

Commentary

QJM

Stravinsky syndrome: giving a voice to chronic stroke disease

D. O'NEILL¹, C.A. MACSWEENEY¹, I.A. CORNELL¹ and H. MOSS²

From the ¹Centre for Ageing, Neuroscience and the Humanities, Trinity College Dublin, Tallaght Hospital, Dublin and ²National Centre for Arts and Health, Tallaght Hospital, Dublin, Ireland

Address correspondence to D. O'Neill, Centre for Ageing, Neuroscience and the Humanities, Trinity Centre for Health Sciences, Tallaght Hospital, Dublin 24, Ireland. email: doneill@tcd.ie

Few areas of medical practice and history generate such lively discussion as the continuing relevance and appropriateness of eponyms.^{1,2} Given controversies of unethical practice by physicians associated with some eponyms,³ misattribution of the original description⁴ and the transformation of progression in medical research from the model of the inspired solo practitioner to that of interdisciplinary team work, it is likely that few future physicians will generate a widely adopted eponym.

In recent times, a different form of eponym has evolved whereby wider awareness and appreciation of relatively neglected illnesses has occurred through identification with a famous person affected by the illness. The cause, and popular understanding, of motor neuron disease in North America was considerably supported by the adoption of the title Lou Gehrig disease, after the famous baseball player.⁵ In a similar fashion, public awareness of adrenoleukodystrophy was increased by its association with Lorenzo's oil, a potential therapy named after a patient.⁶

Chronic stroke disease

Could chronic stroke disease, a complex illness with huge societal impact, benefit in a similar way from identification with a famous sufferer? Although stroke has been relatively neglected in terms of research funding and clinical development,⁷ the welcome focus in the last two decades on acute and hyperacute care—stroke units and thrombolysis—may divert attention from the wider and more chronic manifestations of stroke disease.⁸ These include dementia,⁹ gait disorders,¹⁰ depression¹¹ and

oropharyngeal dysphagia,¹² a combination of which may occur in a single person. The recognition of these stroke syndromes is important at many levels: they represent an important target for primary and secondary prevention,¹³ as well as opportunities for better assessment, treatment and management. Yet even in major international conferences in stroke, these conditions merit scant attention, and knowledge among the general public is likely to be even more circumscribed.

The late illnesses of Igor Stravinsky (1882–1971) could provide a rallying point for chronic stroke syndromes, and highlight the relative lack of curiosity in doctors as to the cause of these syndromes which persists to this day. He is also an inspirational figure for those who have endured stroke disease, as his creativity and vitality in the face of both acute and chronic stroke disease are shining beacons in a narrative of illness that can all too easily tend to despondency and stigma.

A series of strokes

Stravinsky suffered from a significant acute stroke in 1956. This first overt stroke occurred while conducting at a concert in Berlin, and his symptoms included headache, right-sided dysaesthesia with reduced coordination and dysarthria. Reflecting the somewhat more relaxed attitude to stroke of the times (or perhaps elements of denial and anosagnosia), he proceeded to a meal of goose liver and apple strudel, washed down with Sekt before flying to Munich later that evening! There, his fellow composer Karl Amadeus Hartmann called a physician friend, Professor Diehl, and he was admitted to the

Red Cross Hospital. There he was diagnosed with a multi-ocular encephalomalacia, with an 'atheromatous-thrombotic-vasospastic process in the cerebrum' (Professor Diehl, personal communication).

Within 6 weeks he was conducting again, and a stream of compositions flowed: he composed more works in the following decade than he had in the 1930s and 1940s. In addition, he continued to show development and innovation, adopting serial composition during one of his later phases. He had taken up conducting relatively late in life, and was to continue conducting until the age of 85, albeit with a gradually diminishing tempo of engagements and a declining critical reception. At least one further episode, in December of 1967, would appear to have been an overt stroke, marked by transient aphasia and immobility.

In terms of detecting patterns of illness, there is an extensive collection of descriptions of Stravinsky's middle and later years arising from the voluminous correspondence, narratives and diaries collated by his amanuensis, the conductor and writer Robert Craft^{14,15} as well as memoirs from Stravinsky and other sources^{16–24} and authoritative biographies.²⁵ Written from a non-medical perspective, they give many details of Stravinsky's recognized illnesses, including the thrombocytosis which may have been a risk factor for stroke disease, as well as relevant life-style habits including appetites for smoking, alcohol and rich food.

However, these accounts also describe progressive changes in his cognitive function, gait and swallow which suggest a pattern of deterioration consistent with chronic stroke disease. Given that

categorization and expertise in detection and management of this syndrome was not to develop for another three decades, it is not surprising that relatively little analysis was applied to these symptoms at the time. In addition, the preservation of music appreciation and enjoyment developed over many decades, as well as a significant level of financial and human support, allowed a degree of compensation which may have concealed the syndrome from those surrounding him.

Stigmata of progressive chronic stroke disease

Gait dyspraxia

The progressive decline in Stravinsky's mobility was probably the first manifestation of chronic stroke disease (Figure 1). As early as 1962 he was known to use a walking aid, and the need for using a wheelchair over longer distances. His wife, Vera, described him as stooped and frail in correspondence in 1965, walking slowly with a cane, and requiring a wheelchair for distances greater than a few hundred feet. She ascribed his mobility difficulties to a hernia: however, it is unlikely that a hernia would cause this level of difficulty, and she also noted a right-sided weakness and dysaesthesia. Video evidence from a concert that he conducted in Toronto at the age of 85 is revealing: seated at the podium, he shows musical mastery. However, his disequilibrium and magnetic gait on walking to receive an award are typical of a vascular higher-level

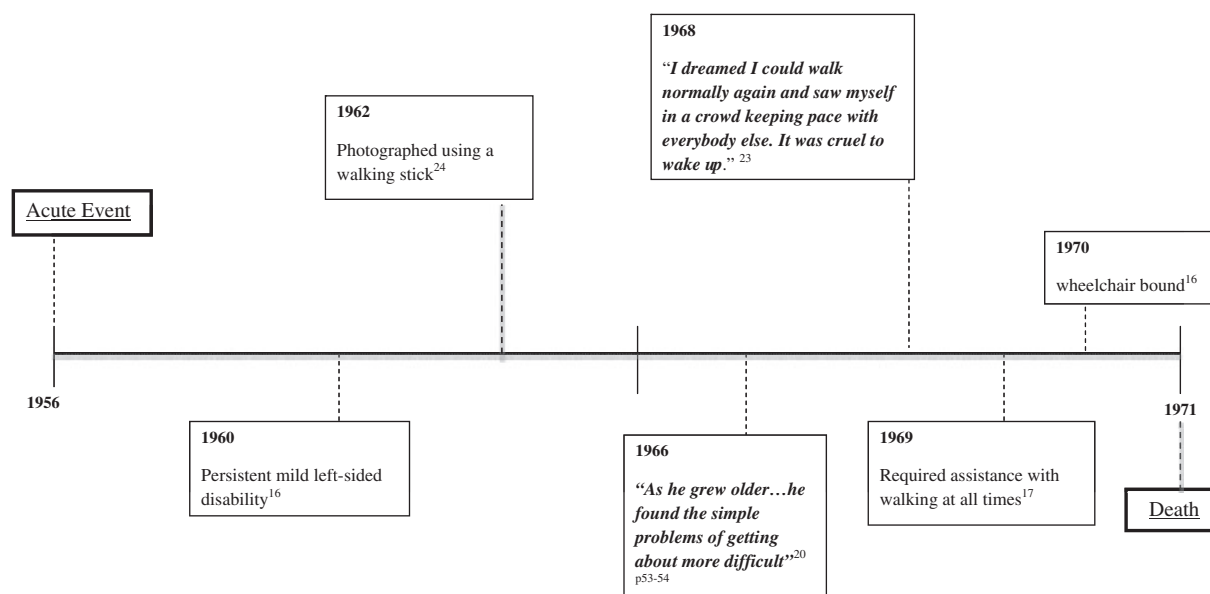


Figure 1. Evidence of gait abnormality 1956–71.

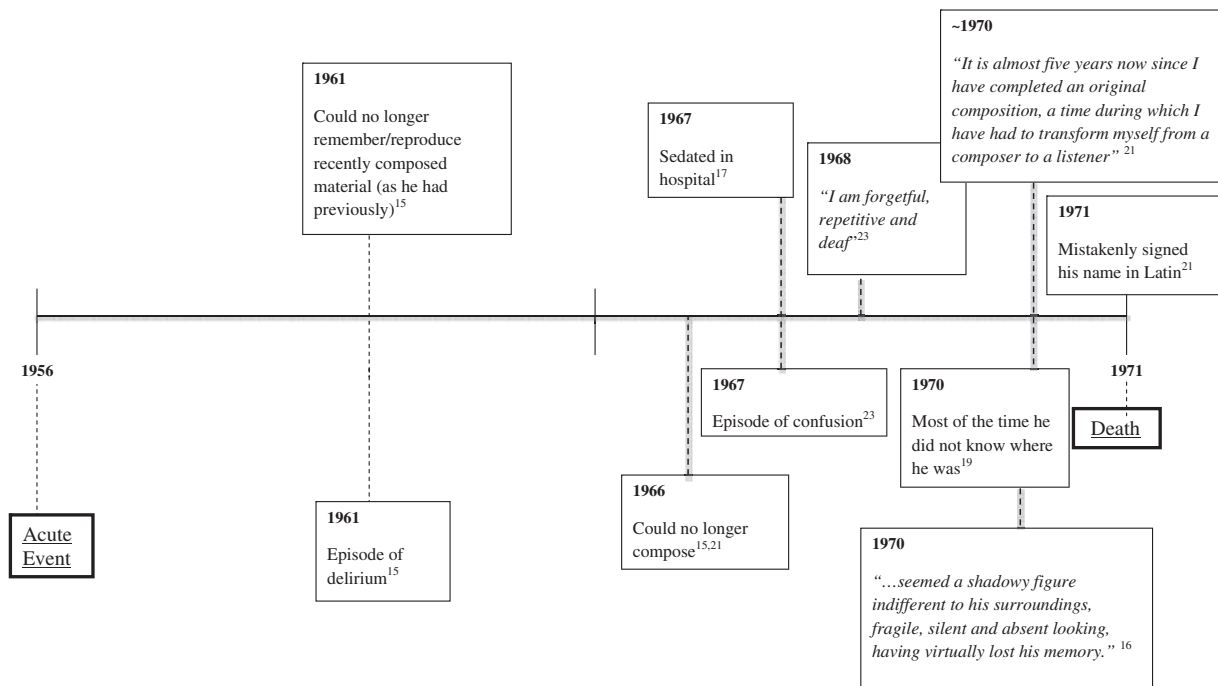


Figure 2. Evidence of cognitive impairment 1956–71.

gait disorder.²⁶ Stravinsky was deeply upset by these changes, and had recurrent dreams in which his gait was unaffected from which he recorded that it was cruel to wake up. By 1969, Horgan was reporting that the composer was bed- and wheelchair-bound.

Cognitive impairment

We now know that gait disorders may be the harbinger of cognitive decline in later life,²⁷ and this seemed also to be the case with Stravinsky (Figure 2). The first indication of cognitive difficulties arose when his wife reported that he was forgetful in 1965, with a loosening of his characteristic economy in time, talk and money—except for matters musical where his specificity was even greater. The next episode identified was a protracted episode of delirium during a hospitalization for pneumonia in November 1967. Following his return to home he now complained of being 'forgetful, repetitive and deaf', in January 1968 could remember where he was married. He was noted by Libman to be forgetful and confused. However, it is notable that he reported no longer being able to compose in his last 5 years, apart from touching arrangements of two songs by Hugo Wolff and four Bach preludes and fugues. There were both a lack of popular awareness and higher levels of tolerance for early dementia in these decades.

Oropharyngeal dysphagia

Stravinsky was treated more than once for pneumonia in the late 1960s and considering a number of accompanying anecdotes describing the composer coughing when drinking fluids (Figure 3). For example, Craft in July 1969 noted that Stravinsky coughed only when swallowing liquids and not solid food, with negative investigations. He records that the doctors could not find a cause for the coughing fits and violent rises in temperature. With the hindsight, these would now be considered to have a high probability of relating to oropharyngeal dysphagia, and further episodes of pneumonia followed with increasing regularity.

Discussion

The timelines of these deficits over a decade highlight the chronic trajectory of the subtle manifestations of chronic stroke disease. Given the neglect of these symptoms in public discourse and current medical practice, outside of geriatric medicine, it is hardly surprising that contemporary biographical accounts did not dwell on further symptoms of the syndrome. However, the consistency across multiple narratives is striking, and although a wide range of other neurological diseases may have caused any one element of the syndrome, the presence and progression of all three against

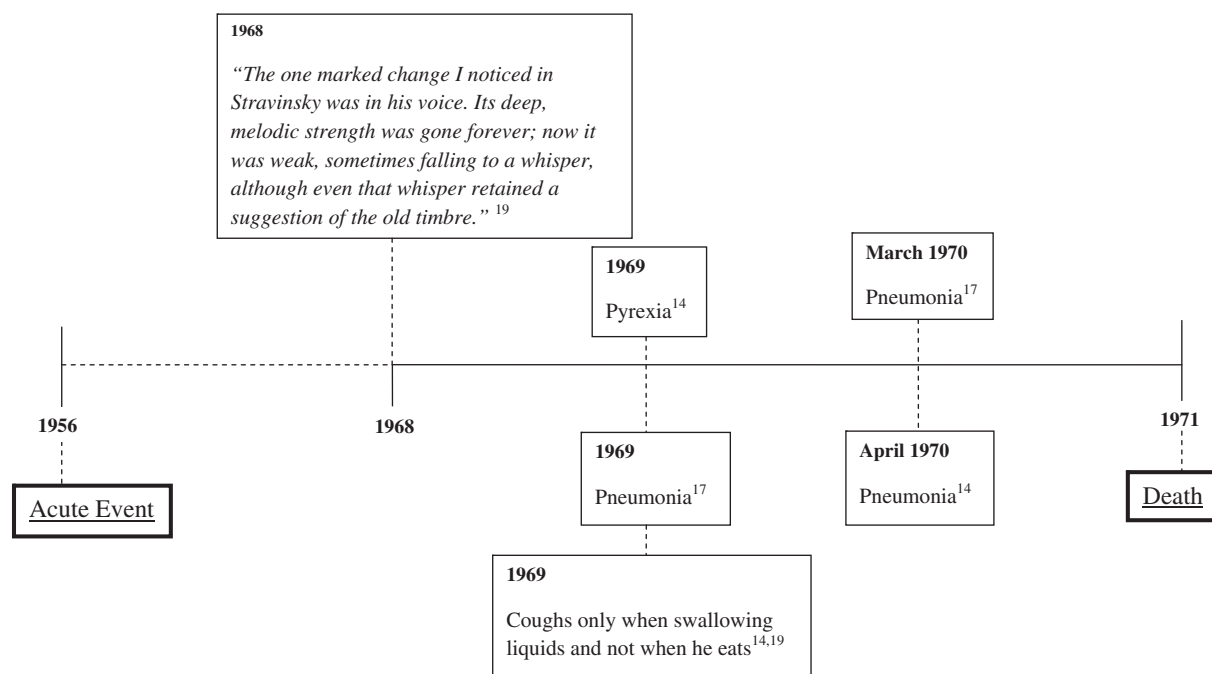


Figure 3. Evidence of dysphagia 1956–71.

a background of stroke disease makes the diagnosis most likely one related to chronic stroke disease, due to a combination of overt and occult stroke, as well as cerebral small vessel disease.²⁸

The prevalence of the syndrome is as yet unclear, but is likely to be substantial given that the prevalence of oropharyngeal dysphagia is 10–15% of older populations,²⁹ and the prevalence of combined gait and cognitive disorders is also high.²⁷

Given that awareness and detection of each of the components of the syndrome remains low,^{30–32} it might be hoped that their identification with one of the iconic figures of the 20th century might prompt a greater awareness among doctors dealing with older people, and a focal point for diagnosis and management. As well as ensuring adequate secondary prevention of vascular disease, strategies exist for managing each aspect, including physiotherapy for the higher level gait disorder,³³ speech and language therapy for oropharyngeal dysphagia,³⁴ and counseling and advance care planning may be of utility in vascular cognitive impairment.

In terms of the general public, the association of Stravinsky with chronic stroke disease could have a very positive impact as well. Given the often negative discourse on stroke and its sequelae, the example of Stravinsky can help to dispel stigma by illuminating his preservation of enjoyment of life and participation in society despite the associated

disabilities. Up to the end, it is clear that Stravinsky enjoyed his beloved music and retained a sharp critical edge in his commentaries. His entourage and supports facilitated a wonderful compensatory life-style: he travelled extensively until his last year despite his growing infirmities and ailments.

Living well is the best revenge, and although all those so affected may not have his formidable intellectual and creative reserves, financial resources and supportive milieu, they can all aspire to sharing his clear desire to live as full a life as possible. For the medical profession, a sharper focus on recognition, assessment and treatment of chronic stroke disease is the least that we owe to the remarkable resilience of this giant of the 20th century, and the very many of our patients with Stravinsky syndrome whose will for fighting back are equally resolute.

Conflict of interest: None declared.

References

1. Whitworth JA. Should eponyms be abandoned? No. *BMJ* 2007; **335**:425.
2. Woywodt A, Matteson E. Should eponyms be abandoned? Yes. *BMJ* 2007; **335**:424.
3. Panush RS, Wallace DJ, Dorff RE, Engleman EP. Retraction of the suggestion to use the term "Reiter's syndrome" sixty-five

- years later: the legacy of Reiter, a war criminal, should not be eponymic honor but rather condemnation. *Arthritis Rheum* 2007; **56**:693–4.
4. Ross IB. The role of Claude Bernard and others in the discovery of Horner's syndrome. *J Am Coll Surg* 2004; **199**:976–80.
 5. Walling AD. Amyotrophic lateral sclerosis: Lou Gehrig's disease. *Am Fam Physician* 1999; **59**:1489–96.
 6. Lerner BH. Complicated lessons: Lorenzo Odone and medical miracles. *Lancet* 2009; **373**:888–9.
 7. Pendlebury ST, Rothwell PM, Algra A, Ariesen MJ, Bakac G, Czlonkowska A, et al. Underfunding of stroke research: a Europe-wide problem. *Stroke* 2004; **35**:2368–71.
 8. O'Neill D, Horgan F, Hickey A, McGee H. Long term outcome of stroke: stroke is a chronic disease with acute events. *BMJ* 2008; **336**:461.
 9. Rockwood K, Howard K, MacKnight C, Darvesh S. Spectrum of disease in vascular cognitive impairment. *Neuroepidemiology* 1999; **18**:248–54.
 10. Martin MP, O'Neill D. Vascular higher-level gait disorders—a step in the right direction? *Lancet* 2004; **363**:8.
 11. Dalby RB, Chakravarty MM, Ahdidan J, Sorensen L, Frandsen J, Jonsdottir KY, et al. Localization of white-matter lesions and effect of vascular risk factors in late-onset major depression. *Psychol Med* 2010; **40**:1389–99.
 12. Buchholz DW. Neurogenic dysphagia: what is the cause when the cause is not obvious? *Dysphagia* 1994; **9**:245–55.
 13. Strandberg TE, Pitkala KH, Tilvis RS, O'Neill D, Erkinjuntti TJ. Geriatric syndromes—vascular disorders? *Ann Med* 2013; **45**:265–73.
 14. Craft R. *Stravinsky: Chronicle of a Friendship, 1948–1971*. 1st edn. New York: A. A. Knopf, 1972.
 15. Stravinsky I, Craft R. *Expositions and Developments*. 1st edn. Garden City, NY: Doubleday, 1962.
 16. Boucourechliev A. *Stravinsky*. New York: Holmes & Meier, 1987.
 17. Craft R. *Stravinsky: Glimpses of a Life*, 1st U.S. edn. New York: St. Martin's Press, 1993.
 18. Horgan P. *Encounters with Stravinsky: A Personal Record*. New York: Farrar Straus and Giroux, 1972.
 19. Libman L. *And Music at the Close: Stravinsky's Last Years, A Personal Memoir*. 1st edn. New York: W. W. Norton, 1972.
 20. McLeish K, McLeish V. *Stravinsky*, vol 83. London: Heinemann, 1978.
 21. Oliver M. *Igor Stravinsky*. London: Phaidon, 2008.
 22. Stravinsky I. *Themes and Conclusions*. London: Faber, 1972.
 23. Stravinsky I, Craft R. *Dialogues and a Diary*. London: Faber, 1968.
 24. Vernon R. *Introducing Stravinsky*. Parsippany, NJ: Silver Burdett Press, 1997.
 25. Walsh S. *Stravinsky: The Second Exile, France and America, 1934–1971*, 1st edn. New York: Alfred A. Knopf, 2006.
 26. *Stravinsky at 85* [DVD]. Kroiter R. Pleasantville, NY: VAI Music, 2009.
 27. Verghese J, Wang C, Lipton RB, Holtzer R, Xue X. Quantitative gait dysfunction and risk of cognitive decline and dementia. *J Neurol Neurosurg Psychiatry* 2007; **78**:929–35.
 28. Pantoni L. Cerebral small vessel disease: from pathogenesis and clinical characteristics to therapeutic challenges. *Lancet Neurol* 2010; **9**:689–701.
 29. Kawashima K, Motohashi Y, Fujishima I. Prevalence of dysphagia among community-dwelling elderly individuals as estimated using a questionnaire for dysphagia screening. *Dysphagia* 2004; **19**:266–71.
 30. Rofes L, Arreola V, Almirall J, Cabre M, Campins L, Garcia-Peris P, et al. Diagnosis and management of oropharyngeal dysphagia and its nutritional and respiratory complications in the elderly. *Gastroenterol Res Pract* 2011; **2011**:1–13.
 31. Griffin E, Martin M, Fitzgerald T, O'Neill D. Gait abnormalities in older people—comparison of inter-professional assessment. *Ir Med J* 2005; **98**:169–70.
 32. Torisson G, Minthon L, Stavenow L, Londo E. Cognitive impairment is undetected in medical inpatients: a study of mortality and recognition amongst healthcare professionals. *BMC Geriatr* 2012; **12**:47.
 33. Mickelborough J, Liston R, Harris B, Pomeroy VM, Tallis RC. Physiotherapy for higher-level gait disorders associated with cerebral multi-infarcts. *Physiother Pract* 1997; **13**:127–38.
 34. Speyer R, Baijens L, Heijnen M, Zwijnenberg I. Effects of therapy in oropharyngeal dysphagia by speech and language therapists: a systematic review. *Dysphagia* 2010; **25**:40–65.