Interventions for promoting participation in shared decision-making for children and adolescents with cystic fibrosis (Protocol)


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Interventions for promoting participation in shared decision-making for children and adolescents with cystic fibrosis

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ABSTRACT

This is a protocol for a Cochrane Review (Intervention). The objectives are as follows:

To determine the effectiveness of interventions that promote SDM for children and adolescents with CF aged four to 18 years.

BACKGROUND

Description of the condition

Cystic fibrosis (CF) is a genetic disease with significant variations in incidence, morbidity and mortality around the world. It is the most common, life-limiting autosomal recessively inherited condition in white populations (Williams 2010). It was reported that in 2012 there were approximately 70,000 people living with CF worldwide (Cystic Fibrosis Foundation 2013). While the condition is most common in white populations, it is also being diagnosed with increasing frequency in other populations, often with variant clinical expression (Cystic Fibrosis Worldwide 2002). Although significantly under-diagnosed in Asia, current evidence indicates that the prevalence of CF is low (WHO 2015). In the USA, the incidence of CF is reported to be one in every 3500 births (WHO 2015), with 30,000 people there living with CF in 2012 (Cystic Fibrosis Foundation 2013). In the European Union, one in 2000 to 3000 newborns are affected by CF (WHO 2015); and the Republic of Ireland has the highest prevalence of CF in the world at 2.98 per 10,000 head of population (Cystic Fibrosis Registry of Ireland 2014; Farrell 2008a). The Australian Cystic Fibrosis Data Registry reported holding records of 3235 people living with CF at the end of 2013 (Cystic Fibrosis Australia 2013).

The condition is caused by mutations in a gene called the cystic fibrosis transmembrane conductance regulator (CFTR) (Bronsveld 2001) which is a type of protein that regulates the transport of chloride ions across epithelial cell membranes (Brice 2007; Harris 1993). Dysfunction of the CFTR dehydrates secretions in the airways, the pancreatic ducts and elsewhere in the body (Cystic Fibrosis Foundation 2015) and results in the accumulation of mucus or thick sticky secretions (Brice 2007; Dyce 2015). In the lungs mucus blocks the airways and traps organisms increasing the risk of infection; and in the pancreatic ducts mucus prevents the re-
lease of enzymes for food breakdown and absorption of essential nutrients (Cystic Fibrosis Foundation 2013). Elevated chloride output in the sweat increases the probability of dehydration and mineral imbalances. These aspects of CF lead to complications, such as poor nutrition and growth, respiratory infections and lung damage (Brice 2007). Abnormal and irreversible dilatation of the lower bronchi (bronchiectasis) permits more mucus build-up and further increases the risk of infection (NORD 2015; Tariq 2015). Newborn screening is increasing internationally, so the majority of individuals with CF are now diagnosed in infancy and as such, both the child and the family are challenged with the condition from the early stages of their relationship (Brice 2007). Advances in treatment over the last three to four decades have led to marked increases in survival for people with CF, with survival now into the fourth decade (Dodge 2007). However, the prevalence of associated medical conditions (such as gastro-oesophageal reflux, CF-related diabetes (CFRD), osteoporosis or osteopenia, and lower body mass index (BMI)) increases with age and more severe impairment of lung function is found in older age groups. Daily therapies usually include nutritional supplements, pancreatic enzymes, daily chest physiotherapy, a regular exercise routine, nebulised medication to assist with airway clearance and antibiotics to treat acute exacerbations. When lung disease worsens, oxygen therapy and non-invasive ventilation therapy may be required (Cystic Fibrosis Australia 2013). This daily treatment regimen places significant responsibility on families, children and young people with CF to ensure optimal health and to slow down disease progression. A multidisciplinary team of trained professionals is required to manage the range of complex issues that can arise in the care of a person with CF (Quon 2012) and it is now well-established that outcomes for individuals cared for in specialist CF centres are better than for those who are not (Conway 2014).

Description of the intervention

Shared decision-making (SDM) is defined as a patient-centred, collaborative process that enables individuals and their healthcare providers to make decisions together (IMDF 2014). It is an approach to decision-making that integrates a person’s values, goals and concerns with the best available evidence about benefits, risks and uncertainties of treatment. The aim is to facilitate the best healthcare decisions that fully incorporate the holistic needs of the individual (IMDF 2014). In partnership with their clinician, individuals are encouraged to consider available treatment or management options (and the likely benefits and harms of each), to communicate their preferences and to work together to choose the course of action that best fits their preference (Drayton 2014; Polaris 2014). Measures of SDM generally focus on the two-way interaction between one individual and one practitioner; however, SDM can also take place in a multi-disciplinary setting with triadic encounters (Clayman 2013).

The process of SDM has been studied extensively in adult healthcare, but to a much lesser extent in paediatric care, where decision making includes other family members (Dokken 2000). Paediatric care includes both children and adolescents and for the purposes of this review we will define a child as up to nine years old and an adolescent as between 10 and 19 years of age (WHO 2016). Early literature focused on the term ‘informed consent’ for individuals involved in medical decision making (President’s Commission Report 1982). Since then, the term SDM has emerged in the literature with its definition being variable, a systematic review resulted in an integrative model of SDM that includes essential elements such as explaining the problem, presenting options, discussion of pros and cons, individual preferences, an individual’s ability or self-efficacy, doctor’s knowledge, checking patient understanding, deferral of decision and follow-up (Makoul 2006). Elwyn has built on this previous work, translating existing models into an easy to remember three-step model suitable for most clinical settings and based on ‘choice talk’, ‘option talk’ and ‘decision talk’ (Elwyn 2012). More recently, Elwyn produced a decision aid (OPTIONS) which covers the following (Elwyn 2013a):

1. provision of explicit explanations that decisions exist which need attention and deliberation;
2. provision of reassurance to patients that the provider will support deliberation;
3. provision of information about treatment or management options;
4. elicitation of the patient’s views, preferences, priorities, at a stage when the patient is better informed (preference elicitation); and
5. integration of the patient’s preferences into the next stage of decision making (preference integrations).

It is generally accepted that children and adolescents have the right to self-determination, dignity, respect, and the right to make informed decisions (Coyne 2008). Patient participation in decision making is well-cited in contemporary publications (Elwyn 2016; Wyatt 2015). It is an accepted approach, particularly when guided by ethical principles (Elwyn 2013b; Kraus 2016). It promotes patient autonomy (Birchley 2014) and has demonstrated positive outcomes, including significant reductions in hospitalisation (Wennerg 2010) and individual satisfaction with healthcare (Quaschning 2013). However, SDM may also have unintended outcomes, such as decision regret (Simcox 2009) and patients require guidance to assess the risks and benefits of an intervention (Elwyn 2016). The competence of a child should be assessed in relation to the decision being made (Fennestra 2014); however, it is still important for children to be involved in SDM because adult proxy views within the healthcare setting may differ markedly from the child’s view (Söderbäck 2011). Also, the level of parental involvement in decision making changes as children grow older and become increasingly capable of informed decision making (Bejarano 2015). Additionally, children with a lifelong condition can benefit from recognising the importance of their
own participation early in life (Alderson 2006). Interventions to promote the adoption of SDM by children and adolescents may include (but are not restricted to): the building of partnerships between physicians, patients and parents (Brand 2013); the provision and exchange of information, verbally, on paper or online (Miller 2008); encouraging the participant to ask questions (Brand 2013); establishing understanding (Joffe 2016); encouraging the expression of preferences (Joffe 2003; Waligora 2016); assisting preparation for a decision (Graham 1995); programmes that embed SDM in clinician training and continuous professional development (The Health Foundation 2012); CF self-management programmes that advocate shared decision-making (Savage 2007); and eliciting post-decision reactions from the patient (either satisfaction (Quaschning 2013) or regret (Brehaut 2003)). Practical examples could include:

- communication training for clinicians (Silverman 2011) - Elwyn demonstrated this significantly increased the involvement of patients in decision making (Elwyn 2004);
- decision supports such as paediatric computerised clinical decision supports (CCDS) (Stultz 2012);
- provision of information - following a diagnosis of their infant with CF, parents suggested fact sheets, brochures and booklets should be used (Jessup 2016); other resources include websites to facilitate self-management of the condition for people with CF and their families (Roehrer 2013), CD-Roms (Davis 2004) or telephone interventions which aim to improve patient-physician communication (Wennberg 2010);
- counselling and dialogue to reduce decision conflict (Westerman 2013).

How the intervention might work

One of the key components of SDM is the provision of information to encourage an individual to have a discussion with their clinician (Elwyn 2013a). Studies have shown that there can be a gap in a person’s knowledge about their condition and how relevant interventions work (Conway 1996; Nolan 1986; Siklosi 2010). One study demonstrated differences between age groups, with gaps in knowledge appearing in adolescence when the individuals are more likely to be involved in SDM (Chomik 2014). If left uncorrected, such gaps could impact on the progression of the illness (Chomik 2014). Research has shown that while adolescents with chronic conditions wish to be involved in decision making-aids can improve the quality of decision-making (Chomik 2014). Research has shown that while adolescents with chronic conditions wish to be involved in decisions about their treatment, they also seek the involvement of parents and physicians (Lipstein 2013). However, parents may consider decision factors differently to adolescents, with adolescents more likely to focus on immediate treatment effects and parents tending to focus on the long-term implications (Lipstein 2016). The building of partnerships between physicians, patients and parents may encourage the exchange of information. Children can make valuable contributions regarding healthcare decisions (Alderson 2006). In practice a SDM intervention can include parents, carers and healthcare providers, but focuses primarily on the child; it is commensurate with the child’s age, cognitive ability and the illness trajectory (Coyne 2013). Prior to engaging the child in SDM, healthcare professionals must first assess the age competency of the child (Gabe 2004). It has been suggested that using SDM between healthcare professionals, individuals with CF and their families may lead to improvements in quality of care (Elf 2015). According to a recent study, five elements contribute to improved care that is co-produced by patients, families and healthcare professionals:

1. mental and emotional readiness to engage;
2. curiosity and the search for insight;
3. re-framing challenges into opportunities for improvement;
4. listening and learning from everyone, bringing home what is relevant; and
5. personal participation (Sabadosa 2014).

Assisting preparation for a decision, e.g. role play, might give a child confidence to voice his or her opinions in a real life situation. Interventions that encourage the asking of questions may enhance understanding, which in turn may alleviate uncertainty. Research has indicated that children’s participation in SDM reduces fears and concerns, gives a sense of competence and results in more satisfaction with healthcare provision (Angst 1996; Runeson 2002). Children with CF face practical challenges in managing their condition on a daily basis, e.g. physiotherapy and nutrition, and solutions tailored to an individual’s needs may be enhanced by their participation in SDM. As the child grows older, age-related CF complications require increasing levels of (self) care and support (Cystic Fibrosis Australia 2015) and inclusion in discussions about treatment decisions may help to develop self-caring and participation skills. It may be that if children observe parents actively engaging in SDM it will be easier for them to engage also. Furthermore, with increasing age and cognitive maturity, children better understand the purpose, risks and benefits of treatment (Fiks 2010). A study of young adults transitioning to adult services showed that the acquisition of independence in everyday life was accompanied by greater autonomy in managing illness (Duguéproux 2008). Programmes that embed SDM in the training of health professionals may increase awareness of SDM as a model of patient-centred care and equip healthcare professionals with the skills to engage competently with SDM. Self-management programmes that advocate SDM may improve patient satisfaction with care. The inclusion of a decision-aid during a consultation with a clinician indicates to the patient that their opinions and preferences are valued. Increasingly, decision aids are being used to assist with the process of SDM and evidence strongly suggests that patient decision-aids can improve the quality of decision-making (O’Connor 2007). A Cochrane Review found that the application of decision aids led to a significant improvement in knowledge regarding a decision, as well as significantly reducing anxiety and decision conflict (Stacey 2014). A literature review concluded that patient-centred care is a key factor in increasing adherence to treatment.
(Robinson 2008) and a systematic review reported evidence for positive influences on patient-centred care and self-management (Rathert 2013).

**Why it is important to do this review**

Advances in treatment continue to extend the life span of young people with CF (MacKenzie 2014). As survival improves, the long-term complications of the disease as well as depression will become increasingly evident adding to the treatment burden (Anton-Paduraru 2014; Buntain 2004; Quon 2012). Furthermore, some adolescents with CF may transition to adult care with near normal lung function (Quon 2012), but in others the transition may be associated with a decline in lung function (Duguépéroux 2008) since transition requires greater responsibility for self-care and taking on more self-advocacy skills (Okumura 2014).

Implementing SDM can help foster patient-centred care and is particularly relevant given the increasing number of healthcare choices (Stacey 2010; Weiner 2014). There is currently a strong emphasis on the importance of SDM in the healthcare setting (De Boer 2013; Kiessling 2013; Klatt 2013; Légare 2014; Muller-Juge 2013). As providers, purchasers, policymakers, and consumers explore opportunities to integrate patient-centred concepts into standards of care, SDM is receiving increased attention (Shafir 2012). The time seems right to engage healthcare professionals and policy makers in devising policies that prioritise SDM for children and adolescents living with CF, especially given the new emerging therapies for CF. Additionally, this systematic review supports and promotes the United Nation’s Convention on the Rights of the Child (UNCRC), which places value and importance on the child’s opinion with the statement: “Parties shall assure to the child who is capable of forming his or her own views the right to express those views freely in all matters affecting the child, the views of the child being given due weight in accordance with the age and maturity of the child” (United Nations 1989).

An annual review is now standard practice for people with CF and consensus documents define standards for routine evaluation, monitoring and treatment (Chuang 2014; Long 2001). While consensus documents such as that by Kermen cite CF as requiring a holistic approach (Kermen 2005), others such as that by Walters make no reference to SDM (Walters 1990). However, the paediatric CF annual review process has the potential to promote SDM in CF care; for example, on its website, the annual review clinic in CF paediatric care at Great Ormond Street Hospital for children is described as “designed to provide more time with members of the CF team than is usually available in the CF clinic. This allows more detailed discussion of relevant issues” (GOSH 2016).

In so far as SDM is associated with beneficial outcomes (Elf 2015), clarification of evidence-based age-appropriate SDM interventions for children and adolescents living with CF becomes increasingly relevant. Healthcare professionals need to know how to involve children and adolescents with CF in SDM and which interventions are most effective (Coyne 2014). Despite the ever-increasing focus on decision aids in the literature (Abhyankar 2013; Stacey 2015), SDM is not routinely applied in clinical practice and limited use of SDM has been observed, despite previous research indicating that parents wish to collaborate in decision making (Lipstein 2014). Studies on children’s experiences of hospitalisation report a lack of self determination regarding personal needs (Coyne 2006). A systematic review of interventions that promote participation in SDM for children and adolescents with CF will aim to identify a body of evidence from which a SDM framework could be established.

**OBJECTIVES**

To determine the effectiveness of interventions that promote SDM for children and adolescents with CF aged four to 18 years.

**METHODS**

**Criteria for considering studies for this review**

**Types of studies**

We will include all randomised controlled trials (RCTs) and cluster RCTs. We will not include trials with a cross-over design since some interventions to promote SDM may have a sustained effect.

**Types of participants**

We will include children and adolescents diagnosed with CF clinically (Farrell 2008), by sweat testing (McKone 2015) or genetically (Martiniano 2014) with or without any concomitant disease or disorder and aged between four and 18 years. Consistent with a previously reported Cochrane Review on SDM for children with cancer, we will exclude children under four years of age as they are potentially too young to adequately engage in the intervention (Coyne 2013). Children between the age of four and 15 years have participated in discussions related to child-parent decision-making roles (Miller 2008). Since SDM is a collaborative process with the child, we will also include parents and carers and any healthcare professionals (doctors, nurses or allied healthcare professionals) who are involved in the care of children and young people with CF. However, this review is about finding out the best way to support the participation of children and adolescents in SDM so that the decision-making is not dominated by parents and healthcare professionals.
Comment: above in background you state ".we will define a child as up to nine years old and an adolescent as between 10 and 19 years of age (WHO 2016)". Address issue of 18 or 19 year olds in inclusion criteria. Also define an adolescent in 'Types of Participants'.

Types of interventions
We will compare interventions for promoting SDM in children or adolescents which are aimed at children or adolescents, their parents or healthcare professionals (or any combinations of these groups) to usual care or to alternative SDM promotion strategies for the same group of people. We will include interventions delivered in a number of formats including a one-to-one basis, a group basis, discussion sessions, role play sessions, blended learning sessions, online learning sessions and the use of hard-copy information resources such as leaflets or workbooks. Interventions may be delivered by professionals or parents or both.

Note: the term parent includes the parent or carer or guardian who is responsible for the parental role; for convenience the term parent will be applied where appropriate.

Types of outcome measures

Primary outcomes
1. Presence of shared decision-making measured by the change in score of any validated tool
   i) the Observing Patient Involvement 12-item (OPTION) Scale (Elwyn 2003)
   ii) the Observer-based Measure Observer 5-item (OPTION) Scale (Elwyn 2013a)
   iii) decision-making instrument facilitation antecedents (e.g. the Preparation for Decision-making Scale (Graham 1995))
   iv) decision process (e.g. the Rochester Participatory Decision-making Scale (Shields 2005))
2. Quality of life (QoL) as measured by e.g. the Cystic Fibrosis Questionnaire-Revised (CFQ-R) (Quittner 2005; Quittner 2009; Quittner 2012) or the CF disease-specific (HRQoL) questionnaire (Gee 2000)
3. Adverse effect such as longer consultation time, increased frequency of hospital admissions, longer hospital stay, increased costs or unanticipated adverse effects as reported by study authors

Secondary outcomes
1. Adherence to CF medication (measured by e.g. electronic monitoring)
2. Anxiety
3. Decision conflict (as measured by the Decision Conflict Scale (O’Connor 1995) or the SURE scale (Légare 2010))
4. Decision regret (as measured by the Decision Regret Scale (Brehaut 2003))
5. Participant satisfaction with decision
6. Depression (measured by e.g. the Generalised Anxiety Disorder 7-item (GAD-7) Scale (Spitzer 2006), or the CES-D Scale (Eaton 2004), or the Hospital Anxiety and Depression Scale (HADS) (Zigmond 1983))
7. Changes in lung function (measured by forced expiratory volume in one second (FEV1))
8. Changes in nutritional indices
   i) weight (kg)
   ii) body mass index (BMI)
   iii) height (cm)
9. Survival

Search methods for identification of studies

Electronic searches
The authors will identify relevant studies from the Cochrane Cystic Fibrosis and Genetic Disorders Group's Cystic Fibrosis Trials Register using the terms: mental health in CF OR *program*.
The Cystic Fibrosis Trials Register is compiled from electronic searches of the Cochrane Central Register of Controlled Trials (CENTRAL) (updated each new issue of The Cochrane Library), weekly searches of MEDLINE, a search of Embase to 1995 and the prospective handsearching of two journals - Pediatric Pulmonology and the Journal of Cystic Fibrosis. Unpublished work is identified by searching the abstract books of three major cystic fibrosis conferences: the International Cystic Fibrosis Conference; the European Cystic Fibrosis Conference and the North American Cystic Fibrosis Conference. For full details of all searching activities for the register, please see the relevant sections of the Cochrane Cystic Fibrosis and Genetic Disorders Group's website.
The systematic review authors will also initially search the following databases.
- PubMed (1946 to present) (Appendix 1)
- CINAHL (EBSCO) (1982 to present) (Appendix 2)
- Embase (OvidSP) (1995 to present) (Appendix 3)
- PsycINFO (EBSCO) (1806 to present) (Appendix 4)
- WHO International Clinical Trials Registry Platform (ICTRP), which includes data from ClinicalTrials.gov and the ISRCTN Register (Appendix 5)
- ASSIA - Applied Social Sciences Index and Abstracts (1987 to present) (Appendix 6)
- ERIC (1966 to present) (Appendix 7)

Searching other resources
Grey literature
We will search ProQuest Dissertation & Theses Global to access digital theses and dissertations that are relevant to this systematic review (ProQuest 2013) (Appendix 8). We will also search Open Grey (Open Grey 2015).

Handsearches
We will search subscription copies of the Medical Decision Making (MDM) journal for rigorous and systematic approaches to decision making.

Reference lists
We will search the reference lists of all included studies for other studies relevant to this systematic review.

Personal communication
We will contact (by email or telephone) experts in the field of CF who have agreed to provide advisory support to this systematic review and (where relevant) authors of included studies for the identification of further published papers of interest to this systematic review.

Data collection and analysis

Selection of studies
We will select studies in accordance with the Cochrane Handbook for Systematic Reviews of Interventions (Higgins 2011a). We will compile a study eligibility form with clear inclusion and exclusion criteria and using this form, the authors will independently select studies for inclusion in the review from those identified by the searches. Where the eligibility of a study is not clear, we will list the study as ‘Awaiting classification’ and contact the investigators for clarification. Studies (not reports) will be our unit of interest (Higgins 2011a). We will use reference management software to remove duplicate records (Lefebvre 2011) and will list all references (both full papers and abstracts) to a single study under a single identity code in Review Manager (RevMan 2014). We will list as excluded any studies that initially appear to be eligible, but which on further inspection are not, and we will record the primary reason for exclusion. We will resolve disagreements regarding the inclusion of a study by discussion leading to consensus (Higgins 2011a). In order to check agreement of consistency between the two authors (HM, IC) in selecting studies, we will generate a Kappa K co-efficient using four categories (agree/agree), (disagree/disagree), (agree/disagree) and (disagree/agree) and will report the number of studies included in this calculation. We will apply a co-efficient value of 0.7 (indicating a good level of agreement) (Harris 2007).

Data extraction and management
The authors will use a study selection, quality assessment and data extraction form template (developed by the Cochrane Cystic Fibrosis and Genetic Disorders Group) modified to meet the objectives of this review. We will also consider the need to develop any additional data extraction tools and will seek guidance for collecting data from the Cochrane Handbook for Systematic Reviews of Interventions (Higgins 2011a). In order to clarify, refine and ensure consistency of data extraction, we will pilot test our data extraction form by taking a representative sample (based on the number of studies to be reviewed) to identify any problems with the tool. We will seek a consensus between authors before we modify the form. Two authors (HM, IC) will independently extract the data. A third author will cross-check the data collection forms. We will resolve disagreements regarding the data extraction by discussion leading to consensus (Higgins 2011a). We will extract the following details from study reports if possible and endeavour to contact study authors for clarification of any missing data.
- Study characteristics: author’s name; year of publication; study title; country; ethical approval; hypothesis and alternative hypothesis; aim of study; study design; phase of RCT; span or duration of the study.
- Participant characteristics: method of CF diagnosis; age; gender; ethnicity; socio-economic group; and education.
- Sample size: method used to calculate sample size; the sample size estimated prior to the trial; numbers approached to participate; numbers who agreed to participate; and numbers that actually participated; numbers in the intervention and control group; final sample size following attrition; loss to follow-up or missing data.
- Inclusion and exclusion criteria.
- Intervention: content; process and comparator; whether the intervention had a training component for participants; and whether the delivery of the training was by a parent or a professional or both.
- Outcome data for our listed outcomes at the following time points: one month, over one month and up to three months, over three months and up to six months (if outcome data are available for other time points, we will consider these for examination).
- Data type, e.g. dichotomous data, continuous data.
- Study author’s conclusions and limitations.
- Duration of the intervention (not the study), e.g. data for a study that has a daily intervention for one week followed by a six-month follow-up will be recorded as a one-week intervention.
- Additional information that may potentially affect the interpretation or applicability of results.

We will present study details in a ‘Characteristics of included studies’ table in the Review Manager software (RevMan 2014). One au-
Dealing with missing data

We will endeavour to differentiate between data which are missing at random and data which are not (e.g. due to publication bias, selective reporting bias, attrition and selection of participants) (Higgins 2011c). For data not missing at random, we will contact the study authors to establish reasons for this and to request any data we require for our analysis (e.g. means and standard deviations (SDs)). If these data are not available, we will consider imputing values either from studies in the same meta-analysis or from studies in other meta-analyses (Furukawa 2006). We will only impute values from studies using the same measurement scale, with the same degree of measurement error and over the same time periods (Higgins 2011c).

We will consider presenting results as an ‘available-case analysis’ (where we analyse data for the participants for whom data are available). If we are able to do so without imputing values, we will present our results using an intention-to-treat (ITT) analysis. If we are not able to obtain data we need for our analysis, we will report the study results narratively and discuss the potential implications of their omission from a meta-analysis (Higgins 2011c).

Assessment of heterogeneity

We plan to assess the heterogeneity (variability) between included studies. Initially, we will assess clinical heterogeneity by comparing study characteristics and participant demographics; we will assess methodological heterogeneity by comparing the different study designs and risk of bias judgements. If we consider studies to be too heterogeneous, we will not combine them in a meta-analysis. If we are able to combine studies in a meta-analysis we will further assess for heterogeneity by visually inspecting the forest plot to see if the CIs overlap. We also plan to assess heterogeneity using the Chi² test and the I² statistic. The I² statistic describes the percentage of total variation across studies due to heterogeneity rather than chance where an I² value of 0% indicates no observed heterogeneity and

Unit of analysis issues

For each included study, we will determine the level at which randomisation took place i.e. by individual or by cluster. If an included study employs a cluster-randomisation design, i.e. participants are randomised to a particular group according to clinician or particular practice, we plan to assess these studies for unit of analysis issues. If only cluster RCTs with unit of analysis issues are available for meta-analysis we will consider carrying out ‘approximate analysis’ of cluster RCTs, following guidance from Donner to estimate the intracluster (or intraclass) correlation coefficient (ICC), which is required for this analysis (Donner 1980; Higgins 2011c). In light of guidance in the Cochrane Handbook for Systematic Reviews of Interventions, we will consider whether to analyse parallel RCTs and cluster RCTs separately or to combine them using the generic inverse variance method of meta-analysis (Deeks 2011).

For studies with multiple treatment groups, we will follow guidance from the Cochrane Handbook for Systematic Reviews of Interventions (Higgins 2011c).
larger values show increasing heterogeneity (Higgins 2003). Since the thresholds for the interpretation of $I^2$ can be misleading, we plan to use the following guide (Deeks 2011):

- 0% to 40%: might not be important;
- 30% to 60%: may represent moderate heterogeneity;
- 50% to 90%: may represent substantial heterogeneity;
- 75% to 100%: considerable heterogeneity.

We will seek advice from a statistical expert for the appraisal of our $I^2$ result.

**Assessment of reporting biases**

We will examine the included studies for any evidence of reporting biases (Sterne 2011a). We will compare any published protocols we identify with the final study reports to ensure that all planned outcomes are reported. If protocols are not available, we plan to compare the 'Methods' section and the 'Results' section of the final published report to identify any discrepancies. If necessary, we will contact study authors for additional clarification. Additionally, if we are able to combine at least 10 studies, we will evaluate each outcome for publication bias using a funnel plot (Sterne 2011a). We will examine these funnel plots and assess any asymmetry present, noting that this is not necessarily due to publication bias, but may have a number of other possible causes (Sterne 2011b).

**Data synthesis**

We will only carry out a meta-analysis if we judge study design, participants, intervention, and outcome to be sufficiently similar to derive a clinically meaningful result. Since there is no single true effect for SDM, we will not estimate a single effect size and plan to apply a random-effects model which makes an adjustment to the study weights according to the extent of variation or heterogeneity (Deeks 2011). Additionally, the random-effects model will award relatively more weight to smaller studies than a fixed-effect model in a heterogeneous set of studies (Deeks 2011). If we are not able to present the study results in a meta-analysis, we will present them narratively by intervention type, population group and setting etc (Schünemann 2011a).

**Subgroup analysis and investigation of heterogeneity**

If searches yield a sufficient number of studies which we are able to combine in a meta-analysis we will conduct a subgroup analysis by:

- age for the following ranges: 4 to 7 years; 8 to 11 years; 12 to 15 years; 16 to 18 years;
- male versus female participants.

**Sensitivity analysis**

In order to check the robustness of the summary statistic, we plan to carry out sensitivity analyses if there are sufficient comparable trials (at least 10) included in the review:

- repeating the analysis using different statistical models (fixed-effect versus random-effects models);
- adding in and taking out trials where there is high risk of bias (rating determined using a 'Risk of Bias' graph) in relation to randomisation, allocation concealment, or blinding of the interventions from participants or trial personnel.

**Summary of findings table**

We will prepare a 'Summary of Findings' table to present the results of our meta-analyses for each comparison, based on the methods described in Chapter 11 of the Cochrane Handbook for Systematic Reviews of Interventions (Schünemann 2011a). We will present a separate 'Summary of findings' table for each comparison and report the outcomes listed below. Findings from interventions that promote shared decision-making too clinically different to be summarised together will be summarised in separate comparisons.

1. Presence of shared decision-making
2. Quality of life
3. Adverse effects (such as longer consultation time, increased frequency of hospital admissions, longer hospital stay, unanticipated adverse effects as reported by study authors)
4. Adherence to CF medication
5. Anxiety
6. Decision conflict
7. Decision regret

We will grade the quality of evidence as high, moderate or low in accordance with the specific evidence grading system described by Schünemann and developed by the Grade Working Group (Atkins 2004; Schünemann 2011b). If meta-analysis is not possible we will present results in a narrative summary in accordance with guidance (Schünemann 2011a).

**Economic issues**

We do not plan to incorporate economic issues in this review since from our knowledge based on a literature review we do not envisage that studies will make explicit comparisons between alternative interventions in terms of either costs (resource use) or consequences (effects).

**Consumer involvement**

Our consumer expert is Susan Biggar, Consumer Partnerships, Health Issues Centre, Melbourne Australia, who is a co-author on this review. The authors also have strong links with the consumer organisation Cystic Fibrosis Ireland and we will send a draft of the
review to our contacts in this organisation for comment, particularly regarding the discussion and recommendations.

ACKNOWLEDGEMENTS

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Anton-Paduraru 2014

Atkins 2004

Bejarano 2015

Birchley 2014

Brand 2013

Brehaut 2003

Brice 2007

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Open Grey 2015

Polaris 2014

President’s Commission Report 1982

ProQuest 2013

Quaschning 2013

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Quiñones 2009

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Robinson 2008

Roehrle 2013

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Sabadosa 2014

Savage 2007

Schünemann 2011a


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Walters 1990


Wennberg 2010

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WHO 2016

Williams 2010

Wyatt 2015

Zigmond 1983

* Indicates the major publication for the study

APPENDICES

Appendix 1. Search strategy for PubMed (NLM)

1. For decision making the following MeSH headings and text words will be used:
   ("attitude of health personnel"[Mesh Terms] OR "attitude to health"[Mesh Terms] OR "choice behavior"[Mesh Terms] OR "communication"[Mesh Terms] OR "consumer participation"[Mesh Terms] OR "cooperative behavior"[Mesh Terms] OR "decision making"[Mesh Terms] OR "decision support techniques"[Mesh Terms] OR "decision theory"[Mesh Terms] OR "educational technology"[Mesh Terms] OR "health education"[Mesh Terms] OR "informed consent"[Mesh Terms] OR "professional-family relations"[Mesh Terms] OR "psychology"[Subheading] OR ((affective OR cognitive OR emotional OR psychosocial OR psychosomatic) AND aspect*) OR (choice OR compliant OR cooperative OR co-operative OR co-operative OR illness) AND (behavior*) OR (clinical support technique* OR collaborati* OR communication* OR consensus OR consent* OR consumer* OR decision* OR dispute* OR dissent* OR ((doctor OR doctors OR physician* OR provider*) AND (attitude OR patient relation*))) OR educational technology OR (health AND (attitude* OR education OR information OR literacy)) OR (informed AND (assent OR choice*)) OR misinformation OR negotiat* OR (nurs* AND role*) OR participation* OR partner* OR (patient AND (acceptance OR adherence OR attitude* OR centered OR centre* OR compliance OR cooperation OR co-operation OR education OR involvement OR nonadherence OR noncompliance OR participation OR preference* OR satisfaction)) OR (professional AND (family disagreement* OR family relation* OR patient disagreement*)) OR refusal participat* OR staff attitude* OR treatment refusal* OR uncertainty)

2. For cystic fibrosis the following MeSH headings and text words will be used:
   ("cystic fibrosis"[Mesh Terms] OR ((cystic OR pancreas OR pancreatic) AND fibrosis) OR fibrocystic OR mucoviscidosis)

3. For RCTs/CCTs the following MeSH headings and text words will be used:
   (((random*[tiab] OR "controlled"[tiab]) AND trial*[tiab]) OR "randomised"[tiab] OR "randomized"[tiab] OR "randomly"[tiab] OR "Randomized Controlled Trial"[Publication Type] OR "Controlled Clinical Trial"[Publication Type] OR "Randomized Controlled Trials as Topic"[Mesh Terms] OR "Placebos"[Mesh Terms] OR placebo)

The final combined search will be: 1 AND 2 AND 3.

Note: On the advice of the Trial Search Co-ordinator, terms for Children 4-18 years will not be used to limit the search, in order to maximize recall of relevant results.

*tiab = title or abstract; * = zero or more characters
Appendix 2. Search strategy for CINAHL (EBSCO)

1. For decision making the following CINAHL subject headings and text words will be used:
   (MH “Attitude of Health Personnel+” OR MH “Attitude to Health+” OR MH “Communication+” OR MH “Consumer Participation” OR MH “Cooperative Behavior” OR MH “Decision Making+” OR MH “Decision Support Techniques+” OR MH “Educational Technology” OR MH “Health Education+” OR MH “Consent+” OR MH “Professional-Family Relations” OR MH “Psychology+” OR MH “Nursing Role” OR (affective OR cognitive OR emotional OR psychosocial OR psychosomatic AND aspect*) OR ((choice OR compliant OR cooperative OR co-operative OR illness) AND behavior*) OR clinical support technique* OR collaboration* OR communication* OR consensus OR consent* OR consumer* OR decision* OR dispute* OR dissent* OR ((doctor OR doctors OR physician* OR provider*) AND (attitude OR patient relation*)) OR educational technology OR (health AND (attitude OR education OR information OR literacy)) OR (informed AND (assent OR choice*)) OR misinformation OR negotiat* OR (nurs* AND role*) OR participat* OR partner* OR (patient AND (acceptance OR adherence OR attitude* OR centered OR centred OR compliance OR cooperation OR co-operation OR education OR involvement OR nonadherence OR noncompliance OR preference* OR satisfaction)) OR (professional AND (family disagreement* OR family relation* OR patient disagreement*)) OR staff attitude* OR treatment refusal* OR uncertainty)

2. For cystic fibrosis the following CINAHL subject headings and text words will be used:
   (MH “Cystic Fibrosis” OR ((cystic OR pancreas OR pancreatic) AND fibrosis) OR fibrocystic OR mucoviscidosis)

3. For RCTs/CCTs the following CINAHL subject headings and text words were used:
   (((random* OR controlled) AND trial*) OR MH “Placebos” OR MH “Clinical Trials” OR (TI randomized OR AB randomized) OR (TI randomly OR AB randomly) OR (TI randomised OR AB randomised) OR placebo*)

The final combined search will be: 1 AND 2 AND 3.

Note: On the advice of the Trial Search Co-ordinator, terms for Children 4-18 years will not be used to limit the search, in order to maximize recall of relevant results.

Appendix 3. Search strategy for Embase (OvidSP)

1. For decision making the following text words will be used:
   1. affective OR cognitive OR emotional OR psychosocial OR psychosomatic
   2. aspect*
   3. #1 AND #2
   4. choice OR compliant OR cooperative OR co-operative OR illness
   5. behavior*
   6. #4 AND #5
   7. clinical support technique* OR collaborati* OR communication* OR consensus OR consent* OR consumer* OR decision* OR dispute* OR dissent*
   8. doctor OR doctors OR physician* OR provider*
   9. attitude OR patient relation*
   10. #8 AND #9
   11. educational technology
   12. health
   13. attitude* OR education OR information OR literacy
   14. #12 AND #13
   15. informed
   16. assent OR choice*
   17. #15 AND #16
   18. misinformation OR negotiat* OR nurs* AND role*
   19. participat* OR partner*
   20. patient
   21. acceptance OR adherence OR attitude* OR centered OR centred OR compliance OR cooperation OR co-operation OR education OR involvement OR nonadherence OR noncompliance OR preference* OR satisfaction
   22. #21 AND #22
   23. #21 AND #22

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24. professional
25. family disagreement* OR family relation* OR patient disagreement*
26. #24 AND #25
27. staff attitude* OR treatment refusal* OR uncertainty
28. #3 OR #6 OR #7 OR #10 OR #11 OR #14 OR #17 OR #18 OR #19 OR #20 OR #23 OR #26 OR #27.
2. For cystic fibrosis the following text words will be used:
29. cystic OR pancreas OR pancreatic
30. fibrosis
31. #30 AND #31
32. fibrocystic OR mucoviscidosis
33. #31 OR #32
3. For RCTs/CCTs the following text words were used:
34. random* OR controlled
35. trial*
36. #34 AND #35
37. randomly OR randomized OR randomised OR placebo*
38. #36 OR #37
The final combined search will be: #28 AND #33 AND #38.

Note: On the advice of the Trial Search Co-ordinator, terms for Children 4-18 years will not be used to limit the search, in order to maximize recall of relevant results.

Appendix 4. Search strategy for PsycINFO (EBSCO)

1. For decision making the following PsycINFO Thesaurus Descriptors subject headings and text words will be used:
   (DE “Decision Making” OR DE “Decision Support Systems” OR DE “Decision Theory” OR DE “Choice Behavior” OR DE “Group Decision Making” OR DE “Health Education” OR DE “Health Behavior” OR DE “Health Personnel Attitudes” OR DE “Health Attitudes” OR DE “Communication” OR DE “Interpersonal Communication” OR DE “Persuasive Communication” OR DE “Choice Behavior” OR DE “Informed Consent” OR ((affective OR cognitive OR emotional OR psychosocial OR psychosomatic) AND aspect*) OR ((choice OR compliant OR cooperative OR co-operative OR ill) AND behavio*) OR clinical support technique* OR collaborati* OR communication* OR consensus OR consent* OR consumer* OR decision* OR disput* OR dissent* OR ((doctor OR doctors OR physician OR provider*) AND (attitude OR patient relation*)) OR educational technology OR (health AND (attitude* OR education OR information OR literacy)) OR (informed AND (assent OR choice*)) OR misinformation OR negotiati* OR (nurs* AND role*) OR participati* OR partner* OR (patient AND (acceptance OR adherence OR attitude* OR centered OR centred OR compliance OR cooperation OR co-operation OR education OR involvement OR nonadherence OR noncompliance OR preference* OR satisfaction)) OR (professional AND (family disagreement* OR family relation* OR patient disagreement*)) OR staff attitude* OR treatment refusal* OR uncertainty)

2. For cystic fibrosis the following PsycINFO Thesaurus Descriptors subject headings and text words will be used:
   (DE “Cystic Fibrosis” OR ((cystic OR pancreas OR pancreatic) AND fibrosis) OR fibrocystic OR mucoviscidosis)

3. For RCTs and CCTs the following PsycINFO Thesaurus Descriptors subject headings and text words were used:
   (DE “Placebo” OR ((random* OR controlled) AND trial*) OR randomly OR randomized OR randomised OR placebo*)
The final combined search will be: 1 AND 2 AND 3.

Note: On the advice of the Trial Search Co-ordinator, terms for Children 4-18 years will not be used to limit the search, in order to maximize recall of relevant results.

[DE = PsycINFO Thesaurus Descriptors; TI = title; AB = abstract; * = zero or more characters]
Appendix 5. Search strategy for WHO International Clinical Trials Registry Platform

For cystic fibrosis the following text words will be used:
cystic fibrosis OR pancreatic fibrosis OR fibrocystic OR mucoviscidosis
The search will be conducted in the title field, limited using the clinical trials in children filter. Both recruiting and non-recruiting trials will be included.

Appendix 6. Search strategy for ASSIA (ProQuest)

1. For decision making the following ProQuest subject headings and text words will be used:
(SU.EXACT.EXPLODE(“Decision Making” OR “Participative Decision Making”) OR SU.EXACT.EXPLODE(“Decision Making Skills”) OR ((affective OR cognitive OR emotional OR psychosocial OR psychosomatic) AND aspect*) OR ((choice OR compliant OR cooperative OR co-operative OR illness) AND behavio*) OR clinical support technique* OR collaborati* OR communication* OR consensus OR consent* OR consumer* OR decision* OR disput* OR dissent* OR ((doctor OR doctors OR physician* OR provider*) AND (attitude OR patient relation*)) OR educational technology OR (health AND (attitude* OR education OR information OR literacy)) OR (informed AND (assent OR choice*)) OR misinformation OR negociati* OR (nurs* AND role*) OR participat* OR partner* OR (patient AND (acceptance OR adherence OR attitude* OR centered OR centred OR compliance OR cooperation OR cooperation OR education OR involvement OR nonadherence OR noncompliance OR preference* OR satisfaction)) OR (professional AND (family disagreement* OR family relation* OR patient disagreement*)) OR staff attitude* OR treatment refusal* OR uncertainty)
2. For cystic fibrosis the following ProQuest subject headings and text words will be used:
(((cystic OR pancreas OR pancreatic) AND fibrosis) OR fibrocystic OR mucoviscidosis)
3. For RCTs and CCTs the following text words were used:
((((random* OR controlled) AND trial*) OR randomly OR randomized OR randomised OR placebo*))
The final combined search will be: 1 AND 2 AND 3.

Note: On the advice of the Trial Search Co-ordinator, terms for Children 4-18 years will not be used to limit the search, in order to maximize recall of relevant results.

Appendix 7. Search strategy for ERIC (ProQuest)

1. For decision making the following ProQuest subject headings and text words will be used:
(SU.EXACT.EXPLODE(“Decision Making” OR “Participative Decision Making”) OR SU.EXACT.EXPLODE(“Decision Making Skills”) OR ((affective OR cognitive OR emotional OR psychosocial OR psychosomatic) AND aspect*) OR ((choice OR compliant OR cooperative OR co-operative OR illness) AND behavio*) OR clinical support technique* OR collaborati* OR communication* OR consensus OR consent* OR consumer* OR decision* OR disput* OR dissent* OR ((doctor OR doctors OR physician* OR provider*) AND (attitude OR patient relation*)) OR educational technology OR (health AND (attitude* OR education OR information OR literacy)) OR (informed AND (assent OR choice*)) OR misinformation OR negociati* OR (nurs* AND role*) OR participat* OR partner* OR (patient AND (acceptance OR adherence OR attitude* OR centered OR centred OR compliance OR cooperation OR cooperation OR education OR involvement OR nonadherence OR noncompliance OR preference* OR satisfaction)) OR (professional AND (family disagreement* OR family relation* OR patient disagreement*)) OR staff attitude* OR treatment refusal* OR uncertainty)
2. For cystic fibrosis the following ProQuest subject headings and text words will be used:
(((cystic OR pancreas OR pancreatic) AND fibrosis) OR fibrocystic OR mucoviscidosis)
3. For RCTs and CCTs the following text words will be used:
((((random* OR controlled) AND trial*) OR randomly OR randomized OR randomised OR placebo*))
The final combined search will be: 1 AND 2 AND 3.

Note: On the advice of the Trial Search Co-ordinator, terms for Children 4-18 years will not be used to limit the search, in order to maximize recall of relevant results.

[SU.EXACT.EXPLODE = ProQuest subject heading (exploded); * = zero or more characters]
Appendix 8. Search strategy for ProQuest Dissertations and Theses (ProQuest)

Cystic fibrosis AND (((random* OR controlled) AND trial*) OR randomized OR randomly OR randomised OR placebo*)

Search will be run in all indexed fields, but not within the full text of theses.

Note: On the advice of the Trial Search Co-ordinator, terms for Children 4-18 years will not be used to limit the search, in order to maximize recall of relevant results.

Contributions of Authors

<table>
<thead>
<tr>
<th>Task</th>
<th>Who undertook task</th>
</tr>
</thead>
<tbody>
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<td>Protocol/Review stage: supervisor</td>
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### DECLARATIONS OF INTEREST

Helen Malone declares no known conflict of interest.

Imelda Coyne declares no known conflict of interest.

Greg Sheaf declares no known conflict of interest.

Sheila Javadpour declares no known conflict of interest.

Susan Biggar declares no known conflict of interest.

Zai Edworthy declares no known conflict of interest.

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| Review stage: analysis of data | Helen Malone |
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| Review stage: providing a methodological perspective | Imelda Coyne |
| Review stage: providing general advice on the review | Imelda Coyne, Susan Bigger, Sheila Javadpour, Zai Edworthy |
| Review stage: writing the review | Helen Malone, Susan Bigger, Imelda Coyne |
| Performing previous work that was the foundation of the current review | Imelda Coyne |
| Update stage: updating review | Helen Malone, Imelda Coyne |