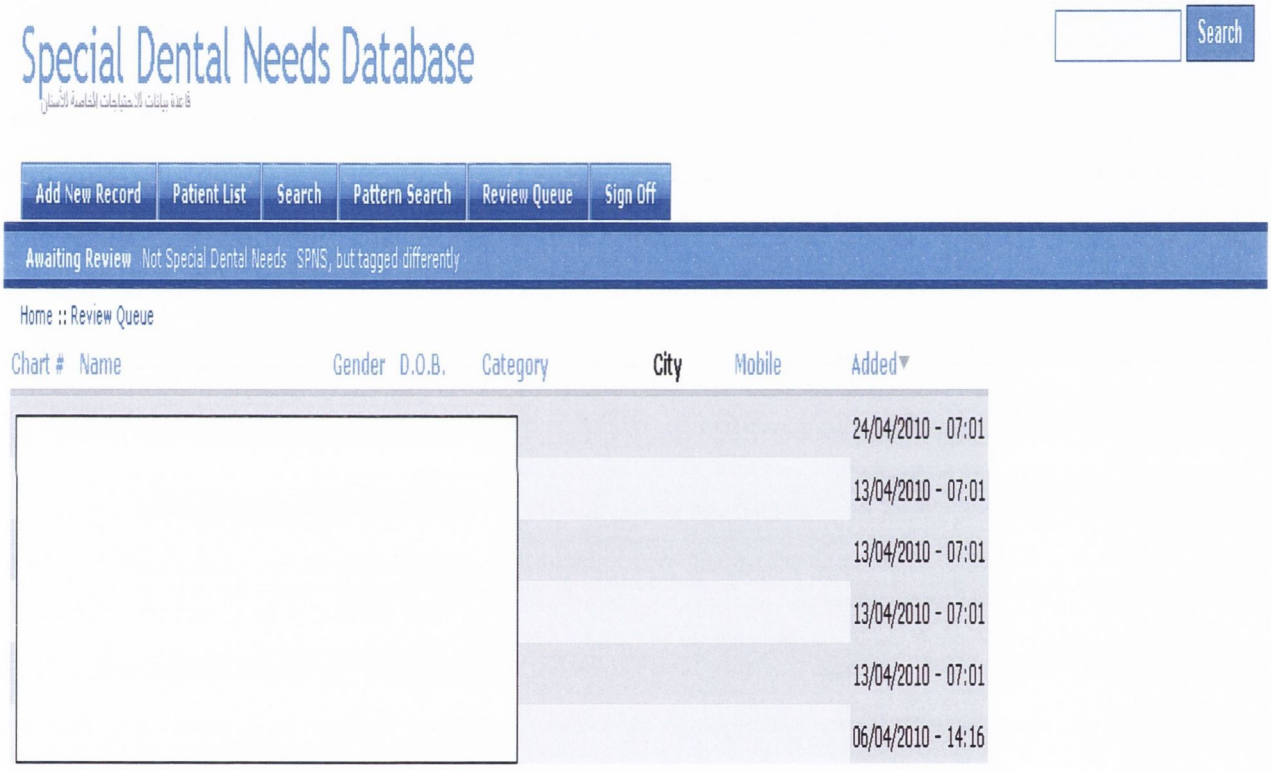
Figure 2-13: Automatic pattern detection



2.5.14 Awaiting review

The Awaiting Review tab shows the details of any new patients who are tagged as SPNS patients by Central Medical Records Staff. These patients are synchronised from Salud and placed in this tab for chart review by the database coordinator. This facilitates approval or rejection of patients according to preset clinical criteria and in consultation with the consultant involved. Patient details are concealed in this figure to ensure privacy.

Figure 2-14: Awaiting Review tab



2.5.15 Implementation problems and solutions

During SPNSDB construction, the following technical problems were encountered:

2.5.15.1 Limited number of fields that could be successfully displayed and filtered simultaneously

To display and filter information, Drupal uses a module called "Views". Views allow information to be presented in various formats, such as tables, lists and titles. It also has a mechanism to allow displayed information to be filtered. Filters can be either hidden or shown to the user. The exposed filters allow the users to customise views and search for records with particular attributes. This mechanism was used to implement SPNDB search functionality, where a user could input desired search parameters in exposed filters with appropriate views to search for specific records.

During implementation, it was discovered that there was a limit on the number of fields which could be simultaneously displayed and filtered by the Views module. This limit was sixty-three fields. This was due to Views using SQL's "LEFT JOIN" command in order to filter records and display only those that match filter requirements. Current MySQL implementation of 'LEFT JOIN' allows only 63 tables to join simultaneously.

There were three possible solutions to this problem:

- Switching from MySQL to other database, such as PostgreSQL:
  Switching to another database could have potentially solved the "LEFT JOIN" problem. PostgreSQL is an excellent, solid and robust DB platform, which is also free and open source, similar to MySQL. However, such a database switch would require all tables to be recreated in a PostgreSQL database. Simple merging of the data was not possible so moving platform would equate to re-building the entire system.

- Rewriting the Drupal Views module to use logical operators for filtering instead of "LEFT JOIN":
  Rewriting the Views module to use resources more efficiently, if they are stored within the same table, could be a preferred solution and be more time efficient. Since Drupal is an open system, this would be possible. However, custom and modified solutions would lead to problems with technical support in the future,

as any updates to the main Views module would require re-application of this custom code. Updates are common practices to address identified system problems or security risks. This would substantially add to the cost of maintaining the system. Local support availability could also not be assured.

- Reducing the amount of data displayed and filtered by splitting it into smaller groups, which would require less than 63 fields displayed and filtered simultaneously:

  Reduction of displayed and filtered fields would not require time-consuming source code modification nor database migration. In addition, it would solve the problem of screen cluttering, where too much information was presented simultaneously. This solution was preferred, which led to search facility being split into "General Search" and "Pattern Search". This was the preferred option for further development, since it allowed introduction of a "Pattern Search" into the following subgroups: "Missing Teeth", "Overretained Teeth", and "Hyperdontic Teeth". The latter search category, "Hyperdontic Teeth", was introduced at a much later stage of database development.

2.5.15.4 Internet Explorer 6 and Cascading Style Sheet (CSS) display formatting:

All web browsers use Hyper Text Markup Language (HTML) to deliver information from websites and interpret its appearance. To ensure that all browsers display information in a similar manner, a World Wide Web Consortium (W3C) was formed in 1994 in Massachusetts Institute of Technology (Birds-Eye.Net, 2005). W3C publishes various standards which are used on the Internet, and browser makers follow these standards to make sure that their browsers display most of the sites on the Internet correctly.

However not all browser makers follow these standards. Some developers:

1. Take "shortcuts" while implementing W3C standards.
2. Feel that their market share is sufficiently high to ignore them and implement standards in their own way.
3. Leave implementation of certain features for later versions of their product, thus intentionally creating incompatibilities with appropriate

web standards (Santos, 2009).

As a result, most of the websites and web-based systems designed today have to undergo extensive testing procedures, where their functionality and presentation is tested in various popular browsers to ensure that standards are upheld and webpages appear as intended.

One of the most problematic browsers that constantly creates problems for web developers is Microsoft Internet Explorer® version 6. Despite the fact that its initial release was in May 2001 and Microsoft® publicly admitting that the browser is "bad" (Microsoft, 2010), it is still in use (mainly in business environments) and at the present time it holds about 7% of the browser market share according to W3C Schools (http://www.w3schools.com/browsers/browsers_stats.asp).

In the case of SPNSDB, these problems were addressed in two ways:

- A Zen Classic theme was used as a formatting base. It is a polished and robust theme for Drupal Content Management System (CMS) that has been proven with various browsers, ensuring that the appearance is consistent. This theme checks and identifies browser versions and installs appropriate fixes into the formatting stream in order to compensate for shortcomings of particular browsers.
- Since SPNSDB introduced a number of fields that required a specific display format, such as dental quadrants, these modifications were added to Zen's Cascading Style Sheets (CSS) code. All of these modifications were tested to ensure that pages were displayed correctly in Microsoft Internet Explorer® Versions 6,7,8; Mozilla Firefox® 2, 3; as well as Google Chrome® and Apple Safari® browsers.

2.5.16 Database Development Methodology

There are various different methodologies to software development. A typical methodology is called "waterfall", where customers and developers construct a set of requirements, agree upon them, and then developers implement the product, test it and present it to customers (Whitgift, 1991). The drawbacks of this approach include lack of interactivity and the inability to dynamically adapt requirements as development takes place.

Initial SPNSDB development started in the "waterfall" mode, but quickly moved into more progressive "eXtreme Programming" (XP) form. Extreme programming form is characterized by the presence of customers during the development process and allows dynamically changing the requirements as the implementation progresses (Beck, 1999). The customer can immediately identify how certain parts of the system are going to appear. It can also quickly address any potential questions that arise from incomplete original requirement specifications. Such direct customer involvement has been shown to lead to much better requirement understanding by developers, and introduction of additional requirements by customer (Beck, 1999). As a result, the final product is much more usable than the one which would have been produced by the more traditional "waterfall" development methodology. As an example, the late addition of fields with features such as "Hyperdontic teeth", as well as integration of patient details with the main hospital database "Salud" came as a result of direct and frequent team engagement.

2.6 Conclusion

This chapter described the development and establishment of an Irish SPNSDB for patients with hereditary developmental dental disorders such as hypodontia, amelogenesis imperfecta, and dentinogenesis imperfecta. The database was developed in collaboration with all disciplines involved in providing patient care in the Special Dental Needs Clinic established in the Dublin Dental University Hospital. This database is the first of its type in the Republic of Ireland and aims to store all relevant clinical and biographical information for this patient population in order to identify the prevalence and geographical distribution of these developmental dental disorders. The information can also be used to help allocate services and outline the resources needed to optimise the management of patients with these conditions. The database allows for a better understanding and facilitates future research to further investigate these developmental dental disorders in an Irish population.

Chapter 3

Database Analysis for Hypodontia Patients

Chapter 3: Database Analysis for Hypodontia Patients

3.1 Introduction

Hypodontia is the term used to describe the developmental absence of one or more primary or secondary teeth, excluding the third molars. It is the most common developmental dental anomaly and can be challenging to manage clinically (Larmour et al., 2005).

Prior to 2001 the treatment of patients with developmental dental disorders in Ireland depended on the region where the patient was resident and was not coordinated.

In December 2001 a Special Dental Needs Restorative Dentistry Clinic was established and funded by the Department of Health and Children to treat this patient population.

The purpose of this study was to identify the prevalence, frequency, severity, and geographic distribution of hypodontia cases referred to the tertiary care centre where an interdisciplinary approach for the management of such cases was available.

3.2 Materials and Methods

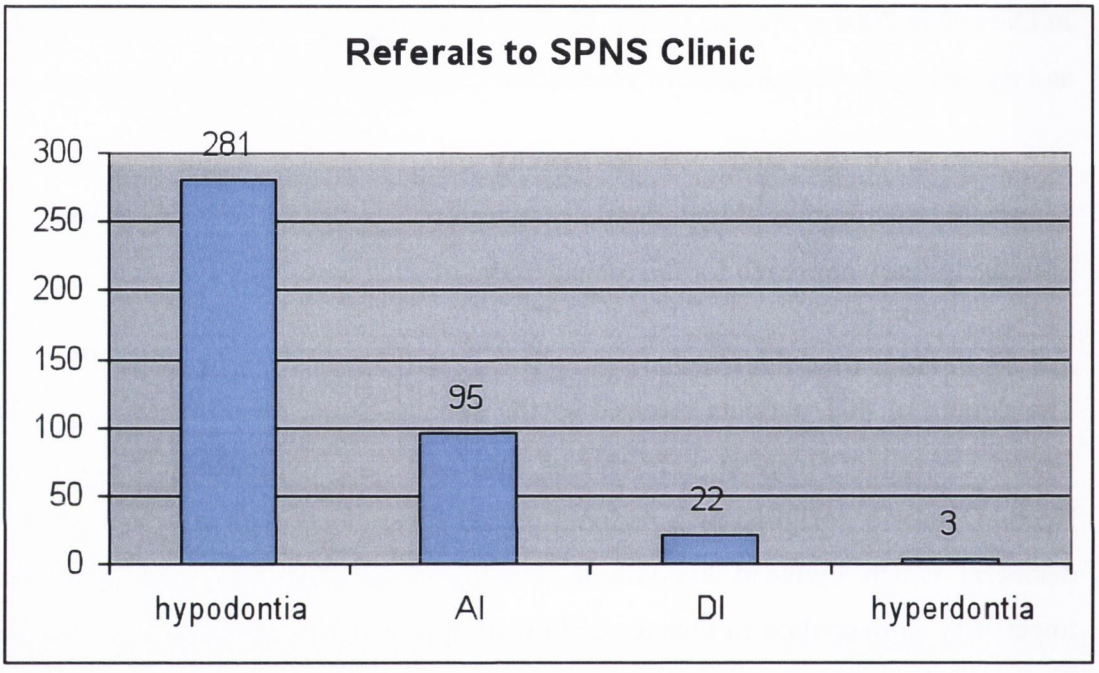
The details of 401 patients referred to the special dental needs clinic in the Dublin Dental University Hospital were entered into web-based purpose built Special Dental Needs Database designed as a national record for patients with developmental dental disorders which included hypodontia, amelogenesis imperfecta, and dentinogenesis imperfecta as described in chapter 2. Ethical approval was obtained from the Faculty Research Committee in Trinity College Dublin (See Appendix) and informed consent obtained from each patient or parent in the case of minors. Teeth charting was completed using both clinical examination records and orthopantomographs. A single investigator entered all data.

3.3 Results

3.3.1 Referred Cases

Of all patients referred to the clinic, hypodontia constituted 70% of referrals, amelogenesis imperfecta represented 23.7% and dentinogenesis imperfecta represented 5.5%, whereas, hyperdontia constituted only 0.75% of the total referrals (Figure 3-1). This chapter will only describe the hypodontia population.

Figure 3-1: Referrals type to the SPNS clinic



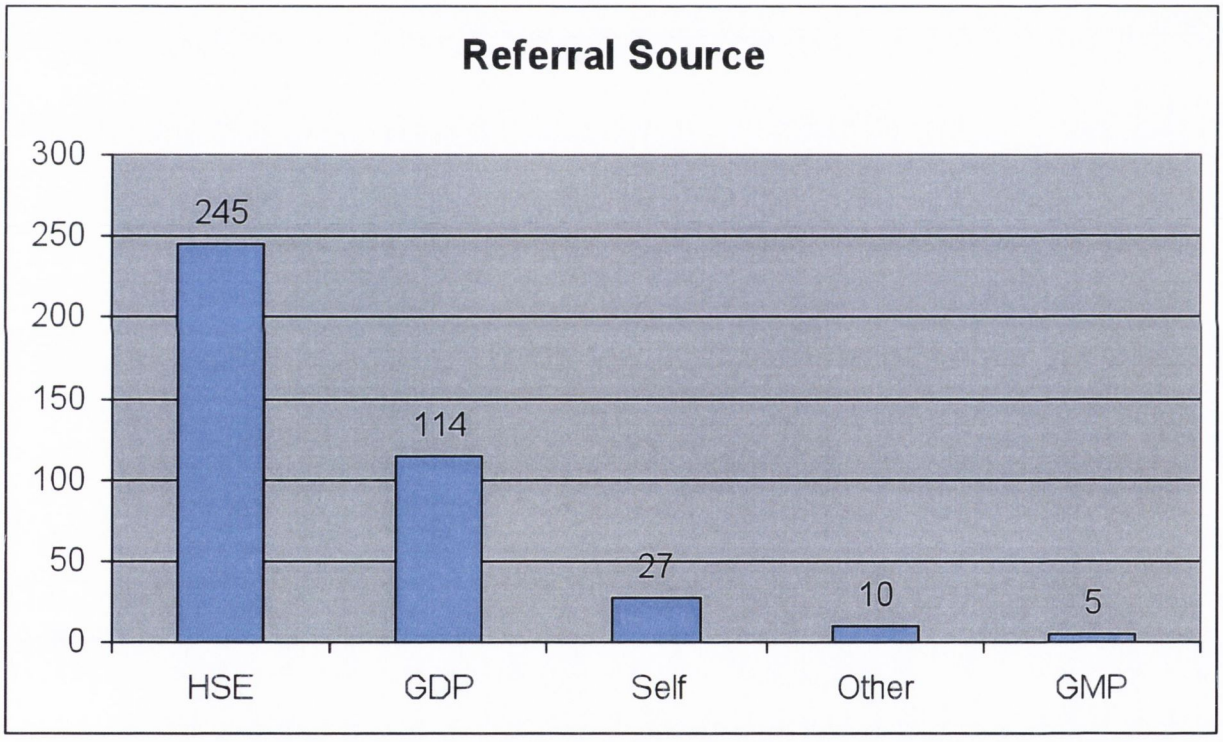
3.3.2 Gender Distribution

Females were referred to the SPNS clinic more frequently than males with a ratio of approximately (1.2: 1).

3.3.3 Source of Referrals

Most referrals were from the Dublin North East and Dublin Mid-Leinster areas, with fewer referrals coming from the west and south of the country. The majority of referrals were from the Health Service Executive (Previously know as Health Board Dental Service), followed by referrals from general dental practitioners (Figure 3-2). Very few cases had been self referred or referred by a general medical practitioner.

Figure 3-2: Referral source for all SPNS patients



3.3.4 Pattern of Tooth Agenesis

The mandibular second premolars showed the highest frequency of tooth agenesis representing 15.6% of the total number of missing teeth, followed by the maxillary second premolars (14.8%) and lateral incisors (13.9%). The distribution of tooth agenesis is shown in Figure 3-3. The total number of missing teeth for all patients was 2240 (Mild = 76, Moderate = 517, Severe 1647), with a range of 1-26. Symmetry of tooth agenesis of the right and left sides was a feature in individual patients and in the whole population with 1123 teeth missing on the right side and 1117 on the left side. More teeth were absent in the maxillary arch (n=1211) as compared to the mandibular arch (n=1029).

When moderate and severe hypodontia cases were analysed as two separate groups it was noteworthy to find that the same pattern of tooth agenesis was maintained but symmetry was less evident. (Figures 3-4 and 3-5).

Figure 3-3: Pattern of tooth agenesis of combined moderate and severe hypodontia patients.

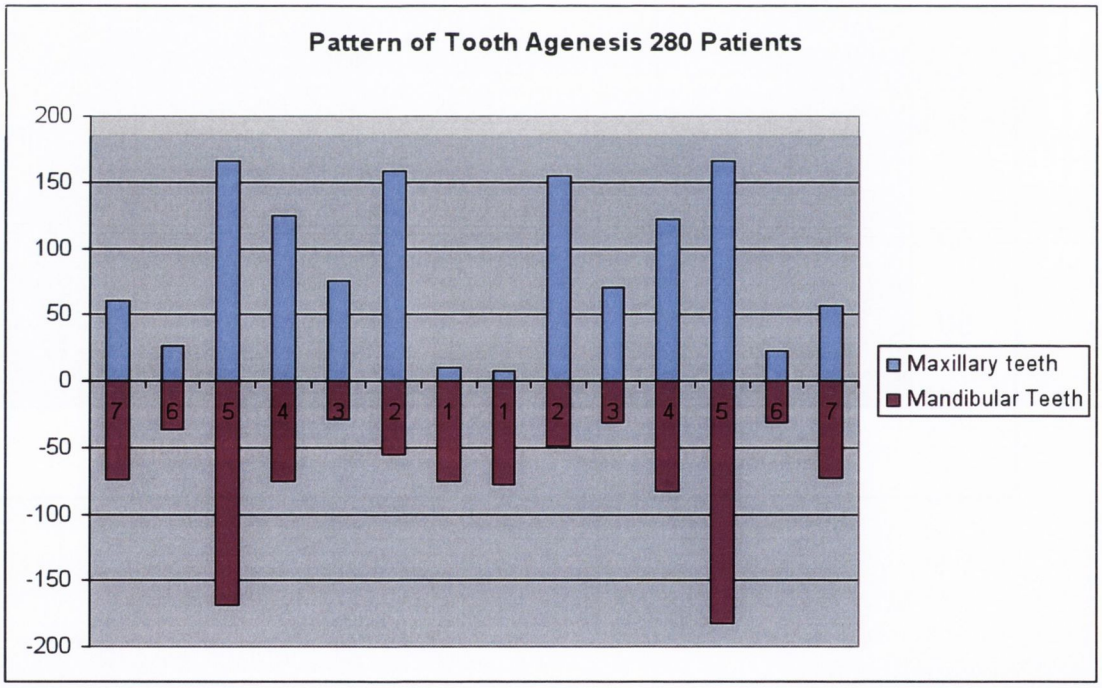


Figure 3-4: Pattern of tooth agenesis of moderate hypodontia patients (N=106 patients).

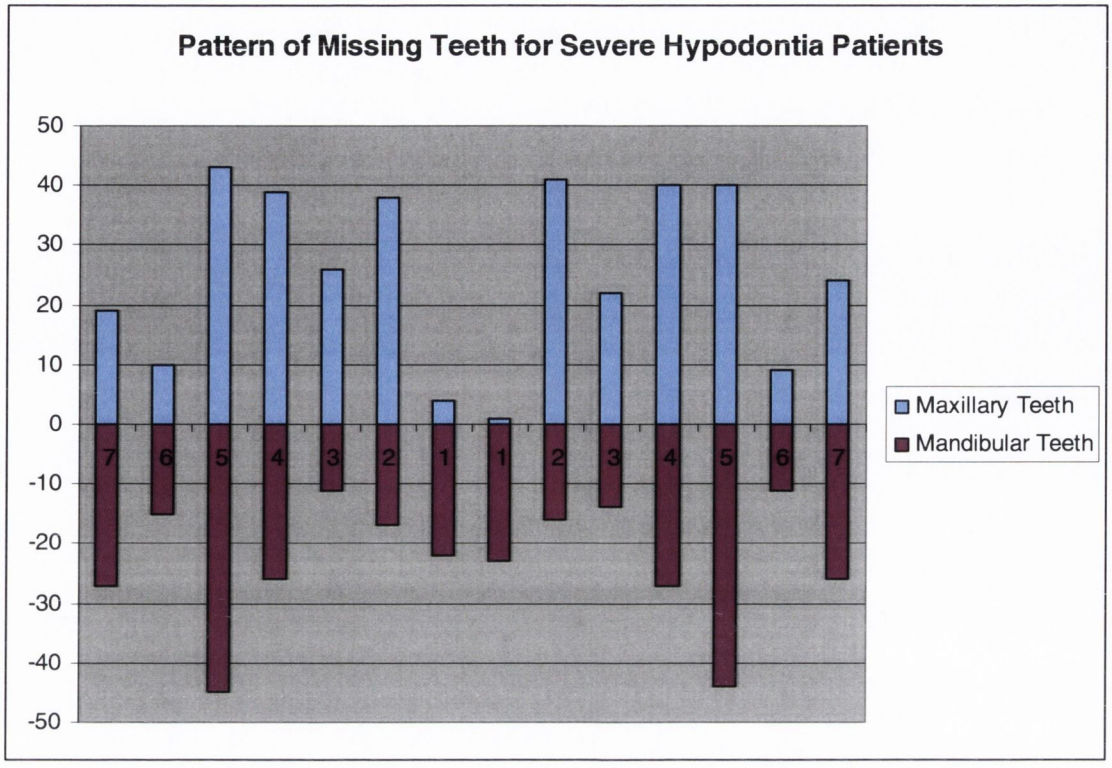
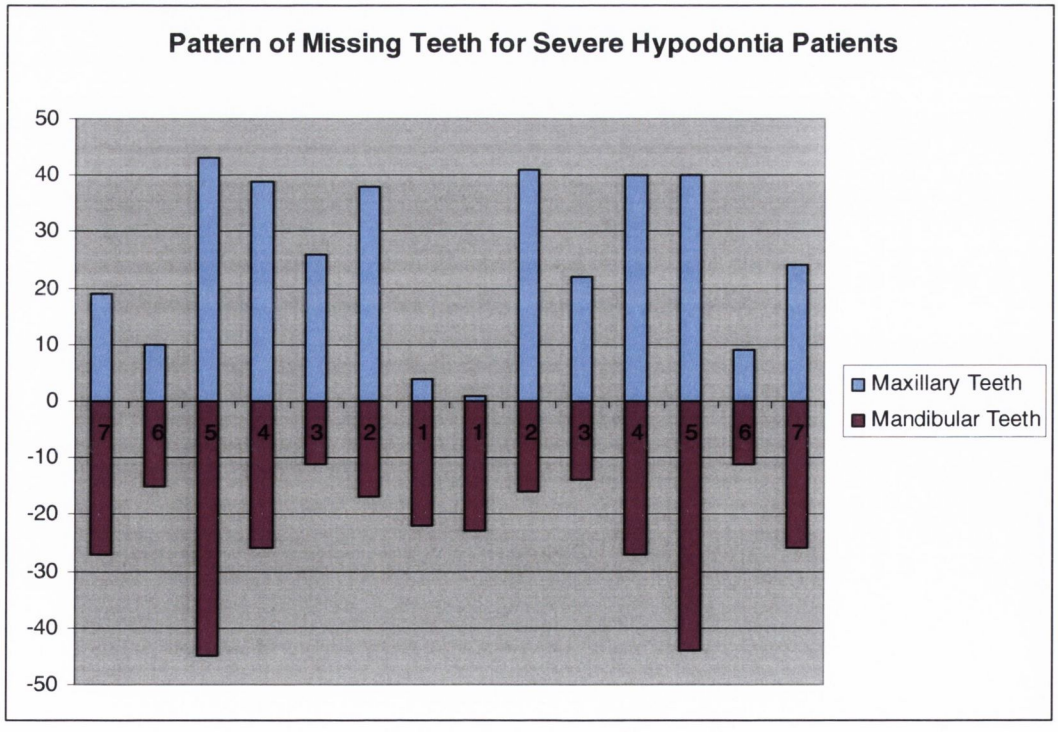
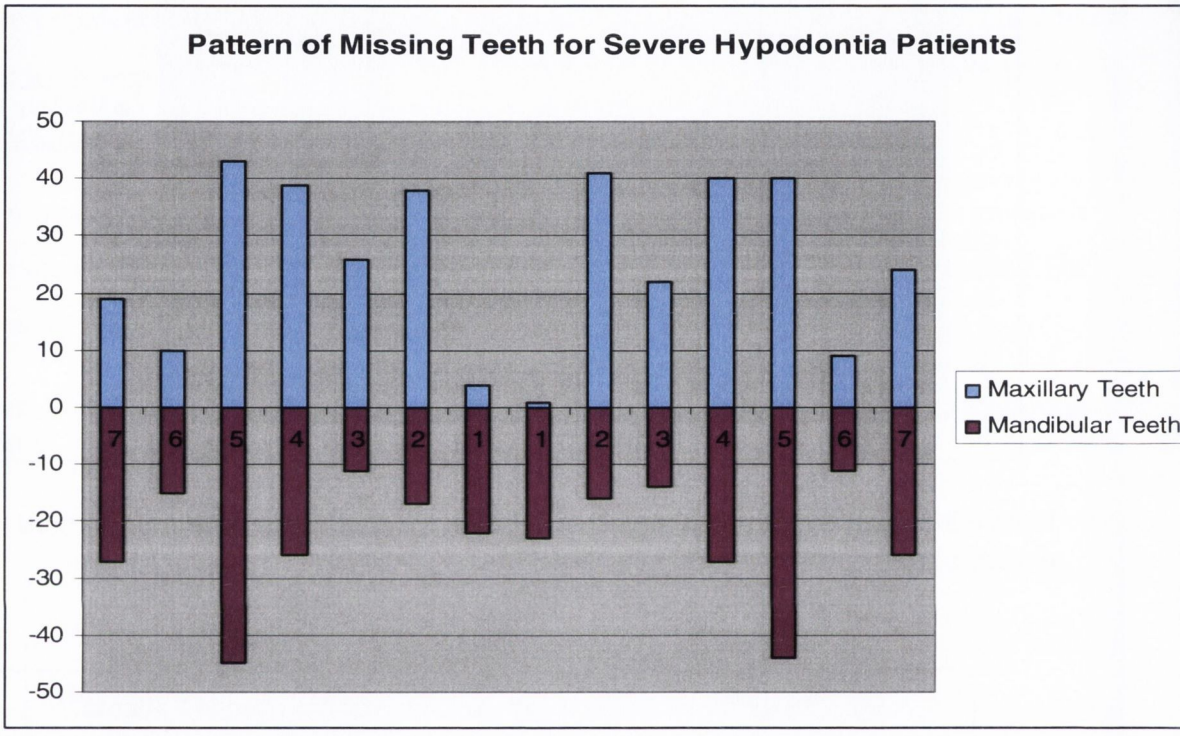


Figure 3-5: Pattern of tooth agenesis of severe hypodontia patients (N= 138 patients).



The most common pattern of tooth agenesis per patient was six missing teeth followed by four missing teeth per patient as shown in Figure 3-6. One hundred and thirty eight patients (49.2%) had severe hypodontia where more than six teeth were developmentally absent, and 106 patients (37.8%) had moderate hypodontia where four to six teeth were developmentally absent.

Figure 3-6: Number of Missing Teeth Per Patient



3.4 Discussion

Hypodontia was the most common developmental dental disorder referred to the Special Dental Needs Clinic since its inception in 2001. By definition this patient population is biased and does not reflect the prevalence and severity of hypodontia for the Irish population. The results should be interpreted bearing this limitation in mind.

The majority of referrals came from the Health Service Executive and were from the greater Dublin area. This may reflect the density of the population in this area as 50.6% of the population live in the Dublin Mid-Leinster and Dublin North East areas, whereas 25.5% reside in the south and 24% reside in the west of the Country (CSOI, 2006).

The age range observed was 7-50 years, with a median age of 19 years old. The age range was greater than that reported in the studies in Table 1-1. The difference was related to the nature of the population under investigation. Some studies investigated school children; whereas other studies investigated orthodontic patients. Only one study examined recruits of the Australian defense forces (Lynham, 1990). None of the previous studies reported the age median. In this project most patients were adults attending the Special Dental Needs clinic for advanced restorative care.

In order to exclude other causes of tooth loss for older patients, clinical notes and referral letters were carefully examined, and compared with previous available radiographs. While recognition of younger patients with hypodontia results either from chance observation or a positive family history, it might be expected that the majority of cases would be identified in the mixed dentition phase. The median age of this patient population was 19 years old; this may suggest either poor recognition by dental practitioners, or low levels of patient demand. For these cases, it is likely that there were difficulties in locating an appropriate clinic to which a referral for advice or treatment would be made. A second possible reason is that the database was established by the Division of Restorative Dentistry and Periodontology which traditionally provides restorative care at the end of the treatment plan, as compared to the interdisciplinary approach which has been implemented more recently. Patients now are being added to the database as soon as their diagnosis is confirmed often on referral from paediatric dentistry, community and orthodontic clinics.

Another reason for late presentation may be the retention of primary teeth in this patient population. Many primary teeth can survive until later in life and as a result patients may not be concerned to attend a dentist until these teeth start to exfoliate. Quality of life research on hypodontia patients has shown that when over-retained primary teeth were accounted for, the impact of hypodontia on the quality of life of this patient population was reduced (Wong et al., 2006).

The frequency and distribution of tooth agenesis (as shown in Figures 3-3) is similar to that reported by other workers (Castaldi et al., 1966; Rose, 1966; Wisth et al., 1974a; Rolling, 1980; Hobkirk et al., 1994). Castaldi and co-workers investigated 457 school children in Canada between the ages of 6 and 9 years old for congenital anomalies of teeth. They found that hypodontia was the most common developmental dental anomaly at 4.1%, and that the mandibular second premolar was the most commonly developmentally missing tooth, followed by maxillary second premolars and maxillary lateral incisors. However, a slightly different findings was reported by Rose who surveyed 6000 consecutive orthodontic patients and found that 4.3% had hypodontia and that the mandibular second premolar was the most commonly absent tooth followed by the maxillary lateral incisor and maxillary second premolar. Hobkirk and co-workers who investigated 451 patients aged 4-28 years attending a hypodontia clinic also confirmed a similar hypodontia pattern to that reported by Rose. Wisth et al investigated 813 Norwegian children aged 7 years of age attending orthodontic department and found similar results with an incidence of hypodontia of 7.1%. Rolling, in Denmark, examined 3325 orthodontic patients aged 9-10 years, and found a comparable prevalence rate of 7.8%.

In this research however, a slight increase in the prevalence of missing maxillary second premolars as compared to maxillary lateral incisors was noticed; this may be attributed to the fact that the majority of these patients were referred because of moderate to severe hypodontia. Hence, mild hypodontia cases, where there were no additional features like microdontia, impactions, or failure of eruption, were not referred but treated locally. Our results were also in agreement with the results of a Swedish study (Bergendal et al., 1996). However that group reported an increased prevalence of mandibular central incisors aplasia and did not report any agenesis of maxillary central incisors.

An increased prevalence of developmentally missing mandibular central incisors was also reported in a study investigating the quality of life of Chinese patients affected by severe hypodontia (Wong et al., 2006). The maxillary lateral incisor was the most commonly missing tooth followed by the mandibular central incisors.

It is worth noticing that the same pattern of tooth agenesis is maintained when moderate and severe hypodontia cases where plotted separately as evident from Figures (3-4 and 3-5). Unfortunately no research that has discussed patterns of moderate and severe hypodontia separately is available in the literature and this makes comparison more difficult.

The most frequent number of missing teeth per individual patient was six, followed by four missing teeth per patient (as shown in Figure 3-6).

The special dental needs clinic is a specialist clinic and hence most of the cases referred had moderate to severe hypodontia with four or more teeth missing; some mild cases were also treated when additional features such as microdontia, impacted teeth, or primary failure of eruption, which may complicate the management of such mild cases, coexisted.

3.5 Conclusion

This chapter describes the profile of hypodontia patients attending a tertiary care clinic in the Dublin Dental School & Hospital. Hypodontia is the most common developmental disorder affecting teeth in this referred population. The range of missing teeth was 2-26 and the mandibular second premolar was the most frequently absent tooth type.

Chapter 4

Impact of Hypodontia and Amelogenesis Imperfecta on the Quality of Life and Self-Esteem for Adult patients

Chapter 4: Impact of Hypodontia and Amelogenesis imperfecta on Quality of Life and Self-Esteem of Adult Patients

4.1 Introduction

Research has emphasised the close interaction during odontogenesis between different components of the developing tooth germ (Thesleff, 2006). That report indicated that defects in amelogenesis were associated with developmental defects or abnormalities in other components of the developing tooth, such as taurodontism, agenesis of teeth, delayed eruption, and development of an anterior open bite.

Hypodontia is the developmental absence of one or multiple teeth. It is the most common developmental anomaly of the permanent dentition. It has been found that, when third molars are excluded, this condition affects between 5.3% and 7.9% of children (Magnusson, 1977; Rolling, 1980; Aasheim and Ogaard, 1993; Dhanrajani, 2002). For an Irish population, agenesis of mandibular second premolars, maxillary lateral incisors and maxillary second premolars were found to occur frequently in a population referred to a tertiary care centre (Hashem et al., 2010). The pattern of tooth absence varies and can be symmetrical or haphazard. Hypodontia is usually associated with other dental anomalies such as reduced size and structural malformation of teeth, late eruption, transposition and crowding (Dermaut et al., 1986; Vastardis, 2000; Arte et al., 2001). The management of hypodontia is complex and requires a multidisciplinary approach, which can involve paediatric dentistry, orthodontics, prosthodontics, surgical and restorative therapy (Carter et al., 2003; Jepson et al., 2003; Meechan et al., 2003; Nunn et al., 2003).

Amelogenesis imperfecta (AI) is defined as a "disorder in which there is incorrect development of the dental enamel owing to agenesis, hypoplasia, or hypocalcification of the enamel. It is marked by enamel that is very thin and friable and frequently stained in various shades of brown" (Dorland, 2000). It occurs in both primary and permanent dentition.

From the patient's perspective, AI may have severe consequences in the form of low aesthetic quality of the dentition, poor mechanical properties of the dental tissues, and extreme dental sensitivity of teeth. All of these features can result in reduced oral health-related quality of life. Furthermore, some forms of AI require extensive

treatment over an extended period of time and may therefore be an economic burden on patients.

Patients affected with AI nearly always require a interdisciplinary approach for the management of their condition, which is concentrated in the first two decades of life and requires ongoing maintenance therapy for life (Ayers et al., 2004).

No simple solution exists for the treatment of AI and the associated clinical problems. Different treatment modalities have been suggested for the management of AI which can range from composite resin build-ups of teeth to stainless steel crowns in early childhood, to onlays, ceramic or porcelain fused to metal crowns later in life. This is usually accompanied by periodontal treatment, endodontic therapy, crown lengthening surgery, orthodontics and in some cases orthognathic surgery.

A team approach is usually adopted to determine the most appropriate treatment for an individual patient. In a well structured and functioning team, there should be good communication, support, and each speciality should interact in a sequential manner (Yip and Smales, 2003; Ayers et al., 2004; Ozturk et al., 2004).

Patients with hypodontia and AI are confronted with a number of problems from the early days of the eruption of their primary and permanent teeth. These problems may include dental pain or sensitivity, and inability to chew food properly. Aesthetic can be a major issue for parents of the young children and becomes an issue for the younger patients as they themselves reach adolescence and young adult years (Hobkirk et al., 1994; Coffield et al., 2005).

An important issue that remains to be investigated further, with respect to hypodontia and AI, is their impact on well-being and quality of life. Increasing emphasis is being placed on patients' perception of the impact of oral disorders. This is particularly important with respect to conditions such as hypodontia and AI whose effects are both functional and psychosocial. Understanding the consequences of such conditions has the potential to contribute to their clinical management and evaluation of the benefits of different therapeutic approaches (Coffield et al., 2005; Wong et al., 2006; Locker et al., 2010; Akram et al., 2011; Meaney et al., 2011).

4.2 Purpose of The Study

To study the psychosocial impact and self-esteem of hypodontia and AI on adult patients referred to a tertiary care clinic in the Dublin Dental University Hospital compared to patients attending the hospital for routine dental care.

4.3 Inclusion Criteria

- Confirmed clinical and radiographical diagnosis of moderate (developmentally missing 4-6 teeth) to severe (developmentally missing more than six teeth) hypodontia or amelogenesis imperfecta.
- Current biographical information.
- Patient of 18 years old or greater.
- Absence of associated syndromes.
- Absence of other systemic health problems.

4.5 Exclusion Criteria

- The presence of associated syndromes.
- Inability of the patient to provide consent, due to medical or mental conditions.
- Patient unwilling to participate.
- For the control group, any patient with multiple missing teeth was excluded.

4.6 Null Hypothesis

The proposed null hypothesis was that no difference in quality of life and self esteem existed between patients with developmental dental disorders such as moderate to severe hypodontia or amelogenesis imperfecta compared to other dental patients attending the Dublin Dental University Hospital for routine dental care.

4.7 Materials and Methods

Adult patients with clinical and radiological diagnoses of moderate to severe hypodontia and amelogenesis imperfecta and matching (age and gender) control patients attending the Dublin Dental University Hospital for routine dental care were recruited. The research proposal was reviewed and approved by the Research Ethics Committee in Trinity College, Dublin (see Appendix).

Ninety-one hypodontia and thirty-three amelogenesis imperfecta patients were identified and recruited from a pool of patients referred to the Special Dental Needs

Clinic for consultations and/or treatment. Patients were also identified from reviews of consultant day schedules in the Department of Restorative Dentistry and Periodontology. Staff and students in the dental hospital were also contacted by email and informed about the research in order to identify suitable patients.

Control patients were identified primarily from the hospital's main patient database "Salud" as described in Chapter 2. Undergraduate and postgraduate students were recruited to identify suitable patients.

Three mailings, which contained information leaflets, consent forms, and prepaid envelopes were sent to all patients to give multiple opportunities for patients to respond. At least one attempt was made to contact participants by telephone informing them about the study and answer any questions or queries that arose.

Seventy-four patients diagnosed with moderate to severe hypodontia, and thirty patients diagnosed clinically and radiographically as having amelogenesis imperfecta agreed to participate in the study. These patients were then age and gender matched with a control group of patients attending the Dublin Dental University Hospital for routine dental care. The response rate from the control groups was initially very poor for the hypodontia cohort (13%) and for the AI population (23%), but recruitment continued until the required numbers were obtained. This involved contacting patients directly in the Hospital or by telephone, either by the researcher or the clinician (staff/student) treating the patient. Patients who declined to participate in the study were not contacted again. Only patients who did not respond were re-contacted to ensure that they had received the letters and determine their willingness or otherwise to participate. If the answer was negative these patients were not contacted again.

A written, informed consent to participate in the study was obtained from all participants before enrolling in the study. The same procedure was followed for the control group.

A postal package of questionnaires, information leaflet, instructions, and prepaid envelopes was sent to all potential participants. Participants were advised to complete all questions and return them in the prepaid envelope provided. If the questionnaires were returned due to an incorrect address and the correct address could not be

determined, or if questionnaires were incomplete or missing, the person was deleted from the study.

To ensure anonymity and to comply with the Ethics Committee guidelines, all questionnaires were given codes and returned to a research assistant to whom patient identity and underlying condition was unknown, as patients had been assured their identity would not be disclosed.

4.8 Questionnaires design

4.8.1 Oral Health Impact profile-49 items

The social impact of dental disease is measured using the Oral Health Impact Profile (OHIP), which was developed in Australia (Slade and Spencer, 1994). It consisted of 49 items (See appendix) that were weighted according to the opinion of 328 people from community groups, dental practitioners and students who became known as the judging panel. They represented a broad range of opinions and whilst it is possible that such a large group is unlikely to be able to reach a definite consensus, the OHIP-49 has performed well in tests of reliability and validity and OHIP-49 questionnaire has been translated to many languages (Slade and Spencer, 1994; Pires et al., 2006; Awad et al., 2008; John et al., 2008).

The OHIP-49 provides a comprehensive measure of self-reported dysfunction, discomfort and disability attributed to oral conditions (Slade and Spencer, 1994). It provides information about the "burden of illness" within populations and the potential of measuring effectiveness of health services in reducing the burden of illness. It is scored in 0 to 4 response format, with total scores ranging from 0 to 196. Higher scores indicate higher levels of dysfunction, discomfort, and disability attributable to oral conditions.

The questionnaire is subdivided into seven domains to measure:

1. Functional limitation (nine questions)
2. Physical pain (nine questions)
3. Psychological discomfort (five questions)
4. Physical disability (nine questions)
5. Psychological disability (six questions)

6. Social disability (five questions)
7. Handicap (six questions)

4.8.2 Rosenberg Self-Evaluation Scale

This scale measures an adult's global feelings of self-worth or self-acceptance. It consists of 10 items (See appendix 2). It is typically scored using a four point response format (strongly agree, agree, disagree, strongly disagree) designated with numerical values of (0, 1, 2, 3), total scores range from 0 to 30 with higher scores indicating higher self-esteem (Rosenberg, 1965b; Blascovich J and J., 1991). The 10 items that make up the Rosenberg Self-Esteem Scale (SES) were designed to optimise ease of administration, economy of time, unidimensionality (set of items forming an instrument and all measure just one trait) (Hattie, 1985), and validity. Self-Esteem Scale items require the respondent to report their feelings directly. The SES is constructed of five positively and five negatively phrased items composed exclusively of first person evaluative statements about the self (the five negatively scored items are reversed before the total score of the scale is calculated) (Rosenberg, 1965a).

These questionnaires have demonstrated good cross-sectional validity and test-retest reliability in clinical and general populations (Slade and Spencer, 1994; Tafarodi and Swann, 1995; Pires et al., 2006; Awad et al., 2008; John et al., 2008). However, in the case of Oral Health Impact Profile-49 (OHIP-49) the Minimal Important Difference (MID) has only recently been established and still requires validation (John et al., 2009). The MID is the minimum difference in scores on a questionnaire that patients perceive as important and would lead them to report an improvement or deterioration in health status over time or as a result of therapy. This allows "clinical" as well as the statistical significance of differences between groups to be appreciated. Validation for these instruments has been established, and the instruments have been used extensively in other studies in the dental literature (Slade, 1997; Berk et al., 2001; Locker and Allen, 2002; Ozturk et al., 2006; Locker and Allen, 2007; Awad et al., 2008; Nicodemo et al., 2008).

4.9 Results

Sixty-five hypodontia patients (87.8%) and twenty-eight AI patients (93%) of the targeted population returned the completed questionnaires. Of these, thirteen

hypodontia patients and one AI patient were eliminated because of incomplete, missing or inappropriately filled questionnaires The remaining 12.2% for hypodontia patients and 7% of AI patients who did not return the questionnaires dropped out of the study for a variety of reasons including inadequate time, unwilling to complete questionnaires, and moving home addresses. These patients were not contacted again as stipulated by the Ethics Committee guidelines.

To ensure that the impact of hypodontia on QoL of affected patients is not confounded by a treatment effect, further eleven patients were eliminated because they have had their treatment completed or nearly completed. None of AI patients have had their treatment completed.

A total of forty-one hypodontia (29 females and 12 males) and twenty-seven AI patients (19 females and 8 males) with their matching controls completed all questionnaires successfully. The age range was from 18-54 years for both groups. A blinded researcher to whom patient identity and clinical details were unknown scored all questionnaires and entered the scores into a Microsoft Excel® spread sheet (Microsoft Corp. USA), and the scores were prepared for statistical analysis.

As data were paired, a paired t-test was used to analyse data with a statistical significance level of ($P \leq 0.05$). Data analysis was carried out using Windows® JMP® statistical package (JMP Statistics and Graphics Guide, Release 7, Copyright© 2007, SAS Institute Inc., Cary NC, USA) (See appendix 3).

4.9.1 Results and data analysis of all hypodontia patients (moderate and severe) compared to their controls

4.9.1.1 Oral Health Impact Profile (OHIP-49)

Total Scores

The means and standard deviations for the test and control groups were 62 ± 39 and 31 ± 27.2 respectively. When total scores for the OHIP-49 were analysed a statistically significant difference between the test and control groups was found ($P = 0.003$) with patients affected by hypodontia scoring significantly higher than their controls.

Functional limitation

The means and standard deviations for the test and control groups were 13.2 ± 7.2 and 9.4 ± 5.4 respectively. When scores for the functional limitations subscale of the OHIP-49 were analysed a statistically significant difference was found between the test and control groups (P = 0.01).

Physical pain

The means and standard deviations for the test and control groups were 12.4 ± 7.9 and 8.6 ± 4.3 respectively. Physical pain was found to be significantly different between the test and control groups (P = 0.008).

Psychological discomfort

The means and standard deviations for the test and control groups were 10 ± 6.2 and 6.6 ± 4.8 respectively. Psychological discomfort was found to be statistically different between the test and control groups (P = 0.007).

Physical disability

The means and standard deviations for the test and control groups were 10.6 ± 8 and 4.9 ± 4.4 respectively. Physical disability was also found to be statistically different between the test and control groups (P = 0.0006).

Psychological disability

The means and standard deviations for the test and control groups were 8.4 ± 6.5 and 3.9 ± 4.6 respectively. Psychological disability was significantly different between the test and control groups (P = 0.002).

Social disability

The means and standard deviations for the test and control groups were 3.4 ± 4.1 and 1.2 ± 2.3 respectively. Social disability was found to be statistically significant between the test and control groups (P = 0.01).

Handicap

The means and standard deviations for the test and control groups were 3.5 ± 3.7 and 2 ± 3.23 respectively. When data of the handicap domain of the OHIP-49 were analysed no statistically significant differences were observed for this domain (P = 0.08).

For brevity reasons and to avoid repetition, a summary of all OHIP-49 data for all other groups is presented in a table format only for the remainder of this chapter.

Table 4-1: Summary of results of OHIP-49 questionnaire for all hypodontia patients.

This table summarises the quality of life of adult patients with moderate/severe hypodontia and their matching (age and gender) controls of patient attending for routine dental care. A (*) indicates significance ($P \leq 0.05$). N=82.

Domain	**Test Group (N=41)**				**Control group (N=41)**				**Paired t- test**
	Mean	**Standard deviation**	**Median**	**Interquartile range**	**Mean**	**Standard deviation**	**Median**	**Interquartile range**	
Total scores	62	39	54	30-93	31	27.2	24	10.50-47.50	0.003*
Functional limitation	13.2	7.2	12	7.50-16.50	9.4	5.4	10	5-12.50	0.01*
Physical pain	12.4	7.9	11	6-18	8.6	4.3	8	5-12.5	0.008*
Psychological discomfort	10	6.2	9	5.5-16.5	6.6	4.8	6	2.5-10	0.007*
Physical disability	10.6	8.0	10	5-14.5	4.9	4.4	4	2-7	0.0006*
Psychological disability	8.4	6.5	8	2-13	3.9	4.6	2	0-6.5	0.002*
Social disability	3.4	4.1	1	0-8.5	1.2	2.3	0	0-1	0.01*
Handicap	3.5	3.7	2	0-6	2	3.23	1	0-3	0.08

4.9.1.2 Impact of Hypodontia on self-esteem

The means and standard deviations for the test and control groups were 22 ± 4.3 and 22.3 ± 5.5 respectively. When the total scores for Rosenberg Scale were analysed using a paired t-test, no statistically significant differences were found between the two groups (P = 0.98).

4.9.2 Results and data analysis for severe hypodontia patients compared to controls:

It was observed from the raw data that some individuals scored much higher within the hypodontia group. In order to identify if any pattern existed, the hypodontia group were separated into moderate (4-6 teeth missing) and severe (greater than 6 teeth missing) and data were subjected to the same analysis.

Data for a total of twenty-nine patients with severe hypodontia with their age and gender matched controls were analysed, of which twenty were females and nine were males. The age range was from 19 to 52 years old.

As data were paired, a paired t-test was used to analyse data with a statistical significance level of ($t \leq 0.05$). Data analysis was carried out using JMP® statistical package (JMP Statistics and Graphics Guide, Release 7, Copyright© 2007, SAS Institute Inc., Cary NC, USA) (See appendix 3).

Table 4-2: Summary of results of OHIP-49 questionnaire for severe hypodontia patients.

This table summarises the quality of life of adult patients with severe hypodontia and their matching (age and gender) controls of patient attending for routine dental care. A (*) indicates significance ($P \leq 0.05$). N=58

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Domain	Test Group (N=29)				Control Group (N=29)				Paired t-test
	Mean	Standard deviation	Median	Interquartile range	Mean	Standard deviation	Median	Interquartile range	
Total scores	72	40.5	65	36.5-104	31.5	28.2	27	10-45.5	0.0003*
Functional limitation	14.8	7.6	12	9-20	9.6	5.6	10	6-12	0.009*
Physical pain	14.4	8.1	14	8-19	8.9	4.5	8	5-13	0.003*
Psychological discomfort	6.1	1.1	12	7.5-18	6.6	4.9	6	2-10	0.002*
Physical disability	12.1	8.6	11	5-19	4.6	3.8	3	2-6	0.0003*
Psychological disability	9.7	6.9	9	3-16	3.6	4.6	2	0-5	0.0009*
Social disability	4.4	4.3	2	0-9	1	2.3	0	0-1	0.002*
Handicap	4.4	3.9	4	1.5-7	1.7	3.1	0	0-3	0.009*

4.9.2.2 Impact of severe hypodontia on patients' self-esteem

The means and standard deviations for the test and control groups were 22.8 ± 5.4 and 22.4 ± 4 respectively. When the total scores for Rosenberg Scale were analysed using a paired t-test no statistically significant differences were found between the two groups ($P = 0.7$).

4.9.3 Results and data analysis for moderate hypodontia patients compared to their controls

A total of twelve moderate hypodontia patients of which nine were females and three were males with the age range of 19-34 years old were compared to their age and gender matching control group

As data were paired, a paired t-test was used to analyse data with a statistical significance level of ($t \leq 0.05$). Data analysis was carried out using JMP® statistical package (JMP Statistics and Graphics Guide, Release 7, Copyright© 2007, SAS Institute Inc., Cary NC, USA). No statistically significant results were found for the OHIP-49 nor for the Rosenberg self-esteem scale.

Table 4-3: Summary of results of OHIP-49 questionnaire for moderate hypodontia patients.

This table summarised the quality of life of adult patients with moderate hypodontia and their matching (age and gender) controls of patients attending for routine dental care. No statistically significant differences were found. N=24

Domains	**Test Group (N=12)**				**Control Group (N=12)**				**Paired t- test**
	Mean	**Standard deviation**	**Median**	**Interquartile range**	**Mean**	**Standard deviation**	**Median**	**Interquartile range**	
Total scores	38.25	21.9	41	16.75-58.25	30.5	25.5	20	10.25-57	0.44
Functional limitation	9.25	4.3	8.5	6-13	9	5.3	8	4.25-13	0.94
Physical pain	7.5	4.9	5.5	3.25-10.75	7.9	3.9	8.5	3.5-10	0.83
Psychological discomfort	6.6	4.9	8	2-10	6.5	4.8	6	2.25-10	0.97
Physical disability	6.9	4.9	7	2.25-11	5.8	5.5	5	1-9	0.66
Psychological disability	5.5	4.6	7.5	0-8	4.6	4.7	3.5	0-9.75	0.72
Social disability	0.9	1.9	0	0-0.75	1.5	2.27	0.5	0-3.5	0.48
Handicap	1.4	2.15	0.5	0-2	2.9	3.3	1.5	0.25-5.25	0.22

4.9.4 Data analysis for patients with amelogenesis imperfecta

4.9.4.1 Oral Health Impact Profile (OHIP-49)

Table 4-4: Summary of results of OHIP-49 questionnaire for amelogenesis imperfecta patients.

This table summarises the quality of life of adult patients with amelogenesis imperfecta and their matching (age and gender) controls of patient attending for routine dental care. N=54

Subclass	**Test group (N= 27)**				**Control group (N=27)**				**Paired t- test**
	Mean	**Standard deviation**	**Median**	**Interquartile range**	**Mean**	**Standard deviation**	**Median**	**Interquartile range**	
Total scores	67.4	27.9	69	48-85	48.8	33.2	37	22-82	0.01*
Functional limitation	13.5	5	14	10-18	11	7.2	11	5-17	0.08
Physical pain	13.6	5.4	15	9-18	11	6.6	9	5-15	0.12
Psychological discomfort	12.3	5	13	8-16	9.2	6.2	10	3-14	0.02*
Physical disability	9.7	4.6	9	6-13	6.4	6.5	3	2-11	0.03*
Psychological disability	10	6.4	10	4-15	5.7	5.5	4	1-11	0.004*
Social disability	4.1	4.1	4	0-6	1.8	2.4	1	0-3	0.02*
Handicap	3.8	4.1	3	1-5	3.2	3.6	2	0-5	0.57

4.9.4.2 Impact of amelogenesis imperfecta on patients' self-esteem

The means and standard deviations for the test and control groups were 21 ± 5.2 and 21 ± 4.2 respectively. When the total scores for Rosenberg Scale were analysed using a paired t-test no statistically significant differences were found between the two groups (P =0.92).

4.10 Discussion

Given the emphasis society places on facial aesthetics, dental health professionals have come to question the affects hereditary developmental defects of teeth have on a person's psychosocial health (Shaw et al., 1980; Shaw, 1981). It is essential that health care providers understand how dental health and appearance impact on the overall health and well-being (Shaw et al., 1985). Yet, to date, there are no data on the psychosocial influence of severe hereditary dental defects. Therefore, this study sought to provide information regarding the importance of teeth and appearance in normal psychosocial development.

Although the number of patients recruited for this study is relatively small, the sample size was comparable with other studies in this field (Armellini et al., 2008; Kende et al., 2008; Lopez-Jornet and Camacho-Alonso, 2008; Mehl et al., 2009; Goshima et al., 2010). To date the impact of hypodontia on the adult patient population has not yet been investigated.

This study provided some new data and attempted to objectively characterise the functional and psychosocial impacts that developmental dental disorders, such as non-syndromic moderate to severe hypodontia and amelogenesis imperfecta, have on the affected individuals in terms of the perceived quality of life and self-esteem. This patient population was compared to a control group of matching age and gender of individuals without the conditions attending the Dublin Dental University Hospital for routine dental care.

As data were paired, a paired t-test was used to analyse data with a statistical significance level of ($P \leq 0.05$).

Measures such as OHIP-49 are useful for comparisons across populations but have limited ability to capture the effects of a particular condition because, there is, as yet, no

validated condition-specific quality of life measure for the impact of different dental disorders including hypodontia and amelogenesis imperfecta on patients (Cunningham et al., 2000). Hence, a general oral health measure (OHIP-49) was used in this study. During the course of this study, Akram and co-workers conducted a qualitative research and developed a new questionnaire to assess the QoL in patients with hypodontia (Akram et al., 2011); however, this questionnaire remains to be tested and validated. Similarly, Meaney and co-workers carried out a qualitative study to investigate the impact of congenital absence of teeth on functional and psychosocial wellbeing and to identify factors considered important to patients which should be incorporated into health status measures for population studies and clinical trials (Meaney et al., 2011).

One of the shortcomings of the OHIP-49 is the large number of items included, which may limit its use in clinical trials, clinical practice and surveys (Awad et al., 2008). However, when the numbers of items on the questionnaire were reduced, its responsiveness was maintained, but there was a decrease in reliability, especially within the "Psychological" and "Social" disabilities and "Handicap" domains for the 35 to 65-year group. Its construct validity was also reduced in "Physical pain", "Psychological" and "Social disabilities" domains for the 35 to 65-year group, as well as on "Physical pain", "Psychological discomfort", Physical" and Psychological" disabilities in the 65 year and older group. This occurred primarily when reducing from one or two items per domain (Awad et al., 2008). A second problem with these OHRQoL questionnaires is the flooring effects of scores compared to the maximum scores i.e. patients tend to score on the lower end of the scale. Only recently John and co-workers defined the Minimum Important difference (MID) for the OHIP-49 as 6 OHIP units lower (95% confidence interval) (John et al., 2009). This is defined as "the smallest difference in score in the domain of interest which patients perceive as beneficial and which would mandate, in the absence of troublesome side effects and excessive cost, a change in the patient's management" (Jaeschke et al., 1989). This allows the clinical as well as the statistical significance of differences between groups to be better appreciated. This novel approach however remains to be validated.

Given the paucity of literature on this subject matter, calculation of sample sizes required to test for differences with appropriate level of power is not straightforward. However, there are clear limitations on the conclusions that can be drawn based on the

numbers in each group in this research. Based on the observed values of the means and standard deviations in this research a post hoc analysis for sample size calculation was performed using the data generated to provide some guidance as to the numbers required to test for differences at an 80% power level. The analysis was carried out using the minimal important difference, which has been recently determined as approximately 6 points lower (better) on the OHIP-49 scale. This will, at the very least, provide information to guide future researchers of the number of subjects required to do similar research.

According to the data generated from this research (N=41 test, and N=41 control), the power to detect 6 units for "all hypodontia" group and "severe hypodontia" group is in excess of 90% based on observed means and standard deviations. Whereas, for the "moderate hypodontia" group, the power is less than 10% and a sample size of around 150 matched pairs would be required to detect a 6 units difference for this group in order to achieve any statistically significantly different results. The reason why the current study was underpowered in the "moderate hypodontia" group could be attributed to the large standard deviations observed which led to large standard errors. Increasing the sample size for the "moderate hypodontia" group to 150 matched pairs was not feasible at the time this research was conducted.

4.10.1 Hypodontia and AI patients versus their controls

Oral Health Impact Profile (OHIP-49)

Upon examination of the impact of hypodontia and AI on quality of life of patients affected by these conditions it was evident that these two conditions had a significant impact on patients' quality of life under the main domains of the oral health impact profile questionnaire.

For combined hypodontia patients (moderate and severe), the total scores, psychological discomfort, physical disability, psychological disability, physical pain and social disability were all significantly different. Only the handicap domain was not significantly different between the test and control groups, which may indicated that hypodontia does not cause severe impairment to the extent that the patient is handicapped physically, psychologically or socially. The overall mean responses of the total scores of affected patients were higher than that for patients without the condition

(μ = 62.1, and μ = 31.2) respectively, and the difference was statistically significant (P = 0.003).

For AI patients, the condition had a significant impact on adult patients' quality of life under five of the seven domains of the oral health impact profile questionnaire, which included: total scores (P = 0.01), psychological discomfort (P = 0.02), physical disability (P = 0.03), psychological disability (P = 0.004), and social disability (P = 0.02). The overall mean responses of the total scores of affected patients were higher than that for patients without the condition (μ =67.4 and μ =48.8) respectively, and the difference was statistically significant (P = 0.01).

These results are in agreement with the single study available in the literature that investigated thirty patients with AI and compared them to unaffected family members who served as controls. It concluded that amelogenesis imperfecta had a great impact on the quality of life of affected individuals (Coffield et al., 2005). Unfortunately, the authors used OHIP-14, which is a short version of OHIP-49, making comparison between that study and the results of this thesis difficult.

Comparing the mean scores between amelogenesis imperfecta (μ = 67.4) and combined moderate and severe hypodontia patients (μ =62) in this study revealed that amelogenesis imperfecta had a far greater impact than hypodontia. However, the mean score of amelogenesis imperfecta was less than those for severe hypodontia (μ = 72), as will be discussed later.

The results of this study were also in agreement with other studies, which compared different conditions affecting the oral cavity to control groups (Armellini et al., 2008; Lopez-Jornet and Camacho-Alonso, 2008; Lopez-Jornet et al., 2008).

Aremellini and co-workers, in a well designed study, investigated the effects of removable partial dentures (RPDs) on oral health related quality of life in one hundred and sixty subjects with shortened dental arches (SDAs), with fully dentate patients serving as controls. The test group patients were further categorised into five subgroups according to the size and distribution of their edentulous spaces. Aremellini concluded that, from a quality of life perspective, patients with SDAs perceive benefits from RPDs only if anterior teeth replacements were included. This was evident from the significantly higher OHIP-49 scores of the test group compared to the controls. These

benefits could not be demonstrated for patients with uninterrupted SDAs where only posterior teeth were replaced by distal extension RPDs.

Lopez-Jornet and colleagues in two separate studies compared quality of life in patients with either Sjögren's Syndrome (SS) (n = 44) (Lopez-Jornet and Camacho-Alonso, 2008) or Burning Mouth Syndrome (BMS) (n = 60) (Lopez-Jornet et al., 2008) to a group of control patients attending for reviews at the dental hospital of the University of Murcia, Spain. In both studies, scores of OHIP-49 for the test group were higher than those of their controls and statistically significantly different ($P = 0.005$ and $P < 0.001$ respectively). The mean scores of all subscales of this study were greater than those reported by Aremellini et al and comparable to those reported by Lopez-Jornet et al. This could be related to the severity of the oral condition, as there was no disease activity in Aremellini's study when compared to Lopez-Jornet et al, where Burning Mouth Syndrome was investigated.

Table 4-5: Comparing OHIP-49 mean scores between different studies

Domain	This research	Aremellini	Lopez-Jornet
Functional limitations	13.2	1.2	6.94
Physical pain	12.4	1.5	3.9
Psychological discomfort	10	1.4	4.03
Physical disability	10.6	0.5	3.45
Psychological disability	8.4	0.6	1.7
Social disability	3.4	0.3	1.8
Handicap	3.5	0.4	2.4
Total scores	62	Not mentioned	24

Rosenberg Self-Esteem Scale

When examining the total scores of Rosenberg self-esteem scale for hypodontia and amelogenesis imperfecta patients, there were no statistically significant differences

between the test and control groups. The mean scores of the test group for AI patients were slightly lower than that of the control group ($\mu = 21.7$ and $\mu = 21.8$). Whereas, the mean scores for the hypodontia patients were similar to those of their controls ($\mu = 22.34$ and $\mu = 22.36$ respectively).

These findings are in agreement with the findings of the one study, which looked at the quality of life of AI patients compared to unaffected family members and reported that patients with AI had a slightly lower self esteem than their controls ($\mu = 23.9$ and $\mu = 24.9$ respectively) (Coffield et al., 2005).

These results are also in agreement with another study by Nicodemo and co-workers (Nicodemo et al., 2008), which investigated the effects of orthognathic surgery on patients with class III malocclusion and reversed over-jet of $\geq$ 4mm. The authors reported that no significant effects were noticed on patients' self esteem before and after the surgery. The study did not describe their mean scores and did not use a control group. The pre-surgical evaluation was carried out in the thirty days preceding the operation and after orthodontic preparation. If the pre-surgical evaluation was carried out before the initiation of orthodontic treatment, they results may have been different as one would expect a more defined improvement.

The results of this study concurred with a study, which looked at the impact of oral-facial clefts (cleft lip and cleft palate) on patients' self esteem by comparing them to unaffected siblings and a control group (Berk et al., 2001). The study found that patients with oral-facial clefts had lower self esteem compared to their unaffected siblings and controls and the mean score were ($\mu = 19$, $\mu = 21.5$ and $\mu = 21$ respectively), but the differences were not statistically significant.

In a psychiatric analysis of patients who attempted suicide and sustained self-inflicted maxillofacial injuries due to gunshot wounds, Ozturk investigated the self esteem of these patients compared to healthy controls using Rosenberg Self Esteem Scale and found a statistically significant difference between the two groups ($P < 0.01$) (Ozturk et al., 2006).

It is worth mentioning that the mean scores for hypodontia and AI patients in this study were higher than subjects with orofacial malformations such as orofacial clefts ($\mu = 19$)

(Berk et al., 2001), or unattractive skin conditions such as severe acne (μ = 19) as reported by Mallon et al (Mallon et al., 1999). Mean scores for patients who sustained maxillofacial injuries were not detailed (Ozturk et al., 2006).

In context, the results of this study may be explained by the fact that hypodontia and AI may not have an impact on patients' self-esteem or the fact that the Rosenberg Self Esteem Scale may not be sensitive enough to changes in dental patients' self esteem as it seems to pick up differences only in severe facial disfigurement cases. This highlights the need for developing condition specific instruments capable of measuring the impact of dental conditions on patient's self esteem. To date there is a paucity of research in this field.

4.10.2 Severe hypodontia versus controls

When the impact of severe hypodontia only on quality of life for hypodontia patients in this study was investigated, it was clearly evident that the condition had a significant impact on adult patients' quality of life under all the domains of the oral health impact profile questionnaire which included: total scores, functional limitations, physical pain, psychological discomfort, physical disability, psychological disability, social disability and handicap. This effect was masked when both moderate and severe hypodontia patients were combined together. However, this did not change the self esteem results when they were investigated separately.

4.10.3 Moderate hypodontia versus controls

By contrast, when the impact of moderate hypodontia alone was investigated, there was no statistically significant difference in any of the domains of the two questionnaires used. These findings concurred with a 15- year follow up study by Helm et al (Helm et al., 1985) in which he showed that severe traits of malocclusion such as severe over-jet, severe deep bite, overall crowding, and maxillary incisor crowding caused more concerns than moderate traits such as malocclusion in lateral segments (cross-bite, scissor bite) and spacing of the maxillary incisors. Due to the small sample size it may be difficult to extrapolate the findings of this research on the impact of moderate hypodontia of affected patients. Increasing the sample size could have resulted in different outcomes but it was not feasible at the time the research was conducted.

As discussed in the previous Chapter, and for the same tooth type, the site-related pattern of tooth agenesis is almost the same when the results of combined, severe, and moderate hypodontia cases were plotted separately. Hence, the impact of hypodontia on the quality of life may not be solely related to the agenesis of a specific tooth type (anterior or posterior) but may be directly related to the severity of the condition (increased number of developmentally missing teeth) as shown from this study. Research investigating and comparing the impact of mild, moderate and severe hypodontia on quality of life is lacking.

The inclusion of patients presenting at the Dublin Dental University Hospital in this study may have introduced an element of bias. It may be that these patients report greater oral health impact in the hope of receiving earlier treatment or that those seeking treatment (routine caries management) report a greater impact than those not seeking treatment. Furthermore, there is a potential bias due to the fact that the study did not fully control for oral health status. Whilst the test group was more than likely to be free of untreated caries and other oral diseases due their interdisciplinary management approach from an early age, the control group had, by definition, some level of dental disease, as it was a convenience sample from patients attending the dental hospital for routine dental care. However, this limitation applies to any possible control group from the normal population.

An additional limitation of this study is the fact that conclusions are valid only if it is accepted that patients being treated for dental diseases are an appropriate comparison group. Other hypodontia studies have used orthodontic patients without hypodontia as controls or did not use control groups.

4.11 Conclusion

In this study the impact of developmental dental disorders such as hypodontia (moderate/severe), and amelogenesis imperfecta on the quality of life and self-esteem of adult patients was investigated. These developmental dental disorders appear to have a great impact on the quality of life of this patient population under most quality of life domains. The psychosocial impact of severe hypodontia on the quality of life of affected patients is greater than that experienced by patients affected with moderate hypodontia, and is similar to the impact of amelogenesis imperfecta.

Hypodontia and amelogenesis imperfecta did not seem to affect the self esteem of this patient population in comparison to their control. This finding is consistent with the findings of previous research in the field.

The overall results of this study reject the null hypothesis and demonstrate that there is a statistically significant difference between patients with hypodontia and amelogenesis imperfecta as compared to their controls. However, no statistically significant differences were noticed when self esteem was investigated and the null hypothesis is accepted.

Chapter 5

Impact of Hypodontia and Amelogenesis Imperfecta on the Quality of Life and Self-Concept of Young patients and their families

Chapter 5: Impact of Hypodontia and Amelogenesis Imperfecta on Quality of Life and Self Concept of Young Patients

5.1 Introduction

Patients with hypodontia and amelogenesis imperfecta are confronted with a number of problems from the early days of the eruption of their primary and subsequently, their permanent teeth. These problems may include dental pain or sensitivity and the inability to chew food properly. Aesthetic can be a major issue for parents of young children and an issue for the younger patients as they reach teenager and young adult years (Hobkirk et al., 1994; Coffield et al., 2005).

In a meta analysis review of cross sectional studies of available literature on bullying, it was concluded that chronic bullying can lead to depression, loneliness, anxiety, low self-esteem and underachievement at school (Hawker and Boulton, 2000). This was further validated and related to dental appearance and the effects of malocclusion (DiBiase and Sandler, 2001).

In school children, deviations in dental appearance have been found to be a target for teasing, although the group of children to whom this applied was also found to be at a higher risk of harassment generally. The greater the deviation of the dental appearance, the greater the implication for the child, and comments about teeth appeared to be more hurtful than those about other features (Shaw et al., 1980).

Developmental dental disorders may impact significantly on dental aesthetics. Factors, which contribute to a decrease in dental aesthetics include:

- The severity of the condition
- Abnormalities in tooth shape and colour i.e. microdontia, pitted enamel, and discoloured teeth
- Abnormal tooth position

Features such as spacing, as in hypodontia, or discolouration, as in amelogenesis imperfecta, may become a focus for bullies and parents may insist upon treatment on this basis. However, research in this area is lacking.

One important issue that remains to be investigated further with respect to hypodontia and (AI) is their impact on well-being and quality of life. Increased emphasis is being placed on patients' perception of the impact of oral disorders. This is particularly important with respect to conditions such as hypodontia and (AI) whose effects are predominantly functional and psychosocial. Understanding the consequences of such conditions has the potential to contribute to clinical management and evaluation of the benefits of different therapeutic approaches (Coffield et al., 2005; Wong et al., 2006; Locker et al., 2010).

5.2 Purpose of the study

The purpose of this study was, first, to examine the impact of hypodontia and amelogenesis imperfecta on self concept of young patients referred to a tertiary care clinic in the Dublin Dental University Hospital, compared to patients attending the hospital for routine dental care. The second purpose was to investigate the perceived effects of these conditions on young patients' quality of life as reported by their parents and to assess the impact these conditions may have on the family of affected individuals.

5.3 Inclusion Criteria

- Confirmed clinical and radiographical diagnoses of moderate to severe hypodontia or amelogenesis imperfecta.
- Current biographical information.
- Children of at least 8 years old and their parents.
- No associated syndromes.
- No other systemic health problems.

5.4 Exclusion Criteria

- The presence of associated syndromes.
- Inability to consent, due to medical or intellectual conditions.
- Patient and/or parents unwilling to participate.

5.5 Null Hypothesis

The null hypothesis was that there is no difference in patients self concept and quality of life "as reported by parents" between patients with developmental dental disorders

such as hypodontia and amelogenesis imperfecta, and other dental patients attending the Dublin Dental University Hospital for routine dental care.

5.6 Materials and Methods

Young patients and their parents, with clinical and radiological diagnoses of moderate to severe hypodontia or amelogenesis imperfecta (AI) and their matching (age and gender) control patients attending the Dublin Dental University Hospital for routine dental care were recruited. The research proposal was reviewed and approved by the Research Ethics Committee in Trinity College, Dublin (see Appendix).

Fifty-two hypodontia and twenty-one AI patients were identified and recruited from a pool of patients referred to the Special Dental Needs Clinic for consultations and/or treatment. Patients were also identified from trawls of consultant day schedules in the Departments of Restorative Dentistry and Periodontology, and Child and Public Dental Health. Staff and students in the dental hospital were also contacted by email and notified about the ongoing research in order to identify suitable patients.

Control patients were identified primarily from the hospital's main patient database "Salud". Undergraduate and postgraduate students were asked to help identify suitable patients.

Three mailings, which contained information leaflets, consent forms (See Appendix), and prepaid envelopes were sent to all patients to give the recruits multiple opportunities to respond. At least one attempt was made to contact participants by telephone informing them about the study and answer any questions or queries they may arose.

Forty-two hypodontia patients (80.7%) and nineteen AI patients (90.4%) and their parents/caregivers agreed to participate in the study. These patients were then age and gender matched with a control group of patients attending the Dublin Dental University Hospital for routine dental care. The response rate for the control group was initially very poor (10%) for hypodontia and (17%) for AI, but recruitment continued until required numbers were obtained. This involved contacting patients directly in the hospital or by telephone, either by the researcher or the clinician (staff/student) treating the patient. Patients who declined to participate in the study were not contacted again. Only patients who did not respond were re-contacted to ensure that they had received

the letters and determine their willingness to participate. If the answer was negative these patients were not contacted again.

A written informed consent to participate in the study was obtained from all participants and their parents before enrolling in the study. The same procedure was followed for the control group.

A postal package of questionnaires, information leaflet, instructions, and prepaid envelopes was sent to all potential participants. Participants were advised to complete all questions and return them in the prepaid envelope provided. If the questionnaires were returned due to an incorrect address and the correct address could not be determined, or if questionnaires were incomplete or missing the person was deleted from the study.

To ensure anonymity and to comply with the Ethic's Committee guidelines, all questionnaires were given codes and returned to a research assistant to whom patient identity and underlying condition was unknown, as patients had been assured their identity would not be disclosed.

5.7 Questionnaires design

5.7.1 Piers Harris Self Concept Scale (Piers and Herzberg, 2007)

This is a 60-item, self-report instrument designed to assess the self-concept of children and adolescents (See appendix). The term "self-concept" refers to a child's or adolescent's attitude toward his/her behaviour and attributes. "Yes" or "No" responses are requested for each descriptive statement and are scored in either a negative or positive direction. The Piers Harris provides an overall score assessing self-concept, with a high score denoting high self-concept and a low score denoting low self-concept. The Piers-Harris also provides six cluster scales measuring: behavioural adjustments, intellectual and school status, physical appearance, freedom from anxiety, popularity, happiness and satisfaction. The Piers Harris is considered a highly reliable and valid instrument with test reliability ranges from 0.42 to 0.96, and internal consistency from 0.88 to 0.92 (Smith and Rogers, 1978; Cooley and Ayres, 1988a; Piers and Herzberg, 2002).

5.7.2 Parental Perception Questionnaire (Jokovic et al., 2003b)

This questionnaire was developed by Locker et al (Locker et al., 2002a). It consists of thirty one questions (See appendix) for the Parental Perception Questionnaire scored separately on a Likert (Likert, 1932) response format ranging from 0 to 4. The total scores for the Parental Perception Questionnaire range from (0 to 124). It also has two Global Rating Questions to evaluate the overall well being of the child (Jokovic et al., 2003b). It evaluates the impact of the condition on the child as perceived by their parents. The responses of parents are scored under four subsets: oral symptoms, functional limitations, emotional well-being, and social well-being.

5.7.3 Family Impact Scale (FIS) (Locker et al., 2002a)

The FIS is included in the PPQ and consists of 14 items (See Appendix) that attempted to capture the effects of a child's oral or oro-facial condition on four domains related to parental and family activities (5 questions), parental emotions (4 questions), family conflict (4 questions) and family finances (1 question). The questions were asked about the frequency of events in the previous 3 months as follows: "During the past 3 months, how often... (has there been disagreement or conflict in your family)... because of your child's teeth, lips, mouth or jaws". Response options for the four domains and the respective scores were: 'Never' (scoring 0); 'Once or twice' (1); 'Sometimes' (2); 'Often' (3); and 'Everyday' or 'Almost everyday' (4). A 'Don't know' (DK) response was also allowed. The FIS scores were computed by summing all of the item scores. Scores for each of the four domains can also be computed. Since there were 14 questions, the final score could vary from 0 to 56, for which a higher score denoted a greater degree of the impact of the child's oral conditions on the functioning of parents-caregivers and the family as a whole.

Parents and caregivers were also asked to give overall or global assessments of the child's oral health and the extent to which the hypodontia or amelogenesis imperfecta conditions affected the child's overall well-being. These global ratings are included in the Parental Perception Questionnaires (PPQ) These questions are: "How would you rate the health of your child's teeth, lips, jaws and mouth?" with a five-point response format ranging from 'excellent' to 'poor' and "How much is your child's overall well-being affected by the condition of his/her teeth, lips, jaws or mouth affect your life

overall?" with a response range from 'not at all' to 'very much'. These ratings did not include a "Do Not Know" (DK) response.

These questionnaires have demonstrated good cross-sectional validity and test-retest reliability in clinical and general populations, and in their discriminatory properties, i.e., their ability to distinguish between groups of patients with different clinical conditions and the varying levels of severity of these conditions (Locker et al., 2002a; Jokovic et al., 2003a; Wong et al., 2006). However, for the Child Oral Health Related Quality of Life (COHRQoL) questionnaires, which include, the Child Perception Questionnaire 8-10 (CPQ 8-10), the Child Perception Questionnaire 11-14 (CPQ 11- 14), Parental Perception Questionnaire (PPQ), and the Family Impact scale (FIS), the minimal important difference has yet to be established. This is the minimum difference in scores on a questionnaire that patients perceive as important and would lead them to report an improvement or deterioration in health status over time or as a result of therapy. This allows "clinical" as well as the statistical significance of differences with and between groups to be appreciated (Locker et al., 2004; Locker et al., 2010).

As the age range for patients recruited was between 9-18 with a very few subjects within the age range of 8-14, two of the four questionnaires comprising the Child Oral Health Related Quality of Life Questionnaire (COHRQoL) were not used (the Child Perception Questionnaire 8-10 and the Child Perception Questionnaire 11 to 14). However, the Parental Perception Questionnaire and the Family Impact Scale questionnaires were used for all subjects under 18 years of age.

Validation for these instruments have been established, and the instruments have been used extensively in other studies in the dental literature (Jones et al., 1978; Kapp, 1979; Leonard et al., 1991; Dann et al., 1995; Mandall et al., 2001; Jokovic et al., 2002; Locker et al., 2002a; Jokovic et al., 2003a; Jokovic et al., 2003b; Coffield et al., 2005; Wong et al., 2005; Barbosa Tde and Gaviao, 2009).

5.8 Results

Thirty-four hypodontia patients (80.9%) and sixteen AI patients (84.2%) returned the completed questionnaires. Of these, five hypodontia and four AI patients were eliminated because of incomplete, missing or inappropriately filled questionnaires (this includes random responding or Yes or No biased answers for the Piers Harris

Questionnaire). The remaining 19.1% of hypodontia patients and 15.8% of AI patients who did not return the questionnaires have dropped out the study for a variety of reasons including inadequate time, a child and/or parent declining to complete questionnaires, moving addresses, and busy with exams and/or work.

A total of twenty-nine hypodontia patients (55.7% of the total number), and twelve amelogenesis imperfecta patients (57.1% of the total number), and their parents with their matching controls completed all questionnaires successfully. The age range for hypodontia and amelogenesis imperfecta patients was 9-17 and 8-17, respectively. A blinded researcher to whom patient identity and clinical details were unknown scored all questionnaires and entered the scores into a Microsoft Excel® spread sheet (Microsoft Corp. USA), and scores were prepared for statistical analysis. For the Parental Perception Questionnaire and Family Impact Scale, the zero value was assigned to each "don't know" (DK) response prior to the calculation of scores as proposed previously (Jokovic et al., 2003a; Barbosa Tde and Gaviao, 2009).

All scores for the Piers Harris Children Self Concept Scale, Parental Perception Questionnaires and Family Impact Scale were entered into a Microsoft Excel® spreadsheet and prepared for statistical analysis using JMP® statistical package (JMP Statistics and Graphics Guide, Release 7, Copyright© 2007, SAS Institute Inc., Cary NC, USA). A paired t-test was used to analyse data (See appendix).

5.9 Data Analysis for all Hypodontia Patients

5.9.1 Piers Harris questionnaire

5.9.1.1 Total scores

The means and standard deviations for the test and control groups were 54.3 ± 10.4 and 51± 11.7 respectively. When the total scores of Piers Harris Questionnaires were analysed, no statistically significant differences were found between the two groups (P = 0.18).

5.9.1.2 Behavioural adjustments

The means and standard deviations for the test and control groups were 49.3 ± 9.9 and 55.7 ± 8.7 respectively. Scores of the behavioural adjustments of Piers Harris Scale

were analysed and a statistically significant difference was noted between the two groups (P = 0.02).

5.9.1.3 Intellectual and school status

The means and standard deviations for the test and control groups were 53.7 ± 9.7 and 50.2 ± 9.8 respectively. Analysis of intellectual and school status scores revealed no statistically significant difference between the two groups (P = 0.09).

5.9.1.4 Physical appearance and attributes

The means and standard deviations for the test and control groups were 51.3 ± 6.8 and 51.8 ± 11.7 respectively. Analysis of physical appearance and attribute scores showed that no statistically significant difference was observed between the two groups (P = 0.84).

5.9.1.5 Freedom from anxiety

The means and standard deviations for the test and control groups were 51.6 ± 10.2 and 50.2 ± 9.9 respectively. Freedom from anxiety was not found to be statistically different between the two groups (P = 0.5).

5.9.1.6 Popularity

The means and standard deviations for the test and control groups were 55.6 ± 10.5 and 52.4 ±11 respectively. Analysis of popularity subscale scores revealed no statistically significant differences were found between the two groups (P = 0.21).

5.9.1.7 Happiness and satisfaction

The means and standard deviations for the test and control groups were 49.3 ± 7.8 and 49.3 ± 8.5 respectively. Scores of the happiness and satisfaction subscale were analysed and it was found that results were not statistically significantly different between the two groups (P = 0.96).

For brevity reasons and to avoid repetition, a summary of all Piers Harris Self Concept Scale data for all other groups will be presented in a table format only for the remainder of this chapter.

Table 5-1: Summary of results of Piers Harris questionnaire for all young hypodontia patients.

This table summarises the Self Concept Scale results of young patients with moderate/severe hypodontia and their matching (age and gender) controls of patient attending for routine dental care (*) indicates significance ($P \leq 0.05$). (N= 58)

Domain	**Test Group (N=29)**		**Control Group (N=29)**		**Paired t-test**
	Mean	**Standard deviation**	**Mean**	**Standard deviation**	
Total scores	54.3	10.4	51	11.7	0.18
Behavioural adjustment	49.3	9.9	55.7	8.7	0.02*
Intellectual and school status	53.7	9.7	50.2	9.8	0.09
Physical appearance and attributes	51.3	6.8	51.8	11.7	0.84
Freedom from anxiety	51.6	10.2	50.2	9.9	0.5
Popularity	55.6	10.5	52.4	11	0.21
Happiness and satisfaction	49.3	7.8	49.3	8.5	0.96

5.9.2 Parental Perception Questionnaire (PPQ) of all hypodontia patients

5.9.2.1 Global Rating Items

The scores of the two global rating questions of the Parental Perception Questionnaire ("How would you rate the health of your child's teeth, lips, jaws and mouth?" with a five-point response format ranging from 'excellent' to 'poor' and "How much is your child's overall well-being affected by the condition of his/her teeth, lips, jaws or mouth affect your life overall?" with a response range from 'not at all' to 'very much') were calculated. Due to the small sample size and to ensure sufficient numbers for each response, data were collapsed for the first question to compare scores of the "poor" response to the collective scores of "Fair, Good, Very good, and Excellent". The same action was carried out for the second question comparing "Not at all" scores to the collective scores of "Very little, Some, A lot, and Very much". Statistical analysis was performed using Fisher's Exact Test. No statistically significant differences were noticed between the test and control groups for the two items (P = 1.38 and P = 1.00) respectively.

5.9.2.2 Total scores

The means and standard deviations for the test and control groups were 27.4 ± 19.4 and 16 ± 12.5 respectively. Analysis of total scores of the Parental Perception Questionnaire revealed a statistically significant difference between the two groups (P = 0.02). This means that parents of patients affected with hypodontia scored higher on this scale indicating a greater impact of hypodontia on their children's quality of life.

5.9.2.3 Oral Symptoms

The means and standard deviations for the test and control groups were 6 ± 4.2 and 4.2 ± 3.7 respectively. Scores of oral symptoms domain of the Parental Perception Questionnaire revealed no statistically significant difference (P = 0.13).

5.9.2.4 Functional limitations

The means and standard deviations for the test and control groups were 8.4 ± 6.4 and 4.2 ± 3.2 respectively. When scores of functional limitations domain were analysed, a

statistically significant difference was observed between test and control groups (P = 0.005).

5.9.2.5 Emotional well being

The means and standard deviations for the test and control groups were 6.1 ± 6.2 and 3.7 ± 4.2 respectively. Analysis of emotional well-being domain of the Parental Perception Questionnaire revealed no statistically significant difference between groups (P = 0.08).

5.9.2.6 Social well being

The means and standard deviations for the test and control groups were 6.7 ± 6.2 and 3.9 ± 3.9 respectively. When social well-being scores were analysed no statistically significant different was found, however it did approach significance (P = 0.07).

For brevity reasons and to avoid repetition, a summary of all the Parental Perception Questionnaire data for all other groups will be presented in a table format only for the remainder of this chapter.

Table 5-2: Summary of results of Parental Perception Questionnaire of all young hypodontia patients.

This table summarises the Parental Perception Questionnaire results of young patients with moderate/severe hypodontia and their matching (age and gender) controls of patient attending for routine dental care. A (*) indicates significance ($P \leq 0.05$). (N=58)

Domain	**Test Group (N=29)**		**Control Group (N=29)**		**Paired t-test**
	Mean	**Standard deviation**	**Mean**	**Standard deviation**	
Total scores	27.4	19.4	16	12.5	0.02*
Oral symptoms	6	4.2	4.2	3.7	0.13
Functional limitations	8.4	6.4	4.2	3.2	0.005*
Emotional well being	6.1	6.2	3.7	4.2	0.08
Social well being	6.7	6.2	3.9	3.9	0.07

5.9.3 Family Impact Scale of all hypodontia patients (FIS)

5.9.3.1 Total Scores

The means and standard deviations for the test and control groups were 6.3 ± 7 and 5 ± 5.3 respectively. Analysis of total scores of the Family Impact Scale showed no significant difference between groups (P = 0.45).

5.9.3.2 Parental/Family activity

The means and standard deviations for the test and control groups were 2.6 ± 2.6 and 2 ± 2 respectively. No statistically significant difference was found (P = 0.35) when data of parental and family activities domain of the family impact scale were analysed.

5.9.3.3 Parental Emotions

The means and standard deviations for the test and control groups were 2.4 ± 3.6 and 1.2 ± 2.2 respectively. Analysis of impact on parental emotions domain of Family Impact Scale showed no statistically significant difference (P = 0.14).

5.9.3.4 Family conflict

The means and standard deviations for the test and control groups were 1.0 ± 1.5 and 1.0 ± 1.3 respectively. Analysis of data in this domain revealed no statistically significant difference between the two groups (P = 0.92).

5.9.3.5 Financial burden

The means and standard deviations for the test and control groups were 0.17 ± 0.6 and 0.65 ± 1.0 respectively. Analysis of scores for financial burden subscale showed that scores of this domain approached significance (P = 0.06).

For brevity reasons and to avoid repetition, a summary of all Family Impact Scale data for all other groups will be presented in a table format only for the remainder of this chapter.

Table 5-3: Summary of results of Family impact scale of all hypodontia patients.

This table summarises the Family Impact Scale results of young patients with moderate/severe hypodontia and their matching (age and gender) controls of patient attending for routine dental care. A (*) indicates significance ($P \leq 0.05$). (N=58)

Domain	**Test Group (N=29)**		**Control Group (N=29)**		**Paired t-test**
	Mean	**Standard deviation**	**Mean**	**Standard deviation**	
Total Scores	6.3	7	5	5.3	0.45
Parental/family activity	2.6	2.6	2	2	0.35
Parental emotions	2.4	3.6	1.2	2.2	0.14
Family conflict	1	1.5	1	1.3	0.92
Financial burden	0.17	0.6	0.65	1	0.06

5.10 Results and Data Analysis for Severe hypodontia patients compared to controls

It was observed from the raw data that some individuals scored much higher within the hypodontia group. In order to identify if any pattern existed the hypodontia group were separated into moderate (4-6 teeth missing) and severe (greater than 6 teeth missing) and statistically analysed using the same statistical analysis.

A total of twenty young hypodontia patients of which twelve were males and eight were females and their parents were compared to their matching controls. The age range was from 11 to 17 years. All scores were entered into a Microsoft Excel® spreadsheet and prepared for statistical analysis using JMP® statistical package (JMP Statistics and Graphics Guide, Release 7, Copyright© 2007, SAS Institute Inc., Cary NC, USA).

As data were paired, a paired t-test was used to analyse data with a statistical significance level of ($t \leq 0.05$).

5.10.1 Piers Harris questionnaire

Table 5-4: Summary of results of Piers Harris questionnaire for young severe hypodontia patients.

This table summarises the Self Concept Scale results of young patients with severe hypodontia and their matching (age and gender) controls of patient attending for routine dental care. A (*) indicates significance ($P \leq 0.05$) (N=40)

Domain	**Test Group (N=20)**		**Control Group (N=20)**		**Paired t- test**
	Mean	**Standard deviation**	**Mean**	**Standard deviation**	
Total scores	57	10	50	13.1	0.07
Behavioural adjustment	58	7	48.2	10	0.02*
Intellectual and school status	55.8	10.1	50	11.1	0.07
Physical appearance and attributes	52.45	7.2	50.2	12.4	0.4
Freedom from anxiety	53.2	9.7	50.6	9.9	0.3
Popularity	56.25	10.3	50.7	12	0.08
Happiness and satisfaction	50.25	7.4	49.5	8.4	0.7

5.10.2 Parental Perception Questionnaire (PPQ) for young severe hypodontia patients

Table 5-5: Summary of results of Parental Perception Questionnaire of young severe hypodontia.

This table summarises the Parental Perception Questionnaire results of young patients with severe hypodontia and their matching (age and gender) controls of patient attending for routine dental care. A (*) indicates significance ($P \leq 0.05$) (N=40)

Domain	**Test Group (N=20)**		**Control Group (N=20)**		**Paired t-test**
	Mean	**Standard deviation**	**Mean**	**Standard deviation**	
Total scores	25.9	20	15	12.6	0.07
Oral symptoms	5.75	3.6	4.2	3.9	0.27
Functional limitations	7.8	6.36	3.6	2.4	0.01*
Emotional well being	5.65	6.7	3.65	4.7	0.30
Social well being	6.65	6.8	3.5	4	0.13

5.10.3 Family Impact Scale (FIS)

Table 5-6: Summary of results of family impact scale of young severe hypodontia patients.

This table summarises the Family Impact Scale results of young patients with severe hypodontia and their matching (age and gender) controls of patient attending for routine dental care. A (*) indicates significance ($P \leq 0.05$) (N=40)

Domain	**Test Group (N=20)**		**Control Group (N=20)**		**Paired t-test**
	Mean	**Standard deviation**	**Mean**	**Standard deviation**	
Total Scores	5.75	6.8	4.05	5.04	0.41
Parental/family activity	2.45	2.6	1.65	1.72	0.28
Parental emotions	2.3	3.1	0.9	1.2	0.14
Family conflict	0.9	1.6	0.9	1.2	1.00
Financial burden	0.1	0.3	0.9	2.1	0.08

5.11 Results and Data Analysis for moderate hypodontia patients compared to their controls

A total of nine moderate hypodontia patients of which four were females and five were males and their parents were compared to matching controls. The age range was from 14-17 years.

As data were paired, a paired t-test was used to analyse data with a statistical significance level of ($t \leq 0.05$). Data analysis was carried out using JMP® statistical package (JMP Statistics and Graphics Guide, Release 7, Copyright© 2007, SAS Institute Inc., Cary NC, USA) and the results were as follow:

When the results of moderate hypodontia patients and their controls were investigated no statistically significant differences were found between the two groups with the exception of the emotional wellbeing domain of the parental perception questionnaire.

5.11.1 The emotional wellbeing domain of the Parental Perception Questionnaire

The interquartile range for the test group was 2.5- 11.5, and the median was 9, whereas that of the control group was 0- 6.5, and the median 3. Analysis of emotional well being subscale of the Parental Perception Questionnaire revealed a statistically significant difference between the two groups ($P = 0.04$).

5.12 Data Analysis for Young Amelogenesis imperfecta Patients

All scores for the Piers Harris children self concept scale and parental report questionnaires were entered into a Microsoft Excel® spreadsheet and prepared for statistical analysis using JMP statistical package (JMP Statistics and Graphics Guide, Release 7, Copyright© 2007, SAS Institute Inc., Cary NC, USA), and a paired t-test was conducted.

5.12.1 Piers Harris questionnaire

The results of the Piers Harris Self Concept Scale analysis are shown in table 5-7

Table 5-7: Summary of results of Piers Harris questionnaire for amelogenesis imperfecta patients

This table summarises the Self Concept Scale results of young patients with amelogenesis imperfecta and their matching (age and gender) controls of patient attending for routine dental care. A (*) indicates significance (P ≤0.05) (N=24).

Subclass	**Control Group (N=12)**		**Test Group (N=12)**		**Paired t- test**
	Mean	**Standard deviation**	**Mean**	**Standard deviation**	
Total scores	48	13.2	54	9.8	0.19
Behavioural adjustment	49.6	7.5	48.6	9.8	0.8
Intellectual and school status	44.5	9.8	54.5	7.5	0.02*
Physical appearance and attributes	47.5	11.7	53.8	8.7	0.18
Freedom from anxiety	48.2	11.9	52	10	0.35
Popularity	53.8	11.2	55.9	9.8	0.64
Happiness and satisfaction	45.4	9.8	50.4	5.1	0.12

5.12.2 Parental Perception Questionnaire (PPQ)

Table 5-8: Summary of results of Parental Perception Questionnaire of amelogenesis imperfecta patients

This table summarises the Parental Report Questionnaire results of young patients with amelogenesis imperfecta and their matching (age and gender) controls of patient attending for routine dental care. A (*) indicates significance (P ≤0.05) (N=24)

Subclass	**Test Group (N=12)**		**Control Group (N=12)**		**Paired t- test**
	Mean	**Standard deviation**	**Mean**	**Standard deviation**	
Total scores	33.8	25.9	17.3	9.6	0.04*
Oral symptoms	8.5	5.8	4.8	2.5	0.02*
Functional limitations	9.75	5.9	5.6	4.8	0.02*
Emotional well being	7.33	7.2	3	2.6	0.09
Social well being	8.25	8.4	3.75	2.4	0.09

5.12.3 Family Impact Scale (FIS)

Global Rating Items

The scores of the two global rating questions of the Family Impact Scale ("How would you rate the health of your child's teeth, lips, jaws and mouth?" with a five-point response format ranging from 'excellent' to 'poor' and "How much is your child's overall well-being affected by the condition of his/her teeth, lips, jaws or mouth affect your life overall?" with a response range from 'not at all' to 'very much') were calculated. Due to the small sample size and to ensure enough numbers for each response, data were collapsed for the first question to compare scores of the "poor" response to the collective scores of "Fair, Good, Very good, and Excellent". The same action was carried out for the second question comparing "Not at all" scores to the collective scores of "Very little, Some, A lot, and Very much". Statistical analysis was performed using Fisher's Exact Test. No statistically significant differences were noticed between the test and control groups for the two items ($P = 0.37$ and $P = 0.68$) respectively.

Table 5-9: Summary of results of Family impact scale of amelogenesis imperfecta patients

This table summarises the Family Impact Scale results of young patients with amelogenesis imperfecta and their matching (age and gender) controls of patient attending. A (*) indicates significance (P ≤0.05) (N=24)

Subclass	**Test Group (N=12)**		**Control Group (N=12)**		**Paired t-test**
	Mean	**Standard deviation**	**Mean**	**Standard deviation**	
Total Scores	9.6	9.6	5.58	3.6	0.15
Parental/family activity	3.3	3.3	2.5	1.7	0.45
Parental emotions	3.5	4.29	1.1	0.93	0.06
Family conflict	2.25	3	1	1.5	0.24
Financial burden	0.5	0.9	0.83	1	0.26

5.13 Discussion

This study attempted to objectively investigate the impacts that developmental dental disorders such as non-syndromic moderate to severe hypodontia and amelogenesis imperfecta can have on the self concept of affected individuals and how parents perceive the impact of these conditions on their children's quality of life (QoL) and on other family members. This patient population was compared to a control group of age and gender matched individuals without the condition, attending the Dublin Dental University Hospital for routine dental care.

As the age range for patients recruited was between 8-17 with very few subjects within the age range of 8-14, and the fact that self-perception oral health has been described to be age dependant (Barbosa et al., 2009), two of the four questionnaires comprising the Child Oral Health Related Quality of Life Questionnaire (COHRQoL) were not used. These were the Child Perception Questionnaire 8-10 (CPQ 8-10) and the Child Perception Questionnaire 11 to 14 (CPQ 11-14) as these questionnaires were specifically designed to be completed by children of these age groups (8-10 and 11-14) and it was thought that using these questionnaires for a different age group may be developmentally and cognitively inappropriate. It would also limit comparison with published research which used the COHRQoL questionnaires. However, the Parental Perception Questionnaire (PPQ) and the Family Impact Scale (FIS) questionnaires were used for all subjects under 18 years of age because parents completed these two questionnaires, and it is thought that parental reporting was not affected by the age of their children. It is accepted that this might make comparison with other studies in this field more challenging.

There is, as yet, no gold standard OHRQoL measure for children (McGrath et al., 2004). Measures such as CPQ and PPQ are useful for comparisons across populations but have limited ability to capture the effects of a particular condition because there is, as yet, no condition specific quality of life measure for the impact of different dental disorders including hypodontia and amelogenesis imperfecta on patients, parents and/or other family members (Cunningham et al., 2000). Hence a general oral health measure was used in this study and this included the Parental Perception Questionnaire, and the Family Impact Scale.

As discussed in the previous chapter, one common problem with OHRQoL questionnaires is the flooring effects of scores compared to the maximum scores, i.e. patients tend to score on the lower end of the scale. This results in low scores, which may sometimes approach zero. Lower score may give the impression that the person's QoL is not affected by the condition whereas the lower scores may be related to the presence of redundant general questions not directly related to the condition under investigation. A second problem with OHRQoL questionnaires is that the Minimal Important Difference (MID) has not been established. This is defined as "the smallest difference in score in the domain of interest which patients perceive as beneficial and which would mandate, in the absence of troublesome side effects and excessive cost, a change in the patient's management" (Jaeschke et al., 1989). This allows the clinical as well as the statistical significance of differences between groups to be better appreciated. The (MID) for the COHRQoL questionnaire remains to be identified as reported by the author who compiled it (Jokovic et al., 2003a).There is no available short version for the Parental Perception Questionnaire or for the Family Impact Scale, hence the decision was made to use these questionnaires in the study.

The overall results of this study cannot be compared with the scarce literature due to wide range of methodology, instruments used, and/or population investigated. At the time of designing this study there were no similar studies on which the sample size and selection for this study could be based. In one recent study (Laing et al., 2010) a sample size calculation was performed with data from the first 33 hypodontia patients and the first 10 routine orthodontic patients who acted as controls. The authors of the study stated that for the CPQ 11-14 and a chi square test (with a 0.05 level of significance) to have 80% power to detect a 25% difference in questionnaire scores, 61 subjects were required in each group (test and control). Although the number of patients recruited for this study is relatively small, the sample size was comparable with other studies in this field (Locker et al., 2005; Wong et al., 2006; Locker et al., 2010).

Three studies (Wong et al., 2005; Laing et al., 2010; Locker et al., 2010) have looked at the impact of hypodontia on young patients' quality of life, and the results presented were in conflict.

The first study compared 123 orthodontic patients with similar orthodontic needs with and without hypodontia (62 with hypodontia and 61 without hypodontia) (Laing et al.,

2010). Subjects were asked to complete the Child Perception Questionnaire (11-14), and to mark two visual analogue scales (VAS) for function and appearance. The functional VAS was anchored with "I find it easy to eat" and "I find it hard to eat". The appearance VAS was anchored with "I like the way my teeth look" and "I hate the way my teeth look".

The study concluded that there were no psychosocial differences between the CPQ scores of the two groups. However, it is unknown whether the impact experienced by both groups was high or low relative to children without a significant malocclusion. The study was well designed but did not quantify the number of missing teeth per patient. Instead the mean number of missing teeth was noted to be 4.52 (SD, 3.33). It would have been more meaningful if the severity of hypodontia was delineated (mild, moderate, or severe) as they may have highlighted differences with the severity of the condition as found in this study. The mean CPQ score for mild, moderate and severe hypodontia combined was 26.82 (SD, 16.4). One would question how orthodontic patients with and without hypodontia used in this study have "similar" orthodontic needs. The use of orthodontic patients without hypodontia as controls may have underestimated the impact of hypodontia on the quality of life of these patients, as routine orthodontic patients may have been equally psychologically affected by their dentition (Laing et al., 2010). Numerous cross-sectional studies have investigated the impact of functional and aesthetic components of malocclusion on quality of life of patients undergoing orthodontic therapy and concluded that malocclusion can significantly affect the quality of life of orthodontic patients and that malocclusion can lead to peer victimisation and bullying. (Kiyak et al., 1984; Helm et al., 1985; Hollister and Weintraub, 1993; Hawker and Boulton, 2000; Kumpulainen and Rasanen, 2000; Zhou et al., 2001; English et al., 2002; Owens et al., 2002; Onyeaso and Aderinokun, 2003; Klages et al., 2004). However, other studies have reported that technical aspects of malocclusion such as dissatisfaction with the ability to chew are less likely to impact on the quality of life among young adults than the more subjective aspects of self perception of dental appearance (Albino et al., 1981; Daniels and Richmond, 2000; de Oliveira and Sheiham, 2004).

In another study it was shown that severe hypodontia considerably impacts on OHRQoL although no control group was used (Wong et al., 2006). This "considerable

impact" was derived from a mean CPQ score of the severe hypodontia group of 29.0 (SD, 16.4) out of a potential maximum score of 148. If that study had used a control group to compare the results of the hypodontia group they might not have reached the same conclusions as a control group may have scored as high as their test group due to the general tendency to score at the lower end in these questionnaires (flooring effects).

In a recent study, the same design as used in the study by Wong et al was used and reported very high proportions of subjects identifying functional and psychosocial impacts as a result of congenitally missing teeth (Locker et al., 2010). The overall mean score of CPQ was 22.3 (SD= 14) out of a potential maximum score of 148. No control group was used in this study and the number of missing teeth ranged from one to fourteen, with 69% of the subjects having six or more missing teeth. It would have been more meaningful if the severity of hypodontia was more clearly described. As shown in this study, the merging of different degrees of hypodontia can mask important differences.

Compiling the results of the three studies above, it may have been possible that the impact of severe hypodontia was masked by the presence of mild/ moderate hypodontia as evidenced from the differences in mean scores and standard deviations of the three studies (mean 29.0 ±16.4; mean 26.82 ± 16.4; and mean 22.3 ± 14 respectively). This was a significant finding in the current study, and hence it was decided to investigate the combined (moderate and severe) hypodontia patients initially, and then investigate the groups separately.

5.13.1 Combined moderate and severe hypodontia versus controls

When the impact of combined moderate and severe hypodontia on the children's self concept was analysed it was found that hypodontia only affected the behavioural adjustment domain of the Piers Harris scale. This domain measures admission or denial of problematic behaviours. Even though the mean score (49.3) of the test groups was within the average range of this domain it is worth noting that patients with hypodontia scored less than that of their controls (55.7). Responders who score in the average range of the behaviour scale perceive themselves as fairly well behaved, but do acknowledge a few difficulties with their conduct. However, the overall levels of these problems are still considered to be within the normal limits proposed by the developers of this scale.

None of the other domains of this scale was significantly different but the overall scores of the test group were less than that of the control. The mean scores for the test and control were similar to the mean scores reported by a study investigating the relationship between normative (clinician measured) orthodontic treatment need (using the Index of Orthodontic Treatment Need), the child's self esteem (using Piers Harris Self concept Scale), and the Child Self Perceived Aesthetic need of young orthodontic patients (Mandall et al., 2001). No statistically significant correlation was found between the normative Index of Orthodontic Treatment Need (IOTN) grade and the Child self esteem as measure by Piers Harris scale.

A further study of young orthodontic patients investigated the self-concept of 208 patients aged between 7 and 15 years, with an increased overjet before and 15 months into treatment (Dann et al., 1995). These authors reported that children with class II malocclusion do not generally present for treatment with low self-concept and that the importance of self-concept as a reason for early treatment may have been exaggerated.

The findings of the present study concurred with the results of a study which showed, when measuring self concept, that patients with cleft lip and/or palate (with the exception of adolescents girls) did as well as, if not better than, a normal population (Leonard et al., 1991). However, no control group was used in that study. Instead the results were compared with the Piers Harris normative group described in the Piers Harris Self Concept Manual (Piers and Herzberg, 2007). The authors speculated that, it is possible that these children were minimising or denying the value of their interpersonal world in a way that allowed them to preserve their self-concept or the fact that their self-concept was a maladaptive response. This was maintained as a result of poor reality testing that compromised an accurate assessment of their functioning within a social context and undermined the formation of a healthier and more reality-based social environment (Leonard et al., 1991). Another explanation proposed is that these children may employ benign defenses by viewing themselves within a range of mild illness, rating their condition to the concept of headache, and perceiving their disability in a benign way that enables them to reduce the impact of stigmatisation, negative feedback, and social rejection (Clifford, 1967).

Results of the total scores of the Parental Perception Questionnaire revealed a statistically significant impact of hypodontia perceived by parents on the quality of life of their children, as compared to parents of non-affected individuals. Functional limitation domain was the only statistically significant domain of this scale (P = 0.01). This domain investigates the possible functional impairments caused by dental diseases on chewing, speaking, eating, drinking, breathing, food selection, and sleep disturbance. The means of the total and domain scores of this study are considerably higher than that of Do and co-workers, who investigated the Parental Perception Questionnaire for a general child/parent population (total score 25.9 Vs 11.6; oral symptoms 5.75 Vs 5.0; functional limitations 7.8 Vs 2.8; emotional wellbeing 5.65 Vs 2.3; Social wellbeing 6.65 Vs 1.5) (Do and Spencer, 2008). This indicated that the Parental Perception Questionnaire was able to discriminate parental perception of the general population and parents of patients with a specific dental condition. The overall scores and the domain scores showed substantial variability, with scores approaching a floor effect (Score= 0), while there was no ceiling effect (maximum score). Higher domain scores were noticed for functional limitation and social well-being. Our findings of parental perceptions were consistent with those reported by Jokovic where a population of paedodontic, orthodontic and orofacial patients were investigated to measure the agreement between parents and their children in reporting on quality of life (Jokovic et al., 2003a). A total of 42 pairs of mothers and children aged 11-14 years with oral and orofacial conditions completed the parental (PPQ) and child (CPQ11-14) components of the Child Oral Health Quality of Life Questionnaire. Agreement between the overall and subscale scores derived from the questionnaire were reviewed in comparative analysis to assess agreement at the level of the group (collective scores of parents and their children), and correlation analysis to assess agreement at the level of the individual family unit. The study found good agreement between parents and children at the group level in the overall scores, oral symptoms and functional limitation domains, which is explained by the concreteness and observability of the events addressed by these questions. A weaker correlation, which may reflect a real disagreement between mothers and children, was observed for the emotional and social wellbeing domains. This is related to the fact that these domains address events into which mothers do not have insight, as 11-14-year-old children are intensely peer-oriented, and the peer group dynamics is central to their lives (Jokovic et al., 2003a).

The authors concluded that children aged 11-14 years can give valid and reliable reports of their oral health-related quality of life and that mothers may be viewed as a suitable alternative to their children in assessing oral health related quality of life in surveys and clinical trails where groups are the unit of analysis. However, parents cannot be used as a proxy for assessment in clinical situations when the ratings for individuals are of concern. Zhang and co-workers, in an investigation of 71 child patients together with both their parents, reported that neither mothers nor fathers know their child's oral health status very well, as there was significant disagreement between mothers', fathers', and childrens' perceptions, at individual levels (Zhang et al., 2007a).

It would have been very interesting in this study to investigate the agreement and/or disagreement between parents and their children in reporting the impact of these conditions on the quality of life but due to the small sample size and the fact that the Child Perception Questionnaires were not used, no attempts were made to investigate this issue.

Analysis of the Family Impact Scale revealed that hypodontia did not seem to have any impact on parents or other family members. However, it did approach significance under the financial burden domain. This may not be related directly to the actual cost of treatment as this may have been covered by the Special Dental Needs Fund, but could be related to time taken off work, travel expenses, or maintenance treatment provided by a local dentist prior to accepting these patients under the Heath Service Executive's Special Dental Needs Scheme. The mean values of the total and domain scores for the family impact scale, for all hypodontia patients, were comparable to the mean scores of the paedodontic patients reported previously (Locker et al., 2002a), with the exception of parental emotions domain where the mean scores ranked above those for the orthodontic patients and less than for patients with oro-facial conditions. This did not change when the impact of severe and moderate hypodontia were investigated separately.

The means and the standard deviations of the total scores and the domain scores of this study were generally lower than those reported by Barbosa and colleagues in a study to evaluate the Brazilian Portuguese version of the Family Impact Scale in a Brazilian school children (Barbosa Tde and Gaviao, 2009). The Decayed, Missing and Filled

Teeth (DMFT) index (WHO, 1987) was used to assess caries status. Malocclusion was also scored using Dental Aesthetic Index (DAI) (Cons et al., 1986), which assessed the relative social acceptability of dental appearance by collecting and weighting data on 10-intraoral measurements. This enabled each individual to be placed on a dental appearance continuum, ranging from 13 (the most socially acceptable) to 100 (the least acceptable), and orthodontic treatment needs be prioritized based on the pre-defined categories of 'minor/none' (scores 13 to 25), 'definite' (26 to 31), 'severe' (32 to 35), or 'handicapping' (36 or more) (Estioko et al., 1994). These differing outcomes may be explained by the fact that meanings of QoL vary between and within individuals (Gregory et al., 2005) according to culture and education (Krause and Jay, 1994), contributing to distinct impacts of malocclusion on child QoL, and consequently on the functioning of the parents and the family as a whole. Krause investigated the responses of people from different races (White, Black, and Hispanic), age, gender, and educational background by interviewing and asking the participant one global health status item "if you were asked to describe your health, would you say that it was excellent, good, fair, or poor?". When a closed-ended response was selected, the study participants were asked the following open-ended probe: "Tell me why you say that?". He concluded that different people might interpret global health items in different ways. Some respondents think about specific health problems when asked to rate their health while other participants use health behaviour as a frame of reference. Younger people tend to use health status (i.e. mobility, overall physical condition) more often, whereas those who are older are more inclined to think in terms of health problems (i.e. hypertension). The same was true with respect to education, but the findings were more tentative (Krause and Jay, 1994). Gender differences, on the other hand, did not seem to affect the outcomes. One limitation in Krause's study was that the subjects were not selected randomly as it was a convenience sample, from ethnic groups in three different cities and deliberate attempts were made to recruit equal numbers of subjects from different race, gender and educational background. This may make generalisation of results to the broader population more challenging.

5.13.2 Severe hypodontia versus controls

The same domains that were statistically different in the combined moderate and severe hypodontia analysis (with the exception of the total scores of the PPQ) were also

significant when the results of severe hypodontia were investigated separately. The two domains, which were statistically different, were the behavioural adjustment domain of the Piers Harris Self Concept Scale and the functional limitation domain of the Parental Perception Questionnaire. These two domains have already been discussed above.

5.13.3 Moderate hypodontia versus controls

When the impact of moderate hypodontia alone was investigated, it was interesting to note that no statistically significantly different results were obtained in any of the domains of all questionnaires used, with only the mean values for the emotional wellbeing domain of the Parental Perception Questionnaire approaching significance (P = 0.06%). This clearly shows that parents of people with severe hypodontia perceive a greater impact of this condition on their child's quality of life than parents of children with moderate hypodontia. According to the findings presented it would be recommend that, when investigating the impact of hypodontia on the quality of life it may be prudent to separate moderate and severe hypodontia patients, in order not to mask the impact on quality of life. Another recommendation would be to investigate the presence or absence of retained primary teeth (relative versus absolute hypodontia) as it may be possible that the presence of retained primary teeth may reduce the impact of hypodontia on the quality of life of affected individuals. Wong et al have suggested the value of retaining primary teeth as they found a strong correlation (0.94) between the overall Child Perception Questionnaire scores when the primary teeth were accounted for, and a weak correlation (0.54) when primary teeth were not accounted for (Wong et al., 2006). However, this remains to be validated.

5.13.4 Amelogenesis Imperfecta

The rarity of AI makes it difficult to obtain large study groups. Current knowledge of the condition is therefore based on case reports and case series with small numbers of patients. There is only one systematic literature review which attempted to identify dental and orofacial anomalies described as being associated with AI, and to describe the impact of AI complications on patients' oral health-related quality of life, as well as economic consequences of this condition (Poulsen et al., 2008). The review concluded that the large majority of studies identified were case reports, which often emphasised restorative treatment and that other effects on the patients' life and qualify of life were

rarely described. Only one study was based on formal assessment of oral health-related quality of life for an adult population (Coffield et al., 2005). To date, the quality of life and self esteem of young amelogenesis imperfecta patients has not been investigated.

5.13.4.1 Piers Harris Self Concept Scale

When the impact of amelogenesis imperfecta on the children's self concept was analysed it was found that amelogenesis imperfecta only affected the intellectual and school status (INT) domain of the Piers Harris Self Concept Scale ($P = 0.02$).

It is of note that the mean scores of both groups (test and controls) were within the average range of this domain (40T to 55T) with the test group scoring at the lower end ($\mu = 44.5$) whereas the controls scored close to the upper end of the average range ($\mu = 54.5$). This domain reflects a child's assessment of his or her abilities with respect to intellectual and academic tasks. The items also cover general satisfaction with school and future expectations about achievement. Some items of this domain address how a child's intellectual capabilities affect key relationships (e.g., item 16, "I am an important member of my family"; item 26, "My friends like my ideas"). The item that is most strongly correlated with this domain's score, is item 5, "I am smart".

Children who score in the average range on INT scale view themselves as performing acceptably well in the academic realm, but acknowledge a few difficulties with school-related tasks. Examining the individual item responses can illuminate the nature of these problems. When the INT score is in the low average range (40 to 44T), the youngster is admitting to more difficulties with academic work than the typical individual in the standardisation sample, but the overall level of these problems is still considered to be within normal limits.

It was of interest to note that the mean scores of patients affected with amelogenesis imperfecta were generally lower than those of their controls but they were all within the "average range" of the corresponding domains. This may indicate that Piers Harris Scale was able to identify differences between the test and control groups but the relatively small sample size may have hindered its ability to show any statistically significant differences, or the questionnaire is not sufficiently sensitive to identify subtle differences between groups.

5.13.4.2 Parental Report Questionnaire

Results of the parental report questionnaire scores revealed a statistically significant impact of amelogenesis imperfecta, as perceived by parents/caregivers, on the quality of life of their children as compared to parents of non-affected individuals. The perceived impact of amelogenesis imperfecta on children's quality of life was evident by the statistically significant differences noted under total scores (P = 0.04), oral symptoms (P = 0.02), and functional limitations (P = 0.02) domains.

The oral symptoms domain consisted of six questions and evaluated the frequency of the following events in the last 3 months: food caught between teeth, pain in teeth/mouth, bad breath, mouth sores, bleeding gums and food stuck to the roof of the mouth (not applicable to AI).

The functional limitations domain consisted of eight questions and measured the frequency of the following events in the last three months: difficulty chewing firm foods, unclear speech, difficulty drinking/eating hot/ cold foods, difficulty eating foods would like to eat, slow eating, breathing through mouth (not applicable to AI), restricted diet, and trouble sleeping.

These results may reflect the fact that parents are more concerned about their children's quality of life as some parents also have amelogenesis imperfecta, and consequently have a better understanding of the efforts, involved in treatment. The impact of amelogenesis imperfecta on quality of life may become more of a concern to patients as they grow older as was evident from its impact on the adult population observed in the previous chapter. This can be explained by the increased awareness of self-concept and body image. Symptoms may also become more evident and function may be more affected with time.

5.13.4.3 Family Impact Scale (FIS)

Contrary to what one would anticipate, analysis of the Family Impact Scale scores revealed that amelogenesis imperfecta did not seem to have any impact on parents or other family members.

A possible explanation to this finding is that the interdisciplinary collaborative approach and early management of the newly diagnosed cases, in addition to available state funding for their complex treatment has begun to reduce the impact experienced

by families of affected patients. It would be interesting to investigate the impact of amelogenesis imperfecta on patients before receiving any treatment (at presentation) and following dental intervention in future studies.

These findings may also be related to the small sample size or the lack of sensitivity of this particular scale to explore the impact of amelogenesis imperfecta on the families of affected individuals.

Of note in this study is that the mean scores of amelogenesis imperfecta patients were generally lower (in case of Piers Harris Questionnaire) and higher (in case of Parental Perception and Family Impact Scale questionnaires), than those of hypodontia patients, indicating a lower self esteem and a greater impact on quality of life (Table 5-10).

Table 5-10: The (mean) total scores for the four groups tested.

Questionnaire (Total Scores)	All hypodontia	Severe hypodontia	Moderate hypodontia	AI
Piers Harris	54.3	57	48.5	48
Parental perception Questionnaire	27.4	25.9	30.7	33.8
Family Impact Scale	6.3	5.75	7.5	9.6

This may be explained by the fact that amelogenesis imperfecta is likely to be more symptomatic (i.e. tooth sensitivity, discolouration, open bite, gingivitis, worn dentition, poor oral hygiene and infections) (Poulsen et al., 2008) than hypodontia, which may not cause any symptoms until late in life when the affects of missing teeth (i.e. spacing, and difficulty chewing) become more evident as primary teeth are lost.

5.14 Conclusion

The impact of moderate to severe hypodontia and amelogenesis imperfecta on young patients' self esteem and on their quality of life as perceived by their parents has been investigated. Within the limitations of this study and taking all the results into account, developmental dental disorders such as moderate/severe hypodontia and amelogenesis imperfecta may not have a detrimental impact on young patients' self esteem. However, when the quality of life of this young population was investigated using their parents as proxies, it was evident that developmental dental disorders have a significant impact on this patient population. Amelogenesis imperfecta appears to have a greater impact on young patients' quality of life than hypodontia, and severe hypodontia has a marginally greater impact than moderate hypodontia.

Chapter 6
General Discussion

Chapter 6: General Discussion

6.1 Database establishment and analysis

In this thesis a cohort of patients attending the Dublin Dental University Hospital and affected by developmental dental disorders such as hypodontia and amelogenesis imperfecta were investigated.

Prior to 2001, there was no co-ordinated approach to treatment of this patient population in the Republic of Ireland and the provision of care was dependant on the region in which the patient was resident.

In December 2001, a Special Dental Needs Clinic was established and funded by the Department of Health and Children to treat this patient population and, since the establishment of this clinic, more patients with developmental dental disorders have been referred to this service.

The Special Dental Needs Clinic is a tertiary multidisciplinary referral clinic, which provides care for patients with developmental dental disorders in a number of ways, including: consultation, treatment planning and multidisciplinary dental treatment. The Special Dental Needs Clinic accepts referrals from the Health Service Executive, general dental practitioners, general medical practitioners, general hospitals, and, on some occasions, self-referrals of affected family members of special dental needs patients.

In 2006 due to the increased number of referrals from different parts of the country and the lack of a national register for patients with developmental dental disorders, a decision was made to develop an Irish National Database of Patients with Developmental Dental Disorders. The proposed aims of this national registry were as follows:

1. To establish an Irish database for people affected by Hypodontia, AI and DI in order to identify the prevalence, frequency, severity and geographical distribution of these developmental dental disorders.
2. To investigate the impact of developmental dental disorders on patients' self-image, and the quality of life of affected individuals and their families.

3. To examine, by way of future studies, whether patients affected by any of these dentofacial anomalies have the same genetic mutation within the same family and across families. This will involve the following:
    - Delineating the primary genetic defects, which are responsible for hypodontia, amelogenesis and dentinogenesis Imperfecta.
    - Characterising the phenotypes and phenotypic variability for these disorders in an Irish population.
    - Defining the genetic loci for Hypodontia, AI and DI traits and to identify the associated genes and gene mutations.

In 2009, a web-based database was established for patients with developmental dental disorders and is hosted in the Dublin Dental University Hospital. The database contains data of 502 patients affected by hypodontia, amelogenesis imperfecta, and dentinogenesis imperfecta. The database is maintained and updated by staff of the Division of Restorative Dentistry and Periodontology.

The database was developed professionally and is a robust and extremely flexible tool that allows compilation and searching of data in multiple ways. It was specifically designed to facilitate searching for patterns of hypodontia with the maximum number of fields searched simultaneously. A facility to automatically identify patterns of hypodontia was also established.

Hypodontia was the most common developmental dental disorder referred to the Special Dental Needs Clinic since its inception in 2001. By definition, this patient population is biased and does not reflect the prevalence and severity of hypodontia for the Irish population. The findings should be interpreted bearing this limitation in mind.

The majority of referrals came from the Health Service Executive Dental Section and were from the greater Dublin area. This may reflect the density of the population in this area as 50.6% of the population live in the Dublin Mid-Leinster and Dublin North East areas, whereas 25.5% live in the South and 24% live in the West of the Country (CSOI, 2006).

The age range was 7-50 years, with a median age of 19 years. The frequency and distribution of tooth agenesis is similar to that reported by other workers (Castaldi et al., 1966; Rose, 1966; Wisth et al., 1974a; Rolling, 1980; Hobkirk et al., 1994); However, a

slight increase in the prevalence of missing maxillary second premolars as compared to maxillary lateral incisors, was noticed. This may be attributed to the fact that the majority of these patients were referred because of moderate to severe hypodontia. Hence, mild hypodontia cases, where there were no additional features like microdontia, impactions, or failure of eruption, were not referred but treated locally. It is worth noting that a similar pattern of tooth agenesis is maintained when moderate and severe hypodontia cases where plotted separately as evident from figures (3-4 and 3-5). The most frequent number of missing teeth per individual patient was six, followed by four missing teeth per patient.

6.2 Impact on quality of life of adult hypodontia and amelogenesis imperfecta

Given the tremendous emphasis society places on facial aesthetics, health care providers have come to question what effects hereditary developmental defects of teeth have on a person's psychosocial health. It is essential these providers understand how dental health and appearance fit into the larger picture of overall health and well-being. Yet, to date, there are no Irish data on the psychosocial influence of severe hereditary dental defects. This study was undertaken to provide information regarding the importance of teeth and oral aesthetics in normal psychosocial development.

This study provided additional data and objectively characterised the functional and psychosocial impacts that developmental dental disorders have on the affected individuals, in terms of the perceived quality of life and self-esteem. This patient population was compared to a control group of matching age and gender of individuals without the condition attending the Dublin Dental University Hospital for routine dental care.

6.2.1 Hypodontia and amelogenesis imperfecta versus their controls

6.2.1.1 Oral Health Impact Profile (OHIP-49)

Upon examination of the impact of moderate and severe hypodontia and AI on quality of life of patients affected by these conditions it was evident that these two conditions had a significant impact on patients' quality of life under the main domains of the Oral Health Impact Profile Questionnaire.

For hypodontia patients, the total scores, functional limitation, physical pain, psychological discomfort, physical disability, psychological disability, and social disability were all significantly different from age and gender matched controls. Only the handicap domain was not significantly different between the test and control groups, which may indicate that hypodontia does not cause sufficient impairment to the extent that the patient is handicapped physically, psychologically or socially. The overall mean responses of the total scores of affected patients were higher than that for patients without the condition and the differences were statistically significant.

For amelogenesis imperfecta patients, the condition had a significant impact on adult patients' quality of life under five of the seven domains of the oral health impact profile questionnaire, which included: total scores, psychological discomfort, physical disability, psychological disability, and social disability. The overall mean responses of the total scores of affected patients were higher than that for patients without the condition and differences were statistically significant.

Comparing the mean scores between amelogenesis imperfecta and combined moderate and severe hypodontia patients of this study revealed that amelogenesis imperfecta had a far greater impact than hypodontia. However, the mean score of amelogenesis imperfecta was comparable with that of severe hypodontia, when patients with moderate hypodontia were excluded.

6.2.1.2 Rosenberg Self-Esteem Scale

When examining the total scores of the Rosenberg self-esteem scale for hypodontia and amelogenesis imperfecta patients, there were no statistically significant differences between the test and control groups. The mean scores of the test group for AI patients was slightly lower than that of the control group. The mean scores for the hypodontia patients were marginally greater but not significantly different from those of their controls.

These results may be explained by the fact that hypodontia and AI may not have an impact on patients' self-esteem or the fact that the Rosenberg Self Esteem Scale may not be sensitive enough to changes in dental patients' self esteem as it seems to identify differences only in cases involving severe facial disfigurement. This highlights the need

for developing condition specific instruments capable of measuring the impact of dental conditions on patient's self esteem. To date there is a paucity of research in this field.

6.2.1.3 Severe hypodontia versus controls

When only the impact of severe hypodontia on quality of life for hypodontia patients was investigated, it was clearly evident that the condition had a significant impact on adult patients' quality of life under all the domains of the oral health impact profile questionnaire which included: total scores, functional limitations, physical pain, psychological discomfort, physical disability, psychological disability, social disability and handicap. The handicap domain was not statistically significantly different when moderate and severe hypodontia were combined together. The impact of hypodontia on the quality of life of affected individuals was masked when both moderate and severe hypodontia patients were combined together. However, this did not alter the self esteem results when they were investigated separately.

6.2.1.4 Moderate hypodontia versus controls

When only the impact of moderate hypodontia was investigated, it was interesting to note that no statistically significant results were obtained in any of the domains of the two questionnaires used (OHIP-49 and Rosenberg Self Esteem Scale).

The site specific pattern of tooth agenesis is almost the same when the results of combined, severe, and moderate hypodontia cases were plotted separately. Hence, the impact of hypodontia on the quality of life may not be solely related to the agenesis of a specific tooth type (anterior or posterior) but may be directly related to the severity of the condition (increased number of developmentally missing teeth) as shown from this study. This conflicts with the findings of two recently published qualitative researches, which reported that aesthetics were prime concerns of patients affected with hypodontia. These two studies investigated the issues of importance for patients affected with developmental dental disorders using focus groups of patients with hypodontia (Akram et al., 2011; Meaney et al., 2011). The two studies interviewed patients affected with hypodontia in small focus groups to identify any emerging themes until saturation (when no new themes were emerging from the participant's responses) was achieved. In the first study, the themes included: treatment, appearance, activities, and other people's reaction (Akram et al., 2011). Whereas those of the second

study included: transition from childhood to adulthood, waiting time for treatment, and importance of aesthetics (Meaney et al., 2011). Research investigating and comparing the impact of mild, moderate and severe hypodontia on quality of life is lacking.

6.3 Impact of hypodontia and amelogenesis imperfecta on quality of life of young patients

This study attempted to objectively investigate the impacts that developmental dental disorders, such as non-syndromic moderate to severe hypodontia and amelogenesis imperfecta, had on the self concept of affected individuals and how parents perceive the impact of these conditions on their children's QoL and on other family members. This patient population was compared to a control group of age and gender matched individuals without the condition attending the Dublin Dental University Hospital for routine dental care.

There is, as yet, no gold standard OHRQoL measure for children (McGrath et al., 2004). Measures such as CPQ and PPQ are useful for comparisons across populations but have limited ability to capture the effects of a particular condition because there is, as yet, no condition specific quality of life measure for the impact of different dental disorders including hypodontia and amelogenesis imperfecta on patients, parents and/or other family members (Cunningham et al., 2000). Hence a general oral health measure was used in this study which included the Parental Perception Questionnaire, and the Family Impact Scale.

The overall results of this study cannot thus be compared with the published literature due to the different methodology, instruments used, and/or population investigated.

6.3.1 Combined severe and moderate hypodontia versus controls

When the impact of combined moderate and severe hypodontia on the children's self concept was analysed it was found that hypodontia only affected the behavioural adjustment domain of this scale. This domain measured admission or denial of problematic behaviours. Even though the mean score (49.3) of the test groups was within the average range of this domain it is worth noting that patients with hypodontia scored less than that of their controls (55.7). However, the overall levels of these problems are still considered to be within normal limits proposed by the developers of

this scale. None of the other domains of this scale was significantly different but the overall scores of the test group were less than that of the control group.

Results of the total scores of the Parental Perception Questionnaire revealed a statistically significant impact of hypodontia perceived by parents on the quality of life of their children as compared to parents of non-affected individuals. The functional limitation domain was the only statistically significant area observed for this scale. This indicated that the Parental Perception Questionnaire was able to discriminate between general population and parents of patients with a specific dental condition. The overall scores and the domain scores showed substantial variability, with scores approaching a floor effect (Score= 0), while there was no ceiling effect (maximum score). Higher domain scores were noticed for functional limitation and social well-being.

Analysis of the Family Impact Scale revealed that hypodontia did not seem to have any impact on parents or other family members. However, it did approach significance under the financial burden domain. This may not be related directly to the actual cost of treatment as this may have been covered by the Health Service Executive Special Dental Needs Fund, but could be related to time taken off work, travel expenses, or maintenance treatment provided by a local dentist prior to accepting these patients under the Special Dental Needs Scheme.

6.3.2 Severe hypodontia versus controls

No marked differences between the severe hypodontia group and controls were identified when questionnaires were analysed, with the exception of the functional limitation domain of Parental Perception Questionnaire of the severe hypodontia group, which was significantly different between the test and control groups, as one would expect more missing teeth would have an impact on dental function.

6.3.3 Moderate hypodontia versus controls

When the impact of moderate only hypodontia was investigated, it was interesting to note that no statistically significant results were obtained in any of the domains of all questionnaires used. However, the emotional wellbeing domain of the Parental Perception Questionnaire approached significance. This could be attributed to a

difference in reporting the impact of the condition between parents of the moderate and severe hypodontia groups.

A second explanation could be related to a sampling error indicating that the sample investigated may not be a true representation of the entire hypodontia population. Increasing the sample size could have minimised this error but it was not feasible at the time the research was conducted.

6.3.4 Amelogenesis Imperfecta

The rarity of AI makes it difficult to obtain large study groups. Current knowledge of the disease is therefore based on case reports and case series with small numbers of patients. Only one comparable study was identified in the literature (Poulsen et al., 2008).

6.3.4.1 Piers Harris Self Concept Scale

When the impact of amelogenesis imperfecta on the children's self concept was analysed it was found that amelogenesis imperfecta only affected the intellectual and school status (INT) domain of the Piers Harris Self Concept Scale ($P = 0.02$).

It is worth mentioning that the mean scores of both groups (test and controls) were within the average range of this domain (40T to 55T) with the test group scoring at the lower end ($\mu = 44.5$) whereas the controls scored close to the upper end of the average range ($\mu = 54.5$).

It was of interest to note that the mean scores from patients affected with amelogenesis imperfecta were generally lower than those of their controls but they were all within the "average range" of the corresponding domains. This may indicate that Piers Harris Scale was able to identify differences between the test and control groups but the relatively small sample size may have hindered its ability to reveal any statistically significant differences.

6.3.4.2 Parental Report Questionnaire

The perceived impact of amelogenesis imperfecta on children's quality of life was evident from the statistically significant differences noted under total scores ($P = 0.04$), oral symptoms ($P = 0.02$), and functional limitations ($P = 0.02$) domains. It is not clear from this study whether the results obtained reflect the parental perception of their

child's condition or the fact that some parents were also affected by amelogenesis imperfecta. The impact of amelogenesis imperfecta on quality of life may become more of a concern to patients as they grow older as was evident from the impact of AI on adult population. This can be explained by the increased awareness of self-concept and body image. Symptoms may also become more evident and function may be more affected with time.

6.3.4.3 Family Impact Scale (FIS)

Analysis of the Family Impact Scale scores revealed that amelogenesis imperfecta did not seem to have any impact on parents or other family members. This is may be explained by the lack of sensitivity of this particular scale to explore the impact of amelogenesis imperfecta on the families of affected individuals or it may be directly related to the small sample size. A Third possible explanation to this finding is that the interdisciplinary collaborative approach and early management of the newly diagnosed cases in addition to available funding of their complex treatment has actually started to reduce the impact experienced by families of affected patients.

It is worth mentioning that in this study the mean scores of amelogenesis imperfecta patients were generally lower (in case of Piers Harris Questionnaire) and higher (in case of Parental Perception and Family Impact Scale questionnaires) than those of hypodontia patients indicating a lower self esteem and a greater impact on quality of life. This may be explained by the fact that amelogenesis imperfecta is likely to be more symptomatic than hypodontia which may not cause any symptoms until late in life were the effects of missing teeth (i.e. spacing, and difficulty chewing) become more evident (Poulsen et al., 2008).

Chapter 7
General Conclusions

Chapter 7: General Conclusion

This thesis discussed the development and analysis of the first Irish National Special Dental Needs Database. This web-based database was developed in collaboration with all disciplines involved in providing patient care in the Special Dental Needs Clinic. The database is hosted in Dublin Dental University Hospital and maintained by staff in the Division of Restorative Dentistry and Periodontology. The database aims to store all the relevant clinical and biographical information for this patient population in order to identify the prevalence and geographical distribution of these developmental dental disorders, to assist in allocation of services and outline the resources needed to optimise the management of these conditions. The database facilitates future research to further investigate these developmental dental disorders.

The database was interrogated to extract data related to hypodontia patients and two studies were completed to investigate the impact of developmental dental disorders such as hypodontia and amelogenesis imperfecta on quality of life, patient self esteem, and the parental perception.

The overall conclusions of this research can be summarised as following:

- Hypodontia is the most common non-syndromic developmental dental disorder affecting teeth in this referred population. The range of missing teeth was 2-26 and the mandibular second premolar was the most frequently absent tooth type.
- Developmental dental disorders appear to have a great impact on the quality of life of this patient population under most QoL domains. The psychosocial impact of severe hypodontia on the quality of life of affected patients is greater than that experienced by patients affected with moderate hypodontia, and resembles the impact of amelogenesis imperfecta.
- Hypodontia and amelogenesis imperfecta did not seem to affect the self esteem of this patient population when compared with controls. This finding is consistent with the findings of previous research in this field.
- When the impact of moderate to severe hypodontia and amelogenesis imperfecta on young patients' self esteem and on their quality of life, as perceived by their parents, was investigated, developmental dental disorders such as moderate/severe hypodontia and amelogenesis imperfecta may not have

a detrimental impact on young patients' self esteem. However, when the quality of life of this young population was investigated using their parents as proxies, it was evident that developmental dental disorders had a remarkable impact on this patient population. Amelogenesis imperfecta appears to have a greater impact on young patients' quality of life than hypodontia, and severe hypodontia has a marginally greater impact than moderate hypodontia.

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Publications

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Tooth agenesis in patients referred to an Irish tertiary care clinic for the developmental dental disorders

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Précis

Hypodontia is the most common developmental disorder affecting teeth, and the mandibular second premolar is the most frequently absent tooth type.

Abstract

Purpose: This study was carried out to determine the prevalence, severity and pattern of hypodontia in Irish patients referred to a tertiary care clinic for developmental dental disorders.

Materials and methods: Details of 168 patients with hypodontia referred during the period 2002-2006 were entered in a database designed as a national record. Tooth charting was completed using clinical and radiographic examinations. The age of patients ranged from 7-50 years, with a median age of 20 years (Mean: 21.79; SD: 8.005).

Results: Hypodontia referrals constituted 65.5% of the total referrals. Females were more commonly affected than males with a ratio of 1.3:1. The number of referrals reflected the population density in this area; the majority were referrals from the public dental service. Mandibular second premolars were the most commonly missing teeth, followed by maxillary second premolars and maxillary lateral incisors; maxillary central incisors were the least affected. Symmetry of tooth agenesis between the right and left sides was an evident feature. Slightly more teeth were missing on the left side (n = 725) than on the right side (n = 706) and in the maxillary arch (n = 768) as compared to the mandibular arch (n = 663). Some 54% of patients had severe hypodontia with more than six teeth missing; 32% had moderate hypodontia, with four to six teeth missing. The most common pattern of tooth agenesis was four missing teeth.

Conclusion: Hypodontia was a common presentation in a population referred to this tertiary care clinic. The pattern and distribution of tooth agenesis in Irish patients appears to follow the patterns reported in the literature.

Introduction

Hypodontia is the term used to describe the developmental absence of one or more primary or secondary teeth, excluding the third molars. It is the most common developmental dental anomaly and can be challenging to manage clinically.[1] Generally, hypodontia refers to the condition where there is absence of one or a few teeth only. Oligodontia is the term usually used to describe six or more missing teeth, and anodontia is the complete absence of teeth (**Figures 1 and 2**).

Hypodontia can also be classified, according to the severity of the condition, as:

- mild: one to three teeth developmentally missing;
- moderate: four to six teeth developmentally missing; and,
- severe: more than six teeth developmentally missing.

Hypodontia can affect both the primary and permanent dentition. It is rare in the primary dentition, with a prevalence of less than 1%

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FIGURE 1: Clinical photograph of a 21-year-old male with severe hypodontia. He is missing 15, 14, 12, 22, 23, 24, 25, 35, 34, 32, 41 and 42 (12 in total). He also has over-retained and submerged primary molars, and a transposition of teeth 44 and 43.

FIGURE 2: Orthopantomogram of the same patient.

in Caucasians. When it does occur in the primary dentition, it most commonly involves the mandibular incisors.[2]

Aetiology

Developmental absence of teeth is a consequence of:

- physical obstruction or disruption of the dental lamina;
- space limitation;
- functional abnormalities of the dental epithelium; or,
- failure of initiation of the underlying mesenchyme.[3]

Hypodontia may arise as a familial condition, with a high proportion of affected individuals coming from families with a previous history of the condition.[4] The nature of the inheritance is complex and not well understood, but it is thought to be related to more than one gene.[5,6] Hypodontia may also arise in individuals with no family history.

A number of systemic conditions, such as hypohidrotic ectodermal dysplasia, Down syndrome and chondroectodermal dysplasia have hypodontia as a feature. The developmental disruption due to the presence of a cleft lip and palate involving the alveolus may also result in an absence of teeth in that region, notably the maxillary lateral incisors.[7,8]

Teeth develop as appendages of the embryonic surface epithelium. The most important events during regulation of the development of all such organs are the so-called inductive interactions.[6] Signal molecules of several different families are used sequentially during the advancing development, and reciprocally from epithelium to mesenchyme and vice versa. Signalling interactions that determine the location, identity, size, and shape of teeth take place during the early stages of tooth development. The first signals are secreted by the oral ectoderm, which initiates the odontogenic programme in the underlying neural crest-derived mesenchyme. The committed mesenchyme signals back to the epithelium and controls the growth and folding of the epithelium. The mesenchymal signals also induce the formation of signalling centres in the epithelium, in which many genes encoding signal molecules are activated. These centres signal back to mesenchyme, as well as within the epithelium, and regulate the advancing development, including cusp development in molars. Numerous transcription factors have been identified, which are turned on in the target tissues as a result of signalling.

Prevalence

Studies assessing hypodontia vary widely in their reports of prevalence in the permanent dentition, as can be seen in **Table 1**. The majority of these studies report prevalence rates varying from 2.6% in Saudi Arabia[9] to 11.3% in Ireland.[10] Studies in the United Kingdom suggest a prevalence rate of 4-4.5%.[11,12] Some of these studies are biased because of the nature of the population studied, i.e., orthodontic patients, and hence these figures cannot be generalised for the whole population.

Management

The developmental absence of teeth can seriously disable a young person, both physically and emotionally, especially during the turbulent years of adolescence. Early management is indicated, yet there are reports of patients with hypodontia being referred late with all that that implies – treatment disrupting the examination years and reluctance to wear appliances because of the impact on the young person's social life.[13]

There is much to be gained from the interdisciplinary management of young people who have hypodontia. Many patients are looked after by multidisciplinary teams, and each clinician does their own treatment in isolation. True interdisciplinary working involves the close working of a committed team where each member contributes their expertise to achieve an optimum outcome for the patient and their family.[3,13,14]

Table 1: Previous hypodontia prevalence studies.[1]

Country	Author	Population type	Age range (yr)	Number of patients	Prevalence (%)	Most frequently absent tooth
Malaysia	Nik-Hussein, 1989	Children attending the dental hospital	6-15	1,583	2.8%	Maxillary lateral Incisor
Saudi Arabia	Salama and Abdel-Megid, 1994	Children attending the dental hospital.	5-10	1,300	2.6%	Mandibular second premolar
Australia	Lynham, 1990	Australian defence force recruits	16-26	662	6.3%	Maxillary lateral incisor
Norway	Aasheim and Ogaard, 1993	Schoolchildren	7.8-10.4	1,953	6.5%	Mandibular second premolar
Iceland	Johannsdottir *et al*, 1997	Schoolchildren	6	396	5%	Mandibular second premolar
Denmark	Rolling, 1980	Schoolchildren	9-10	3,325	7.8%	Maxillary second premolar
Hong Kong	Davis, 1987	Schoolchildren	12	1,093	6.9%	Mandibular incisor
Ireland	O'Dowling and McNamara, 1990	Orthodontic patients	7-17	3,056	11.3%	Mandibular second premolar
England	Rose, 1966	Orthodontic patients	7-14	6,000	4.3%	Mandibular second premolar
England	Brook, 1974	Nursery and schoolchildren	3-5 & 11-14	958 & 1,183	4.4%	Mandibular second premolar
United States	Muller *et al*, 1970	School students	11-15	14,940	3.5%	Mandibular second premolar
Sweden	Bergstrom, 1977	Schoolchildren	8-9	2,589	7.4%	Mandibular second premolar

Background

Prior to 2001 the treatment of patients with developmental dental disorders in Ireland depended on the region where the patient was resident and was not co-ordinated. In December 2001 a Special Dental Needs Restorative Dentistry Clinic was established and funded by the Department of Health and Children to treat this patient population.

The purpose of this study is to identify the prevalence, frequency, severity, and geographic distribution of hypodontia cases referred to a tertiary care centre for developmental dental disorders, where an interdisciplinary approach for the management of such cases is currently available.

Materials and methods

The details of 168 patients with hypodontia referred to the Special Dental Needs Clinic in the Dublin Dental School and Hospital were entered into an Access Microsoft database designed as a national record for patients with developmental dental disorders, which included hypodontia, amelogenesis imperfecta, and dentinogenesis imperfecta. Ethical approval was obtained from the Faculty Research Committee in Trinity College Dublin and informed consent obtained from each patient or parent. Teeth charting was completed using both clinical examination and orthopantomographs. All data were entered by one investigator.

The Special Dental Needs Clinic is specifically for the treatment of patients with developmental craniofacial and dental anomalies. The main categories include:

1. Dental anomalies:
- moderate and severe developmental hypodontia;
- amelogenesis imperfecta;
- dentinogenesis imperfecta;
- microdontia/macrodontia (anomalies of tooth structure, size or eruption); and,
- failures of eruption.

2. Developmental disorders with associated craniofacial/dental anomalies:
- osteogenesis imperfecta;
- epidermolysis bullosa – recessive dystrophic type;
- ectodermal dysplasias; and,
- cleidocranial dysplasia.

Results

Referred cases

Of all patients referred to the clinic, hypodontia cases constituted 65.5% of referrals, amelogenesis imperfecta cases represented 28.5%, and dentinogenesis imperfecta cases represented 6% of the total referrals. This paper will only describe the hypodontia population.

Sex distribution

Females were more affected than males with a ratio of approximately 1.3:1.

Source of referrals

Most referrals were from the Dublin North East and Dublin Mid-Leinster areas, with fewer referrals coming from the west and south of the country (**Figure 3**). The majority of referrals were from the Health Service Executive (previously known as the Health Board Dental Service), followed by referrals from general dental practitioners (**Figure 4**). Very few cases had been self-referred or referred by a general medical practitioner.

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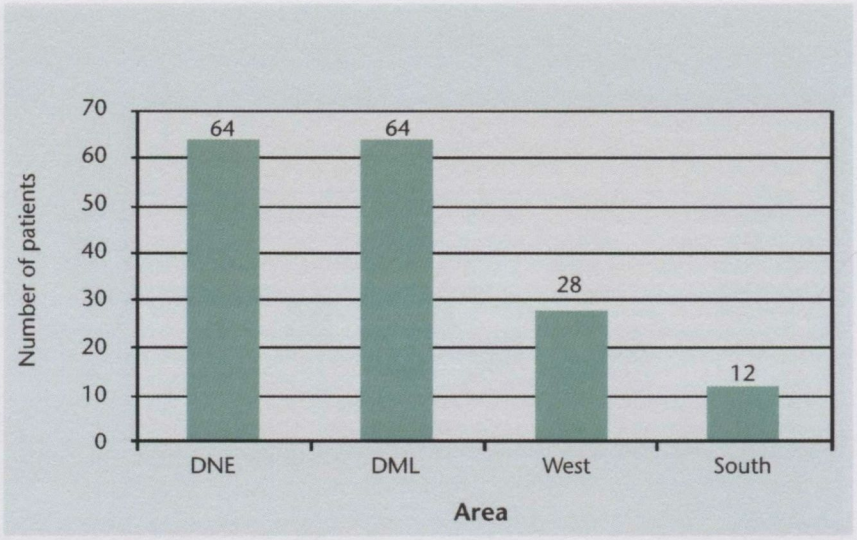

FIGURE 3: Geographic distribution of total referrals of hypodontia cases. DNE: Dublin North-East; DML: Dublin Mid-Leinster; West: West of Ireland; South: South of Ireland.

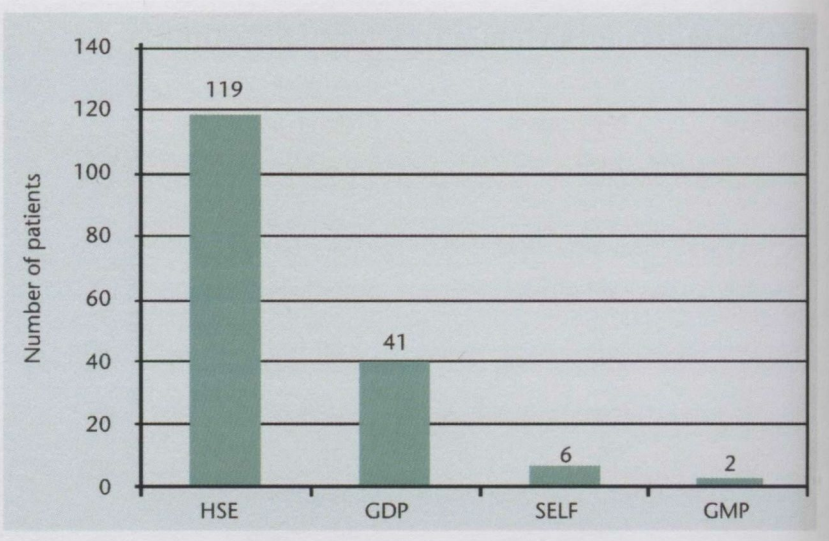

FIGURE 4: Referral sources of hypodontia cases. HSE: Health Service Executive; GDP: general dental practitioner; GMP: general medical practitioner.

Pattern of tooth agenesis

The mandibular second premolars showed the highest frequency of tooth agenesis, representing 15% of the total number of missing teeth, followed by the maxillary second premolars (14.4%) and the lateral incisors (13%). The distribution of tooth agenesis is shown in **Figure 5**. The total number of missing teeth for all patients was 1,431, with a range of two to 26. Symmetry of tooth agenesis of the right and left sides was a feature in individual patients; however, more teeth were missing on the left side (n = 725) as compared to the right side (n = 706), and in the maxillary arch (n = 768) as compared to the mandibular arch (n = 663). The most common pattern of tooth agenesis per patient was four missing teeth, followed by two missing teeth per patient. A total of 91 patients (54%) had severe hypodontia, where more than six teeth were developmentally missing, and 54 patients (32%) had moderate hypodontia, where four to six teeth were developmentally missing.

Discussion

Hypodontia was the most common developmental dental disorder referred to the Special Dental Needs Clinic since its inception in 2001. By definition, this patient population is biased and does not reflect the prevalence and severity of hypodontia for the Irish population. The results should be interpreted bearing this limitation in mind.

In agreement with other studies,[10-13,16-18] females were more commonly affected than males; this may suggest a referral bias, as females are more likely to seek dental treatment than their male counterparts.

The majority of referrals came from the Health Service Executive and were from in and around the Dublin area. This may reflect the density of the population in this area as 50.6% of the population lives in the Dublin Mid-Leinster and Dublin North East areas, whereas 25.5% lives in the south and 24% lives in the west of the country.[15]

The age range was seven to 50 years, with a median age of 20 years (mean: 21.79; SD: 8.005). To exclude other causes of tooth loss for older patients, clinical notes and referral letters were checked carefully, and compared with previous radiographs available in the charts. While recognition of the younger patients with hypodontia results either from chance observation or a positive family history, it might be expected that the majority of cases would be identified in the mixed dentition phase. The median age of this patient population is 20 years; this may suggest either poor recognition by dental practitioners, or low levels of patient demand due to the prohibitively expensive cost of treatment. For these cases, it is likely that there were difficulties in locating an appropriate clinic to which a referral for advice or treatment would be made. A second possible reason is that the database was established by the Division of Restorative Dentistry and Periodontology, which traditionally provides restorative care at the end of the treatment plan, as compared to the interdisciplinary approach that has been implemented recently. Patients are now being added to the database as soon as their diagnosis is confirmed, often on referral from paediatric dentistry, community and orthodontic clinics, and a treatment plan drawn up in interdisciplinary clinics.

The frequency and distribution of tooth agenesis (as shown in **Figure 5**) is similar to that reported by other workers;[12,13,16-18] however, a slight increase in the prevalence of missing maxillary second premolars as compared to maxillary lateral incisors was noticed. This may be attributed to the fact that the majority of these patients are referred because of moderate to severe hypodontia and that mild hypodontia cases, where there were no additional features like microdontia, impactions, or failure of eruption, were not referred but treated locally. Our results were also in agreement with the results of a Swedish study.[14] However, that group reported an increased prevalence of mandibular central incisor aplasia and did not report any agenesis of maxillary central incisors.

The most frequently reported number of missing teeth was four missing teeth per patient, followed by two missing teeth per patient (as shown in **Figure 6**).

The Special Dental Needs Clinic is a specialist clinic and hence most of the cases referred had moderate to severe hypodontia with four or more teeth missing (**Figure 7**); some mild cases were also treated when additional features such as microdontia, impacted teeth, or primary failure of eruption, which may complicate the management of such mild cases, co-existed.

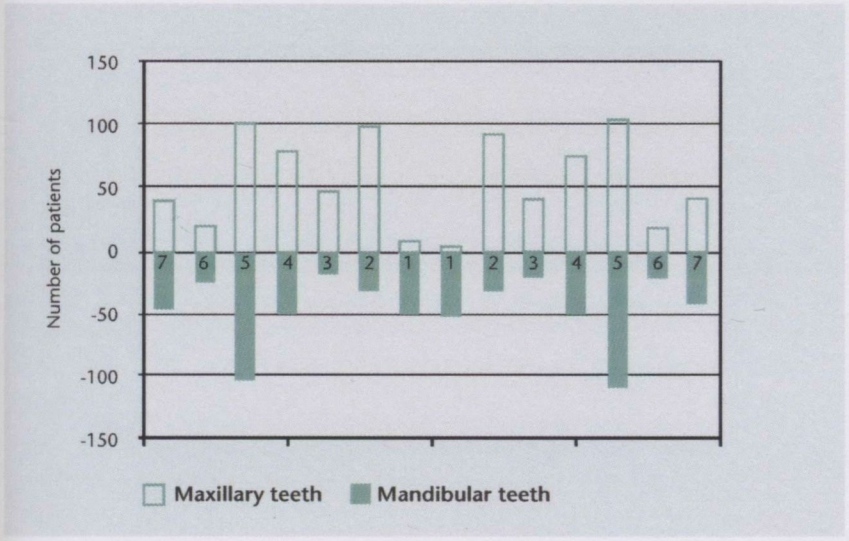

FIGURE 5: Frequency and distribution of congenitally missing teeth.

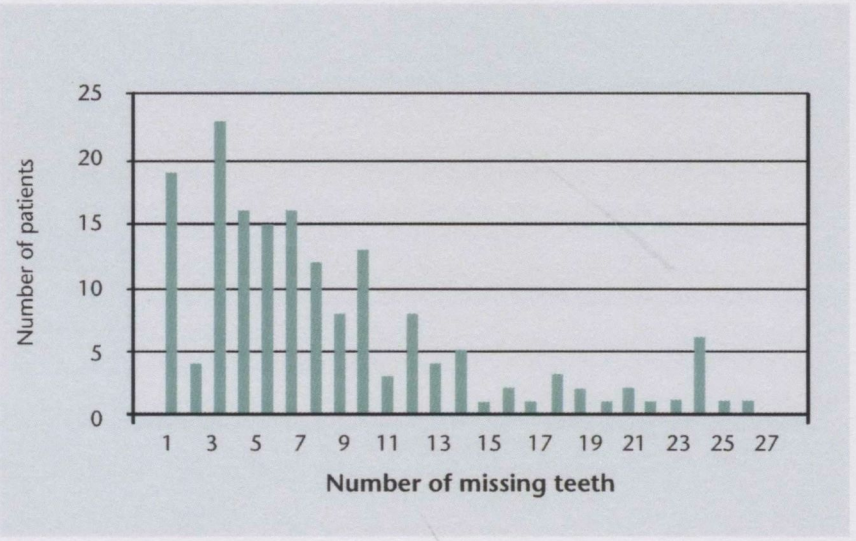

FIGURE 6: Number of missing teeth per patient.

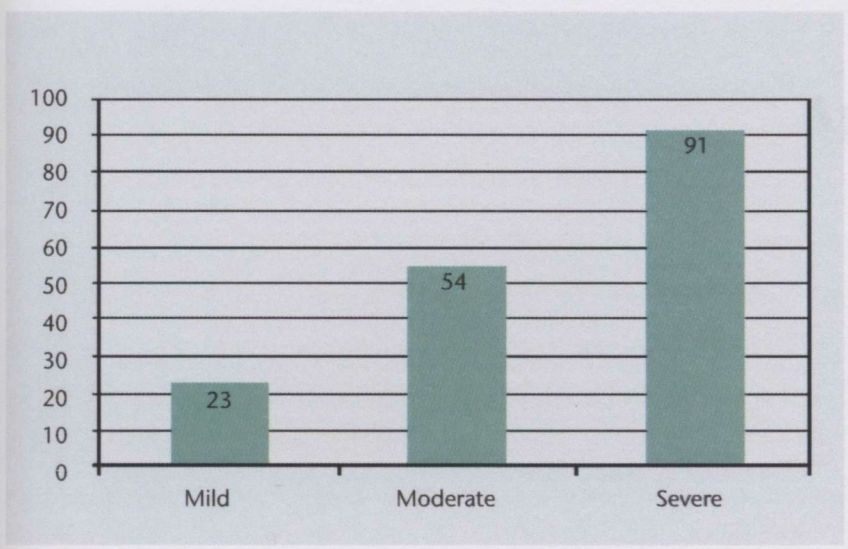

FIGURE 7: Number of cases in relation to severity of tooth agenesis.

Conclusion

This paper describes the profile of patients with hypodontia attending a tertiary care clinic for developmental dental disorders in the Dublin Dental School & Hospital. Hypodontia is the most common developmental disorder affecting teeth in this referred population. The range of missing teeth was two to 26, and the mandibular second premolar was the most frequently absent tooth type.

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Appendix

Appendix content

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    - Moderate young hypodontia patients versus controls
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    - Young Amelogenesis Imperfecta patients versus controls
        - Piers Harris Self Concept Scale
        - Parental Perception Questionnaire
        - Family Impact Scale

Appendix 1

Research Tools Used in the Project

Oral Health Impact Profile-49 (OHIP-49)

Instructions: this questionnaire asks how trouble with your teeth or mouth may have caused problems in your daily life. Please completely fill in ONE circle for each statement that best describes how often you have had each of the following problems.

	Very often	Fairly often	Occas-ionally	Hardly ever	Never	Don't Know
1. Have you had difficulty chewing any foods because of problems with your teeth, mouth or dentures?	○	○	○	○	○	○
2. Have you had trouble pronouncing any words because of problems with your teeth, mouth or dentures?	○	○	○	○	○	○
3. Have you noticed a tooth which doesn't look right?	○	○	○	○	○	○
4. Have you felt that your appearance has been affected because of problems with your teeth, mouth or dentures?	○	○	○	○	○	○
5. Have you felt that your breath has been stale because of problems with your teeth, mouth or dentures?	○	○	○	○	○	○
6. Have you felt that your sense of taste has worsened because of problems with your teeth, mouth or dentures?	○	○	○	○	○	○
7. Have you had food catching in your teeth?	○	○	○	○	○	○
8. Have you felt that your digestion has worsened because of problems with your teeth, mouth or dentures?	○	○	○	○	○	○
9. Have you felt that your dentures have not been fitting properly?	○	○	○	○	○	○
10. Have you had painful aching in your mouth?	○	○	○	○	○	○
11. Have you had a sore jaw?	○	○	○	○	○	○
12. Have you had headaches because of problems with your teeth, mouth or dentures?	○	○	○	○	○	○
13. Have you had sensitive teeth, for example, due to hot or cold foods or drinks?	○	○	○	○	○	○
14. Have you had toothache?	○	○	○	○	○	○

	Very often	Fairly often	Occas-ionally	Hardly ever	Never	Don't know
15. Have you had painful gums?	○	○	○	○	○	○
16. Have you found it uncomfortable to eat any foods because of problems with your teeth, mouth or dentures?	○	○	○	○	○	○
17. Have you had sore spots in your mouth?	○	○	○	○	○	○
18. Have you had uncomfortable dentures?	○	○	○	○	○	○
19. Have you been worried by dental problems?	○	○	○	○	○	○
20. Have you been self conscious because of your teeth, mouth or dentures?	○	○	○	○	○	○
21. Have dental problems made you miserable?	○	○	○	○	○	○
22. Have you felt uncomfortable about the appearance of your teeth, mouth or dentures?	○	○	○	○	○	○
23. Have you felt tense because of problems with your teeth, mouth or dentures?	○	○	○	○	○	○
24. Has your speech been unclear because of problems with your teeth, mouth or dentures?	○	○	○	○	○	○
25. Have people misunderstood some of your words because of problems with your teeth, mouth or dentures?	○	○	○	○	○	○
26. Have you felt that there has been less flavour in your food because of problems with your teeth, mouth or dentures?	○	○	○	○	○	○
27. Have you been unable to brush your teeth properly because of problems with your teeth, mouth or dentures?	○	○	○	○	○	○
28. Have you had to avoid eating some foods because of problems with your teeth, mouth or dentures?	○	○	○	○	○	○
29. Has your diet been unsatisfactory because of problems with your teeth, mouth or dentures?	○	○	○	○	○	○

	Very often	Fairly often	Occas-ionally	Hardly ever	Never	Don't know
30. Have you been unable to eat with your dentures because of problems with them?	○	○	○	○	○	○
31. Have you avoided smiling because of problems with your teeth, mouth or dentures?	○	○	○	○	○	○
32. Have you had to interrupt meals because of problems with your teeth, mouth or dentures?	○	○	○	○	○	○
33. Has your sleep been interrupted because of problems with your teeth, mouth or dentures?	○	○	○	○	○	○
34. Have you been upset because of problems with your teeth, mouth or dentures?	○	○	○	○	○	○
35. Have you found it difficult to relax because of problems with your teeth, mouth or dentures?	○	○	○	○	○	○
36. Have you felt depressed because of problems with your teeth, mouth or dentures?	○	○	○	○	○	○
37. Has your concentration been affected because of problems with your teeth, mouth or dentures?	○	○	○	○	○	○
38. Have you been a bit embarrassed because of problems with your teeth, mouth or dentures?	○	○	○	○	○	○
39. Have you avoided going out because of problems with your teeth, mouth or dentures?	○	○	○	○	○	○
40. Have you been less tolerant of your spouse or family because of problems with your teeth, mouth or dentures?	○	○	○	○	○	○
41. Have you had trouble getting on with other people because of problems with your teeth, mouth or dentures?	○	○	○	○	○	○
42. Have you been a bit irritable with other people because of problems with your teeth, mouth or dentures?	○	○	○	○	○	○
43. Have you had difficulty doing your usual jobs because of problems with your teeth, mouth or dentures?	○	○	○	○	○	○

	Very often	Fairly often	Occas-ionally	Hardly ever	Never	Don't know
44. Have you felt that your general health has worsened because of problems with your teeth, mouth or dentures?	○	○	○	○	○	○
45. Have you suffered any financial loss because of problems with your teeth, mouth or dentures?	○	○	○	○	○	○
46. Have you been unable to enjoy other people's company as much because of problems with your teeth, mouth or dentures?	○	○	○	○	○	○
47. Have you felt that life in general was less satisfying because of problems with your teeth, mouth or dentures?	○	○	○	○	○	○
48. Have you been totally unable to function because of problems with your teeth, mouth or dentures?	○	○	○	○	○	○
49. Have you been unable to work to your full capacity because of problems with your teeth, mouth or dentures?	○	○	○	○	○	○

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ROSENBERG SELF-ESTEEM SCALE

NAME.....DATE.....

Please place a tick in the appropriate box to say whether you strongly agree, agree, disagree, or strongly disagree with the statements below.

	Strongly Agree	Agree	Disagree	Strongly Disagree
1. On the whole I am satisfied with myself				
2. At times I think I am no good at all				
3. I feel I have a number of good qualities				
4. I am able to do things as well as most other people				
5. I feel I do not have much to be proud of				
6. I certainly feel useless at times				
7. I feel that I am a person of worth at least on an equal plane with others				
8. I wish I could have more respect for myself				
9. All in all I am inclined to feel that I am a failure				
10. I take a positive attitude towards myself				

COMMUNITY DENTAL HEALTH SERVICES RESEARCH
UNIT
FACULTY OF DENTISTRY
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CHILD ORAL HEALTH QUESTIONNAIRE

Parental report

6-14 years

SUPPORTED BY
THE HOSPITAL FOR SICK CHILDREN FOUNDATION

INSTRUCTIONS TO PARENTS

1. This questionnaire is about the effects of oral conditions on children's well-being and everyday life, and the effects on their families. We are interested in any condition that involves teeth, lips, mouth or jaws. **Please answer each question.**

2. To answer the question please put an ☒ **in the box by the response**.

3. Please give the response that **best describes your child's experience**. If the question does not apply to your child, please answer with "Never".

Example: How often has your child had a hard time paying attention in school?

If your child has had a hard time paying attention in school because of problems with his/her teeth, lips, mouth or jaws, choose the appropriate response. If it has happened for other reasons, choose "Never".

❑	❑	❑	❑	❑	❑
Never	Once or twice	Sometimes	Often	Everyday or almost everyday	Don't know

4. Please **do not discuss the questions with your child,** as we are interested only in the parents' perspective in this questionnaire.

SECTION 1: Child's oral health and wellbeing

1. How would you rate the health of your child's teeth, lips, jaws and mouth?

❑ Excellent ❑ Very good ❑ Good ❑ Fair ❑ Poor

2. How much is your child's overall wellbeing affected by the condition of his/her teeth, lips, jaws or mouth?

❑ Not at all ❑ Very little ❑ Some ❑ A lot ❑ Very much

SECTION 2: The following questions ask about symptoms and discomfort that children may experience due to the condition of their teeth, lips, mouth and jaws

During the last 3 months, how often has your child had:

3. Pain in the teeth, lips, jaws or mouth?

❑ Never ❑ Once or twice ❑ Sometimes ❑ Often ❑ Everyday or almost everyday ❑ Don't know

4. Bleeding gums?

❑	❑	❑	❑	❑	❑
Never	Once or twice	Sometimes	Often	Everyday or almost everyday	Don't know

5. Sores in the mouth?

❑	❑	❑	❑	❑	❑
Never	Once or twice	Sometimes	Often	Everyday or almost everyday	Don't know

6. Bad breath?

❑	❑	❑	❑	❑	❑
Never	Once or twice	Sometimes	Often	Everyday or almost everyday	Don't know

7. Food stuck in the roof of the mouth?

❑	❑	❑	❑	❑	❑
Never	Once or twice	Sometimes	Often	Everyday or almost everyday	Don't know

8. Food caught in or between the teeth?

❑	❑	❑	❑	❑	❑
Never	Once or twice	Sometimes	Often	Everyday or almost everyday	Don't know

9. Difficulty biting or chewing foods such as fresh apple, corn on the cob or firm meat?

❑	❑	❑	❑	❑	❑
Never	Once or twice	Sometimes	Often	Everyday or almost everyday	Don't know

During the last 3 months, because of his/her teeth, lips, mouth, or jaws, how often has your child:

10. Breathed through the mouth?

❑	❑	❑	❑	❑	❑
Never	Once or twice	Sometimes	Often	Everyday or almost everyday	Don't know

11. Had trouble sleeping?

❑	❑	❑	❑	❑	❑
Never	Once or twice	Sometimes	Often	Everyday or almost everyday	Don't know

12. Had difficulty saying any words?

❑	❑	❑	❑	❑	❑
Never	Once or twice	Sometimes	Often	Everyday or almost everyday	Don't know

13. Taken longer than others to eat a meal?

❑	❑	❑	❑	❑	❑
Never	Once or twice	Sometimes	Often	Everyday or almost everyday	Don't know

14. Had difficulty drinking or eating hot or cold foods?

❑	❑	❑	❑	❑	❑
Never	Once or twice	Sometimes	Often	Everyday or almost everyday	Don't know

15. Had difficulty eating foods he/she would like to eat?

❑	❑	❑	❑	❑	❑
Never	Once or twice	Sometimes	Often	Everyday or almost everyday	Don't know

16. Had diet restricted to certain types of food (e.g. soft food)?

❑	❑	❑	❑	❑	❑
Never	Once or twice	Sometimes	Often	Everyday or almost everyday	Don't know

SECTION 3: The following questions ask about the effects that <u>the condition of children's teeth, lips, mouth and jaws</u> may have on their <u>feelings</u> and <u>everyday activities</u>

During the <u>last 3 months</u>, because of his/her <u>teeth, lips, mouth or jaws</u>, how often has your child been:

17. Upset?

❑	❑	❑	❑	❑	❑
Never	Once or twice	Sometimes	Often	Everyday or almost everyday	Don't know

18. Irritable or frustrated?

❑	❑	❑	❑	❑	❑
Never	Once or twice	Sometimes	Often	Everyday or almost everyday	Don't know

19. Anxious or fearful?

❑	❑	❑	❑	❑	❑
Never	Once or twice	Sometimes	Often	Everyday or almost everyday	Don't know

During the last 3 months, because of his/her teeth, lips, mouth or jaws, how often has your child:

20. Missed school (e.g. pain, appointments, surgery)?

❑	❑	❑	❑	❑	❑
Never	Once or twice	Sometimes	Often	Everyday or almost everyday	Don't know

21. Had a hard time paying attention in school?

❑	❑	❑	❑	❑	❑
Never	Once or twice	Sometimes	Often	Everyday or almost everyday	Don't know

22. Not wanted to speak or read out loud in class?

❑	❑	❑	❑	❑	❑
Never	Once or twice	Sometimes	Often	Everyday or almost everyday	Don't know

23. Not wanted to talk to other children?

❑	❑	❑	❑	❑	❑
Never	Once or twice	Sometimes	Often	Everyday or almost everyday	Don't know

24. Avoided smiling or laughing when around other children?

❑	❑	❑	❑	❑	❑
Never	Once or twice	Sometimes	Often	Everyday or almost everyday	Don't know

During the last 3 months, because of his/her teeth, lips, mouth or jaws, how often has your child:

25. Worried that he/she is not as healthy as other people?

❑	❑	❑	❑	❑	❑
Never	Once or twice	Sometimes	Often	Everyday or almost everyday	Don't know

26. Worried that he/she is different than other people?

❑	❑	❑	❑	❑	❑
Never	Once or twice	Sometimes	Often	Everyday or almost everyday	Don't know

27. Worried that he/she is not as good-looking as other people?

❑	❑	❑	❑	❑	❑
Never	Once or twice	Sometimes	Often	Everyday or almost everyday	Don't know

28. Acted shy or embarrassed?

❑	❑	❑	❑	❑	❑
Never	Once or twice	Sometimes	Often	Everyday or almost everyday	Don't know

29. Been teased or called names by other children?

❑	❑	❑	❑	❑	❑
Never	Once or twice	Sometimes	Often	Everyday or almost everyday	Don't know

30. Been left out by other children?

❑	❑	❑	❑	❑	❑
Never	Once or twice	Sometimes	Often	Everyday or almost everyday	Don't know

31. Not wanted or been unable to spend time with other children?

❑	❑	❑	❑	❑	❑
Never	Once or twice	Sometimes	Often	Everyday or almost everyday	Don't know

32. Not wanted or been unable to participate in activities such as sports, clubs, drama, music, school trips?

❑	❑	❑	❑	❑	❑
Never	Once or twice	Sometimes	Often	Everyday or almost everyday	Don't know

33. Worried that he/she has fewer friends?

❑	❑	❑	❑	❑	❑
Never	Once or twice	Sometimes	Often	Everyday or almost everyday	Don't know

During the last 3 months, how often has your child been:

34. Concerned what other people think about his/her teeth, lips, mouth or jaws?

❑	❑	❑	❑	❑	❑
Never	Once or twice	Sometimes	Often	Everyday or almost everyday	Don't know

35. Asked questions by other children about his/her teeth, lips, mouth or jaws?

❑	❑	❑	❑	❑	❑
Never	Once or twice	Sometimes	Often	Everyday or almost everyday	Don't know

SECTION 4: The following questions ask about effects that a child's oral condition may have on PARENTS AND OTHER FAMILY MEMBERS

During the last 3 months, because of your child's teeth, lips, mouth or jaws, how often have you or another family member:

36. Been upset?

❑	❑	❑	❑	❑	❑
Never	Once or twice	Sometimes	Often	Everyday or almost everyday	Don't know

37. Had sleep disrupted?

❑	❑	❑	❑	❑	❑
Never	Once or twice	Sometimes	Often	Everyday or almost everyday	Don't know

38. Felt guilty?

❑	❑	❑	❑	❑	❑
Never	Once or twice	Sometimes	Often	Everyday or almost everyday	Don't know

39. Taken time off work (e.g. pain, appointments, surgery)?

❑	❑	❑	❑	❑	❑
Never	Once or twice	Sometimes	Often	Everyday or almost everyday	Don't know

40. Had less time for yourself or the family?

❑	❑	❑	❑	❑	❑
Never	Once or twice	Sometimes	Often	Everyday or almost everyday	Don't know

41. Worried that your child will have fewer life opportunities (e.g. for dating, getting married, having children, getting a job he/she will like)?

❑	❑	❑	❑	❑	❑
Never	Once or twice	Sometimes	Often	Everyday or almost everyday	Don't know

42. Felt uncomfortable in public places (e.g. stores, restaurants) with your child?

❑	❑	❑	❑	❑	❑
Never	Once or twice	Sometimes	Often	Everyday or almost everyday	Don't know

During the last 3 months, because of his/her teeth, lips, mouth, or jaws, how often has your child:

43. Been jealous of you or others in the family?

❑	❑	❑	❑	❑	❑
Never	Once or twice	Sometimes	Often	Everyday or almost everyday	Don't know

44. Blamed you or another person in the family?

❑	❑	❑	❑	❑	❑
Never	Once or twice	Sometimes	Often	Everyday or almost everyday	Don't know

45. Argued with you or others in the family?

❑	❑	❑	❑	❑	❑
Never	Once or twice	Sometimes	Often	Everyday or almost everyday	Don't know

46. Required more attention from you or others in the family?

❑	❑	❑	❑	❑	❑
Never	Once or twice	Sometimes	Often	Everyday or almost everyday	Don't know

During the <u>last 3 months</u>, how often has the condition of your child's <u>teeth, lips, mouth or jaws</u>:

47. Interfered with family activities at home or elsewhere?

❑	❑	❑	❑	❑	❑
Never	Once or twice	Sometimes	Often	Everyday or almost everyday	Don't know

48. Caused disagreement or conflict in your family?

❑	❑	❑	❑	❑	❑
Never	Once or twice	Sometimes	Often	Everyday or almost everyday	Don't know

49. Caused financial difficulties for your family?

❑	❑	❑	❑	❑	❑
Never	Once or twice	Sometimes	Often	Everyday or almost everyday	Don't know

SECTION 5: Child's gender and age

a. Your child is:

- ❑ MALE
- ❑ FEMALE

b. Your child's age is: _____YEARS

Questionnaire completed by:

- ❑ MOTHER
- ❑ FATHER
- ❑ OTHER _____

Date completed: _____ / _____ / _____
DAY MONTH YEAR

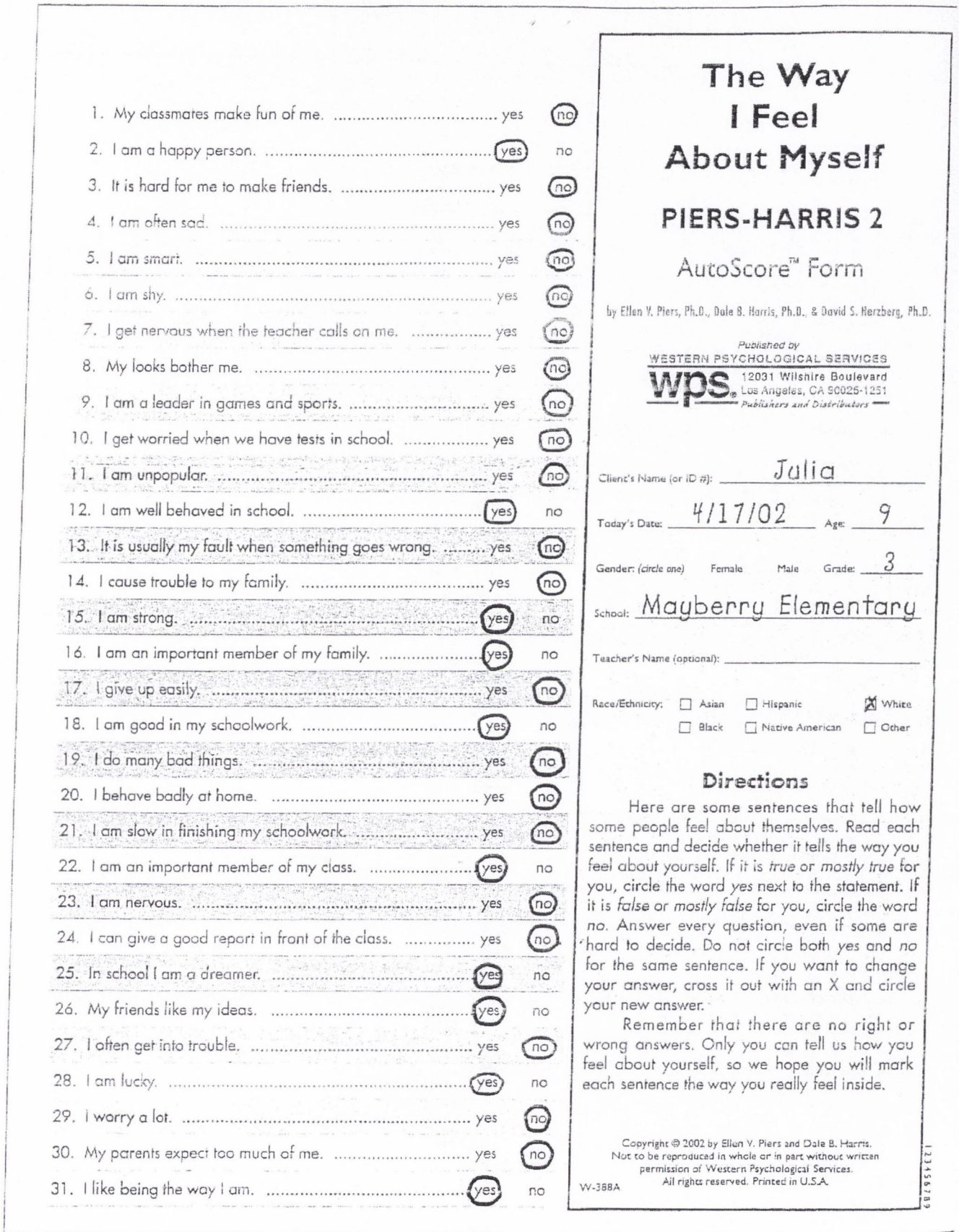
To test how good this questionnaire is at giving us the information we need, we would like a group of parents to complete it again.

Would you be willing to complete another copy of the questionnaire in the next 2 weeks?

☐

Yes

THANK YOU FOR YOUR PARTICIPATION !



1. My classmates make fun of me. ..... yes (no)
2. I am a happy person. ..... (yes) no
3. It is hard for me to make friends. ..... yes (no)
4. I am often sad. ..... yes (no)
5. I am smart. ..... yes (no)
6. I am shy. ..... yes (no)
7. I get nervous when the teacher calls on me. ..... yes (no)
8. My looks bother me. ..... yes (no)
9. I am a leader in games and sports. ..... yes (no)
10. I get worried when we have tests in school. ..... yes (no)
11. I am unpopular. ..... yes (no)
12. I am well behaved in school. ..... (yes) no
13. It is usually my fault when something goes wrong. ..... yes (no)
14. I cause trouble to my family. ..... yes (no)
15. I am strong. ..... (yes) no
16. I am an important member of my family. ..... (yes) no
17. I give up easily. ..... yes (no)
18. I am good in my schoolwork. ..... (yes) no
19. I do many bad things. ..... yes (no)
20. I behave badly at home. ..... yes (no)
21. I am slow in finishing my schoolwork. ..... yes (no)
22. I am an important member of my class. ..... (yes) no
23. I am nervous. ..... yes (no)
24. I can give a good report in front of the class. ..... yes (no)
25. In school I am a dreamer. ..... (yes) no
26. My friends like my ideas. ..... (yes) no
27. I often get into trouble. ..... yes (no)
28. I am lucky. ..... (yes) no
29. I worry a lot. ..... yes (no)
30. My parents expect too much of me. ..... yes (no)
31. I like being the way I am. ..... (yes) no

The Way I Feel About Myself

PIERS-HARRIS 2

AutoScore™ Form

by Ellen V. Piers, Ph.D., Dale B. Harris, Ph.D., & David S. Herzberg, Ph.D.

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Los Angeles, CA 90025-1251
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Client's Name (or ID #): Julia

Today's Date: 4/17/02 Age: 9

Gender: (circle one) Female Male Grade: 3

School: Mayberry Elementary

Teacher's Name (optional): _____

Race/Ethnicity: ☐ Asian ☐ Hispanic ☒ White ☐ Black ☐ Native American ☐ Other

Directions

Here are some sentences that tell how some people feel about themselves. Read each sentence and decide whether it tells the way you feel about yourself. If it is *true* or *mostly true* for you, circle the word *yes* next to the statement. If it is *false* or *mostly false* for you, circle the word *no*. Answer every question, even if some are hard to decide. Do not circle both *yes* and *no* for the same sentence. If you want to change your answer, cross it out with an X and circle your new answer.

Remember that there are no right or wrong answers. Only you can tell us how you feel about yourself, so we hope you will mark each sentence the way you really feel inside.

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W-388A

1 2 3 4 5 6 7 8 9

Figure 1
Completed Piers-Harris 2 AutoScore™ Form

32. I feel left out of things. ..... yes (no)

33. I have nice hair. ..... yes (no)

34. I often volunteer in school. ..... yes (no)

35. I wish I were different. ..... yes (no)

36. I hate school. ..... yes (no)

37. I am among the last to be chosen for games and sports. ..... yes (no)

38. I am often mean to other people. ..... yes (no)

39. My classmates in school think I have good ideas. ..... (yes) no

40. I am unhappy. ..... yes (no)

41. I have many friends. ..... (yes) no

42. I am cheerful. ..... (yes) no

43. I am dumb about most things. ..... yes (no)

44. I am good-looking. ..... (yes) no

45. I get into a lot of fights. ..... yes (no)

46. I am popular with boys. ..... yes (no)

47. People pick on me. ..... yes (no)

48. My family is disappointed in me. ..... yes (no)

49. I have a pleasant face. ..... (yes) no

50. When I grow up, I will be an important person. ..... (yes) no

51. In games and sports, I watch instead of play. ..... yes (no)

52. I forget what I learn. ..... yes (no)

53. I am easy to get along with. ..... (yes) no

54. I am popular with girls. ..... (yes) no

55. I am a good reader. ..... (yes) no

56. I am often afraid. ..... yes (no)

57. I am different from other people. ..... (yes) no

58. I think bad thoughts. ..... yes (no)

59. I cry easily. ..... yes (no)

60. I am a good person. ..... (yes) no

Figure 1 *(continued)*
Completed Piers-Harris 2 AutoScore™ Form

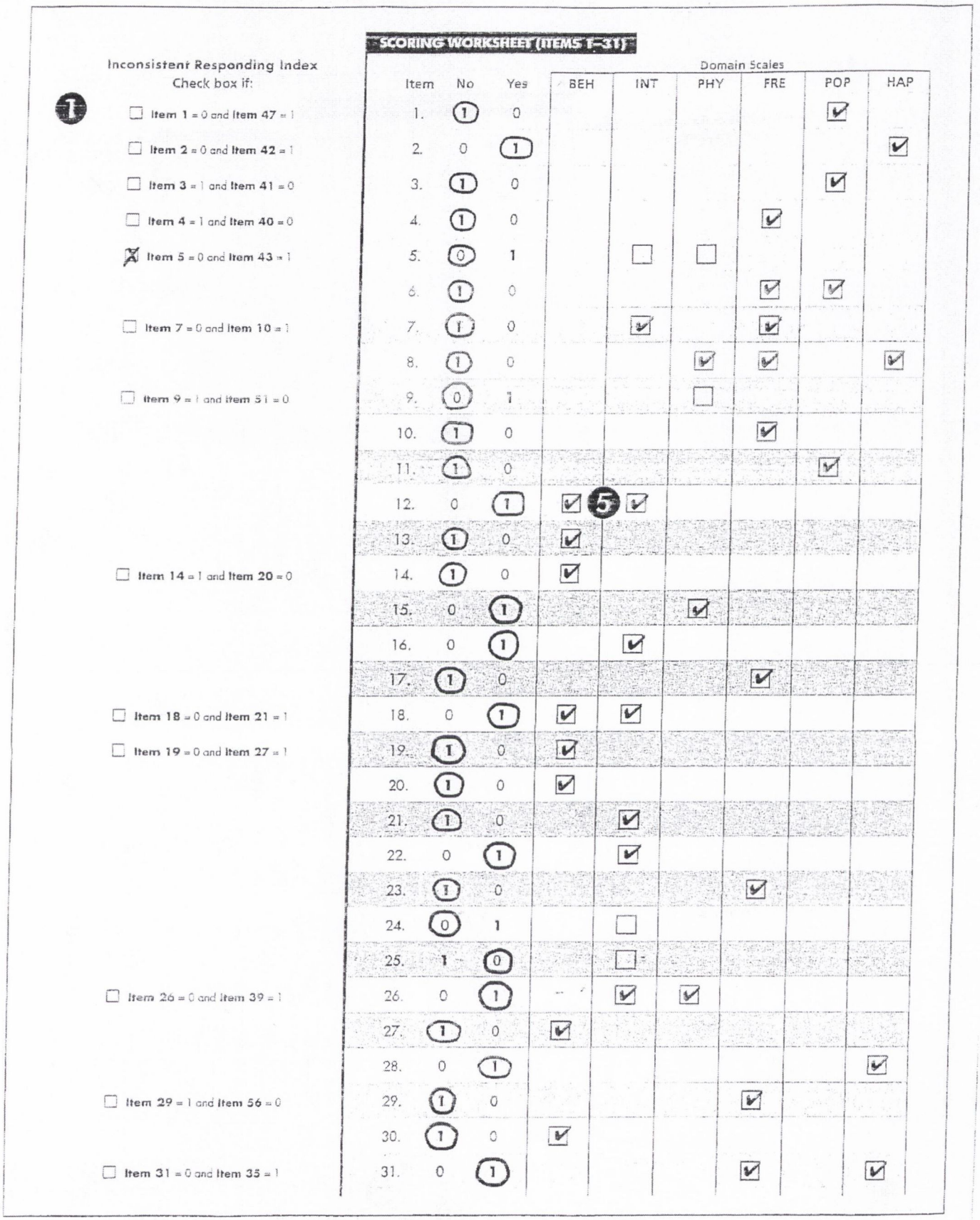


Figure 1 *(continued)*
Completed Piers-Harris 2 AutoScore™ Form

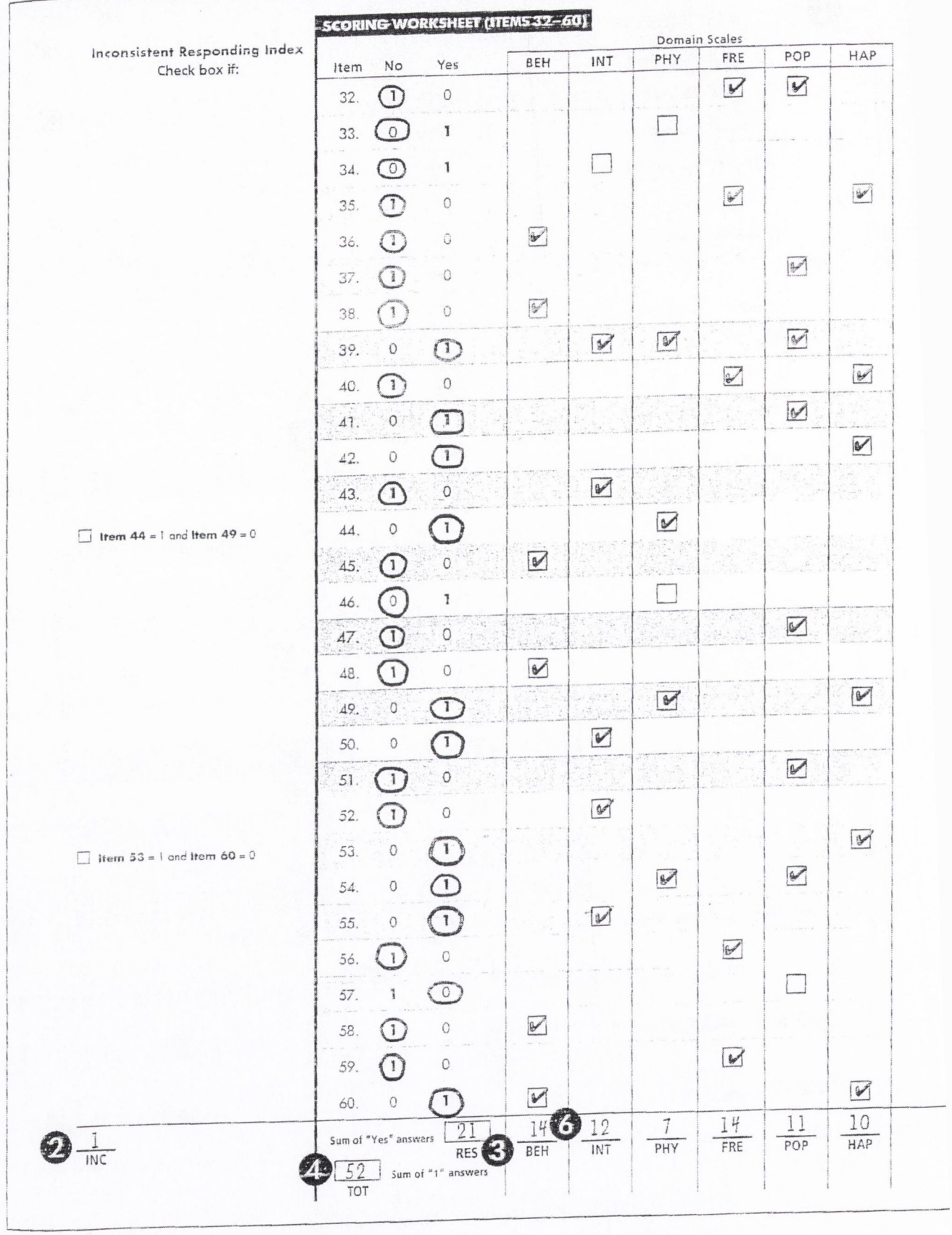


Figure 1 *(continued)*
Completed Piers-Harris 2 AutoScore™ Form

Scoring Instructions

1. Calculate the Validity Scores

To determine the Inconsistent Responding (INC) index raw score, review the 15 INC item pairs listed in the left column of the Scoring Worksheet. Make a check mark in the box next to each pair for which the inconsistency conditions are met. For example, for the first INC pair listed, you mark the box *only if* Item 1 is scored "0" and Item 47 is scored "1." Count the number of check marks in these boxes, and enter the total in the space labeled *INC* at the bottom of the Scoring Worksheet. To calculate the Response Bias (RES) index raw score, count the number of circles that appear in the "Yes" column. Enter this number in the space labeled *RES* at the bottom of the Scoring Worksheet.

2. Calculate the Self-Concept Scores

The Self-Concept raw scores include the Piers-Harris 2 Total (TOT) score and the six domain scale scores: Behavioral Adjustment (BEH), Intellectual and School Status (INT), Physical Appearance and Attributes (PHY), Freedom From Anxiety (FRE), Popularity (POP), and Happiness and Satisfaction (HAP). To obtain the raw TOT score, count the number of items for which "1" is circled on the Scoring Worksheet. Enter this number in the space labeled *TOT* at the bottom of the Scoring Worksheet. To determine the raw scores for the six domain scales, locate each item for which a "1" has been circled and make a check mark in the box(es) in the same row as that item. Then count the number of check marks you have made in the columns that correspond to each domain scale. Enter these totals in the appropriate spaces at the bottom of the Scoring Worksheet. **Note:** Do not calculate the TOT score by summing the raw scores from the six domain scales. Because some items appear on more than one scale, the TOT raw score is not equivalent to the sum of the domain scale raw scores.

3. Complete the Piers-Harris 2 Profile Sheet

Transfer the Validity and Self-Concept raw scores from the Scoring Worksheet to the corresponding spaces at the bottom of the Profile Sheet. Circle the value in each column that corresponds to the raw score you have entered at the bottom. Then connect the circled scores to plot the profile. The *T*-score and percentile rank for each raw score can be found along the left and right margins of the Profile Sheet. Enter the *T*-scores for the Validity and Self-Concept scales in the appropriate spaces at the bottom of the Profile Sheet. Please refer to chapter 3 of the *Piers-Harris 2 Manual* for complete instructions on how to interpret the scores.

Figure 1 *(continued)*
Completed Piers-Harris 2 AutoScore™ Form

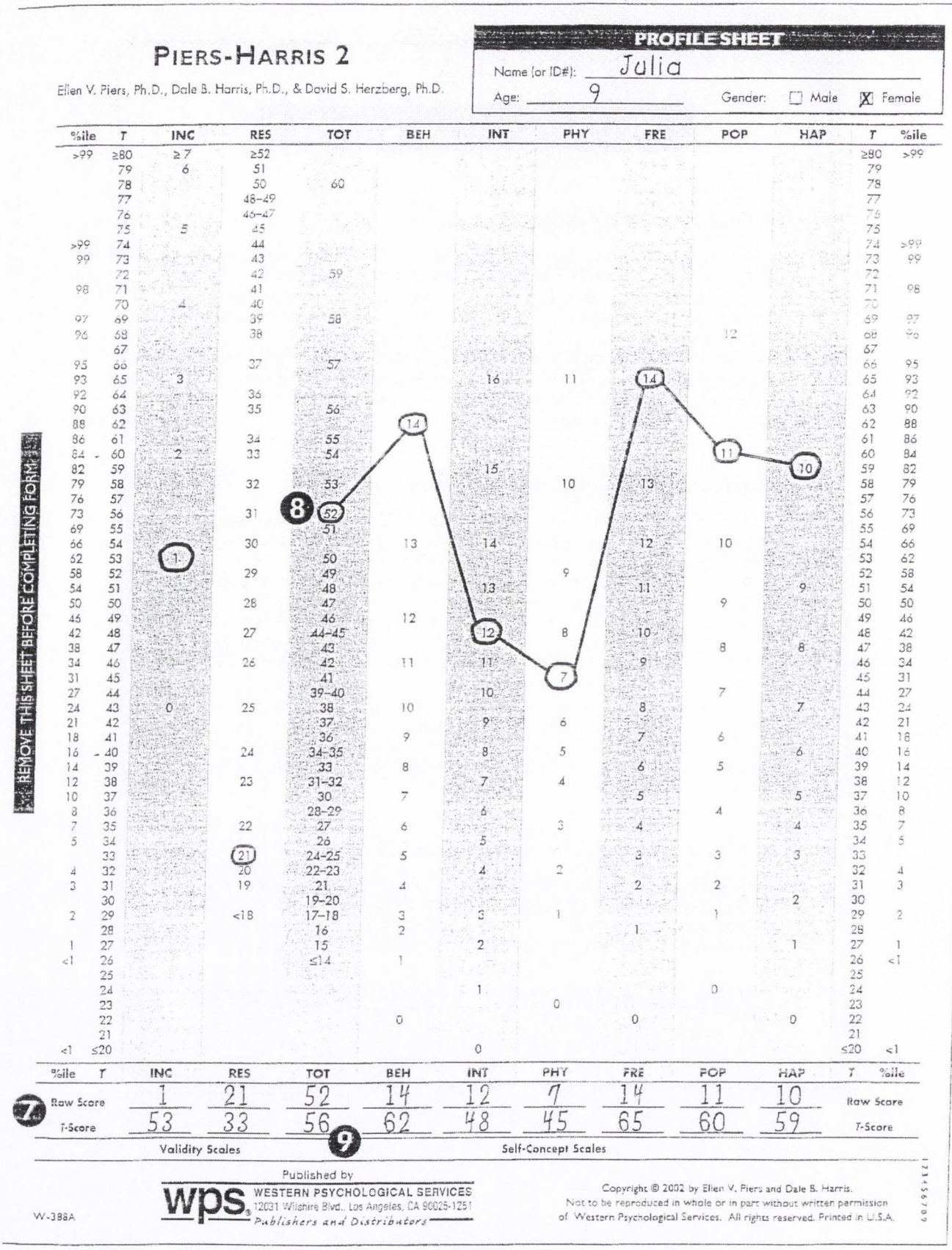


Figure 1 *(continued)*
Completed Piers-Harris 2 AutoScore™ Form

Appendix 2
Excel Spread Sheets of Raw Data for Test and Control Groups

Adult Population

All hypodontia patients versus controls

Oral Health Impact Profile

Rosenberg Self-Esteem Scale

OHIP-49 raw data for combined hypodontia patients

Type	Functional Limitation	Physical Pain	Psychological Discomfort	Physical Disability	Psychological Disability	Social Disability	Handicapped	Total Scores
M	6	3	0	6	0	0	0	15
M	14	5	10	5	8	0	0	42
M	6	4	8	0	3	0	1	22
M	15	10	16	11	7	0	0	59
M	2	3	2	0	0	0	1	8
M	7	6	8	8	11	0	0	40
M	15	16	10	12	8	4	4	69
M	11	5	9	15	8	6	7	61
M	8	11	10	10	13	1	2	55
M	5	3	0	2	0	0	0	10
M	13	17	5	11	8	0	2	56
M	9	8	2	3	0	0	0	22
S	8	14	1	2	0	0	0	25
S	21	21	13	23	12	4	9	103
S	6	8	2	0	1	0	0	17
S	16	14	6	6	0	0	0	42
S	10	8	8	8	8	1	4	47
S	7	9	3	6	1	1	0	27
S	15	20	5	8	3	1	2	54
S	23	19	18	22	15	6	7	110
S	10	6	16	9	16	9	7	73
S	22	28	20	27	20	10	16	143
S	6	8	8	3	7	0	2	34
S	12	18	20	14	18	10	4	96
S	9	3	12	6	6	1	2	39
S	31	31	15	28	18	11	3	137
S	10	2	7	12	0	0	2	33
S	16	14	18	13	16	9	5	91
S	10	10	9	11	6	0	0	46
S	23	16	18	20	13	9	6	105
S	5	4	5	5	3	1	2	25
S	12	18	8	11	9	2	5	65
S	6	7	8	5	1	0	0	27
S	19	20	20	18	19	10	12	118
S	17	22	17	11	13	6	7	93
S	29	19	19	21	13	9	5	115
S	33	35	20	32	21	10	10	161
S	17	7	19	16	18	10	6	93
S	12	13	13	13	13	8	8	80
S	11	11	9	2	6	1	4	44
S	15	13	11	1	6	0	1	47
C	21	10	15	18	7	4	1	76
C	6	10	3	1	1	0	0	21

C	13	13	9	12	10	4	10	71
C	4	3	3	0	1	0	0	11
C	10	9	10	8	6	0	3	46
C	4	5	7	2	0	2	2	22
C	13	15	13	5	9	1	6	62
C	4	3	5	5	0	1	0	18
C	5	8	2	0	0	0	1	16
C	5	3	0	1	0	0	1	10
C	10	9	2	9	11	7	3	51
C	14	7	10	9	11	0	8	59
C	11	5	2	6	0	0	0	24
C	25	13	18	13	18	6	3	96
C	10	10	6	5	1	0	0	32
C	15	6	11	7	5	2	1	47
C	12	2	11	3	2	0	1	31
C	4	14	14	7	8	3	7	57
C	14	17	0	3	5	0	1	40
C	6	11	4	4	4	0	0	29
C	7	12	7	3	4	0	0	33
C	3	13	1	2	0	0	0	19
C	10	8	5	3	1	0	0	27
C	7	6	3	0	0	0	1	17
C	21	18	17	17	15	10	14	112
C	4	3	5	5	0	1	0	18
C	10	8	7	2	1	1	0	29
C	8	7	3	2	1	0	0	21
C	0	4	2	1	0	0	0	7
C	11	13	7	5	3	0	3	42
C	4	2	2	2	2	1	0	13
C	12	8	12	6	6	0	4	48
C	17	16	13	11	9	6	4	76
C	12	15	7	4	5	0	1	44
C	11	5	2	6	0	0	0	24
C	9	4	4	2	0	0	0	19
C	7	10	10	3	4	0	3	37
C	0	4	1	0	0	0	0	5
C	14	7	10	9	11	0	8	59
C	10	8	7	2	1	1	0	29
C	6	10	3	1	1	0	0	21

Rosenberg Scale raw data for Hypodontia patients

Type (T)	total scores (T)	Type ©	total scores ©
M	18	C	17
M	10	C	23
M	21	C	30
M	14	C	23
M	29	C	28
M	26	C	28
M	23	C	17
M	20	C	17
M	27	C	18
M	21	C	20
M	28	C	24
M	16	C	20
S	24	C	18
S	28	C	19
S	14	C	18
S	17	C	19
S	30	C	24
S	20	C	20
S	15	C	27
S	23	C	21
S	16	C	19
S	19	C	18
S	25	C	21
S	22	C	20
S	22	C	25
S	25	C	30
S	23	C	30
S	26	C	30
S	13	C	30
S	28	C	21
S	23	C	23
S	30	C	17
S	15	C	25
S	16	C	23
S	30	C	25
S	20	C	24
S	30	C	18
S	27	C	18
S	26	C	21
S	27	C	19
S	30	C	28

Severe hypodontia patients versus controls

Oral Health Impact Profile

OHIP raw data (severe hypodontia)

Type	Functional Limitation	OHIP (Physical Pain)	Psychological Discomfort	Physical Disability	Psychological Disability	Social Disability	Handicapped	Total Scores
S	8	14	1	2	0	0	0	25
S	21	21	13	23	12	4	9	103
S	6	8	2	0	1	0	0	17
S	16	14	6	6	0	0	0	42
S	10	8	8	8	8	1	4	47
S	7	9	3	6	1	1	0	27
S	15	20	5	8	3	1	2	54
S	23	19	18	22	15	6	7	110
S	10	6	16	9	16	9	7	73
S	22	28	20	27	20	10	16	143
S	6	8	8	3	7	0	2	34
S	12	18	20	14	18	10	4	96
S	9	3	12	6	6	1	2	39
S	31	31	15	28	18	11	3	137
S	10	2	7	12	0	0	2	33
S	16	14	18	13	16	9	5	91
S	10	10	9	11	6	0	0	46
S	23	16	18	20	13	9	6	105
S	5	4	5	5	3	1	2	25
S	12	18	8	11	9	2	5	65
S	6	7	8	5	1	0	0	27
S	19	20	20	18	19	10	12	118
S	17	22	17	11	13	6	7	93
S	29	19	19	21	13	9	5	115
S	33	35	20	32	21	10	10	161
S	17	7	19	16	18	10	6	93
S	12	13	13	13	13	8	8	80
S	11	11	9	2	6	1	4	44
S	15	13	11	1	6	0	1	47
C	11	5	2	6	0	0	0	24
C	25	13	18	13	18	6	3	96
C	10	10	6	5	1	0	0	32
C	15	6	11	7	5	2	1	47
C	12	2	11	3	2	0	1	31
C	4	14	14	7	8	3	7	57
C	14	17	0	3	5	0	1	40
C	6	11	4	4	4	0	0	29
C	7	12	7	3	4	0	0	33
C	3	13	1	2	0	0	0	19
C	10	8	5	3	1	0	0	27
C	7	6	3	0	0	0	1	17
C	21	18	17	17	15	10	14	112

C	4	3	5	5	0	1	0	18
C	10	8	7	2	1	1	0	29
C	8	7	3	2	1	0	0	21
C	0	4	2	1	0	0	0	7
C	11	13	7	5	3	0	3	42
C	4	2	2	2	2	1	0	13
C	12	8	12	6	6	0	4	48
C	17	16	13	11	9	6	4	76
C	12	15	7	4	5	0	1	44
C	11	5	2	6	0	0	0	24
C	9	4	4	2	0	0	0	19
C	7	10	10	3	4	0	3	37
C	0	4	1	0	0	0	0	5
C	14	7	10	9	11	0	8	59
C	10	8	7	2	1	1	0	29
C	6	10	3	1	1	0	0	21

Moderate hypodontia patients versus controls

Oral Health Impact Profile

OHIP raw data (moderate) hypodontia

Type	Functional Limitation	Physical Pain	Psychological Discomfort	Physical Disability	Psychological Disability	Social Disability	Handicapped	Total Scores
M	6	3	0	6	0	0	0	15
M	14	5	10	5	8	0	0	42
M	6	4	8	0	3	0	1	22
M	15	10	16	11	7	0	0	59
M	2	3	2	0	0	0	1	8
M	7	6	8	8	11	0	0	40
M	15	16	10	12	8	4	4	69
M	11	5	9	15	8	6	7	61
M	8	11	10	10	13	1	2	55
M	5	3	0	2	0	0	0	10
M	13	17	5	11	8	0	2	56
M	9	8	2	3	0	0	0	22
C	21	10	15	18	7	4	1	76
C	6	10	3	1	1	0	0	21
C	13	13	9	12	10	4	10	71
C	4	3	3	0	1	0	0	11
C	10	9	10	8	6	0	3	46
C	4	5	7	2	0	2	2	22
C	13	15	13	5	9	1	6	62
C	4	3	5	5	0	1	0	18
C	5	8	2	0	0	0	1	16
C	5	3	0	1	0	0	1	10
C	10	9	2	9	11	7	3	51
C	14	7	10	9	11	0	8	59

Amelogenesis Imperfecta patients versus controls

Oral Health Impact Profile

Rosenberg Self-Esteem Scale

OHIP-49 raw data for AI patients

Type	Functional Limitation	Physical Pain	Psychological Discomfort	Physical Disability	Psychological Disability	Social Disability	Handicapped	Total Scores
T	13	7	0	4	0	0	0	24
T	16	15	5	12	0	0	1	49
T	12	17	6	9	10	5	4	63
T	0	6	15	8	14	2	3	48
T	13	17	15	6	9	3	3	66
T	10	14	10	6	6	0	2	48
T	15	19	8	18	9	0	0	69
T	7	6	8	2	0	0	0	23
T	4	2	8	0	3	0	0	17
T	16	14	13	7	4	5	5	64
T	9	16	9	8	4	2	0	48
T	17	18	14	16	18	10	8	101
T	12	20	14	8	13	1	4	72
T	17	9	18	13	16	15	12	100
T	12	14	19	14	16	8	5	88
T	7	9	6	6	5	2	2	37
T	19	10	17	10	15	5	7	83
T	9	15	16	6	14	6	4	70
T	21	20	16	13	10	4	1	85
T	15	16	10	14	9	5	5	74
T	11	15	13	10	18	4	1	72
T	18	10	18	8	14	4	5	77
T	18	17	11	10	9	0	2	67
T	23	19	20	17	21	14	18	132
T	14	3	8	7	2	1	0	35
T	19	22	20	18	21	7	8	115
T	19	19	16	12	14	8	5	93
C	20	15	13	8	12	6	8	82
C	2	12	2	1	0	0	0	17
C	10	8	5	3	1	0	0	27
C	4	14	14	7	8	3	7	57
C	6	12	9	2	10	1	6	46
C	10	8	7	2	1	1	0	29
C	12	8	6	2	1	1	0	30
C	12	8	12	6	6	0	4	48
C	5	3	0	1	0	0	1	10
C	5	9	7	2	3	1	1	28
C	9	26	12	6	13	3	5	74
C	8	7	3	2	1	0	0	21
C	2	12	0	0	1	0	0	15
C	2	5	0	0	1	0	0	8
C	4	5	7	2	0	2	2	22
C	0	4	1	0	0	0	0	5
C	18	16	11	16	11	3	10	85
C	21	10	20	18	13	4	4	90
C	12	2	11	3	2	0	1	31
C	11	5	2	6	0	0	0	24
C	16	13	17	18	15	1	5	85
C	11	8	10	2	6	0	0	37
C	22	21	13	11	4	2	4	77
C	26	24	15	20	16	4	5	110
C	17	19	14	17	11	9	13	100
C	24	23	20	15	13	8	9	112
C	12	5	19	4	5	1	3	49

Rosenberg Raw Data for AI patients

Type	TOTAL SCORE	Type	TOTAL SCORE
T	20	C	19
T	14	C	19
T	25	C	30
T	26	C	28
T	24	C	23
T	19	C	18
T	24	C	19
T	24	C	27
T	27	C	21
T	25	C	18
T	30	C	15
T	8	C	20
T	18	C	28
T	20	C	20
T	19	C	25
T	29	C	30
T	17	C	22
T	27	C	16
T	19	C	24
T	30	C	21
T	26	C	22
T	19	C	23
T	19	C	20
T	14	C	16
T	24	C	26
T	19	C	19
T	22	C	22

All young hypodontia patients versus controls

Piers Harris Self Concept Scale

Parental Perception Questionnaire

Family Impact Scale

Piers Harris raw data for Hypodontia patients

TYPE	T-TOT	T-BEH	T-INT	T-PHY	T-FRE	T-POP	T-HAP
T	58	62	59	58	54	50	59
T	53	62	65	65	39	60	43
T	72	62	65	58	65	68	59
T	69	62	59	58	65	68	59
T	72	62	65	58	58	68	51
T	66	62	65	58	54	68	59
T	58	62	59	52	54	50	51
T	49	62	54	45	46	47	47
T	60	54	59	58	54	68	51
T	50	62	48	52	43	47	59
T	55	62	48	45	65	60	59
T	48	54	54	45	39	39	37
T	53	49	42	65	58	60	51
T	44	54	46	40	39	47	37
T	60	62	65	42	58	68	51
T	38	35	46	52	35	60	33
T	60	62	59	52	54	50	51
T	58	54	59	52	58	54	51
T	49	41	48	52	51	68	51
T	31	46	38	40	28	31	33
T	55	62	48	48	54	50	47
T	63	62	51	58	65	60	59
T	48	43	51	52	46	50	51
T	63	62	65	52	54	60	51
T	60	62	65	53	51	60	43
T	37	46	40	45	33	41	40
T	63	62	65	45	65	68	51
T	38	37	32	45	58	39	47
T	47	49	40	45	54	54	51

TYPE	T-TOT	T-BEH	T-INT	T-PHY	T-FRE	T-POP	T-HAP
C	69	49	65	65	65	68	59
C	66	54	65	65	58	68	47
C	69	54	65	58	65	68	59
C	32	33	42	32	37	36	37
C	56	62	54	58	46	54	51
C	56	62	54	58	46	54	51
C	41	46	36	35	46	41	47
C	48	54	46	57	39	50	47
C	61	62	49	58	65	68	59
C	47	37	40	57	51	53	51
C	53	35	54	65	65	54	59
C	32	33	42	32	37	36	37
C	55	54	49	52	48	60	59
C	48	37	44	57	57	60	51
C	45	43	46	52	39	50	51
C	32	46	40	38	35	40	33
C	61	62	59	58	51	60	51
C	69	54	65	65	68	47	47
C	51	62	54	45	43	60	37

C	47	46	38	48	51	54	59
C	32	43	40	29	35	29	30
C	60	54	62	58	58	60	59
C	56	54	51	65	51	60	51
C	63	62	65	58	58	60	51
C	31	33	38	35	41	31	43
C	56	62	59	58	51	50	59
C	51	43	51	45	58	50	59
C	40	49	34	35	48	39	43
C	53	46	51	65	46	60	47

Parental Perception Questionnaire (Hypodontia)

Type	Total scores	oral symptoms	functional limitation	emotional well-being	Social well-being
T	23	4	14	0	5
T	30	11	9	6	4
T	5	4	0	0	1
T	4	2	1	0	1
T	3	0	0	0	3
T	44	6	20	9	9
T	11	4	3	0	4
T	15	2	4	6	3
T	6	1	4	0	1
T	56	7	13	18	18
T	9	4	4	0	1
T	37	6	13	10	8
T	18	6	6	0	6
T	66	12	18	22	14
T	25	8	8	6	3
T	33	6	5	13	9
T	31	5	13	10	3
T	15	11	0	1	3
T	63	18	23	9	13
T	48	8	12	14	14
T	22	1	4	11	6
T	4	2	2	0	0
T	28	3	8	9	8
T	51	11	12	10	18
T	14	3	10	0	1
T	17	7	1	7	2
T	61	8	15	13	25
T	12	2	8	0	2
T	44	13	16	5	10

Type	Total scores	oral symptoms	functional limitation	emotional well-being	Social well-being
C	6	2	2	1	1
C	6	3	2	0	1
C	14	3	6	0	5
C	9	0	3	3	3
C	9	0	3	3	3
C	15	5	4	3	3
C	8	5	1	1	1
C	31	7	10	7	7
C	6	4	0	0	2
C	4	2	2	0	0
C	32	10	13	3	6
C	14	3	3	4	4
C	7	3	2	1	1
C	4	2	2	0	0
C	42	15	9	13	5
C	33	6	6	10	11
C	7	0	4	3	0
C	2	2	0	0	0
C	18	2	7	6	3

C	28	6	9	4	9
C	35	8	5	8	14
C	33	12	9	5	7
C	2	0	0	0	2
C	17	3	2	11	1
C	43	10	3	16	14
C	14	3	6	1	4
C	9	2	3	2	2
C	10	3	5	1	1
C	11	3	2	2	4

Family Impact Scale raw data (Hypodontia patients)

Type	FIS total scores	Parental and family activity	Impact on parental emotions	Impact on family conflict	Financial burden
T	4	0	4	0	0
T	8	1	4	3	0
T	0	0	0	0	0
T	2	1	1	0	0
T	1	1	0	0	0
T	12	4	5	2	1
T	7	6	1	0	0
T	6	3	3	0	0
T	0	0	0	0	0
T	13	4	6	2	1
T	7	3	2	2	0
T	4	2	2	0	0
T	3	3	0	0	0
T	29	9	13	7	0
T	3	0	3	0	0
T	3	0	1	2	0
T	2	0	1	1	0
T	2	1	0	1	0
T	3	2	1	0	0
T	26	7	15	4	0
T	8	5	2	1	0
T	0	0	0	0	0
T	8	6	1	1	0
T	5	5	0	0	0
T	1	0	0	1	0
T	0	0	0	0	0
T	11	6	4	1	0
T	2	1	1	0	0
T	13	6	2	2	3

Type	FIS total scores	Parental and family activity	Impact on parental emotions	Impact on family conflict	Financial burden
C	1	1	0	0	0
C	0	0	0	0	0
C	1	1	0	0	0
C	5	2	0	0	3
C	0	0	0	0	0
C	1	0	1	0	0
C	3	2	0	1	0
C	6	2	2	0	2
C	2	2	0	0	0
C	0	0	0	0	0
C	14	5	3	5	1
C	7	3	0	1	3
C	3	2	0	1	0
C	0	0	0	0	0
C	13	5	2	3	3
C	16	8	4	2	2

C	3	1	0	2	0
C	0	0	0	0	0
C	11	1	7	1	2
C	9	5	2	2	0
C	12	4	4	3	1
C	0	0	0	0	0
C	1	1	0	0	0
C	9	1	2	4	2
C	17	5	9	3	0
C	2	1	0	1	0
C	5	4	0	1	0
C	2	2	0	0	0
C	3	2	0	1	0

Young Amelogenesis Imperfecta patients versus controls

Piers Harris Self Concept Scale

Parental Perception Questionnaire

Family Impact Scale

Piers Harris raw data for Amelogenesis Imperfecta Patients

TYPE	T-TOT	T-BEH	T-INT	T-PHY	T-FRE	T-POP	T-HAP
T	48	43	44	52	48	54	59
T	26	35	29	32	28	29	33
T	58	46	51	65	65	60	47
T	34	33	32	38	41	50	37
T	53	54	51	48	54	54	47
T	58	43	54	65	65	60	51
T	45	62	38	40	43	50	43
T	48	62	44	40	46	68	37
T	78	62	65	65	65	68	59
T	44	46	38	38	46	60	40
T	38	49	44	40	35	39	33
T	47	49	44	48	43	54	59

TYPE	T-TOT	T-BEH	T-INT	T-PHY	T-FRE	T-POP	T-HAP
C	49	54	51	40	58	47	51
C	63	49	59	65	55	68	47
C	63	54	59	58	65	60	51
C	49	54	54	48	39	47	51
C	32	35	42	38	37	36	40
C	61	54	65	48	65	68	51
C	49	54	48	57	39	50	51
C	60	49	59	58	53	60	47
C	56	43	48	65	48	54	47
C	61	62	59	58	54	60	59
C	46	39	46	52	46	54	51
C	66	49	65	59	65	67	59

Parental Perception raw data (AI)

Type	Total scores	oral symptoms	functional limitation	emotional well-being	Social well-being
T	18	6	6	4	2
T	87	22	17	21	27
T	6	2	4	0	0
T	50	13	18	6	13
T	27	6	12	4	5
T	31	8	14	3	6
T	17	5	7	3	2
T	38	9	8	12	9
T	5	4	0	0	1
T	16	3	6	4	3
T	76	16	18	21	21
T	35	8	7	10	10

Type	Total scores	oral symptoms	functional limitation	emotional well-being	Social well-being
C	7	2	1	3	1
C	12	6	2	1	3
C	6	2	2	1	1
C	24	10	12	0	2
C	15	3	5	3	4
C	19	4	6	2	7
C	36	8	12	8	8
C	14	6	4	2	2
C	11	2	4	1	4
C	21	4	5	8	4
C	33	7	15	4	7
C	10	4	0	4	2

Appendix 3

Statistical Analysis

Adult Population

All hypodontia patients versus controls

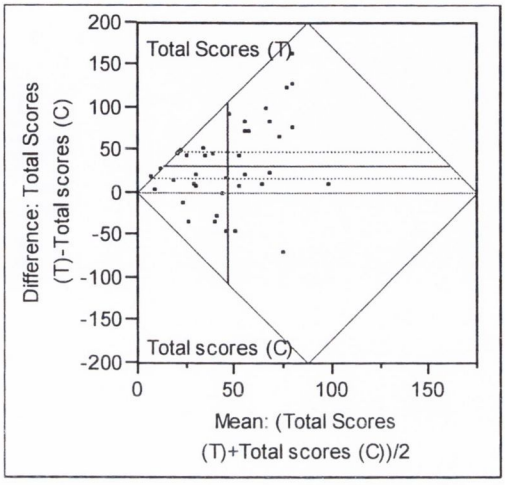
Oral Health Impact Profile

Rosenberg Self-Esteem Scale

Adult hypodontia, combined (Severe + Moderate)

Matched Pairs (Total Scores)

Difference: Total Scores (T)-Total scores (C)



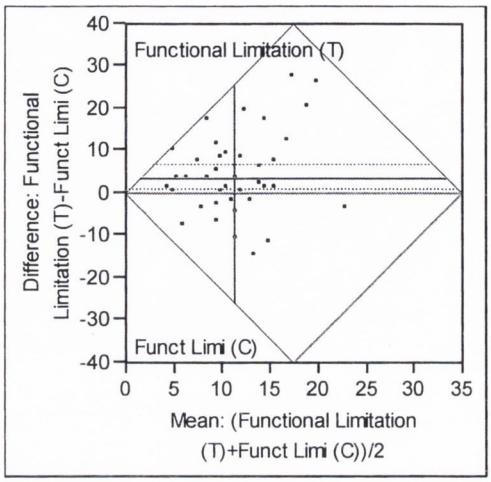
Total Scores (T)	62.1707	t-Ratio	3.994002
Total scores (C)	31.2195	DF	40
Mean Difference	30.9512	Prob > |t|	0.0003
Std Error	7.74943	Prob > t	0.0001
Upper95%	46.6134	Prob < t	0.9999
Lower95%	15.289		
N	41		
Correlation	-0.092		

Wilcoxon Sign-Rank

	Total Scores (T)-Total scores (C)
Test Statistic	263.000
Prob > |z|	0.0001
Prob > z	<.0001
Prob < z	0.9999

Matched Pairs (Functionai Limitations)

Difference: Functional Limitation (T)-Funct Limi (C)



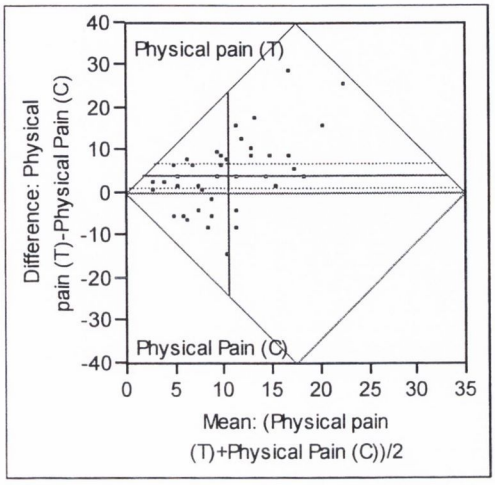
Functional Limitation (T)	13.2195	t-Ratio	2.500698
Funct Limi (C)	9.4878	DF	40
Mean Difference	3.73171	Prob > |t|	0.0166
Std Error	1.49227	Prob > t	0.0083
Upper95%	6.74769	Prob < t	0.9917
Lower95%	0.71572		
N	41		
Correlation	-0.1015		

Wilcoxon Sign-Rank

	Functional Limitation (T)-Funct Limi (C)
Test Statistic	149.500
Prob > |z|	0.0280
Prob > z	0.0140
Prob < z	0.9860

Matched Pairs (physical pain)

Difference: Physical pain (T)-Physical Pain (C)



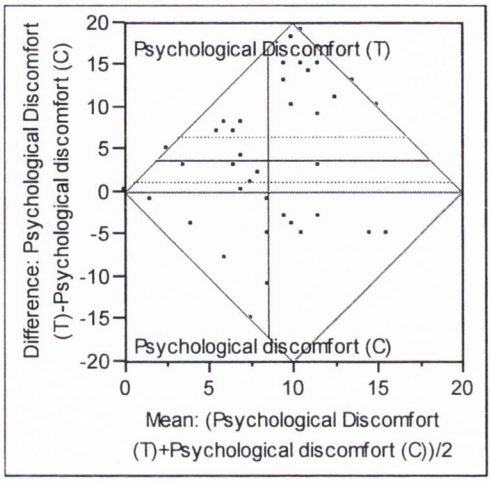
Physical pain (T)	12.4146	t-Ratio	2.778086
Physical Pain (C)	8.63415	DF	40
Mean Difference	3.78049	Prob > |t|	0.0083
Std Error	1.36082	Prob > t	0.0041
Upper95%	6.53082	Prob < t	0.9959
Lower95%	1.03016		
N	41		
Correlation	0.0982		

Wilcoxon Sign-Rank

	Physical pain (T)-Physical Pain (C)
Test Statistic	183.500
Prob > |z|	0.0085
Prob > z	0.0043
Prob < z	0.9957

Matched Pairs (Psychological Discomfort)

Difference: Psychological Discomfort (T)-Psychological discomfort (C)



Psychological Discomfort (T)	10.439	t-Ratio	2.82691
Psychological discomfort (C)	6.65854	DF	40
Mean Difference	3.78049	Prob > |t|	0.0073
Std Error	1.33732	Prob > t	0.0037
Upper95%	6.48332	Prob < t	0.9963
Lower95%	1.07766		
N	41		
Correlation	-0.1824		

Wilcoxon Sign-Rank

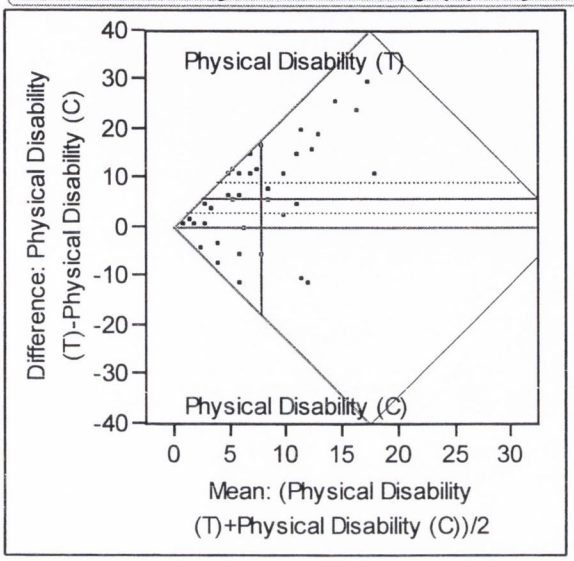
	Psychological Discomfort (T)-Psychological discomfort (C)
Test Statistic	175.000
Prob > |z|	0.0125
Prob > z	0.0062
Prob < z	0.9938

Matched Pairs (Physical Disability)

Difference: Physical Disability (T)- Physical Disability (C)

Matched Pairs

Difference: Physical Disability (T)-Physical Disability (C)



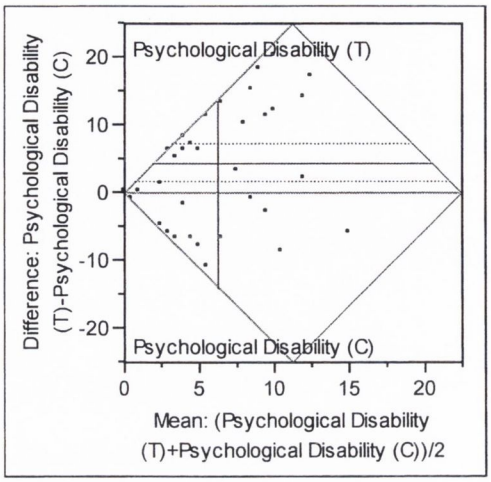
Physical Disability (T)	10.6341	t-Ratio	3.699284
Physical Disability (C)	4.97561	DF	40
Mean Difference	5.65854	Prob > |t|	0.0006*
Std Error	1.52963	Prob > t	0.0003*
Upper95%	8.75003	Prob < t	0.9997
Low er95%	2.56704		
N	41		
Correlation	-0.1574		

Wilcoxon Sign-Rank

	Physical Disability (T)- Physical Disability (C)
Test Statistic	215.500
Prob > |z|	0.0010*
Prob > z	0.0005*
Prob < z	0.9995

Matched Pairs (Psychological Disability)

Difference: Psychological Disability (T)-Psychological Disability (C)



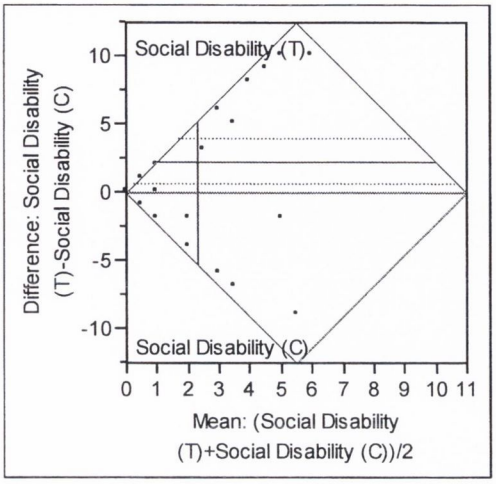
Psychological Disability (T)	8.4878	t-Ratio	3.289233
Psychological Disability (C)	3.97561	DF	40
Mean Difference	4.5122	Prob > |t|	0.0021
Std Error	1.37181	Prob > t	0.0011
Upper95%	7.28472	Prob < t	0.9989
Lower95%	1.73967		
N	41		
Correlation	-0.1992		

Wilcoxon Sign-Rank

	Psychological Disability (T)-Psychological Disability (C)
Test Statistic	193.500
Prob > |z|	0.0036
Prob > z	0.0018
Prob < z	0.9982

Matched Pairs (Social Disability)

Difference: Social Disability (T)-Social Disability (C)



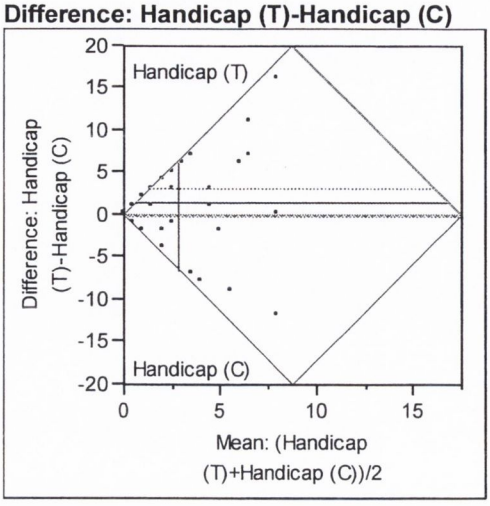
Social Disability (T)	3.41463	t-Ratio	2.658119
Social Disability (C)	1.21951	DF	40
Mean Difference	2.19512	Prob > |t|	0.0112
Std Error	0.82582	Prob > t	0.0056
Upper95%	3.86416	Prob < t	0.9944
Lower95%	0.52608		
N	41		
Correlation	-0.2892		

Wilcoxon Sign-Rank

	Social Disability (T)-Social Disability (C)
Test Statistic	111.000
Prob > |z|	0.0133
Prob > z	0.0066
Prob < z	0.9934

Matched Pairs (Handicap)

Difference: Handicap (T)-Handicap (C)



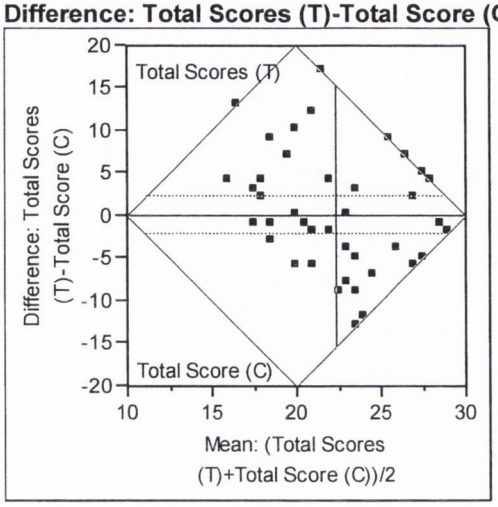
Handicap (T)	3.56098	t-Ratio	1.791604
Handicap (C)	2.09756	DF	40
Mean Difference	1.46341	Prob > |t|	0.0808
Std Error	0.81682	Prob > t	0.0404
Upper95%	3.11427	Prob < t	0.9596
Lower95%	-0.1874		
N	41		
Correlation	-0.1069		

Wilcoxon Sign-Rank

	Handicap (T)-Handicap (C)
Test Statistic	118.500
Prob > |z|	0.0501
Prob > z	0.0250
Prob < z	0.9750

Adult Combined Hypodontia (Rosenberg)

Matched Pairs
Difference: Total Scores (T)-Total Score (C)



Total Scores (T)	22.3415	t-Ratio	-0.0223
Total Score (C)	22.3659	DF	40
Mean Difference	-0.0244	Prob > |t|	0.9823
Std Error	1.09349	Prob > t	0.5088
Upper95%	2.18563	Prob < t	0.4912
Lower95%	-2.2344		
N	41		
Correlation	0.0134		

Wilcoxon Sign-Rank

	Total Scores (T)-Total Score (C)
Test Statistic	-12.000
Prob > |z|	0.8694
Prob > z	0.5653
Prob < z	0.4347

Severe hypodontia patients versus controls

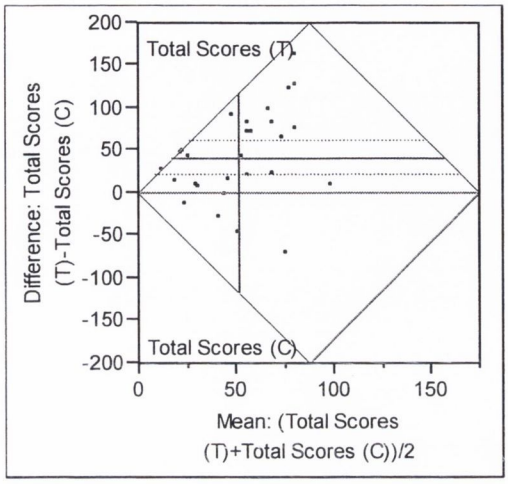
Oral Health Impact Profile

Rosenberg Self-Esteem Scale

Adult Severe hypodontia (OHIP-49)

Matched Pairs (Total Scores)

Difference: Total Scores (T)-Total Scores (C)



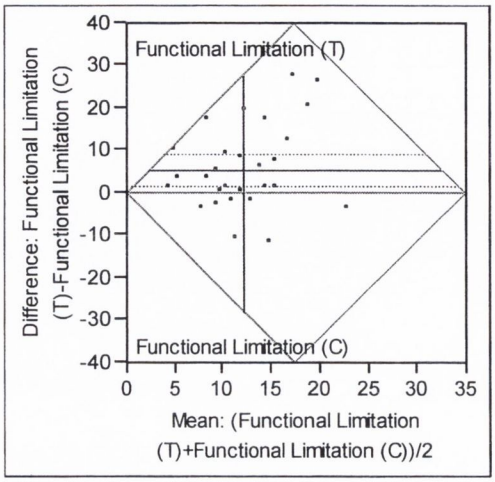
Total Scores (T)	72.069	t-Ratio	4.169687
Total Scores (C)	31.5172	DF	28
Mean Difference	40.5517	Prob > |t|	0.0003
Std Error	9.72537	Prob > t	0.0001
Upper95%	60.4732	Prob < t	0.9999
Lower95%	20.6302		
N	29		
Correlation	-0.13		

Wilcoxon Sign-Rank

	Total Scores (T)-Total Scores (C)
Test Statistic	159.500
Prob > |z|	0.0001
Prob > z	<.0001
Prob < z	0.9999

Matched Pairs (Functional Limitation)

Difference: Functional Limitation (T)-Functional Limitation (C)



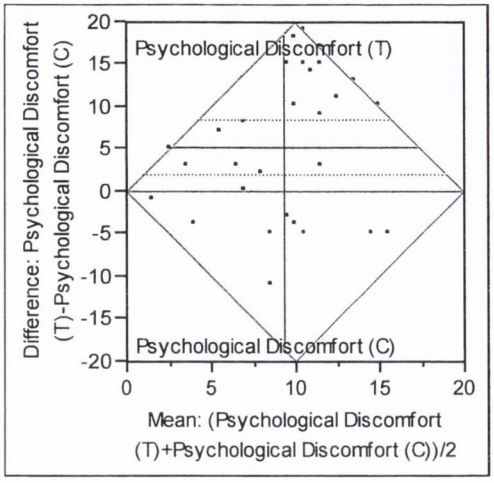
Functional Limitation (T)	14.8621	t-Ratio	2.799624
Functional Limitation (C)	9.65517	DF	28
Mean Difference	5.2069	Prob > |t|	0.0092
Std Error	1.85986	Prob > t	0.0046
Upper95%	9.01664	Prob < t	0.9954
Lower95%	1.39716		
N	29		
Correlation	-0.1137		

Wilcoxon Sign-Rank

	Functional Limitation (T)-Functional Limitation (C)
Test Statistic	97.500
Prob > |z|	0.0160
Prob > z	0.0080
Prob < z	0.9920

Matched Pairs (Psychological Discomfort)

Difference: Psychological Discomfort (T)-Psychological Discomfort (C)



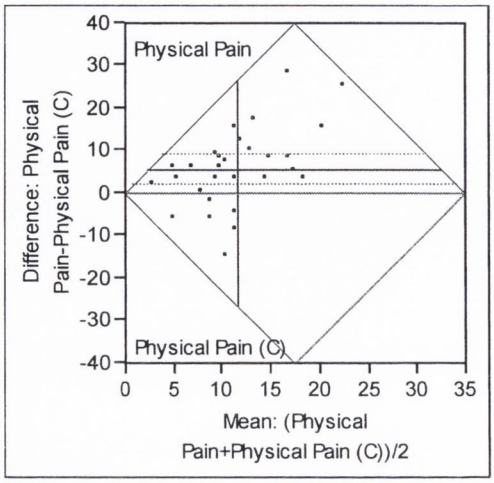
Psychological Discomfort (T)	12	t-Ratio	3.358386
Psychological Discomfort (C)	6.68966	DF	28
Mean Difference	5.31034	Prob > |t|	0.0023
Std Error	1.58122	Prob > t	0.0011
Upper95%	8.54933	Prob < t	0.9989
Lower95%	2.07136		
N	29		
Correlation	-0.1764		

Wilcoxon Sign-Rank

	Psychological Discomfort (T)-Psychological Discomfort (C)
Test Statistic	119.000
Prob > |z|	0.0045
Prob > z	0.0022
Prob < z	0.9978

Matched Pairs (Physical Pain)

Difference: Physical Pain-Physical Pain (C)



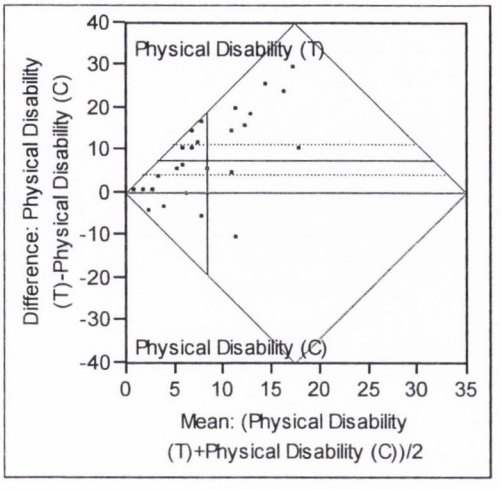
Physical Pain	14.4138	t-Ratio	3.168569
Physical Pain (C)	8.93103	DF	28
Mean Difference	5.48276	Prob > |t|	0.0037
Std Error	1.73036	Prob > t	0.0018
Upper95%	9.02724	Prob < t	0.9982
Lower95%	1.93828		
N	29		
Correlation	0.01788		

Wilcoxon Sign-Rank

	Physical Pain-Physical Pain (C)
Test Statistic	125.500
Prob > |z|	0.0024
Prob > z	0.0012
Prob < z	0.9988

Matched Pairs (Physical Disability)

Difference: Physical Disability (T)-Physical Disability (C)



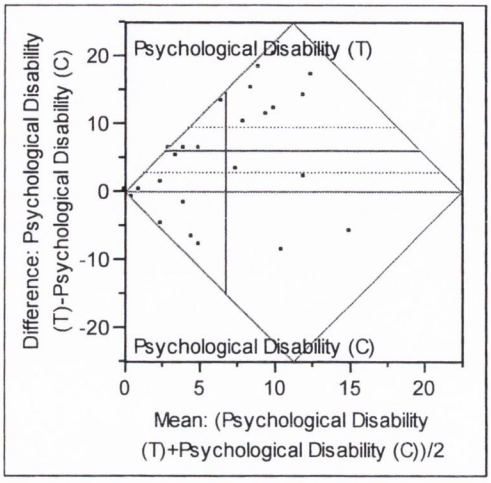
Physical Disability (T)	12.1724	t-Ratio	4.155868
Physical Disability (C)	4.62069	DF	28
Mean Difference	7.55172	Prob > |t|	0.0003
Std Error	1.81712	Prob > t	0.0001
Upper95%	11.2739	Prob < t	0.9999
Lower95%	3.82952		
N	29		
Correlation	-0.0827		

Wilcoxon Sign-Rank

	Physical Disability (T)-Physical Disability (C)
Test Statistic	132.000
Prob > |z|	0.0002
Prob > z	<.0001
Prob < z	0.9999

Matched Pairs (Psychological Disability)

Difference: Psychological Disability (T)-Psychological Disability (C)



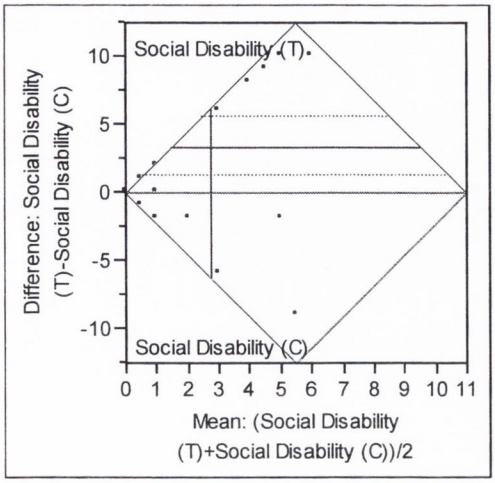
Psychological Disability (T)	9.72414	t-Ratio	3.699084
Psychological Disability (C)	3.68966	DF	28
Mean Difference	6.03448	Prob > |t|	0.0009
Std Error	1.63135	Prob > t	0.0005
Upper95%	9.37614	Prob < t	0.9995
Lower95%	2.69282		
N	29		
Correlation	-0.1171		

Wilcoxon Sign-Rank

	Psychological Disability (T)-Psychological Disability (C)
Test Statistic	124.500
Prob > |z|	0.0013
Prob > z	0.0007
Prob < z	0.9993

Matched Pairs (Social Disability)

Difference: Social Disability (T)-Social Disability (C)



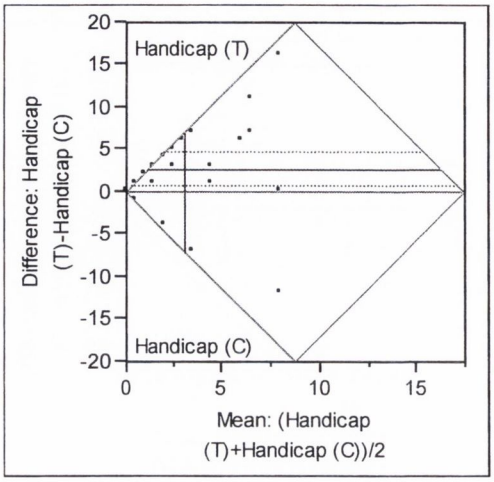
Social Disability (T)	4.44828	t-Ratio	3.274472
Social Disability (C)	1.06897	DF	28
Mean Difference	3.37931	Prob > |t|	0.0028
Std Error	1.03202	Prob > t	0.0014
Upper95%	5.4933	Prob < t	0.9986
Lower95%	1.26532		
N	29		
Correlation	-0.3049		

Wilcoxon Sign-Rank

	Social Disability (T)-Social Disability (C)
Test Statistic	85.000
Prob > |z|	0.0030
Prob > z	0.0015
Prob < z	0.9985

Matched Pairs (Handicap)

Difference: Handicap (T)-Handicap (C)



Handicap (T)	4.44828	t-Ratio	2.77612
Handicap (C)	1.75862	DF	28
Mean Difference	2.68966	Prob > |t|	0.0097
Std Error	0.96885	Prob > t	0.0048
Upper95%	4.67426	Prob < t	0.9952
Lower95%	0.70505		
N	29		
Correlation	-0.0504		

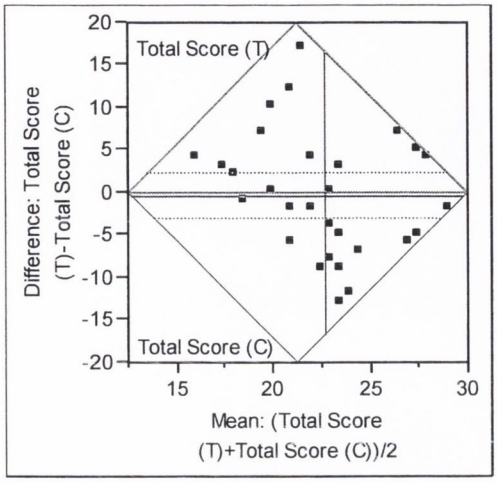
Wilcoxon Sign-Rank

	Handicap (T)-Handicap (C)
Test Statistic	103.500
Prob > |z|	0.0030
Prob > z	0.0015
Prob < z	0.9985

Severe Hypodontia (Rosenberg)

Matched Pairs

Difference: Total Score (T)-Total Score (C)



Total Score (T)	22.4483	t-Ratio	-0.3344
Total Score (C)	22.8966	DF	28
Mean Difference	-0.4483	Prob > |t|	0.7406
Std Error	1.34053	Prob > t	0.6297
Upper95%	2.29767	Prob < t	0.3703
Lower95%	-3.1942		
N	29		
Correlation	-0.097		

Wilcoxon Sign-Rank

	Total Score (T)-Total Score (C)
Test Statistic	-20.500
Prob > |z|	0.6310
Prob > z	0.6845
Prob < z	0.3155

Moderate hypodontia patients versus controls

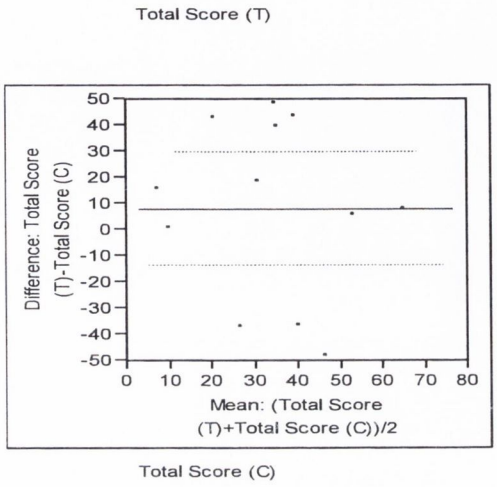
Oral Health Impact Profile

Rosenberg Self-Esteem Scale

Adult Moderate Hypodontia (OHIP-49)

Matched Pairs (Total Scores)

Difference: Total Score (T)-Total Score (C)



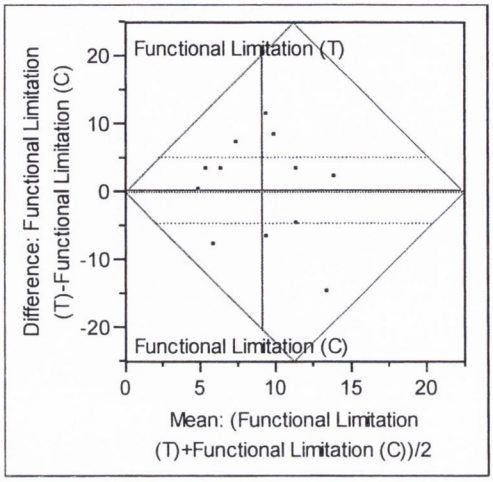
Total Score (T)	38.25	t-Ratio	0.794682
Total Score (C)	30.5	DF	11
Mean Difference	7.75	Prob > |t|	0.4436
Std Error	9.75233	Prob > t	0.2218
Upper95%	29.2147	Prob < t	0.7782
Lower95%	-13.715		
N	12		
Correlation	-0.0049		

Wilcoxon Sign-Rank

	Total Score (T)-Total Score (C)
Test Statistic	11.000
Prob > |z|	0.3652
Prob > z	0.1826
Prob < z	0.8174

Matched Pairs (Functional Limitation)

Difference: Functional Limitation (T)-Functional Limitation (C)



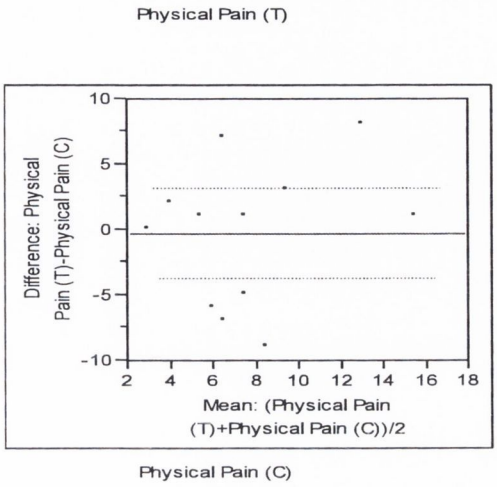
Functional Limitation (T)	9.25	t-Ratio	0.076431
Functional Limitation (C)	9.08333	DF	11
Mean Difference	0.16667	Prob > |t|	0.9404
Std Error	2.18061	Prob > t	0.4702
Upper95%	4.96615	Prob < t	0.5298
Lower95%	-4.6328		
N	12		
Correlation	-0.2047		

Wilcoxon Sign-Rank

	Functional Limitation (T)-Functional Limitation (C)
Test Statistic	2.000
Prob > |z|	0.8838
Prob > z	0.4419
Prob < z	0.5581

Matched Pairs (Physical Pain)

Difference: Physical Pain (T)-Physical Pain (C)



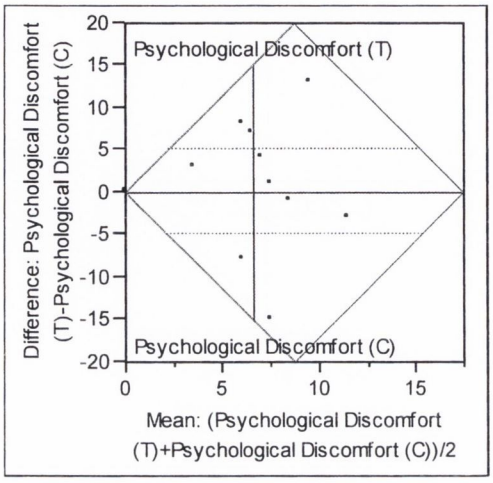
Physical Pain (T)	7.58333	t-Ratio	-0.21453
Physical Pain (C)	7.91667	DF	11
Mean Difference	-0.3333	Prob > |t|	0.8341
Std Error	1.55375	Prob > t	0.5830
Upper95%	3.08645	Prob < t	0.4170
Lower95%	-3.7531		
N	12		
Correlation	0.27956		

Wilcoxon Sign-Rank

	Physical Pain (T)-Physical Pain (C)
Test Statistic	0.500
Prob > |z|	0.9893
Prob > z	0.4946
Prob < z	0.5054

Matched Pairs (Psychological Discomfort)

Difference: Psychological Discomfort (T)-Psychological Discomfort (C)



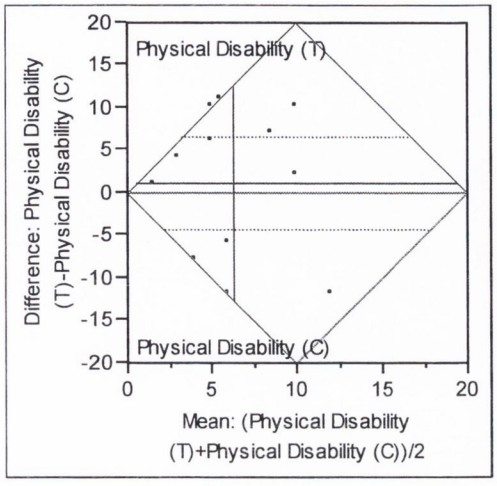
Psychological Discomfort (T)	6.66667	t-Ratio	0.036963
Psychological Discomfort (C)	6.58333	DF	11
Mean Difference	0.08333	Prob > |t|	0.9712
Std Error	2.25448	Prob > t	0.4856
Upper95%	5.04542	Prob < t	0.5144
Lower95%	-4.8788		
N	12		
Correlation	-0.2913		

Wilcoxon Sign-Rank

	Psychological Discomfort (T)-Psychological Discomfort (C)
Test Statistic	1.000
Prob > |z|	0.9521
Prob > z	0.4761
Prob < z	0.5239

Matched Pairs (Physical Disability)

Difference: Physical Disability (T)-Physical Disability (C)



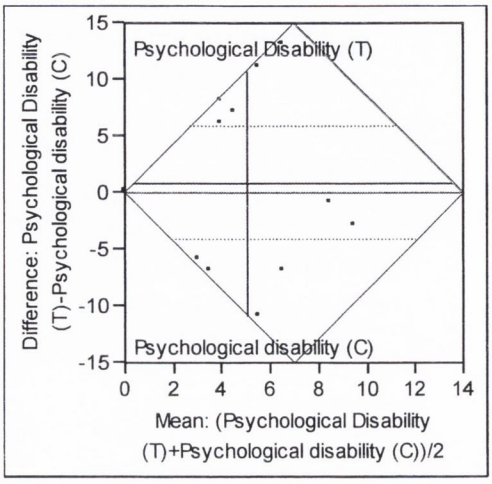
Physical Disability (T)	6.91667	t-Ratio	0.4398
Physical Disability (C)	5.83333	DF	11
Mean Difference	1.08333	Prob > |t|	0.6686
Std Error	2.46324	Prob > t	0.3343
Upper95%	6.50489	Prob < t	0.6657
Lower95%	-4.3382		
N	12		
Correlation	-0.3007		

Wilcoxon Sign-Rank

	Physical Disability (T)-Physical Disability (C)
Test Statistic	4.500
Prob > |z|	0.7495
Prob > z	0.3748
Prob < z	0.6252

Matched Pairs (Psychological Disability

Difference: Psychological Disability (T)-Psychological disability (C)



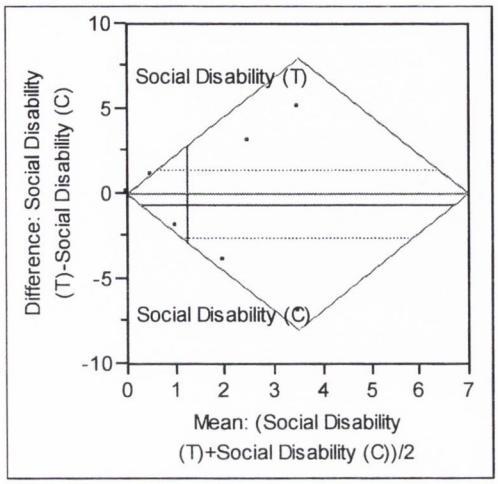
Psychological Disability (T)	5.5	t-Ratio	0.362999
Psychological disability (C)	4.66667	DF	11
Mean Difference	0.83333	Prob > |t|	0.7235
Std Error	2.29569	Prob > t	0.3617
Upper95%	5.88612	Prob < t	0.6383
Lower95%	-4.2195		
N	12		
Correlation	-0.4207		

Wilcoxon Sign-Rank

	Psychological Disability (T)-Psychological disability (C)
Test Statistic	5.000
Prob > |z|	0.6846
Prob > z	0.3423
Prob < z	0.6577

Matched Pairs (Social Disability)

Difference: Social Disability (T)-Social Disability (C)



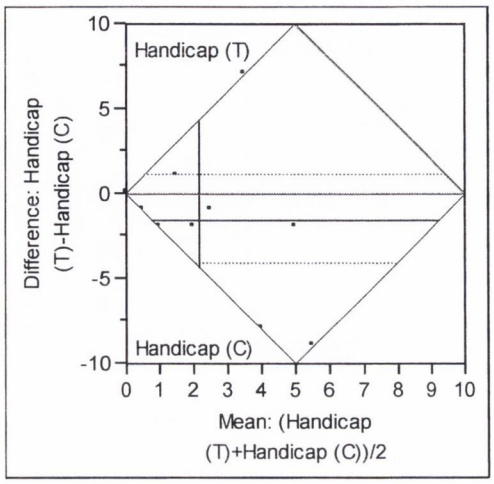
Social Disability (T)	0.91667	t-Ratio	-0.71528
Social Disability (C)	1.58333	DF	11
Mean Difference	-0.6667	Prob > |t|	0.4893
Std Error	0.93203	Prob > t	0.7553
Upper95%	1.38473	Prob < t	0.2447
Lower95%	-2.7181		
N	12		
Correlation	-0.1501		

Wilcoxon Sign-Rank

	Social Disability (T)-Social Disability (C)
Test Statistic	-4.000
Prob > |z|	0.5469
Prob > z	0.7266
Prob < z	0.2734

Matched Pairs (Handicap)

Difference: Handicap (T)-Handicap (C)



Handicap (T)	1.41667	t-Ratio	-1.27395
Handicap (C)	2.91667	DF	11
Mean Difference	-1.5	Prob > |t|	0.2289
Std Error	1.17744	Prob > t	0.8855
Upper95%	1.09153	Prob < t	0.1145
Lower95%	-4.0915		
N	12		
Correlation	-0.0579		

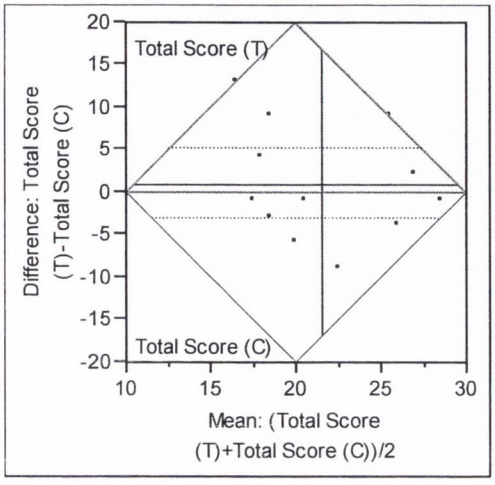
Wilcoxon Sign-Rank

	Handicap (T)-Handicap (C)
Test Statistic	-17.000
Prob > |z|	0.0742
Prob > z	0.9629
Prob < z	0.0371

Moderate Hypodontia (Rosenberg)

Matched Pairs

Difference: Total Score (T)-Total Score (C)



Total Score (T)	22.0833	t-Ratio	0.522233
Total Score (C)	21.0833	DF	11
Mean Difference	1	Prob > |t|	0.6119
Std Error	1.91485	Prob > t	0.3059
Upper95%	5.21457	Prob < t	0.6941
Lower95%	-3.2146		
N	12		
Correlation	0.23019		

Wilcoxon Sign-Rank

	Total Score (T)-Total Score (C)
Test Statistic	3.500
Prob > |z|	0.8091
Prob > z	0.4045
Prob < z	0.5955

Amelogenesis Imperfecta patients versus controls

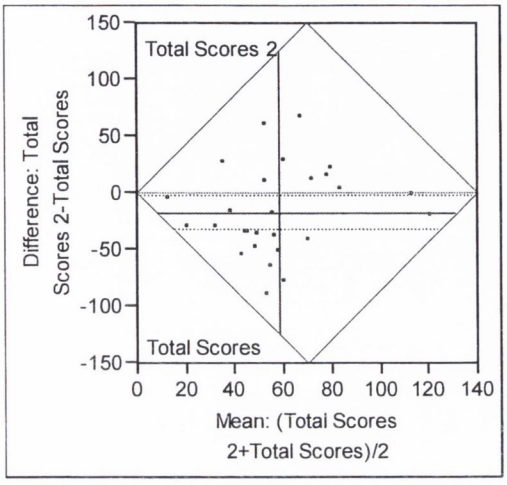
Oral Health Impact Profile

Rosenberg Self-Esteem Scale

Adult Amelogenesis Imperfecta OHIP-49

Matched Pairs (Total Scores)

Difference: Total Scores 2-Total Scores



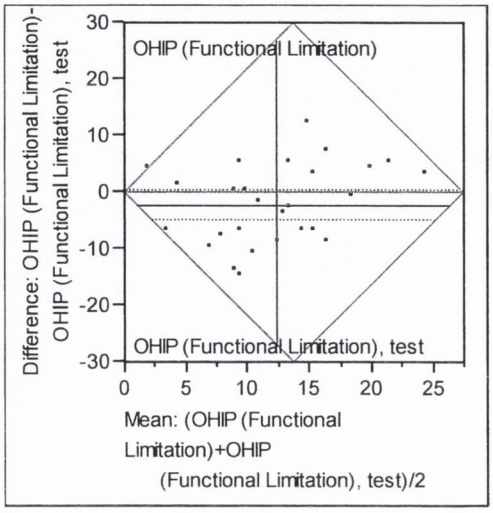
Total Scores 2	48.8519	t-Ratio	-2.4821
Total Scores	67.4074	DF	26
Mean Difference	-18.556	Prob > |t|	0.0198
Std Error	7.47576	Prob > t	0.9901
Upper95%	-3.1889	Prob < t	0.0099
Lower95%	-33.922		
N	27		
Correlation	0.20343		

Wilcoxon Sign-Rank

	Total Scores 2-Total Scores
Test Statistic	-94.500
Prob > |z|	0.0201
Prob > z	0.9900
Prob < z	0.0100

Matched Pairs (Functional Limitation)

Difference: OHIP (Functional Limitation)-OHIP (Functional Limitation), test



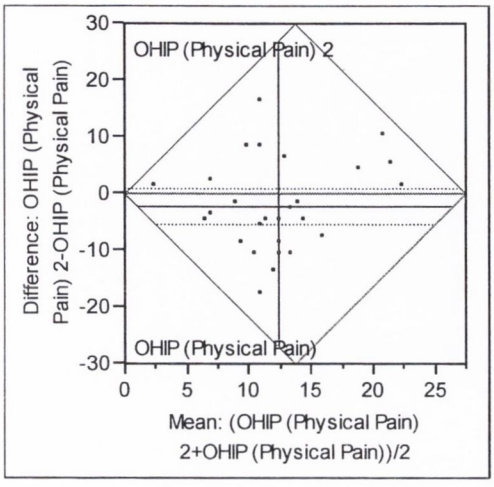
OHIP (Functional Limitation)	11.1481	t-Ratio	-1.79942
OHIP (Functional Limitation), test	13.5556	DF	26
Mean Difference	-2.4074	Prob > |t|	0.0836
Std Error	1.33788	Prob > t	0.9582
Upper95%	0.34264	Prob < t	0.0418
Lower95%	-5.1575		
N	27		
Correlation	0.42894		

Wilcoxon Sign-Rank

	OHIP (Functional Limitation)-OHIP (Functional Limitation), test
Test Statistic	-64.000
Prob > |z|	0.0844
Prob > z	0.9578
Prob < z	0.0422

Matched Pairs (Physical Pain)

Difference: OHIP (Physical Pain) 2-OHIP (Physical Pain)



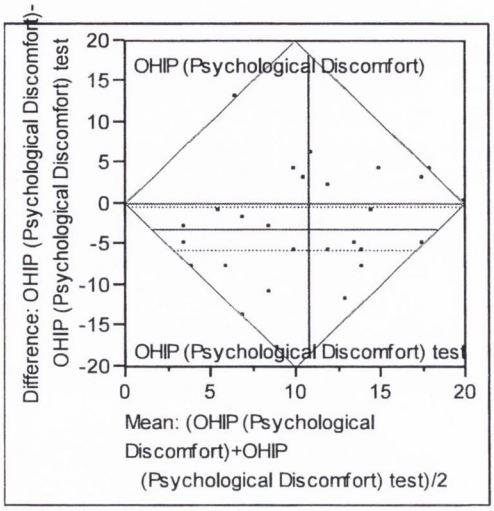
OHIP (Physical Pain) 2	11.1852	t-Ratio	-1.60468
OHIP (Physical Pain)	13.6667	DF	26
Mean Difference	-2.4815	Prob > |t|	0.1206
Std Error	1.54641	Prob > t	0.9397
Upper95%	0.6972	Prob < t	0.0603
Lower95%	-5.6602		
N	27		
Correlation	0.14091		

Wilcoxon Sign-Rank

	OHIP (Physical Pain) 2-OHIP (Physical Pain)
Test Statistic	-68.000
Prob > |z|	0.1027
Prob > z	0.9486
Prob < z	0.0514

Matched Pairs (Psychological Discomfort)

Difference: OHIP (Psychological Discomfort)-OHIP (Psychological Discomfort) test



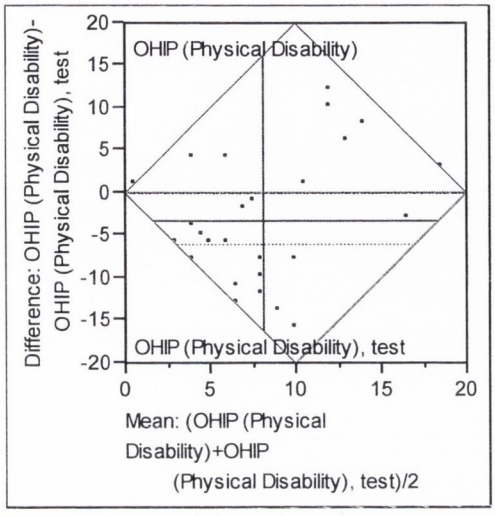
OHIP (Psychological Discomfort)	9.25926	t-Ratio	-2.34745
OHIP (Psychological Discomfort) test	12.3333	DF	26
Mean Difference	-3.0741	Prob > |t|	0.0268
Std Error	1.30954	Prob > t	0.9866
Upper95%	-0.3823	Prob < t	0.0134
Lower95%	-5.7659		
N	27		
Correlation	0.30636		

Wilcoxon Sign-Rank

	OHIP (Psychological Discomfort)-OHIP (Psychological Discomfort) test
Test Statistic	-88.500
Prob > |z|	0.0212
Prob > z	0.9894
Prob < z	0.0106

Matched Pairs (Physical Disability)

Difference: OHIP (Physical Disability)-OHIP (Physical Disability), test



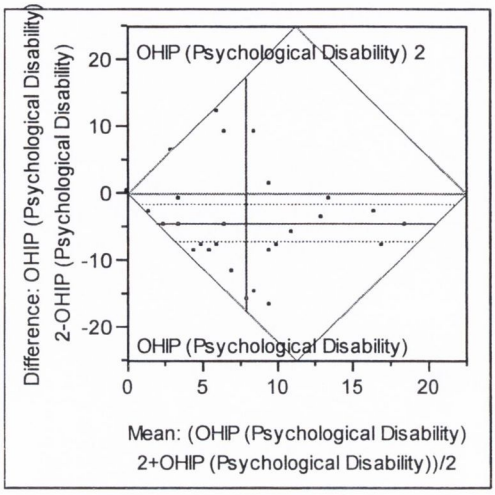
OHIP (Physical Disability)	6.44444	t-Ratio	-2.2622
OHIP (Physical Disability), test	9.7037	DF	26
Mean Difference	-3.2593	Prob > |t|	0.0323
Std Error	1.44075	Prob > t	0.9839
Upper95%	-0.2978	Prob < t	0.0161
Lower95%	-6.2208		
N	27		
Correlation	0.13469		

Wilcoxon Sign-Rank

	OHIP (Physical Disability)-OHIP (Physical Disability), test
Test Statistic	-87.500
Prob > |z|	0.0325
Prob > z	0.9838
Prob < z	0.0162

Matched Pairs (Psychological Disability)

Difference: OHIP (Psychological Disability) 2-OHIP (Psychological Disability)



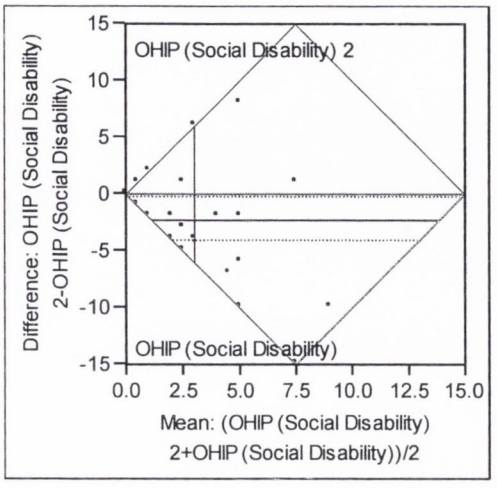
OHIP (Psychological Disability) 2	5.7037	t-Ratio	-3.15319
OHIP (Psychological Disability)	10.1481	DF	26
Mean Difference	-4.4444	Prob > |t|	0.0040
Std Error	1.40951	Prob > t	0.9980
Upper95%	-1.5472	Prob < t	0.0020
Lower95%	-7.3417		
N	27		
Correlation	0.25344		

Wilcoxon Sign-Rank

	OHIP (Psychological Disability) 2-OHIP (Psychological Disability)
Test Statistic	-101.50
Prob > |z|	0.0070
Prob > z	0.9965
Prob < z	0.0035

Matched Pairs (Social Disability)

Difference: OHIP (Social Disability) 2-OHIP (Social Disability)



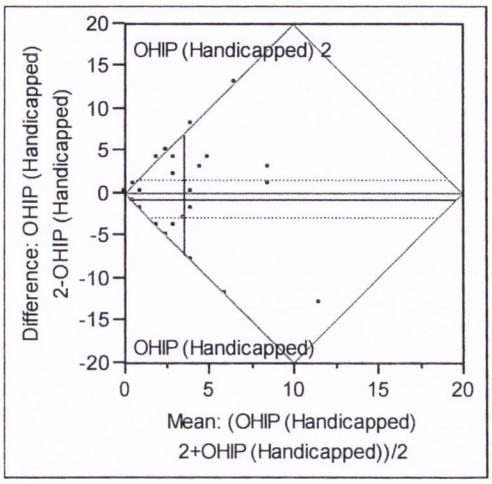
OHIP (Social Disability) 2	1.85185	t-Ratio	-2.43327
OHIP (Social Disability)	4.11111	DF	26
Mean Difference	-2.2593	Prob > |t|	0.0221
Std Error	0.92849	Prob > t	0.9889
Upper95%	-0.3507	Prob < t	0.0111
Lower95%	-4.1678		
N	27		
Correlation	0.00908		

Wilcoxon Sign-Rank

	OHIP (Social Disability) 2-OHIP (Social Disability)
Test Statistic	-84.000
Prob > |z|	0.0126
Prob > z	0.9937
Prob < z	0.0063

Matched Pairs (Handicap)

Difference: OHIP (Handicap) 2-OHIP (Handicap)



OHIP (Handicapped) 2	3.25926	t-Ratio	-0.58389
OHIP (Handicapped)	3.88889	DF	26
Mean Difference	-0.6296	Prob > |t|	0.5643
Std Error	1.07834	Prob > t	0.7178
Upper95%	1.58693	Prob < t	0.2822
Lower95%	-2.8462		
N	27		
Correlation	-0.0285		

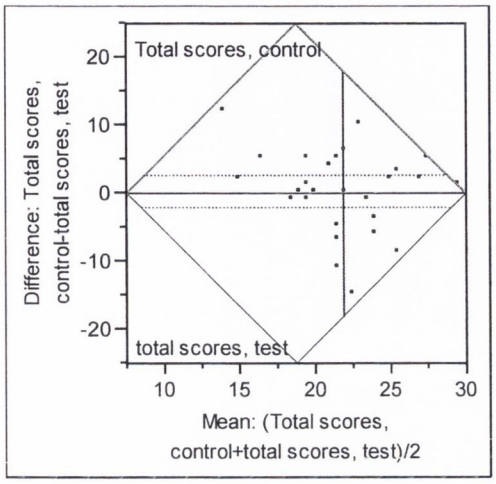
Wilcoxon Sign-Rank

	OHIP (Handicapped) 2-OHIP (Handicapped)
Test Statistic	-20.000
Prob > |z|	0.5776
Prob > z	0.7112
Prob < z	0.2888

Adult Amelogenesis Imperfecta (Rosenberg)

Matched Pairs

Difference: Total scores, control-total scores, test



Total scores, control	21.8889	t-Ratio	0.094297
total scores, test	21.7778	DF	26
Mean Difference	0.11111	Prob > |t|	0.9256
Std Error	1.17831	Prob > t	0.4628
Upper95%	2.53316	Prob < t	0.5372
Lower95%	-2.3109		
N	27		
Correlation	0.1765		

Wilcoxon Sign-Rank

	Total scores, control-total scores, test
Test Statistic	14.000
Prob > |z|	0.6973
Prob > z	0.3487
Prob < z	0.6513

All young hypodontia patients versus controls

Piers Harris Self Concept Scale

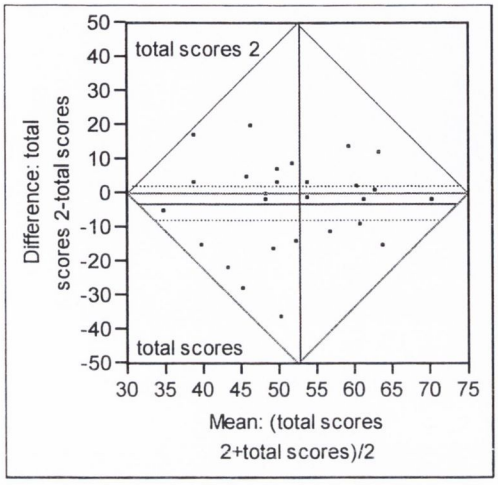
Parental Perception Questionnaire

Family Impact Scale

Piers Harris Self Concept Scale for combined hypodontia patients

Matched Pairs (Total Scores)

Difference: total scores 2-total scores



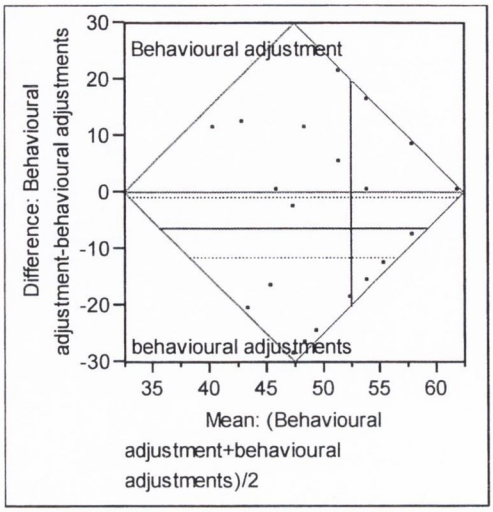
total scores 2	51.0345	t-Ratio	-1.36015
total scores	54.3793	DF	28
Mean Difference	-3.3448	Prob > |t|	0.1846
Std Error	2.45916	Prob > t	0.9077
Upper95%	1.69254	Prob < t	0.0923
Lower95%	-8.3822		
N	29		
Correlation	0.29716		

Wilcoxon Sign-Rank

	total scores 2-total scores
Test Statistic	-46.000
Prob > |z|	0.3029
Prob > z	0.8485
Prob < z	0.1515

Matched Pairs (Behavioural adjustment)

Difference: Behavioural adjustment-behavioural adjustments



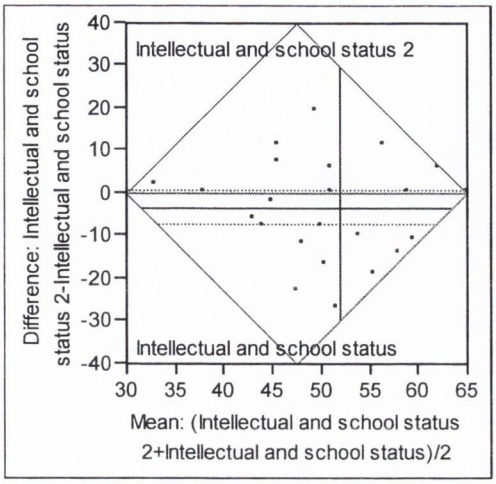
Behavioural adjustment	49.3448	t-Ratio	-2.44431
behavioural adjustments	55.7241	DF	28
Mean Difference	-6.3793	Prob > |t|	0.0211
Std Error	2.60986	Prob > t	0.9895
Upper95%	-1.0333	Prob < t	0.0105
Lower95%	-11.725		
N	29		
Correlation	-0.1368		

Wilcoxon Sign-Rank

	Behavioural adjustment-behavioural adjustments
Test Statistic	-73.000
Prob > |z|	0.0225
Prob > z	0.9888
Prob < z	0.0112

Matched Pairs (Intellectual and School Status)

Difference: Intellectual and school status 2-Intellectual and school status



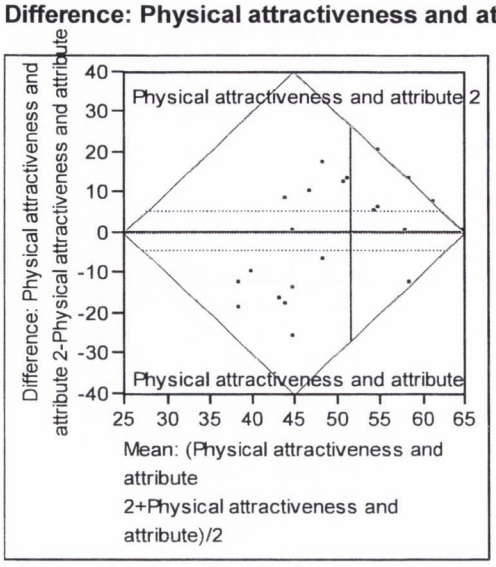
Intellectual and school status 2	50.2759	t-Ratio	-1.75451
Intellectual and school status	53.7931	DF	28
Mean Difference	-3.5172	Prob > |t|	0.0903
Std Error	2.00469	Prob > t	0.9549
Upper95%	0.58917	Prob < t	0.0451
Lower95%	-7.6237		
N	29		
Correlation	0.39756		

Wilcoxon Sign-Rank

	Intellectual and school status 2-Intellectual and school status
Test Statistic	-59.000
Prob > |z|	0.0710
Prob > z	0.9645
Prob < z	0.0355

Matched Pairs (Physical Attractiveness and Attribute)

Difference: Physical attractiveness and attribute 2-Physical attractiveness and attribute



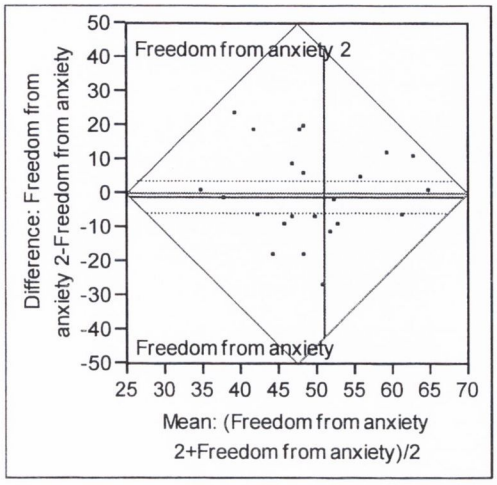
Physical attractiveness and attribute 2	51.8276	t-Ratio	0.193075
Physical attractiveness and attribute	51.3793	DF	28
Mean Difference	0.44828	Prob > |t|	0.8483
Std Error	2.32177	Prob > t	0.4241
Upper95%	5.2042	Prob < t	0.5759
Lower95%	-4.3077		
N	29		
Correlation	0.18325		

Wilcoxon Sign-Rank

	Physical attractiveness and attribute 2-Physical attractiveness and attribute
Test Statistic	0.000
Prob > |z|	1.0000
Prob > z	0.5000
Prob < z	0.5000

Matched Pairs (Freedom from anxiety)

Difference: Freedom from anxiety 2-Freedom from anxiety



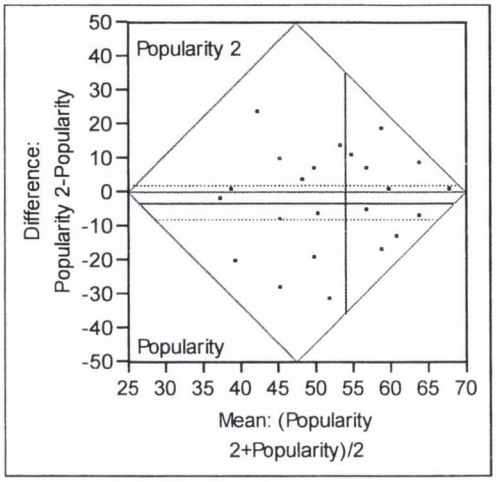
Freedom from anxiety 2	50.2759	t-Ratio	-0.5845
Freedom from anxiety	51.6207	DF	28
Mean Difference	-1.3448	Prob > |t|	0.5636
Std Error	2.30082	Prob > t	0.7182
Upper95%	3.36819	Prob < t	0.2818
Lower95%	-6.0578		
N	29		
Correlation	0.2466		

Wilcoxon Sign-Rank

	Freedom from anxiety 2-Freedom from anxiety
Test Statistic	-20.000
Prob > |z|	0.6205
Prob > z	0.6898
Prob < z	0.3102

Matched Pairs (Popularity)

Difference: Popularity 2-Popularity



Popularity 2	52.4138	t-Ratio	-1.27527
Popularity	55.6207	DF	28
Mean Difference	-3.2069	Prob > |t|	0.2127
Std Error	2.51468	Prob > t	0.8937
Upper95%	1.94418	Prob < t	0.1063
Lower95%	-8.358		
N	29		
Correlation	0.22519		

Wilcoxon Sign-Rank

	Popularity 2-Popularity
Test Statistic	-37.500
Prob > |z|	0.2628
Prob > z	0.8686
Prob < z	0.1314

Matched Pairs (Happiness and satisfaction)

Difference: Happiness and satisfaction 2-Happiness and satisfaction

Happiness and satisfaction 2	49.4483	t-Ratio	0.038527
Happiness and satisfaction	49.3793	DF	28
Mean Difference	0.06897	Prob > |t|	0.9695
Std Error	1.79005	Prob > t	0.4848
Upper95%	3.73571	Prob < t	0.5152
Lower95%	-3.5978		
N	29		
Correlation	0.31236		

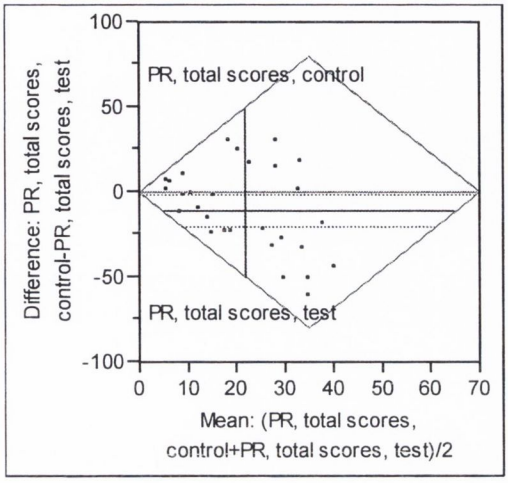
Wilcoxon Sign-Rank

	Happiness and satisfaction 2-Happiness and satisfaction
Test Statistic	0.500
Prob > |z|	0.9926
Prob > z	0.4963
Prob < z	0.5037

Parental Perception of Combined Hypodontia Patients

Matched Pairs (total scores)

Difference: PR, total scores, control-PR, total scores, test



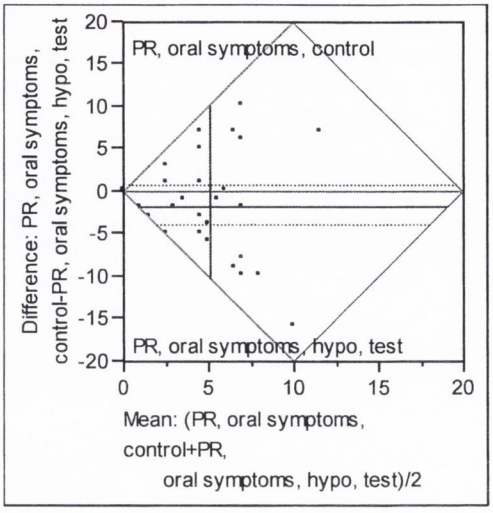
PR, total scores, control	16.1724	t-Ratio	-2.44796
PR, total scores, test	27.4138	DF	28
Mean Difference	-11.241	Prob > |t|	0.0209
Std Error	4.59215	Prob > t	0.9896
Upper95%	-1.8348	Prob < t	0.0104
Lower95%	-20.648		
N	29		
Correlation	-0.1544		

Wilcoxon Sign-Rank

	PR, total scores, control-PR, total scores, test
Test Statistic	-89.500
Prob > |z|	0.0285
Prob > z	0.9857
Prob < z	0.0143

Matched Pairs (Oral Symptoms)

Difference: PR, oral symptoms, control-PR, oral symptoms, hypo, test



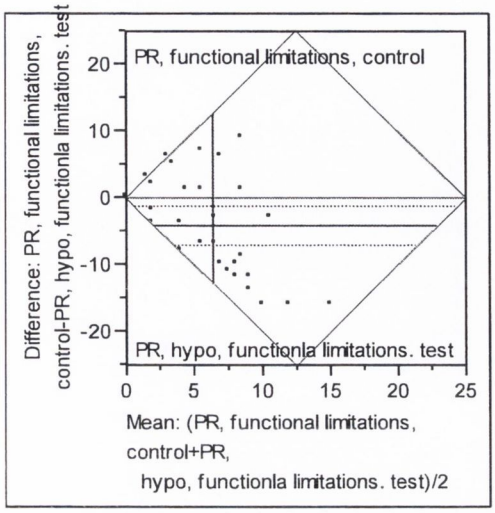
PR, oral symptoms, control	4.27586	t-Ratio	-1.55595
PR, oral symptoms, hypo, test	6.03448	DF	28
Mean Difference	-1.7586	Prob > |t|	0.1310
Std Error	1.13025	Prob > t	0.9345
Upper95%	0.5566	Prob < t	0.0655
Lower95%	-4.0738		
N	29		
Correlation	-0.1686		

Wilcoxon Sign-Rank

	PR, oral symptoms, control-PR, oral symptoms, hypo, test
Test Statistic	-62.000
Prob > |z|	0.1386
Prob > z	0.9307
Prob < z	0.0693

Matched Pairs (Functional Limitations)

Difference: PR, functional limitations, control-PR, hypo, functional limitations. Test



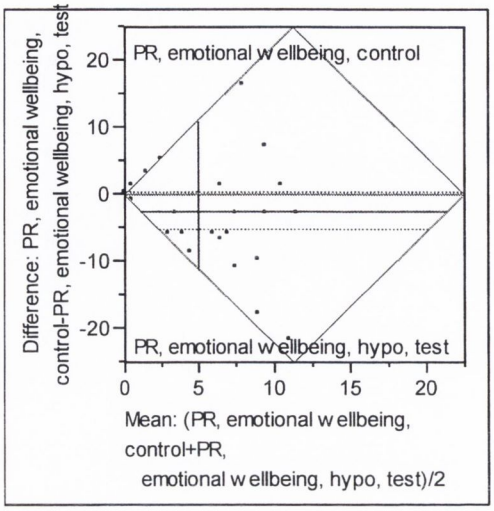
PR, functional limitations, control	4.24138	t-Ratio	-3.02976
PR, hypo, functionla limitations. test	8.48276	DF	28
Mean Difference	-4.2414	Prob > |t|	0.0052
Std Error	1.39991	Prob > t	0.9974
Upper95%	-1.3738	Prob < t	0.0026
Lower95%	-7.109		
N	29		
Correlation	-0.1157		

Wilcoxon Sign-Rank

	PR, functional limitations, control-PR, hypo, functionla limitations. test
Test Statistic	-116.00
Prob > |z|	0.0058
Prob > z	0.9971
Prob < z	0.0029

Matched Pairs (Emotional Well-being)

Difference: PR, emotional well-being, control-PR, emotional well-being, hypo, test



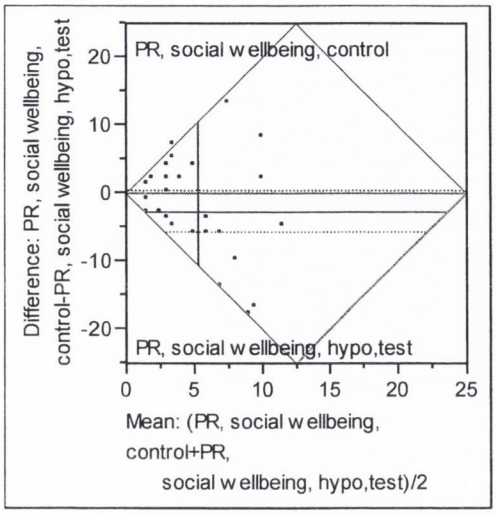
PR, emotional wellbeing, control	3.72414	t-Ratio	-1.78406
PR, emotional wellbeing, hypo, test	6.17241	DF	28
Mean Difference	-2.4483	Prob > |t|	0.0853
Std Error	1.3723	Prob > t	0.9574
Upper95%	0.36276	Prob < t	0.0426
Lower95%	-5.2593		
N	29		
Correlation	0.04229		

Wilcoxon Sign-Rank

	PR, emotional wellbeing, control-PR, emotional wellbeing, hypo, test
Test Statistic	-71.500
Prob > |z|	0.0843
Prob > z	0.9579
Prob < z	0.0421

Matched Pairs (Social Well-being)

Difference: PR, social wellbeing, control-PR, social wellbeing, hypo,test



PR, social wellbeing, control	3.93103	t-Ratio	-1.87872
PR, social wellbeing, hypo,test	6.72414	DF	28
Mean Difference	-2.7931	Prob > |t|	0.0707
Std Error	1.48671	Prob > t	0.9646
Upper95%	0.25228	Prob < t	0.0354
Lower95%	-5.8385		
N	29		
Correlation	-0.1909		

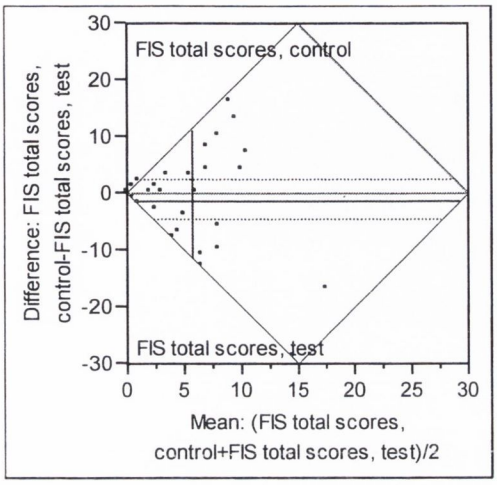
Wilcoxon Sign-Rank

	PR, social wellbeing, control-PR, social wellbeing, hypo,test
Test Statistic	-75.500
Prob > |z|	0.0851
Prob > z	0.9575
Prob < z	0.0425

Family Impact Scale of Combined Hypodontia Patients

Matched Pairs (FIS total scores)

Difference: FIS total scores, control-FIS total scores, test



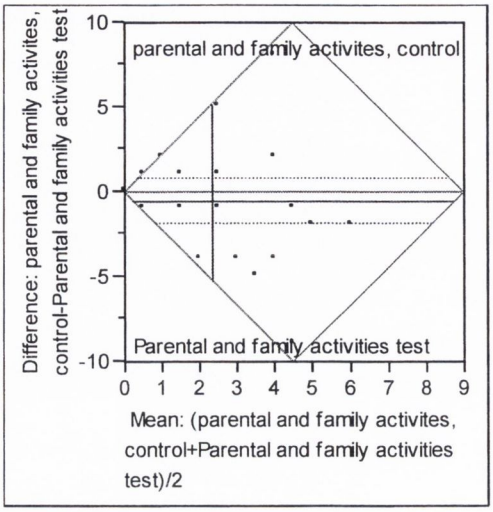
FIS total scores, control	5.03448	t-Ratio	-0.75001
FIS total scores, test	6.31034	DF	28
Mean Difference	-1.2759	Prob > |t|	0.4595
Std Error	1.70113	Prob > t	0.7702
Upper95%	2.20874	Prob < t	0.2298
Lower95%	-4.7605		
N	29		
Correlation	-0.0754		

Wilcoxon Sign-Rank

	FIS total scores, control-FIS total scores, test
Test Statistic	-16.500
Prob > |z|	0.6659
Prob > z	0.6670
Prob < z	0.3330

Matched Pairs (Parental and Family Activities)

Difference: parental and family activities, control-Parental and family activities test



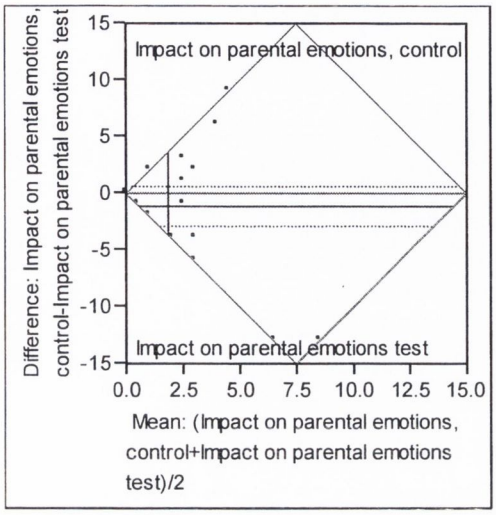
parental and family activites, control	2.06897	t-Ratio	-0.87727
Parental and family activities test	2.62069	DF	28
Mean Difference	-0.5517	Prob > |t|	0.3878
Std Error	0.62891	Prob > t	0.8061
Upper95%	0.73655	Prob < t	0.1939
Lower95%	-1.84		
N	29		
Correlation	-0.0281		

Wilcoxon Sign-Rank

	parental and family activites, control-Parental and family activities test
Test Statistic	-40.500
Prob > |z|	0.3580
Prob > z	0.8210
Prob < z	0.1790

Matched Pairs (Impact on Parental Emotions)

Difference: Impact on parental emotions, control-Impact on parental emotions test



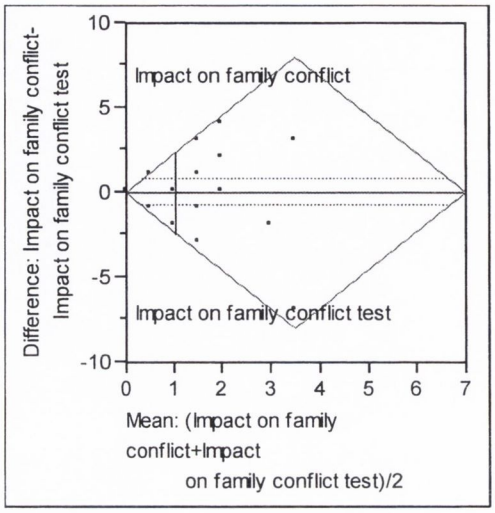
Impact on parental emotions, control	1.24138	t-Ratio	-1.5176
Impact on parental emotions test	2.48276	DF	28
Mean Difference	-1.2414	Prob > |t|	0.1403
Std Error	0.81799	Prob > t	0.9298
Upper95%	0.43419	Prob < t	0.0702
Lower95%	-2.9169		
N	29		
Correlation	-0.076		

Wilcoxon Sign-Rank

	Impact on parental emotions, control-Impact on parental emotions test
Test Statistic	-49.500
Prob > |z|	0.1069
Prob > z	0.9466
Prob < z	0.0534

Matched Pairs (Impact on Family Conflict)

Difference: Impact on family conflict-Impact on family conflict test



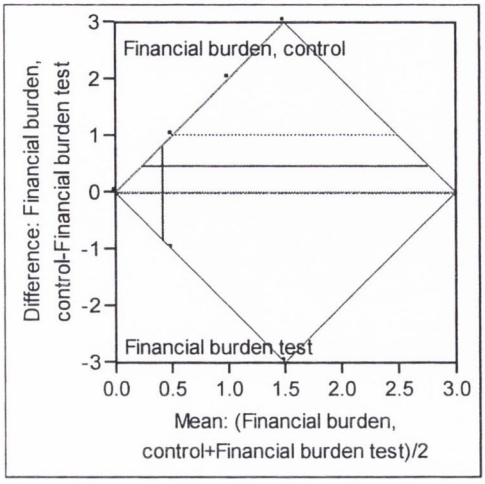
Impact on family conflict	1.06897	t-Ratio	0.089341
Impact on family conflict test	1.03448	DF	28
Mean Difference	0.03448	Prob > |t|	0.9294
Std Error	0.38597	Prob > t	0.4647
Upper95%	0.82511	Prob < t	0.5353
Lower95%	-0.7561		
N	29		
Correlation	0.01528		

Wilcoxon Sign-Rank

	Impact on family conflict-Impact on family conflict test
Test Statistic	9.000
Prob > |z|	0.7357
Prob > z	0.3678
Prob < z	0.6322

Matched Pairs (Financial Burden)

Difference: Financial burden, control-Financial burden test



Financial burden, control	0.65517	t-Ratio	1.960392
Financial burden test	0.17241	DF	28
Mean Difference	0.48276	Prob > |t|	0.0600
Std Error	0.24626	Prob > t	0.0300
Upper95%	0.98719	Prob < t	0.9700
Lower95%	-0.0217		
N	29		
Correlation	-0.1803		

Wilcoxon Sign-Rank

	Financial burden, control-Financial burden test
Test Statistic	23.500
Prob > |z|	0.0732
Prob > z	0.0366
Prob < z	0.9634

Severe young hypodontia patients versus controls

Piers Harris Self Concept Scale

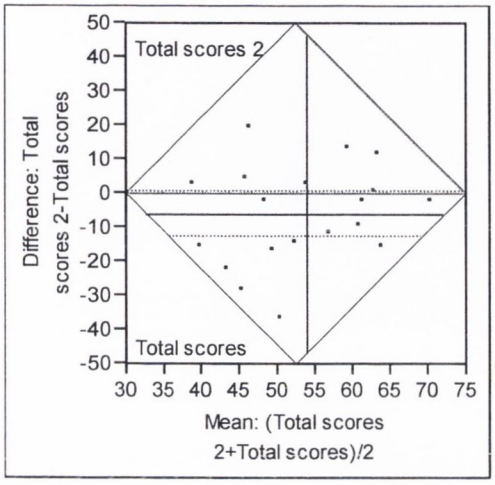
Parental Perception Questionnaire

Family Impact Scale

Piers Harris Self Concept Scale for Severe Hypodontia patients

Matched Pairs

Difference: Total scores 2-Total scores



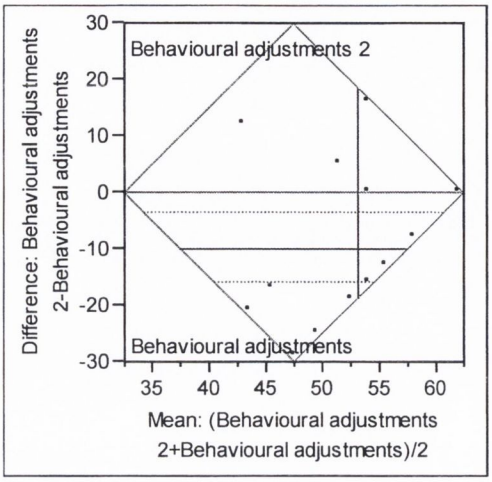
Total scores 2	50.9	t-Ratio	-1.85985
Total scores	57	DF	19
Mean Difference	-6.1	Prob > |t|	0.0785
Std Error	3.27984	Prob > t	0.9608
Upper95%	0.76479	Prob < t	0.0392
Lower95%	-12.965		
N	20		
Correlation	0.21839		

Wilcoxon Sign-Rank

	Total scores 2-Total scores
Test Statistic	-42.000
Prob > |z|	0.0932
Prob > z	0.9534
Prob < z	0.0466

Matched Pairs (Behavioural adjustments)

Difference: Behavioural adjustments 2-Behavioural adjustments



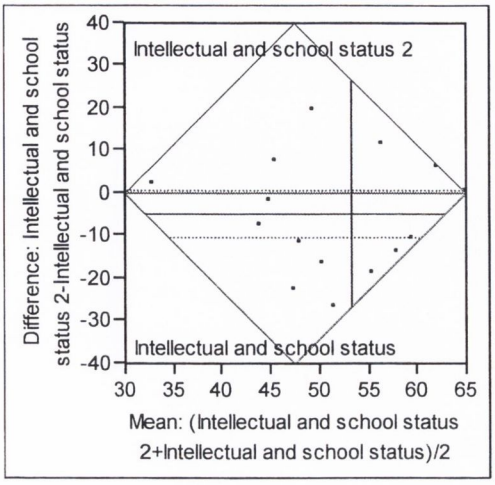
Behavioural adjustments 2	48.2	t-Ratio	-3.41059
Behavioural adjustments	58.1	DF	19
Mean Difference	-9.9	Prob > |t|	0.0029
Std Error	2.90272	Prob > t	0.9985
Upper95%	-3.8245	Prob < t	0.0015
Lower95%	-15.975		
N	20		
Correlation	-0.1116		

Wilcoxon Sign-Rank

	Behavioural adjustments 2-Behavioural adjustments
Test Statistic	-54.500
Prob > |z|	0.0027
Prob > z	0.9986
Prob < z	0.0014

Matched Pairs (Intellectual and School Status)

Difference: Intellectual and school status 2-Intellectual and school status



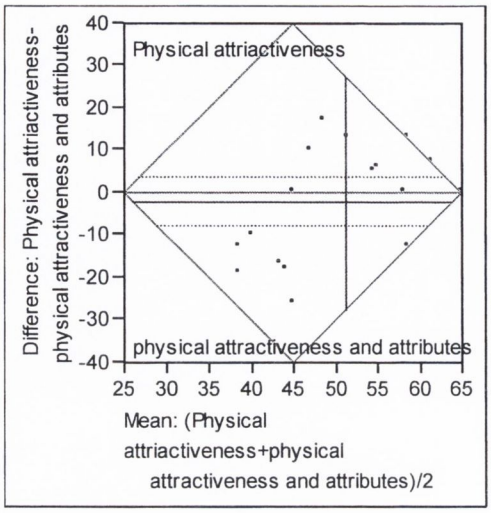
Intellectual and school status 2	50.8	t-Ratio	-1.88792
Intellectual and school status	55.85	DF	19
Mean Difference	-5.05	Prob > |t|	0.0744
Std Error	2.67491	Prob > t	0.9628
Upper95%	0.54865	Prob < t	0.0372
Lower95%	-10.649		
N	20		
Correlation	0.37143		

Wilcoxon Sign-Rank

	Intellectual and school status 2-Intellectual and school status
Test Statistic	-39.500
Prob > |z|	0.0617
Prob > z	0.9691
Prob < z	0.0309

Matched Pairs (Physical attractiveness)

Difference: Physical attractiveness-physical attractiveness and attributes



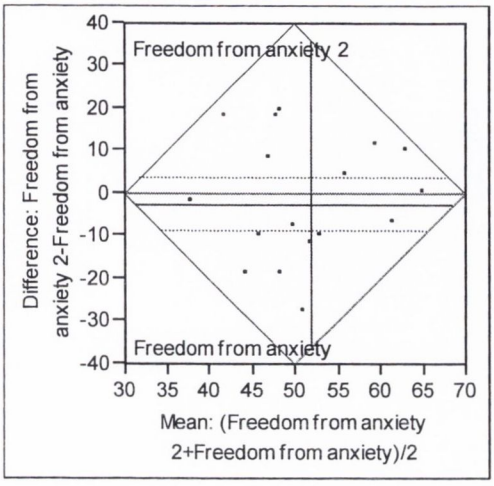
Physical attriactiveness	50.2	t-Ratio	-0.82539
physical attractiveness and attributes	52.45	DF	19
Mean Difference	-2.25	Prob > |t|	0.4194
Std Error	2.72597	Prob > t	0.7903
Upper95%	3.45552	Prob < t	0.2097
Lower95%	-7.9555		
N	20		
Correlation	0.3295		

Wilcoxon Sign-Rank

	Physical attriactiveness-physical attractiveness and attributes
Test Statistic	-16.500
Prob > |z|	0.3154
Prob > z	0.8423
Prob < z	0.1577

Matched Pairs (Freedom from Anxiety)

Difference: Freedom from anxiety 2-Freedom from anxiety



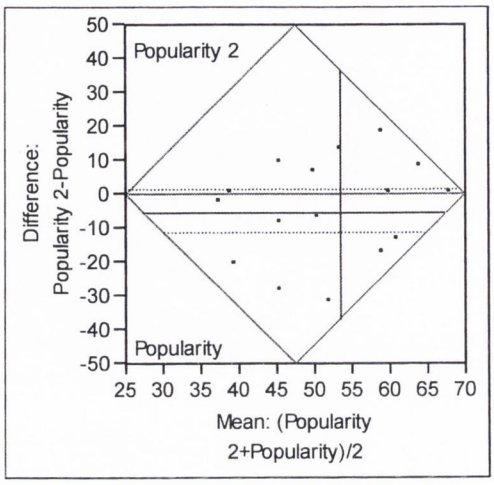
Freedom from anxiety 2	50.6	t-Ratio	-0.87505
Freedom from anxiety	53.2	DF	19
Mean Difference	-2.6	Prob > |t|	0.3925
Std Error	2.97127	Prob > t	0.8038
Upper95%	3.61893	Prob < t	0.1962
Lower95%	-8.8189		
N	20		
Correlation	0.09136		

Wilcoxon Sign-Rank

	Freedom from anxiety 2-Freedom from anxiety
Test Statistic	-19.500
Prob > |z|	0.4469
Prob > z	0.7766
Prob < z	0.2234

Matched Pairs(Popularity)

Difference: Popularity 2-Popularity



Popularity 2	50.7	t-Ratio	-1.81762
Popularity	56.25	DF	19
Mean Difference	-5.55	Prob > |t|	0.0849
Std Error	3.05345	Prob > t	0.9575
Upper95%	0.84094	Prob < t	0.0425
Lower95%	-11.941		
N	20		
Correlation	0.26978		

Wilcoxon Sign-Rank

	Popularity 2-Popularity
Test Statistic	-30.500
Prob > |z|	0.0855
Prob > z	0.9572
Prob < z	0.0428

Matched Pairs (Happiness and satisfaction)

Difference: Happiness and satisfaction 2-Happiness and satisfaction

Happiness and satisfaction 2	49.55	t-Ratio	-0.33693
Happiness and satisfaction	50.25	DF	19
Mean Difference	-0.7	Prob > |t|	0.7399
Std Error	2.07757	Prob > t	0.6301
Upper95%	3.64841	Prob < t	0.3699
Lower95%	-5.0484		
N	20		
Correlation	0.31823		

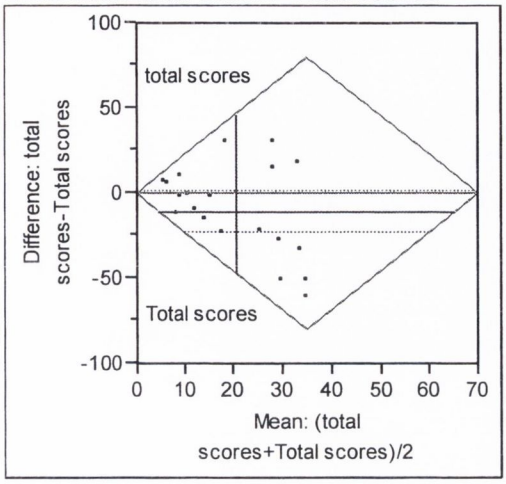
Wilcoxon Sign-Rank

	Happiness and satisfaction 2-Happiness and satisfaction
Test Statistic	-3.500
Prob > |z|	0.8027
Prob > z	0.5986
Prob < z	0.4014

Parental Perception for Severe Hypodontia Patients

Matched Pairs (Total Scores)

Difference: total scores-Total scores



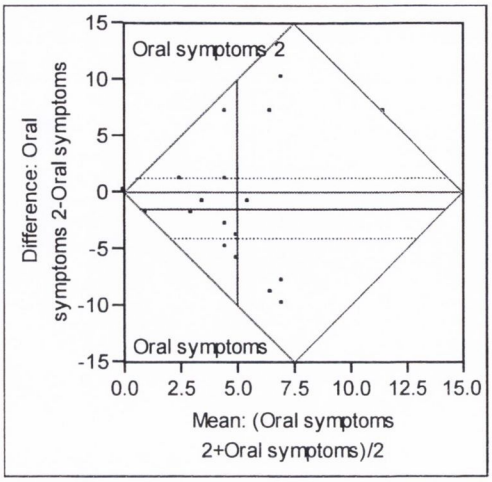
total scores	15.05	t-Ratio	-1.86346
Total scores	25.9	DF	19
Mean Difference	-10.85	Prob > |t|	0.0779
Std Error	5.82249	Prob > t	0.9610
Upper95%	1.33662	Prob < t	0.0390
Lower95%	-23.037		
N	20		
Correlation	-0.2293		

Wilcoxon Sign-Rank

	total scores-Total scores
Test Statistic	-41.000
Prob > |z|	0.1300
Prob > z	0.9350
Prob < z	0.0650

Matched Pairs (Oral symptoms)

Difference: Oral symptoms 2-Oral symptoms



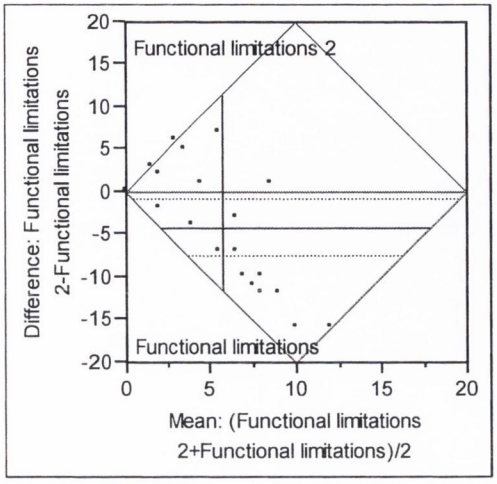
Oral symptoms 2	4.3	t-Ratio	-1.13431
Oral symptoms	5.75	DF	19
Mean Difference	-1.45	Prob > |t|	0.2708
Std Error	1.27831	Prob > t	0.8646
Upper95%	1.22554	Prob < t	0.1354
Lower95%	-4.1255		
N	20		
Correlation	-0.1379		

Wilcoxon Sign-Rank

	Oral symptoms 2-Oral symptoms
Test Statistic	-32.500
Prob > |z|	0.1984
Prob > z	0.9008
Prob < z	0.0992

Matched Pairs (Functional Limitations)

Difference: Functional limitations 2-Functional limitations



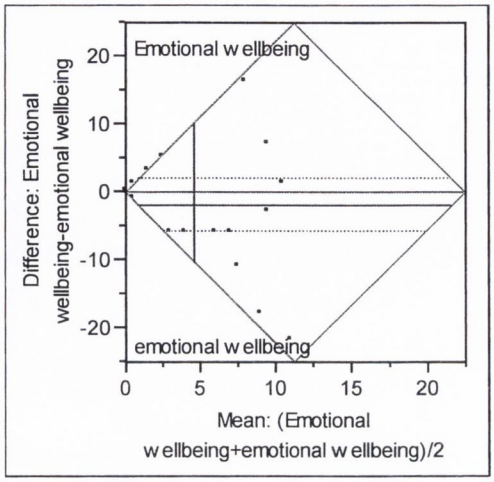
Functional limitations 2	3.6	t-Ratio	-2.60462
Functional limitations	7.85	DF	19
Mean Difference	-4.25	Prob > |t|	0.0174
Std Error	1.63172	Prob > t	0.9913
Upper95%	-0.8348	Prob < t	0.0087
Lower95%	-7.6652		
N	20		
Correlation	-0.2231		

Wilcoxon Sign-Rank

	Functional limitations 2-Functional limitations
Test Statistic	-55.000
Prob > |z|	0.0250
Prob > z	0.9875
Prob < z	0.0125

Matched Pairs (Emotional well-being)

Difference: Emotional wellbeing-emotional wellbeing



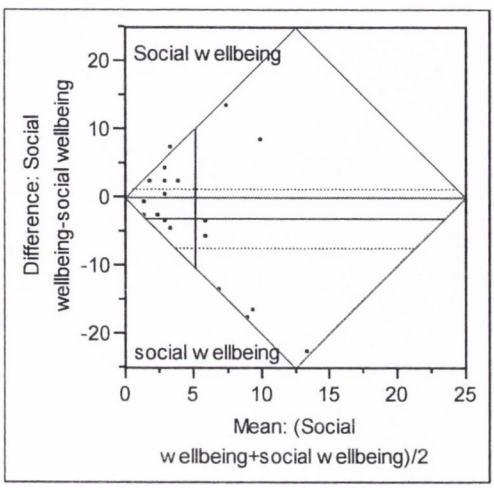
Emotional wellbeing	3.65	t-Ratio	-1.05875
emotional wellbeing	5.65	DF	19
Mean Difference	-2	Prob > |t|	0.3030
Std Error	1.88903	Prob > t	0.8485
Upper95%	1.95378	Prob < t	0.1515
Lower95%	-5.9538		
N	20		
Correlation	-0.0605		

Wilcoxon Sign-Rank

	Emotional wellbeing-emotional wellbeing
Test Statistic	-19.500
Prob > |z|	0.4445
Prob > z	0.7777
Prob < z	0.2223

Matched Pairs (Social Well-being)

Difference: Social well-being-social well-being



Social wellbeing	3.5	t-Ratio	-1.54945
social wellbeing	6.65	DF	19
Mean Difference	-3.15	Prob > |t|	0.1378
Std Error	2.03298	Prob > t	0.9311
Upper95%	1.10509	Prob < t	0.0689
Lower95%	-7.4051		
N	20		
Correlation	-0.3378		

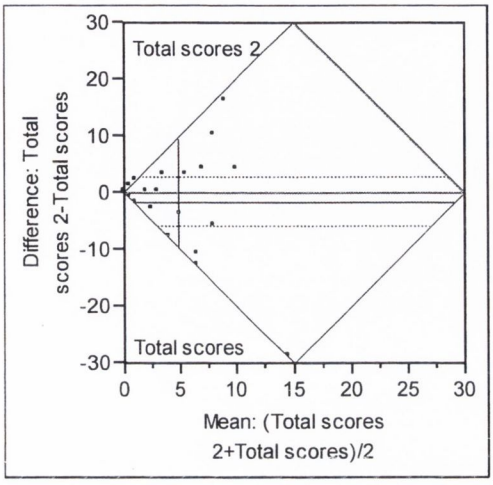
Wilcoxon Sign-Rank

	Social wellbeing-social wellbeing
Test Statistic	-35.000
Prob > |z|	0.1683
Prob > z	0.9159
Prob < z	0.0841

Family Impact Scale for Moderate Hypodontia Patients

Matched Pairs

Difference: Total scores 2-Total scores



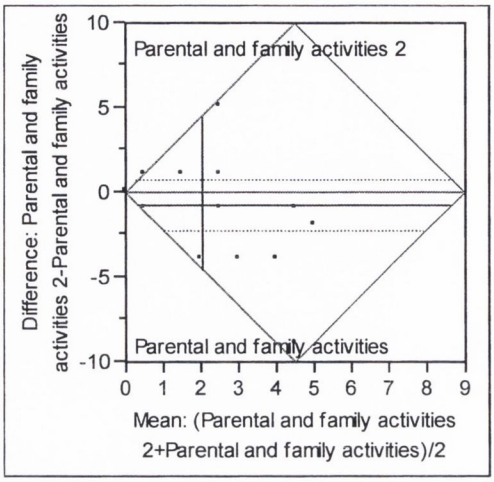
Total scores 2	4.05	t-Ratio	-0.82483
Total scores	5.75	DF	19
Mean Difference	-1.7	Prob > |t|	0.4197
Std Error	2.06104	Prob > t	0.7901
Upper95%	2.61381	Prob < t	0.2099
Lower95%	-6.0138		
N	20		
Correlation	-0.1929		

Wilcoxon Sign-Rank

	Total scores 2-Total scores
Test Statistic	-12.500
Prob > |z|	0.5707
Prob > z	0.7146
Prob < z	0.2854

Matched Pairs (Parental and Family Activities)

Difference: Parental and family activities 2-Parental and family activities



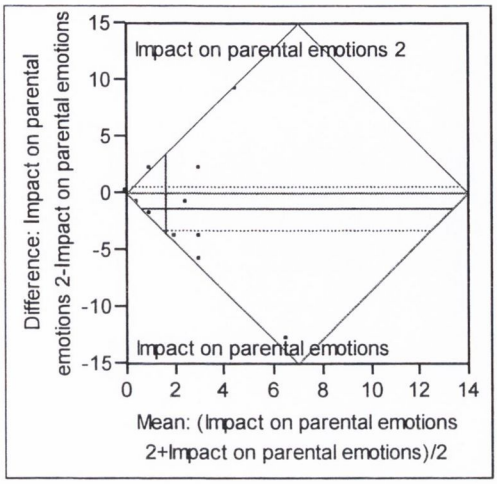
Parental and family activities 2	1.65	t-Ratio	-1.11052
Parental and family activities	2.45	DF	19
Mean Difference	-0.8	Prob > |t|	0.2806
Std Error	0.72038	Prob > t	0.8597
Upper95%	0.70777	Prob < t	0.1403
Lower95%	-2.3078		
N	20		
Correlation	-0.0444		

Wilcoxon Sign-Rank

	Parental and family activities 2-Parental and family activities
Test Statistic	-24.000
Prob > |z|	0.3467
Prob > z	0.8266
Prob < z	0.1734

Matched Pairs (Impact on parental emotions)

Difference: Impact on parental emotions 2-Impact on parental emotions



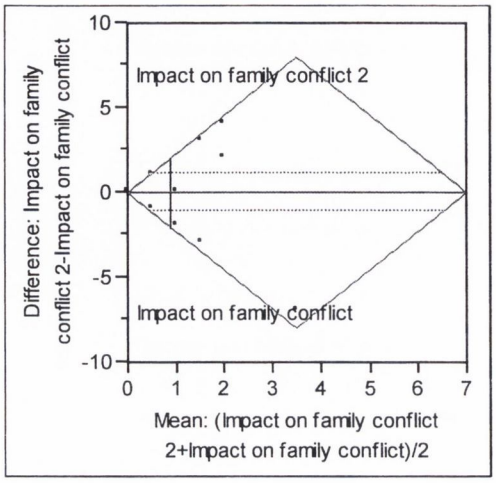
Impact on parental emotions 2	0.9	t-Ratio	-1.50966
Impact on parental emotions	2.3	DF	19
Mean Difference	-1.4	Prob > |t|	0.1476
Std Error	0.92736	Prob > t	0.9262
Upper95%	0.54099	Prob < t	0.0738
Lower95%	-3.341		
N	20		
Correlation	-0.1705		

Wilcoxon Sign-Rank

	Impact on parental emotions 2-Impact on parental emotions
Test Statistic	-27.500
Prob > |z|	0.0865
Prob > z	0.9567
Prob < z	0.0433

Matched Pairs (Impact on family conflict)

Difference: Impact on family conflict 2-Impact on family conflict



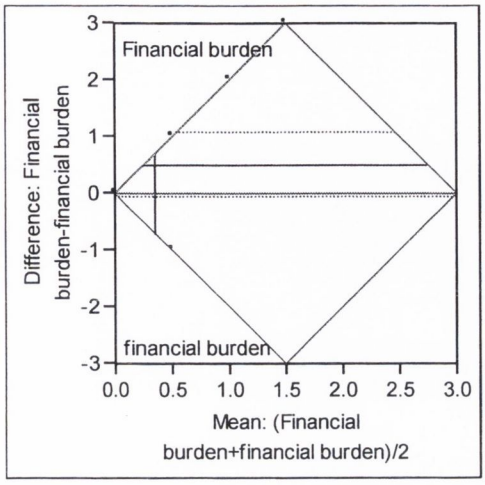
Impact on family conflict 2	0.9	t-Ratio	0
Impact on family conflict	0.9	DF	19
Mean Difference	0	Prob > |t|	1.0000
Std Error	0.52315	Prob > t	0.5000
Upper95%	1.09496	Prob < t	0.5000
Lower95%	-1.095		
N	20		
Correlation	-0.2224		

Wilcoxon Sign-Rank

	Impact on family conflict 2-Impact on family conflict
Test Statistic	4.000
Prob > |z|	0.7944
Prob > z	0.3972
Prob < z	0.6028

Matched Pairs (Financial burden)

Difference: Financial burden-financial burden



Financial burden	0.6	t-Ratio	1.809934
financial burden	0.1	DF	19
Mean Difference	0.5	Prob > |t|	0.0862
Std Error	0.27625	Prob > t	0.0431
Upper95%	1.0782	Prob < t	0.9569
Lower95%	-0.0782		
N	20		
Correlation	-0.1796		

Wilcoxon Sign-Rank

	Financial burden-financial burden
Test Statistic	10.000
Prob > |z|	0.1250
Prob > z	0.0625
Prob < z	0.9375

Moderate young hypodontia patients versus controls

Piers Harris Self Concept Scale

Parental Perception Questionnaire

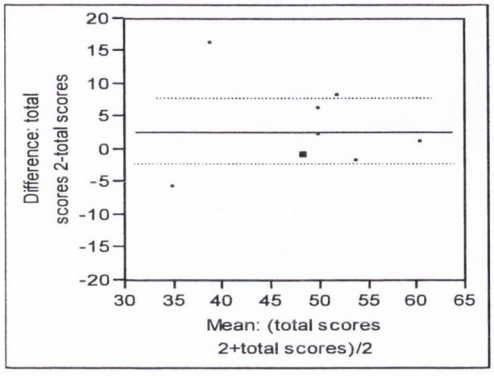
Family Impact Scale

Piers Harris Self Concept Scale for Moderate Hypodontia

Matched Pairs (Total Score)

Difference: total scores 2-total scores

total scores 2



total scores

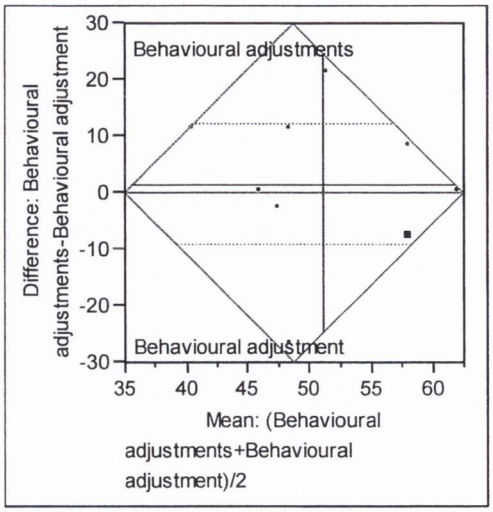
total scores 2	51.3333	t-Ratio	1.290564
total scores	48.5556	DF	8
Mean Difference	2.77778	Prob > |t|	0.2329
Std Error	2.15237	Prob > t	0.1165
Upper95%	7.74116	Prob < t	0.8835
Lower95%	-2.1856		
N	9		
Correlation	0.75431		

Wilcoxon Sign-Rank

	total scores 2-total scores
Test Statistic	9.500
Prob > |z|	0.3125
Prob > z	0.1563
Prob < z	0.8438

Matched Pairs (Behavioural adjustments)

Difference: Behavioural adjustments-Behavioural adjustment



Behavioural adjustments	51.8889	t-Ratio	0.313321
Behavioural adjustment	50.4444	DF	8
Mean Difference	1.44444	Prob > |t|	0.7620
Std Error	4.61011	Prob > t	0.3810
Upper95%	12.0754	Prob < t	0.6190
Lower95%	-9.1865		
N	9		
Correlation	-0.0047		

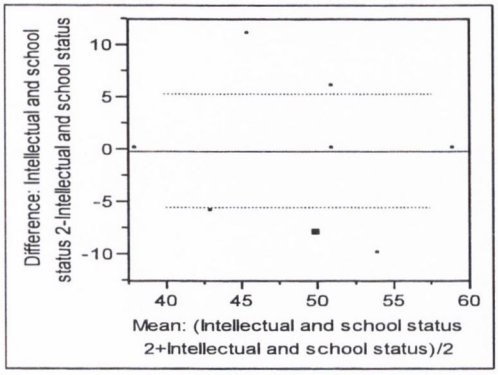
Wilcoxon Sign-Rank

	Behavioural adjustments-Behavioural adjustment
Test Statistic	3.500
Prob > |z|	0.6250
Prob > z	0.3125
Prob < z	0.6875

Matched Pairs (Intellectual and school status)

Difference: Intellectual and school status 2-Intellectual and school status

Intellectual and school status 2



Intellectual and school status

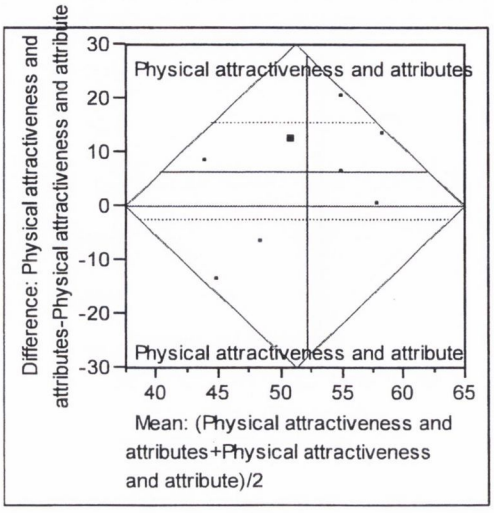
Intellectual and school status 2	49.1111	t-Ratio	-0.04757
Intellectual and school status	49.2222	DF	8
Mean Difference	-0.1111	Prob > |t|	0.9632
Std Error	2.33598	Prob > t	0.5184
Upper95%	5.27566	Prob < t	0.4816
Lower95%	-5.4979		
N	9		
Correlation	0.5173		

Wilcoxon Sign-Rank

	Intellectual and school status 2-Intellectual and school status
Test Statistic	-0.500
Prob > |z|	1.0000
Prob > z	0.5000
Prob < z	0.5000

Matched Pairs (Physical attractiveness and attributes)

Difference: Physical attractiveness and attributes-Physical attractiveness and attribute



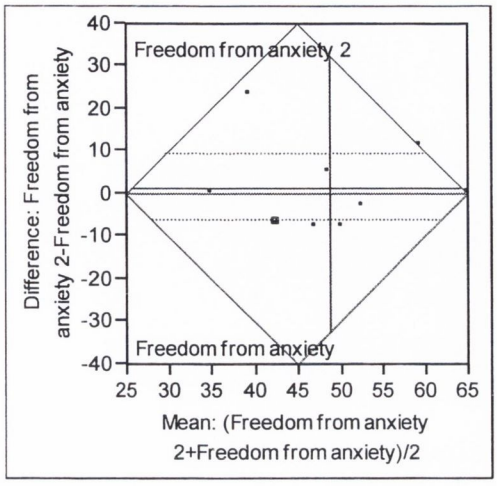
Physical attractiveness and attributes	55.4444	t-Ratio	1.660705
Physical attractiveness and attribute	49	DF	8
Mean Difference	6.44444	Prob > |t|	0.1354
Std Error	3.88055	Prob > t	0.0677
Upper95%	15.393	Prob < t	0.9323
Lower95%	-2.5041		
N	9		
Correlation	-0.0926		

Wilcoxon Sign-Rank

	Physical attractiveness and attributes-Physical attractiveness and attribute
Test Statistic	10.000
Prob > |z|	0.1875
Prob > z	0.0938
Prob < z	0.9063

Matched Pairs (Freedom from anxiety)

Difference: Freedom from anxiety 2-Freedom from anxiety



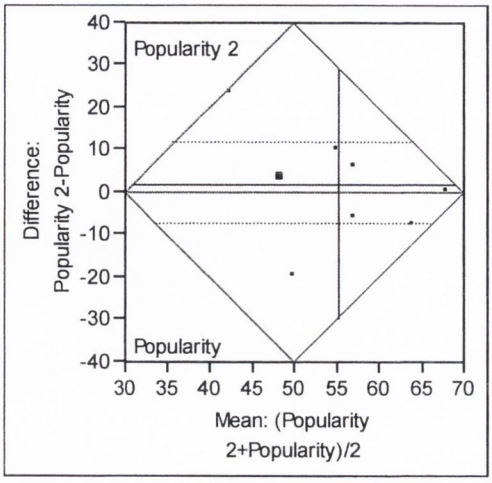
Freedom from anxiety 2	49.5556	t-Ratio	0.422332
Freedom from anxiety	48.1111	DF	8
Mean Difference	1.44444	Prob > |t|	0.6839
Std Error	3.42017	Prob > t	0.3420
Upper95%	9.33136	Prob < t	0.6580
Lower95%	-6.4425		
N	9		
Correlation	0.5447		

Wilcoxon Sign-Rank

	Freedom from anxiety 2-Freedom from anxiety
Test Statistic	1.000
Prob > |z|	0.8906
Prob > z	0.4453
Prob < z	0.5547

Matched Pairs (Popularity)

Difference: Popularity 2-Popularity



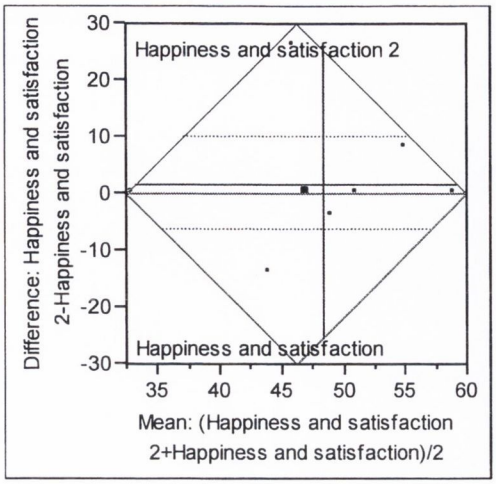
Popularity 2	56.2222	t-Ratio	0.482321
Popularity	54.2222	DF	8
Mean Difference	2	Prob > |t|	0.6425
Std Error	4.14662	Prob > t	0.3212
Upper95%	11.5621	Prob < t	0.6788
Lower95%	-7.5621		
N	9		
Correlation	0.23241		

Wilcoxon Sign-Rank

	Popularity 2-Popularity
Test Statistic	4.500
Prob > |z|	0.5859
Prob > z	0.2930
Prob < z	0.7070

Matched Pairs (Happiness and satisfaction)

Difference: Happiness and satisfaction 2-Happiness and satisfaction



Happiness and satisfaction 2	49.2222	t-Ratio	0.496378
Happiness and satisfaction	47.4444	DF	8
Mean Difference	1.77778	Prob > |t|	0.6330
Std Error	3.5815	Prob > t	0.3165
Upper95%	10.0367	Prob < t	0.6835
Lower95%	-6.4812		
N	9		
Correlation	0.30637		

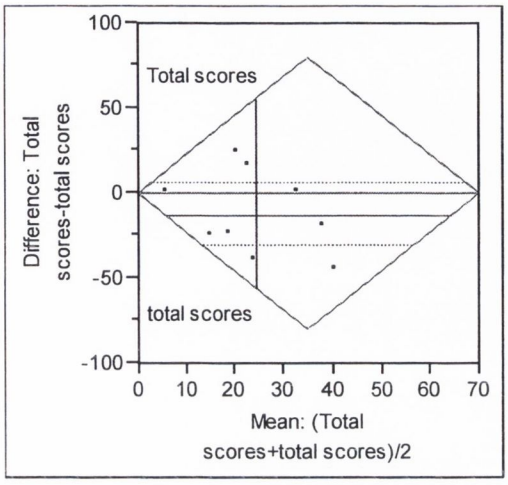
Wilcoxon Sign-Rank

	Happiness and satisfaction 2-Happiness and satisfaction
Test Statistic	1.000
Prob > |z|	0.8750
Prob > z	0.4375
Prob < z	0.5625

Parental Perception for Moderate Hypodontia Patients

Matched Pairs

Difference: Total scores-total scores



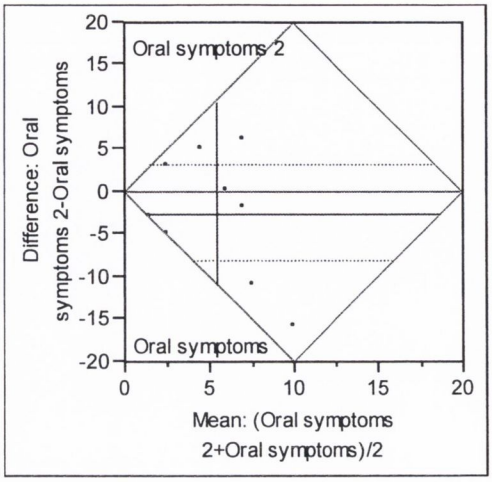
Total scores	17.8889	t-Ratio	-1.61835
total scores	30.7778	DF	8
Mean Difference	-12.889	Prob > |t|	0.1442
Std Error	7.96423	Prob > t	0.9279
Upper95%	5.47667	Prob < t	0.0721
Lower95%	-31.254		
N	9		
Correlation	-0.0749		

Wilcoxon Sign-Rank

	Total scores-total scores
Test Statistic	-10.000
Prob > |z|	0.1094
Prob > z	0.9453
Prob < z	0.0547

Matched Pairs

Difference: Oral symptoms 2-Oral symptoms



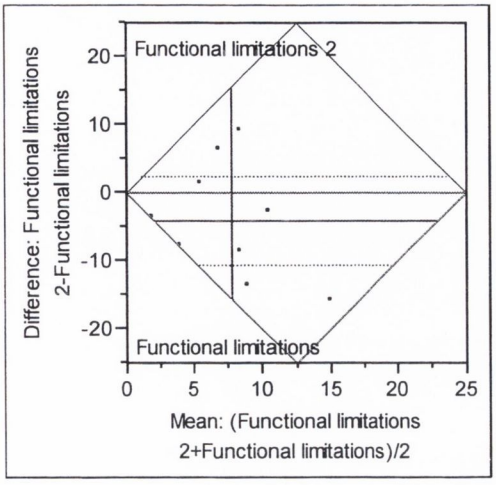
Oral symptoms 2	4.11111	t-Ratio	-1.05035
Oral symptoms	6.66667	DF	8
Mean Difference	-2.5556	Prob > |t|	0.3242
Std Error	2.43305	Prob > t	0.8379
Upper95%	3.05508	Prob < t	0.1621
Lower95%	-8.1662		
N	9		
Correlation	-0.2811		

Wilcoxon Sign-Rank

	Oral symptoms 2-Oral symptoms
Test Statistic	-5.000
Prob > |z|	0.5313
Prob > z	0.7344
Prob < z	0.2656

Matched Pairs (Functional limitations)

Difference: Functional limitations 2-Functional limitations



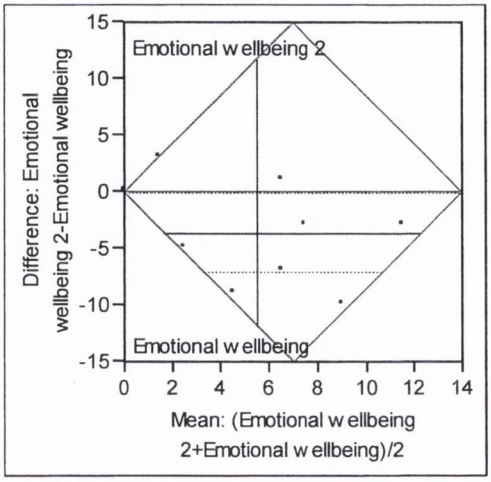
Functional limitations 2	5.66667	t-Ratio	-1.48819
Functional limitations	9.88889	DF	8
Mean Difference	-4.2222	Prob > |t|	0.1750
Std Error	2.83714	Prob > t	0.9125
Upper95%	2.32024	Prob < t	0.0875
Lower95%	-10.765		
N	9		
Correlation	-0.1209		

Wilcoxon Sign-Rank

	Functional limitations 2-Functional limitations
Test Statistic	-11.000
Prob > |z|	0.2148
Prob > z	0.8926
Prob < z	0.1074

Matched Pairs

Difference: Emotional wellbeing 2-Emotional wellbeing



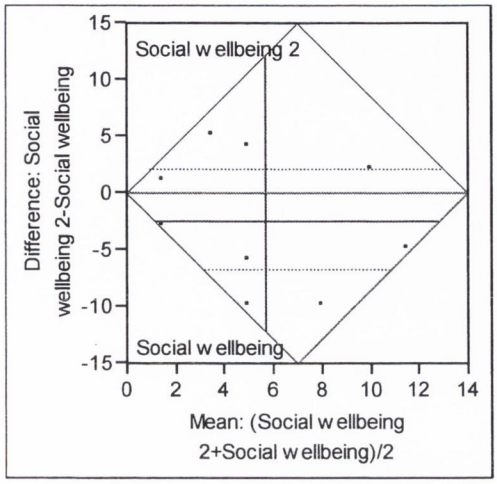
Emotional wellbeing 2	3.66667	t-Ratio	-2.44444
Emotional wellbeing	7.33333	DF	8
Mean Difference	-3.6667	Prob > |t|	0.0403
Std Error	1.5	Prob > t	0.9799
Upper95%	-0.2077	Prob < t	0.0201
Lower95%	-7.1257		
N	9		
Correlation	0.49507		

Wilcoxon Sign-Rank

	Emotional wellbeing 2-Emotional wellbeing
Test Statistic	-14.000
Prob > |z|	0.0625
Prob > z	0.9688
Prob < z	0.0313

Matched Pairs (Social Well-being)

Difference: Social wellbeing 2-Social wellbeing



Social wellbeing 2	4.44444	t-Ratio	-1.28089
Social wellbeing	6.88889	DF	8
Mean Difference	-2.4444	Prob > |t|	0.2361
Std Error	1.9084	Prob > t	0.8819
Upper95%	1.95632	Prob < t	0.1181
Lower95%	-6.8452		
N	9		
Correlation	0.20899		

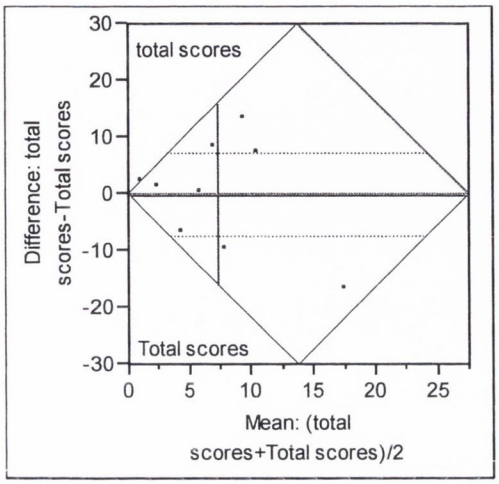
Wilcoxon Sign-Rank

	Social wellbeing 2-Social wellbeing
Test Statistic	-10.000
Prob > |z|	0.2695
Prob > z	0.8652
Prob < z	0.1348

Family Impact Scale of Moderate Hypodontia Patients

Matched Pairs

Difference: total scores-Total scores



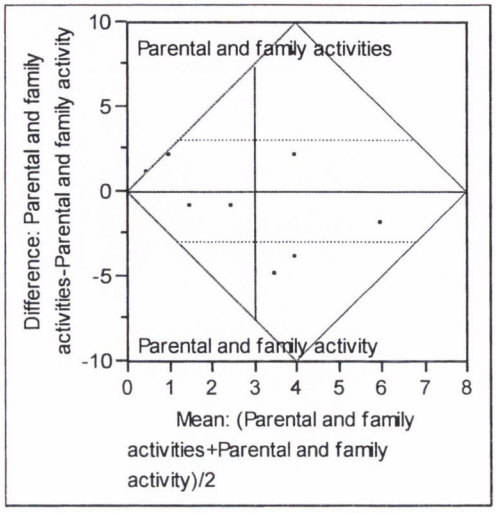
total scores	7.22222	t-Ratio	-0.10512
Total scores	7.55556	DF	8
Mean Difference	-0.3333	Prob > |t|	0.9189
Std Error	3.17105	Prob > t	0.5406
Upper95%	6.97912	Prob < t	0.4594
Lower95%	-7.6458		
N	9		
Correlation	0.03113		

Wilcoxon Sign-Rank

	total scores-Total scores
Test Statistic	0.500
Prob > |z|	0.9844
Prob > z	0.4922
Prob < z	0.5078

Matched Pairs (Parental and family activities)

Difference: Parental and family activities-Parental and family activity



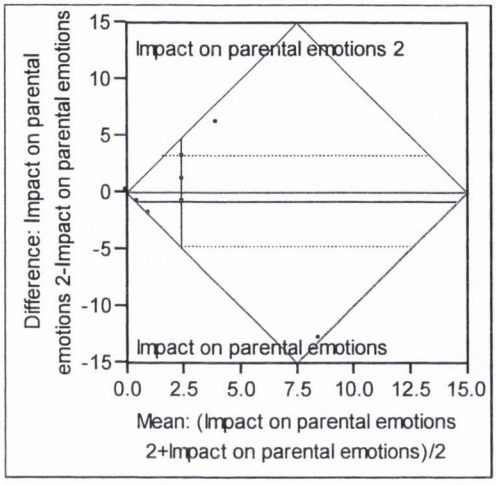
Parental and family activities	3	t-Ratio	0
Parental and family activity	3	DF	8
Mean Difference	0	Prob > |t|	1.0000
Std Error	1.29099	Prob > t	0.5000
Upper95%	2.97704	Prob < t	0.5000
Lower95%	-2.977		
N	9		
Correlation	-0.0917		

Wilcoxon Sign-Rank

	Parental and family activities-Parental and family activity
Test Statistic	-1.500
Prob > |z|	0.9102
Prob > z	0.5449
Prob < z	0.4551

Matched Pairs (Impact on parental emotions)

Difference: Impact on parental emotions 2-Impact on parental emotions



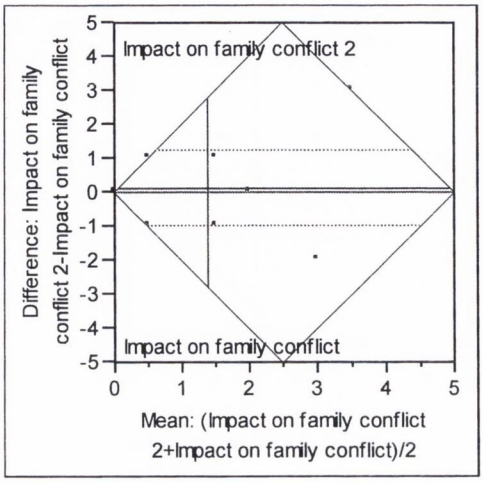
Impact on parental emotions 2	2	t-Ratio	-0.51453
Impact on parental emotions	2.88889	DF	8
Mean Difference	-0.8889	Prob > |t|	0.6208
Std Error	1.72759	Prob > t	0.6896
Upper95%	3.09494	Prob < t	0.3104
Lower95%	-4.8727		
N	9		
Correlation	0.01128		

Wilcoxon Sign-Rank

	Impact on parental emotions 2-Impact on parental emotions
Test Statistic	-2.500
Prob > |z|	0.7734
Prob > z	0.6133
Prob < z	0.3867

Matched Pairs (Impact on family conflict)

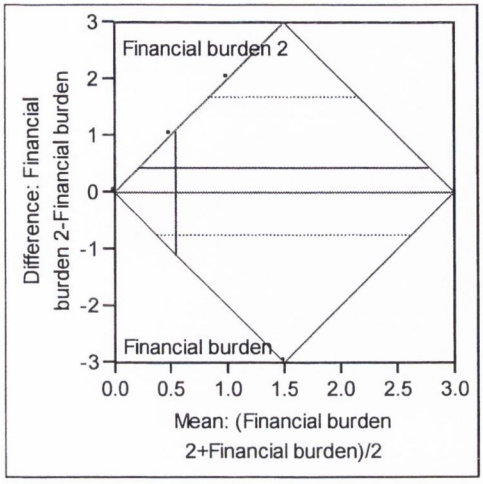
Difference: Impact on family conflict 2-Impact on family conflict



Impact on family conflict 2	1.44444	t-Ratio	0.229416
Impact on family conflict	1.33333	DF	8
Mean Difference	0.11111	Prob > |t|	0.8243
Std Error	0.48432	Prob > t	0.4122
Upper95%	1.22796	Prob < t	0.5878
Lower95%	-1.0057		
N	9		
Correlation	0.51508		

Matched Pairs (Financial burden)

Difference: Financial burden 2-Financial burden



Financial burden 2	0.77778	t-Ratio	0.838628
Financial burden	0.33333	DF	8
Mean Difference	0.44444	Prob > |t|	0.4260
Std Error	0.52997	Prob > t	0.2130
Upper95%	1.66655	Prob < t	0.7870
Lower95%	-0.7777		
N	9		
Correlation	-0.3001		

Wilcoxon Sign-Rank

	Financial burden 2-Financial burden
Test Statistic	2.500
Prob > |z|	0.5625
Prob > z	0.2813
Prob < z	0.7188

Young Amelogenesis Imperfecta patients versus controls

Piers Harris Self Concept Scale

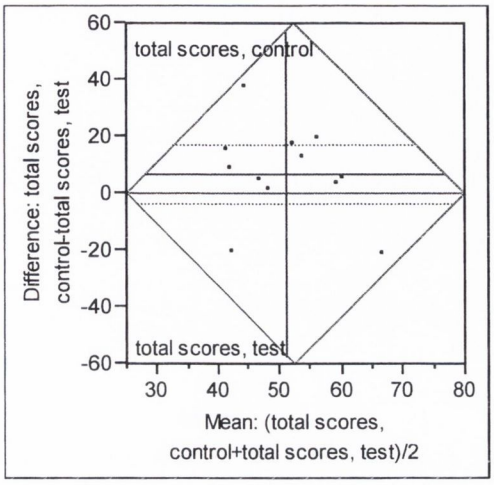
Parental Perception Questionnaire

Family Impact Scale

Piers Harris Self Evaluation Scale (AI)

Matched Pairs (total scores)

Difference: total scores, control-total scores, test



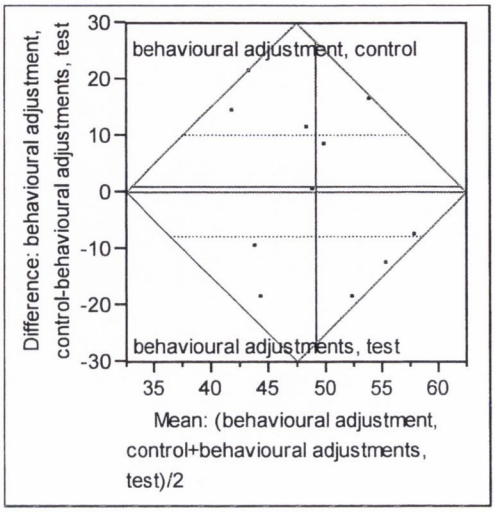
total scores, control	54.5833	t-Ratio	1.381766
total scores, test	48.0833	DF	11
Mean Difference	6.5	Prob > |t|	0.1945
Std Error	4.70412	Prob > t	0.0972
Upper95%	16.8537	Prob < t	0.9028
Lower95%	-3.8537		
N	12		
Correlation	0.02542		

Wilcoxon Sign-Rank

	total scores, control-total scores, test
Test Statistic	18.000
Prob > |z|	0.1763
Prob > z	0.0881
Prob < z	0.9119

Matched Pairs (behavioural adjustment)

Difference: behavioural adjustment, control-behavioural adjustments, test



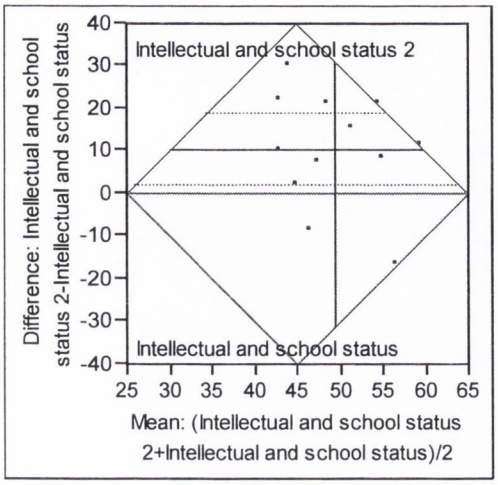
behavioural adjustment, control	49.6667	t-Ratio	0.242644
behavioural adjustments, test	48.6667	DF	11
Mean Difference	1	Prob > |t|	0.8127
Std Error	4.12127	Prob > t	0.4064
Upper95%	10.0708	Prob < t	0.5936
Lower95%	-8.0708		
N	12		
Correlation	-0.3362		

Wilcoxon Sign-Rank

	behavioural adjustment, control-behavioural adjustments, test
Test Statistic	3.500
Prob > |z|	0.7842
Prob > z	0.3921
Prob < z	0.6079

Matched Pairs (Intellectual and School Status)

Difference: Intellectual and school status 2-Intellectual and school status



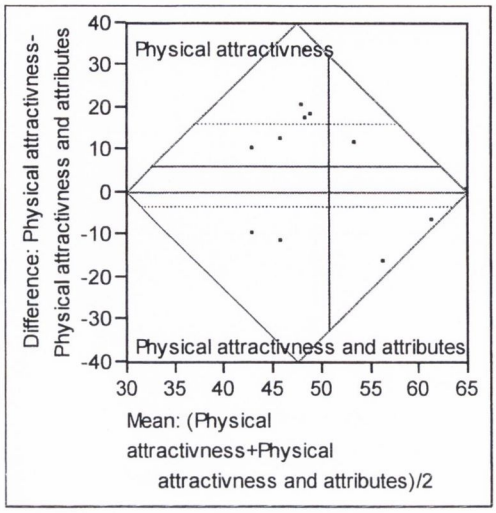
Intellectual and school status 2	54.5833	t-Ratio	2.604218
Intellectual and school status	44.5	DF	11
Mean Difference	10.0833	Prob > |t|	0.0245
Std Error	3.87192	Prob > t	0.0123
Upper95%	18.6054	Prob < t	0.9877
Lower95%	1.56129		
N	12		
Correlation	-0.1673		

Wilcoxon Sign-Rank

	Intellectual and school status 2-Intellectual and school status
Test Statistic	27.000
Prob > |z|	0.0332
Prob > z	0.0166
Prob < z	0.9834

Matched Pairs (Physical attractiveness)

Difference: Physical attractiveness-Physical attractiveness and attributes



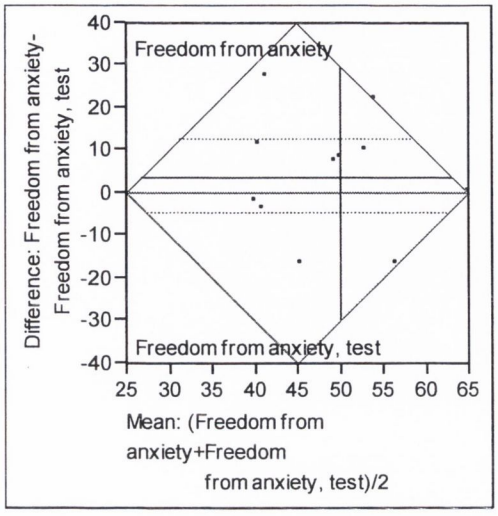
Physical attractivness	53.8333	t-Ratio	1.413631
Physical attractivness and attributes	47.5833	DF	11
Mean Difference	6.25	Prob > |t|	0.1851
Std Error	4.42124	Prob > t	0.0926
Upper95%	15.9811	Prob < t	0.9074
Lower95%	-3.4811		
N	12		
Correlation	-0.0875		

Wilcoxon Sign-Rank

	Physical attractivness-Physical attractivness and attributes
Test Statistic	16.500
Prob > |z|	0.1563
Prob > z	0.0781
Prob < z	0.9219

Matched Pairs (Freedom from anxiety)

Difference: Freedom from anxiety-Freedom from anxiety, test



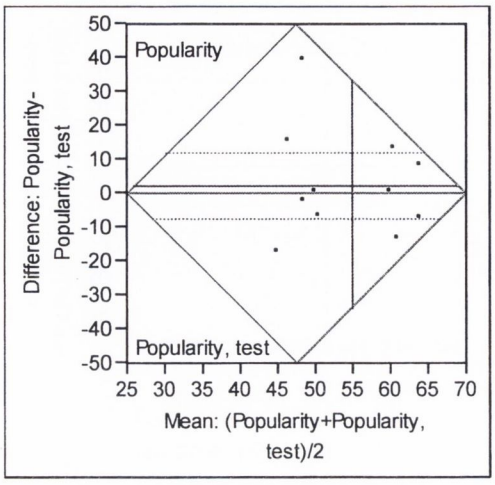
Freedom from anxiety	52	t-Ratio	0.969164
Freedom from anxiety, test	48.25	DF	11
Mean Difference	3.75	Prob > |t|	0.3533
Std Error	3.86931	Prob > t	0.1766
Upper95%	12.2663	Prob < t	0.8234
Lower95%	-4.7663		
N	12		
Correlation	0.28455		

Wilcoxon Sign-Rank

	Freedom from anxiety-Freedom from anxiety, test
Test Statistic	9.500
Prob > |z|	0.3613
Prob > z	0.1807
Prob < z	0.8193

Matched Pairs (Popularity)

Difference: Popularity-Popularity, test



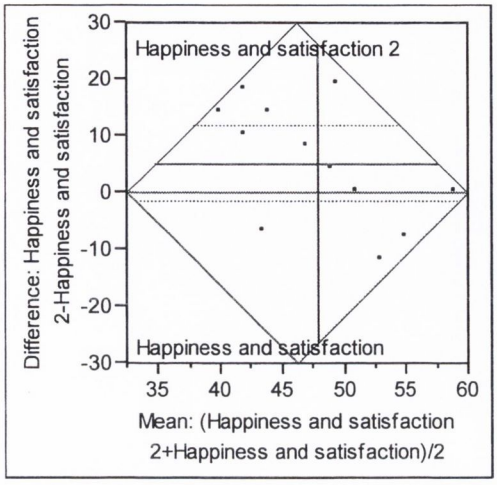
Popularity	55.9167	t-Ratio	0.472249
Popularity, test	53.8333	DF	11
Mean Difference	2.08333	Prob > |t|	0.6460
Std Error	4.41152	Prob > t	0.3230
Upper95%	11.793	Prob < t	0.6770
Lower95%	-7.6264		
N	12		
Correlation	-0.0513		

Wilcoxon Sign-Rank

	Popularity-Popularity, test
Test Statistic	2.000
Prob > |z|	0.8438
Prob > z	0.4219
Prob < z	0.5781

Matched Pairs (Happiness and Satisfaction)

Difference: Happiness and satisfaction 2-Happiness and satisfaction



Happiness and satisfaction 2	50.4167	t-Ratio	1.648723
Happiness and satisfaction	45.4167	DF	11
Mean Difference	5	Prob > |t|	0.1274
Std Error	3.03265	Prob > t	0.0637
Upper95%	11.6748	Prob < t	0.9363
Lower95%	-1.6748		
N	12		
Correlation	0.13708		

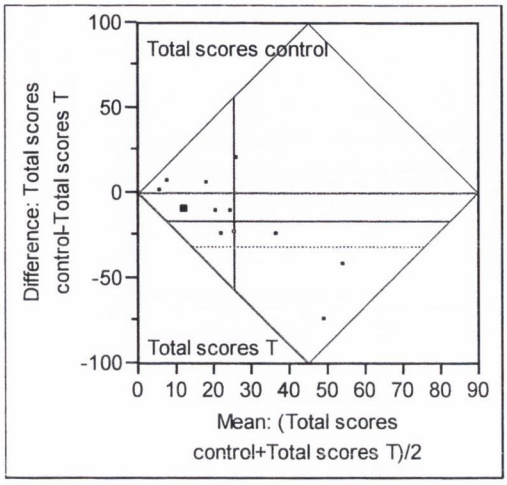
Wilcoxon Sign-Rank

	Happiness and satisfaction 2-Happiness and satisfaction
Test Statistic	16.000
Prob > |z|	0.1094
Prob > z	0.0547
Prob < z	0.9453

Parental Report of Amelogenesis Imperfecta patients

Matched Pairs (Total scores)

Difference: Total scores control-Total scores T



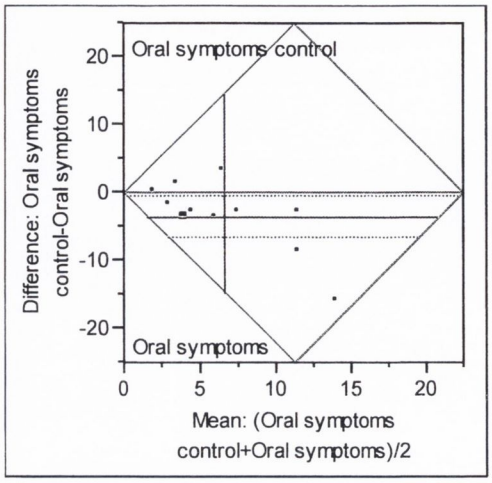
Total scores control	17.3333	t-Ratio	-2.27968
Total scores T	33.8333	DF	11
Mean Difference	-16.5	Prob > |t|	0.0436
Std Error	7.23784	Prob > t	0.9782
Upper95%	-0.5696	Prob < t	0.0218
Lower95%	-32.43		
N	12		
Correlation	0.27393		

Wilcoxon Sign-Rank

	Total scores control-Total scores T
Test Statistic	-24.000
Prob > |z|	0.0313
Prob > z	0.9844
Prob < z	0.0156

Matched Pairs (Oral symptoms)

Difference: Oral symptoms control-Oral symptoms



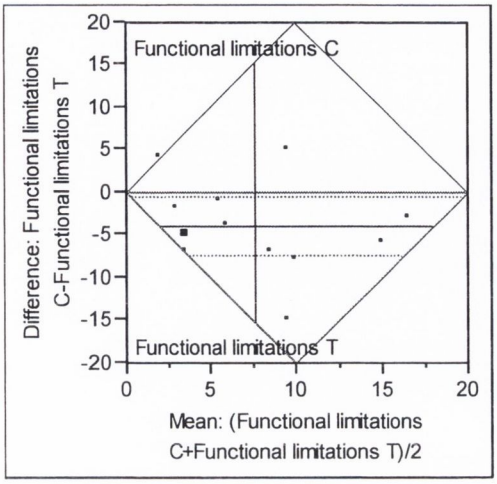
Oral symptoms control	4.83333	t-Ratio	-2.58946
Oral symptoms	8.5	DF	11
Mean Difference	-3.6667	Prob > |t|	0.0252
Std Error	1.416	Prob > t	0.9874
Upper95%	-0.5501	Prob < t	0.0126
Lower95%	-6.7833		
N	12		
Correlation	0.55805		

Wilcoxon Sign-Rank

	Oral symptoms control-Oral symptoms
Test Statistic	-27.500
Prob > |z|	0.0117
Prob > z	0.9941
Prob < z	0.0059

Matched Pairs (Functional Limitations)

Difference: Functional limitations C-Functional limitations T



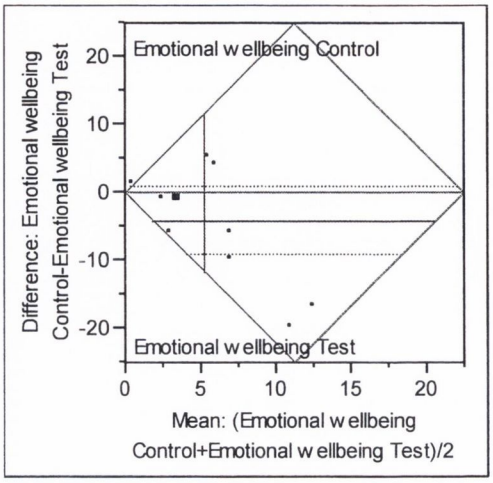
Functional limitations C	5.66667	t-Ratio	-2.62702
Functional limitations T	9.75	DF	11
Mean Difference	-4.0833	Prob > |t|	0.0235
Std Error	1.55436	Prob > t	0.9882
Upper95%	-0.6622	Prob < t	0.0118
Lower95%	-7.5045		
N	12		
Correlation	0.51334		

Wilcoxon Sign-Rank

	Functional limitations C-Functional limitations T
Test Statistic	-28.000
Prob > |z|	0.0259
Prob > z	0.9871
Prob < z	0.0129

Matched Pairs (Emotional wellbeing)

Difference: Emotional wellbeing Control-Emotional wellbeing Test



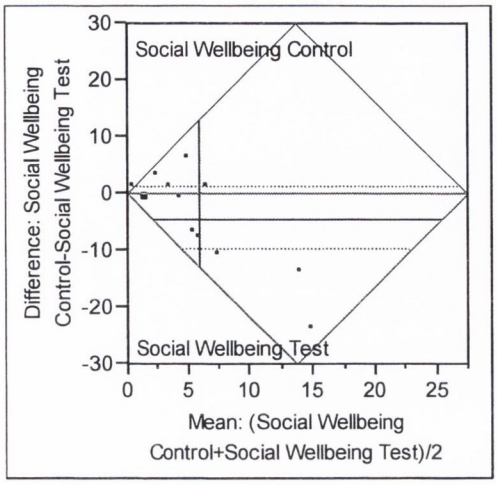
Emotional wellbeing Control	3.08333	t-Ratio	-1.85854
Emotional wellbeing Test	7.33333	DF	11
Mean Difference	-4.25	Prob > |t|	0.0900
Std Error	2.28674	Prob > t	0.9550
Upper95%	0.78307	Prob < t	0.0450
Lower95%	-9.2831		
N	12		
Correlation	-0.0782		

Wilcoxon Sign-Rank

	Emotional wellbeing Control-Emotional wellbeing Test
Test Statistic	-20.000
Prob > |z|	0.1260
Prob > z	0.9370
Prob < z	0.0630

Matched Pairs (Social Wellbeing)

Difference: Social Wellbeing Control-Social Wellbeing Test



Social Wellbeing Control	3.75	t-Ratio	-1.81324
Social Wellbeing Test	8.25	DF	11
Mean Difference	-4.5	Prob > |t|	0.0971
Std Error	2.48175	Prob > t	0.9514
Upper95%	0.9623	Prob < t	0.0486
Lower95%	-9.9623		
N	12		
Correlation	0.07471		

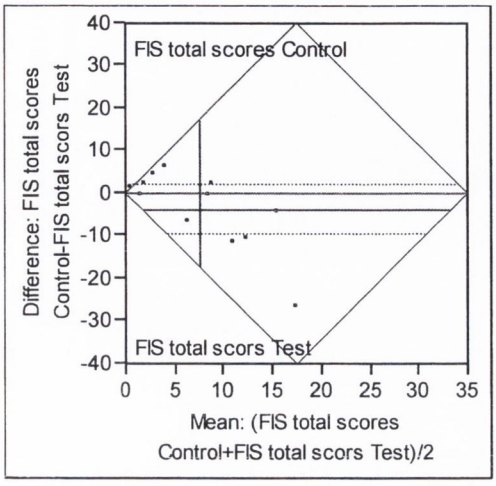
Wilcoxon Sign-Rank

	Social Wellbeing Control-Social Wellbeing Test
Test Statistic	-17.000
Prob > |z|	0.1992
Prob > z	0.9004
Prob < z	0.0996

Family Impact Scale of Amelogenesis Imperfecta patients

Matched Pairs (total scores)

Difference: FIS total scores Control-FIS total scores Test



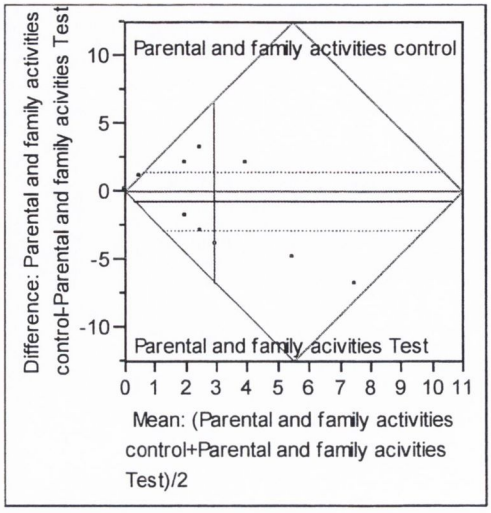
FIS total scores Control	5.58333	t-Ratio	-1.53761
FIS total scors Test	9.66667	DF	11
Mean Difference	-4.0833	Prob > |t|	0.1524
Std Error	2.65564	Prob > t	0.9238
Upper95%	1.76168	Prob < t	0.0762
Lower95%	-9.9283		
N	12		
Correlation	0.31384		

Wilcoxon Sign-Rank

	FIS total scores Control-FIS total scors Test
Test Statistic	-14.000
Prob > |z|	0.2900
Prob > z	0.8550
Prob < z	0.1450

Matched Pairs (Parental and family activities)

Difference: Parental and family activities control-Parental and family activities Test



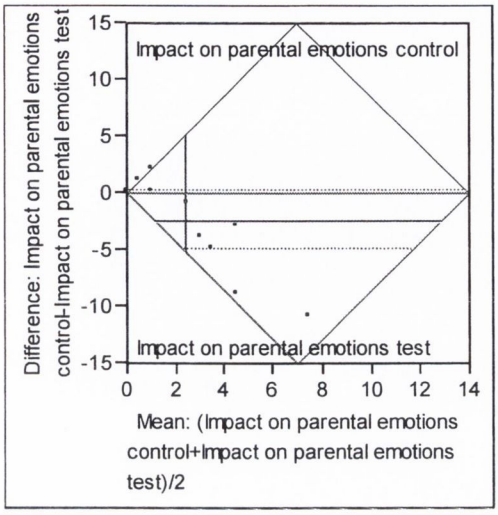
Parental and family activities control	2.58333	t-Ratio	-0.77933
Parental and family acivities Test	3.33333	DF	11
Mean Difference	-0.75	Prob > |t|	0.4522
Std Error	0.96236	Prob > t	0.7739
Upper95%	1.36814	Prob < t	0.2261
Lower95%	-2.8681		
N	12		
Correlation	0.26229		

Wilcoxon Sign-Rank

	Parental and family activities control-Parental and family acivities Test
Test Statistic	-8.500
Prob > |z|	0.4785
Prob > z	0.7607
Prob < z	0.2393

Matched Pairs (parental emotions)

Difference: Impact on parental emotions control-Impact on parental emotions test



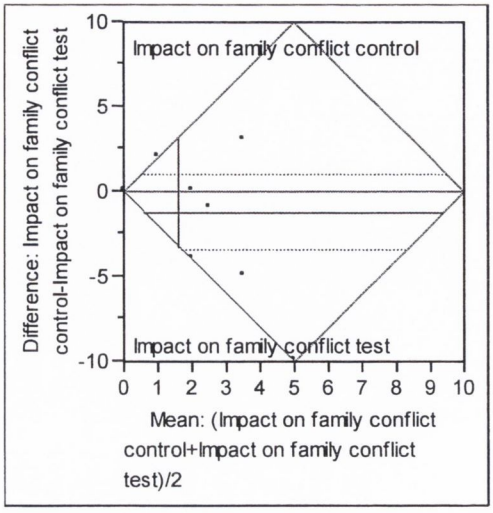
Impact on parental emotions control	1.16667	t-Ratio	-2.02008
Impact on parental emotions test	3.58333	DF	11
Mean Difference	-2.4167	Prob > |t|	0.0684
Std Error	1.19632	Prob > t	0.9658
Upper95%	0.21642	Prob < t	0.0342
Lower95%	-5.0498		
N	12		
Correlation	0.26718		

Wilcoxon Sign-Rank

	Impact on parental emotions control-Impact on parental emotions test
Test Statistic	-14.500
Prob > |z|	0.0977
Prob > z	0.9512
Prob < z	0.0488

Matched Pairs (family conflict)

Difference: Impact on family conflict control-Impact on family conflict test



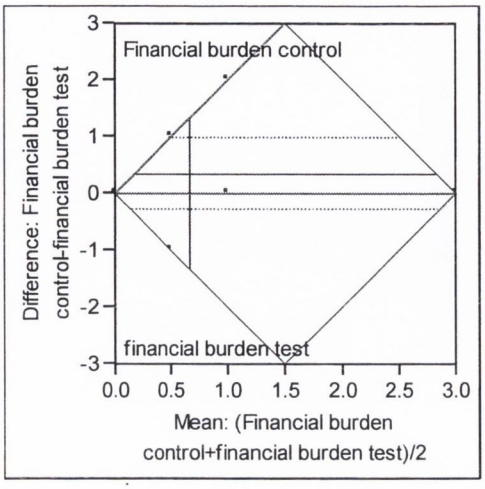
Impact on family conflict control	1	t-Ratio	-1.23035
Impact on family conflict test	2.25	DF	11
Mean Difference	-1.25	Prob > |t|	0.2442
Std Error	1.01597	Prob > t	0.8779
Upper95%	0.98614	Prob < t	0.1221
Lower95%	-3.4861		
N	12		
Correlation	-0.0188		

Wilcoxon Sign-Rank

	Impact on family conflict control-Impact on family conflict test
Test Statistic	-5.500
Prob > |z|	0.3125
Prob > z	0.8438
Prob < z	0.1563

Matched Pairs (Financial burden)

Difference: Financial burden control-financial burden test



Financial burden control	0.83333	t-Ratio	1.172604
financial burden test	0.5	DF	11
Mean Difference	0.33333	Prob > |t|	0.2657
Std Error	0.28427	Prob > t	0.1329
Upper95%	0.959	Prob < t	0.8671
Lower95%	-0.2923		
N	12		
Correlation	0.48795		

Wilcoxon Sign-Rank

	Financial burden control-financial burden test
Test Statistic	5.500
Prob > |z|	0.3438
Prob > z	0.1719
Prob < z	0.8281