

Institution: University of Sheffield		
Unit of Assessment: A-03 Allied Health Professions, Dentistry, Nursing and Pharmacy		
Title of case study: Improving nutritional management for patients with amyotrophic lateral sclerosis/motor neuron disease		
Period when the underpinning research was undertaken: 2011–2015		
Details of staff conducting the underpinning research from the submitting unit:		
Name(s):	Role(s) (e.g. job title):	Period(s) employed by submitting HEI:
Christopher J. McDermott	Professor of Neurology	2000–present
Dame Pamela J. Shaw	Professor of Neurology	2000–present
Haris Stavroulakis	Lecturer in Clinical Neuroscience, Co-investigator	2010–present
Period when the claimed impact occurred: August 2013 onwards		
Is this case study continued from a case study submitted in 2014? N		
<p>1. Summary of the impact (indicative maximum 100 words)</p> <p>Weight management is critical for survival in amyotrophic lateral sclerosis/motor neuron disease (ALS/MND). Gastrostomy feeding directly into the stomach is recommended to support patients with ALS/MND that develop severe dysphagia. Notwithstanding, there was a lack of evidence of optimal timing and method for gastrostomy insertion. Furthermore, the lack of authoritative lay information meant that patients tended to put off having the procedure until it was too late despite the opposite advice by health professionals. The ProGas study led by Sheffield researchers determined that gastrostomy should be done at 5% weight loss rather than 10% as was in the guidelines at the time, and that radiologically inserted gastrostomy (RIG) tubes should not be used. This has led to updated national and international clinical guidelines on nutritional support and the increased use of gastrostomy in ALS/MND disease patients. This research has also informed the creation of a unique online resource (MyTube, https://mytube.mymnd.org.uk/) for patients, carers, health professionals and the general public to inform and assist decision making for patients. As a direct result the lives of patients and carers were improved.</p>		
<p>2. Underpinning research (indicative maximum 500 words)</p> <p>Amyotrophic lateral sclerosis (ALS), known in the UK as motor neuron disease (MND) is a progressive degenerative fatal neuromuscular disease that is caused by degeneration of motor neurones. ALS/MND sufferers typically die within 2-3 years from diagnosis due to respiratory arrest. As the disease is incurable, the efforts to support patients are heavily focused on sustaining a maximum quality of life (QoL). Worldwide an estimated 450,000 patients are living with ALS/MND at any given time. Dysphagia (swallowing difficulties) is a common problem in patients with ALS/MND. Patients with severe swallowing difficulty experience malnutrition, dehydration, choking and an increased risk of chest infections. Traditional use of nasogastric tubes for feeding is problematic for ALS/MND patients and is not compatible with using non-invasive ventilation (NIV), the current standard therapy to manage respiratory insufficiency.</p> <p>Long-term nutritional support of patients with severe swallowing difficulty can be achieved by placing a feeding tube, known as a gastrostomy, directly into the stomach; both the American</p>		

Academy of Neurology and the European Federation of Neurological Societies recommend gastrostomy feeding in ALS/MND patients who develop severe dysphagia.

A meta-analysis and survey of the practice of gastrostomy use in ALS/MND conducted by the Sheffield team, in which they reviewed the three main methods of gastrostomy insertion (percutaneous endoscopic gastrostomy (PEG); radiologically inserted gastrostomy (RIG); per-oral image-guided gastrostomy (PIG)) and conducted a meta-analysis of mortality data following gastrostomy, revealed a lack of high-quality evidence to indicate the optimal method and timing for gastrostomy insertion in patients with ALS/MND and indicated that RIG and PIG methods may be safer than PEG [R1].

A retrospective, qualitative exploration using semi-structured interviews was conducted with 27 paired patients and carers, out of which 23 underwent gastrostomy, in order to elicit their perceptions in relation to gastrostomy timing. It concluded that the advice for early insertion does not outweigh the personal perceptions and psychosocial factors for patients and their carers. It also found that understanding the factors that influence decision-making on an individual basis is important for information and care provision by healthcare professionals in aiding patients and their carers, to make informed decisions in relation to gastrostomy timing [R2, R3].

Following on from these studies, the Sheffield team initiated a world-first large, longitudinal, prospective cohort study (ProGas) that aimed to compare gastrostomy insertion approaches in terms of safety and clinical outcomes in ALS/MND. The ProGas study team worked with the Sheffield Motor Neurone Disorders Research Advisory Group to enable the public, patients and carers the opportunity to provide feedback on all aspects of ProGas and to contribute to shaping the final ProGas protocol. The membership of this group includes patients and carers who have experience of motor system disorders including ALS/MND. Between Nov 2, 2010, and Jan 31, 2014, 345 patients were recruited in 24 participating UK ALS/MND care centres and clinics of whom 330 had gastrostomy. 163 (49%) patients underwent PEG, 121 (37%) underwent RIG, 43 (13%) underwent PIG, and three (1%) underwent surgical gastrostomy. The study showed that while the three methods of gastrostomy seemed to be as safe as each other in relation to survival and procedural complications, the hazard of death after gastrostomy insertion was significantly affected by the percentage of weight loss from diagnosis to gastrostomy, and patients who received RIG had a significantly higher rate of tube-related complications, leakage and replacement. Accordingly, the ProGas Study recommended that gastrostomy should be done at 5% weight loss rather than 10% as was in the guidelines at the time, and that RIG tubes should not be used [R4].

3. References to the research (indicative maximum of six references)

- R1. Stavroulakis, T., Walsh, T., Shaw, P. J., & McDermott, C. J.** and the Progas Study (2013). Gastrostomy use in motor neurone disease (MND): A review, meta-analysis and survey of current practice. *Amyotrophic Lateral Sclerosis and Frontotemporal Degeneration*, 14(2), 96–104. <https://doi.org/10.3109/17482968.2012.723722>
- R2. Stavroulakis, T., Baird, W. O., Baxter, S. K., Walsh, T., Shaw, P. J., & McDermott, C. J.** (2014). Factors influencing decision-making in relation to timing of gastrostomy insertion in patients with motor neurone disease. *BMJ Supportive & Palliative Care*, 4(1), 57–63. <https://doi.org/10.1136/bmjspcare-2013-000497>
- R3. Stavroulakis, T., Baird, W. O., Baxter, S. K., Walsh, T., Shaw, P. J., & McDermott, C. J.** (2016). The impact of gastrostomy in motor neurone disease: challenges and benefits

from a patient and carer perspective. *BMJ Supportive & Palliative Care*, 6(1), 52–59.
<https://doi.org/10.1136/bmjspcare-2013-000609>

- R4. McDermott, C. J., Shaw, P. J., Stavroulakis, T., et al.,** on behalf of the ProGas Study Group (2015). Gastrostomy in patients with amyotrophic lateral sclerosis (ProGas): a prospective cohort study. *The Lancet Neurology*, 14, 702-709.
[https://doi.org/10.1016/S1474-4422\(15\)00104-0](https://doi.org/10.1016/S1474-4422(15)00104-0)

4. Details of the impact (indicative maximum 750 words)

Impacts include: health and welfare, public policy and services, practitioners and services, commercial.

Main beneficiaries include: patients, carers, practitioners, NHS, NICE, general public, industry.

Gastrostomy feeding directly into the stomach is recommended to support patients with ALS/MND that develop severe dysphagia. These patients struggle to maintain weight, and without a tube they essentially cannot take in enough calories to survive. Prior to the ProGas study the practice of gastrostomy feeding was largely based on consensus and expert opinion rather than robust evidence. Furthermore, the lack of accessible information meant that patients tended to put off having the procedure until it was too late despite the opposite advice by health professionals. The ProGas study was stimulated by the research priorities identified by both the Dementias and Neurodegenerative Diseases Research Network (DeNDRoN) Clinical Studies Group for MND and the MND Association. ProGas has provided scientific underpinning for the practice of gastrostomy in ALS/MND and provided an award-winning online information resource which have directly improved the lives of patients and carers.

Impact on UK and international guidelines

The ProGas Study has led to updated national and international guidelines on nutrition support and the use of gastrostomy in patients with ALS/MND.

In the UK, the British Medical Journal used the results from the ProGas study as evidence that gastrostomy before substantial weight loss might be beneficial as part of ongoing treatment options, in its BMJ Best Practice on ALS/MND publication [S1]. The Motor Neurone Disease Association, the only national ALS/MND charity in the UK and founding member of the international Alliance of ALS/MND associations, which has 40 members, included the recommendations of the ProGas study in its Guide for GPs and primary care teams [S2].

The results have been reflected in current National Institute for Health and Care Excellence (NICE) guidelines for MND, which state that the role of gastrostomy feeding should be discussed at an early stage but that it must be recognised that not all individuals will wish to proceed [S3]. Internationally, the European Society for Clinical Nutrition and Metabolism (ESPEN) has used the recommendations from the ProGas study in its guidelines on clinical nutrition in *Neurology* [S4].

Impact by public participation, involvement, and engagement: direct impact on the lives of MND/ALS patients

As a result of the ProGas study findings the MyTube website (<https://mytube.mymnd.org.uk/>) was created in collaboration with the NHS and the Sheffield MND Care Centre and with support from the MND Association. Built for and by patients, the site offers insights into living with a tube

or alternatives available through the use of short videos and information about tube feeding for people living with ALS/MND with the clearly stated objective to use patient stories to encourage change; myTube was launched in March 2017 [S5]. The myTube website has become the go to information resource for patients and carers: it has become the recommended information source by patient organisations such as the Motor Neurone Disease Association, the Parenteral and Enteral Nutrition Group (PENG) and the British Association of Parenteral and Enteral Nutrition (BAPEN) [S6]. Its influence and impact have further been recognised by several awards, including two British Medical Association (BMA) Patient Information Awards [S7a] describing it as “*a perfect medium for instructional information*” and the Nutrition Resource of the Year Award from Complete Nutrition Magazine, the UK’s number one clinical, medical and health nutrition publication [S7b].

The myTube resource has helped patients and carers worldwide to make informed decisions; one patient commented “*I have just been diagnosed with MND and have been asked to start considering getting a PEG. This is fantastic information and very timely. I've already watched the first two 'videos' and they're great. Thanks to all participants...you're already helping someone in Australia.*” While a carer said “*I'm sure this will help put people's minds at ease regarding PEGs and other things. Wish [husband's name] could have seen this early on. He might not have resisted having his fitted till it was desperate*” [S8].

Feedback was also received from clinics around the UK that the website is used regularly in their clinics to support patients [S8]. The use of myTube, in patient decision making, has been suggested as a reason for the increase in patients in Scotland undergoing gastrostomy ‘*More people could be undergoing gastrostomy because of the increasing amounts of readily accessible information via social media and the internet for example <https://mytube.mymnd.org.uk/>.*’ [S9].

Commercial impact

The myTube resource was designed by Ammba Digital architects (<http://ammba.co.uk/>), a consultancy and digital product development company, in collaboration with Optical Jukebox (the film makers for myTube), the Sheffield team and ALS/MND patients and carers. Ammba’s involvement in the project team that delivered myTube has led to ongoing work for the company in the digital healthcare space. The myTube project allowed the company to take their specialism and skills from the education learning sector into the health sector. Ongoing collaboration with Optical Jukebox led to securing a £50K funding award from the Ufi VocTech programme to develop a digital learning resource to support carers working with those living with a neurological condition; a first for this sector [S10].

5. Sources to corroborate the impact (indicative maximum of 10 references)

- S1. BMJ best practice amyotrophic lateral sclerosis (<https://bestpractice.bmj.com/topics/en-us/330>) pp.22, 26.
- S2. Motor neurone disease: a guide for GPs and primary care teams (<https://www.mndassociation.org/app/uploads/px016-motor-neurone-disease-a-guide-for-gps-and-primary-care-teams.pdf>) pp.32, 40, reference 44 on p.76
- S3. National Institute for Health and Care Excellence (NICE). Motor neurone disease: assessment and management 2016

(<https://www.nice.org.uk/guidance/ng42/chapter/recommendations#nutrition-and-gastrostomy>) 1.10.6, 1.10.7, 1.10.8, p.243 of NG42

- S4.** ESPEN Guideline. Burgos, R., Bretón, I., Cereda, E., Desport, J. C., Dziewas, R., Genton, L., Gomes, F., Jesús, P., Leischker, A., Muscaritoli, M., Poulia, K.-A., Preiser, J. C., Van der Marck, M., Wirth, R., Singer, P., & Bischoff, S. C. (2018). ESPEN guideline clinical nutrition in neurology. *Clinical Nutrition*, 37(1), 354–396.
<https://doi.org/10.1016/j.clnu.2017.09.003>. Clinical Question 12 and 13, Recommendation 17, 18,19.
- S5.** Web link to the myTube website (<http://mytube.mymnd.org.uk/>) combined with media coverage following the launch: patient with MND shares his story, with a view to helping others, on myTube on ITV News, 31-Mar-2017 (https://www.youtube.com/watch?v=OJu6FSf0zQg&ab_channel=JasonLiversidge); Living with the cruelty of motor neurone disease: Brave Yorkshire families share their stories. The Yorkshire Post, 3 April, 2017 (<https://www.yorkshirepost.co.uk/news/living-cruelty-motor-neurone-disease-brave-yorkshire-families-share-their-stories-55332>).
- S6.** MyTube as a recommended information source by patient organisations: MNDA Resources: Living with MND: swallowing, eating and drinking (<https://www.mndassociation.org/support-and-information/living-with-mnd/eating-and-drinking/>) and Dysphagia information sheet for health professionals (p.1, <https://z9t2c4x9.stackpathcdn.com/app/uploads/information-sheet-p8-dysphagia-in-motor-neurone-disease.pdf>); PENG Resources for Patients and HCPs (<https://www.peng.org.uk/publications-resources/resources-for-patients-hcps.php>); BAPEN e-Learning Portal (<https://www.bapen.org.uk/71-resources-and-education/education-research-and-science/e-learning-portal>).
- S7.** Combined 2017 award information: BMA Patient Information Award and Nutrition Magazine Award. PDF available.
- S8.** MyTube website metrics, feedback – report to MND 2017 by Optical Jukebox. Written by Registered Nurse filmmaker and videographer, Optical Jukebox.
- R5.** Gorrie, G. H., Chandran, S., Colville, S., Newton, J., Leighton, D., McDonald, M., Pal, S., Forbes, R., Hair, M., & Swingler, R. (2019). Improved survival and 30-day mortality after gastrostomy in Scottish motor neurone disease patients: evidence from a national retrospective cohort study using STROBE criteria. *Amyotrophic Lateral Sclerosis and Frontotemporal Degeneration*, 20(3–4), 165–171.
<https://doi.org/10.1080/21678421.2019.1570271>
- S9.** AMMBA Collaboration information and correspondence (<http://amma.co.uk/neurocare-knowhow-awarded-ufi-voctech-seed-2020-funding/>).