Article:
Essat, M., Coates, E., Clowes, M. et al. (2022) Understanding the current nutritional management for people with amyotrophic lateral sclerosis - a mapping review. Clinical Nutrition ESPEN, 49. pp. 328-340. ISSN: 2405-4577

https://doi.org/10.1016/j.clnesp.2022.03.026

Understanding the current nutritional management for people with amyotrophic lateral sclerosis - A mapping review

Authors: Munira Essat[a], Elizabeth Coates[a], Mark Clowes[a], Daniel Beever[a], Gemma Hackney[a], Sean White[b], Theocharis Stavroulakis[c], Vanessa Halliday[a*], Christopher McDermott[c] on behalf of the HighCALS group

Affiliations:

[a]School of Health and Related Research (ScHARR), The University of Sheffield, Sheffield, UK.

[b]Sheffield Teaching Hospitals NHS Foundation Trust, Sheffield, UK.

[c]Sheffield Institute for Translational Neuroscience (SITraN), The University of Sheffield, Sheffield, UK.

*Corresponding author: Vanessa Halliday, Senior Lecturer, Section of Public Health, School of Health and Related Research (ScHARR), The University of Sheffield, Regent Court, 30 Regent Street, Sheffield, S1 4DA, UK

email: vanessa.halliday@sheffield.ac.uk

Tel: +44 (0) 114 222 4268

Fax: +44 (0) 114 222 0749

Sheffield, UK.

Abbreviations: ALS, Amyotrophic lateral sclerosis; BMJ BP, British Medical Journal Best Practice; ENCALS, European Network to Cure Amyotrophic Lateral Sclerosis; EPPI-Centre, Evidence for Policy and Practice Information and Co-ordinating Centre; FEESST, Flexible Endoscopic Evaluation of Swallowing with Sensory Testing; FVC, Forced vital capacity; MDT, Multidisciplinary team; MND, Motor Neuron Disease; MNDA, Motor Neuron Disease Association ; NCG, NICE Clinical Guideline; NGT, Nasogastric tube feeding; NICE, National Institute for Health and Care Excellence; NIV, Non-invasive ventilation; NuPEG, Nasal unsedated seated percutaneous endoscopic gastrostomy; PEG, Percutaneous endoscopic gastrostomy; PIG, Per-oral image-guided gastrostomy; pwALS, people with Amyotrophic Lateral Sclerosis; PENG, Parenteral and Enteral Nutrition Group; RIG, Radiologically inserted gastrostomy; RCN, Royal College of Nursing ; SLT, Speech and language therapist;

Abstract

Background & Aims

Poor nutritional outcomes are observed in people with Amyotrophic Lateral Sclerosis (pwALS) including weight loss and poor dietary intake. Surveys of healthcare professionals have highlighted the lack of evidence and knowledge regarding nutritional management of ALS throughout the disease course. Furthermore, national evidence-based guidance is lacking. This mapping review aims to understand the structure and input of nutritional management services for pwALS.

Methods

Systematic searches were conducted across eight electronic databases to identify qualitative and quantitative research on structure and input of nutritional care in ALS. Supplementary searches included grey literature, citation and reference list searching of included studies and key reviews, web searching and contacting experts and organisations that provide ALS services to identify guidelines. Study selection and data extraction were undertaken independently by at least two reviewers. Data was synthesised using a narrative approach.

Results

One hundred and nine documents were identified. These consisted of journal articles, guidelines and related documents that contributed evidence towards mapping of nutritional management of pwALS. No evidence on commissioning of nutritional care was identified. Guidelines provided high-level overviews and gave general guidance or recommendations for care; however, these typically focused on gastrostomy with limited guidance on broader aspects, including oral nutrition support. Evidence from primary studies found nutritional care delivery in ALS consisted of multiple types of nutritional management, at different time points during the disease course and involving a range of professionals. There was little evidence relating to proactive nutritional care. Details of healthcare setting, number of professionals involved in care, team composition and how services were delivered in community settings were sparse. Although the role of the speech and language therapist in swallowing assessment and provision of advice on the management of swallowing difficulties was consistent; there was limited evidence on care provided by dietitians. In addition, a small number of studies reported on the use of

screening tools. Overall, evidence was consistent that weight management, including monitoring of weight change by professionals and patients, was central and recommended that this should be part of nutritional assessment and follow-up.

Conclusions

The evidence identified in this mapping review has highlighted the requirement for further primary research providing specific details on how nutritional management of pwALS is structured and delivered.

Keywords

Nutrition, mapping review, amyotrophic lateral sclerosis, diet, management

Introduction

Amyotrophic lateral sclerosis (ALS), also known as motor neuron disease (MND), is a devastating neurodegenerative disorder with usual survival of around 3 years[1 2] and no curative treatment.[3] Affected individuals experience a progressive weakness of limb muscles, loss of speech, chewing and swallowing, eventually succumbing to the consequences of respiratory failure due to respiratory muscle weakness.[4]

Malnutrition and weight loss are well recognised poor prognostic factors in ALS.[5-7] Current National Institute for Health and Care Excellence (NICE), European and American ALS nutritional guidelines are not evidence-based and only focus on ameliorating dysphagia and the timing and method of gastrostomy insertion.[8-12] Surveys of healthcare professions have highlighted the lack of evidence and knowledge regarding the nutritional management of ALS throughout the disease course.[13-15] The poor evidence base explains the poor nutritional outcomes observed including weight loss and low consumption of high calorie supplements in those losing weight.[16] The recent NICE guidance on ALS management identified a lack of evidence on the nutritional management of ALS patients and made a priority research recommendation in this area.[17]

The aim of this mapping review is to understand how the nutritional needs of pwALS are currently managed by identifying which healthcare professionals are involved in delivering this care and how nutritional management services are structured.

Methods

A systematic mapping review was conducted to find quantitative, qualitative and mixed method published literature on structure and input of nutritional care in ALS and to identify tendencies and gaps.

Data sources and searches

Systematic searches were conducted in eight electronic databases including Medline, EMBASE, CINAHL, the Cochrane Library and Web of Science. A combination of subject headings and free text

terms structured around the condition (amyotrophic lateral sclerosis (including synonyms such as ALS, motor neurone disease and related conditions) and nutritional care process were used. The broadest possible terms were incorporated with a view to including studies about identification of nutritional need, nutritional assessment and monitoring, along with any form of nutrition-related support, advice or intervention. Databases were searched from inception to April 2018. Additional, supplementary searches included grey literature, citation and reference list searching of included studies and key reviews, web searching and contacting experts and organisations that provide ALS services to identify guidelines. An example Medline search strategy is provided in Supplementary Appendix 2.

Study selection

All titles were examined for inclusion by one reviewer and any citations that clearly did not meet the inclusion criteria were excluded. All abstracts and full text articles were then examined independently by two reviewers. Any disagreements in the selection process were resolved by discussion, involving a third reviewer when necessary. In order to maintain relevance to current service delivery, only literature published from January 2008 to April 2018 were included. A summary of the eligibility criteria is available in Appendix 3.

Data extraction

A data extraction template was developed using Evidence for Policy and Practice Information and Co-ordinating Centre (EPPI-Centre) core key wording strategy version 0.9.4[18] and a coding guide developed and applied to extracted data as recommended.[19 20] Data relating to study design, patient characteristics and outcomes were extracted by one reviewer, and independently checked for accuracy by a second reviewer. Any discrepancies were resolved by discussion, with involvement of a third reviewer, if required.

Data synthesis

The presentation of data was informed by existing publications and guidance.[20-24] Evidence was characterised according to study design, country of origin, service approach and structure. Data items were summarised in narrative textual description and tabular format.

Results

A total of 3,122 records were screened from the searches of electronic databases and an additional 18 articles from other sources. Of these, 591 full text articles were considered potentially eligible for inclusion. Following detailed screening, 109 documents were included in the mapping review. The evidence consisted of 10 conference abstracts, 86 studies and 13 guidance documents. As this review was UK focussed, only guidelines and conference abstracts related to the UK were included. The majority of the excluded articles did not relate to nutritional management or were intervention feasibility studies. Figure 1 presents a summary of the process of identifying and selecting the relevant literature.

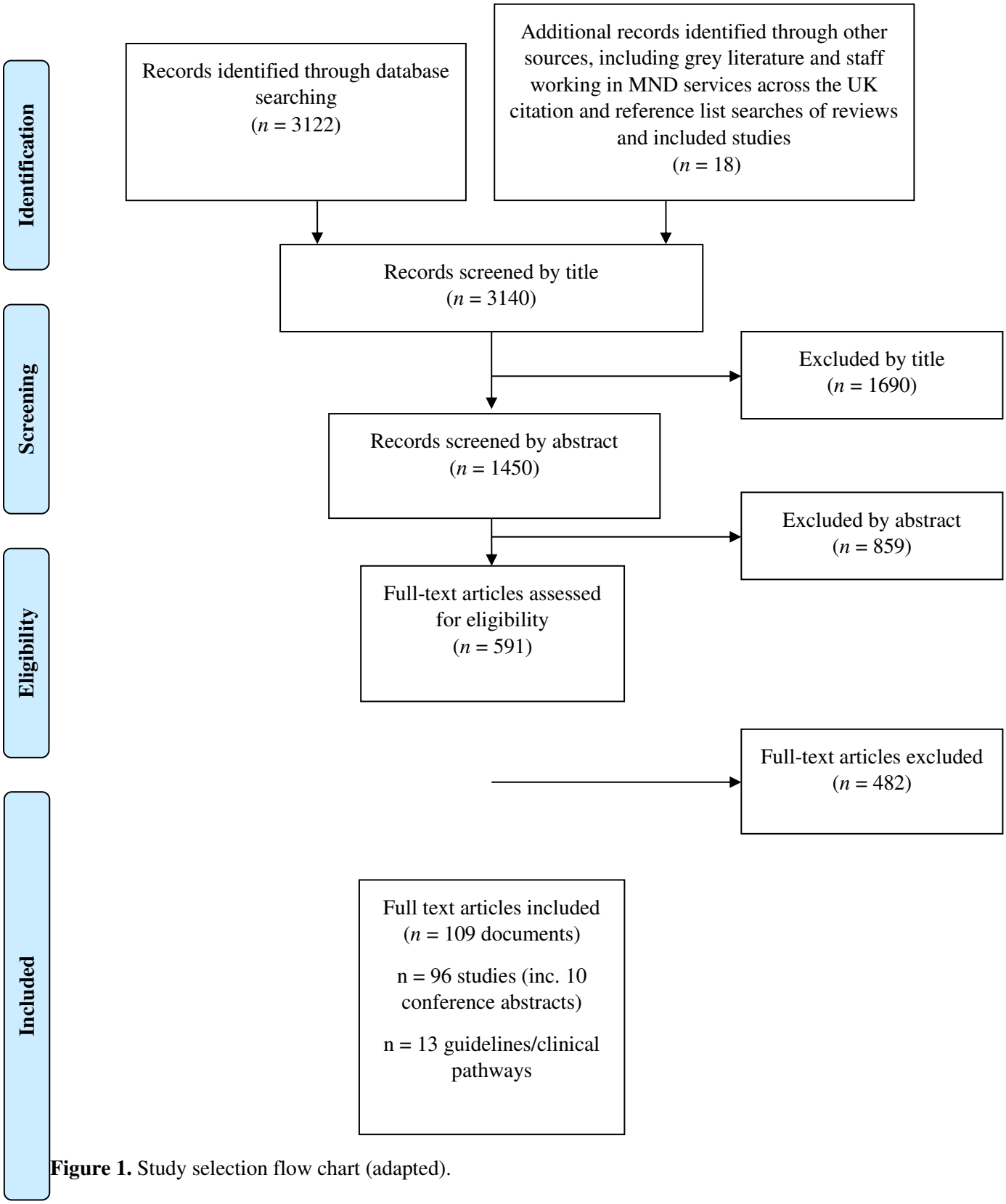

Figure 1. Study selection flow chart (adapted).

Characteristics of included studies

Characteristics of the included studies are presented in Table 2. In summary, articles were published between 2008 and 2018 with studies peaking in 2013, and UK guidelines and related documents peaking

in 2016 and 2017. The most frequently observed study types were reviews of case notes (n=39) followed by prospective cohort studies (n=21). Non-peer reviewed literature included guidelines/clinical pathways (n=7), patient information leaflets (n=4) and summaries of guidelines (n=2). The majority of evidence came from the UK (n= 33) and USA (n=18).

Author Year	**Country of Study**	**Type of Publication**	**Study Design/Data Type**	**Study Population (n)**
Abdelnour-Mallet 2011[25]	France	Journal	Review of case notes	73
Ahmed 2015[26]	USA	Journal	Review of case notes	300
Allen 2013[27]	USA	Journal	Review of case notes	108
Ammar 2010[28]	UK	Journal	Case series	104
Aridegbe 2013[29]	UK	Journal	Review of case notes	417
Astrow 2008[30]	USA	Journal	Before and after study	32
Atassi 2011[31]	USA	Journal	Clinical trial data	300
Bach 2009[32]	USA	Journal	Case series	44
Beggs 2010[33]	Canada	Journal	Review of case notes	69
Benstead 2016[34]	Canada	Journal	Survey	10 clinics
Boylan 2016[35]	USA	Journal	Cost study	1117
Chavada 2010[36]	UK	Journal	Review of case notes	35
Chhetri 2016[37]	UK	Journal	Review of case notes	340
Chio 2012[38]	Italy	Journal	Prospective cohort study	128
Clavelou 2013[39]	France	Journal	Prospective cohort	382

Author Year	Country of Study	Type of Publication	Study Design/Data Type	Study Population (n)
Cleary 2008[40]	Canada	Journal	Case report	1
Cocks 2013[41]	UK	Journal	Survey	68
Connolly 2015[42]	Republic of Ireland	Journal	Cost study	119
Czell 2013[43]	Switzerland	Journal	Retrospective data analysis	26
Datta 2016[44]	UK	Conference abstract	Review of case notes	22
de Bucourt 2012[45]	Germany	Journal	Case series	31
Dorst 2015[46]	Germany	Journal	Prospective cohort study	89
Evans 2010[47]	UK	Conference abstract	Prospective cohort study	85
Faria 2011[48]	Portugal	Journal	Review of case notes	17
Fasano 2017[49]	Italy	Journal	Prospective cohort study	193
Fini 2014[50]	Italy	Journal	Prospective cohort study	193
Fitton 2010[51]	UK	Conference abstract	Review of case notes	24
Foley 2014[52]	Republic of Ireland	Journal	Qualitative study	34
Foster 2009[53]	Ireland	Journal	Case series	41
Georgoulopoulou 2013[54]	Italy	Journal	Prospective cohort study	193
Graziani 2015[55]	Italy	Journal	Case report	1
Greenaway 2015[56]	UK	Journal	Qualitative study	21
Gundogan 2014[57]	Turkey	Journal	Review of case notes	13
Jackson-Tarlton 2016[58]	Canada	Journal	Review of case notes	635

Author Year	Country of Study	Type of Publication	Study Design/Data Type	Study Population (n)
Jesus 2012[59]	France	Journal	Prospective cohort study	40
Johnson 2012[60]	UK	Journal	Prospective cohort study	35
Kak 2017[61]	USA	Journal	Review of case notes	102
Kalava 2016[62]	USA	Journal	Case series	10
Kara 2016[63]	Turkey	Journal	Review of case notes	500
Kawa, 2012[64]	USA	Journal	Review of case notes	29
Kehyayan 2014[65]	Canada	Journal	Cross-sectional study	2092
Kirbis 2015[66]	Slovenia	Journal	Review of case notes	271
Kirstein 2018[67]	Germany	Journal	Review of case notes	43
Kurien 2017[68]	UK	Journal	Prospective cohort study	100
Lavernhe 2017[69]	France	Journal	Prospective cohort study	203
Lewis 2009[70]	UK	Journal	Case series	104
Limousin 2010[71]	France	Journal	Review of case notes	63
Lopez-Gomez 2018[72]	Spain	Journal	Prospective cohort study	43
Lowe 2012[73]	UK	Journal	Survey	684 patients, 45 radiologists
Marin 2016[74]	France	Journal	Cross-sectional study	376
Martin 2014[75]	UK	Journal	Prospective cohort study	78
Martin 2016[76]	UK	Journal	Qualitative study	19
Murphy 2008[77]	New Zealand	Journal	Review of case notes	244

Author Year	Country of Study	Type of Publication	Study Design/Data Type	Study Population (n)
Nunes 2016[78]	Portugal	Journal	Review of case notes	37
Oliver 2011[79] (1)	UK	Journal	Review of case notes	84
Oliver 2011[80] (2)	UK	Journal	Review of case notes	62
Onesti 2017[81]	Italy	Journal	Review of case notes	145
Pena 2012[82]	Portugal	Journal	Review of case notes	151
Phippen 2017[83]	UK	Journal	Case report	1
Plowman 2017[84]	USA	Journal	Survey	38
Pols 2016[85]	Netherlands	Journal	Qualitative study	43
Power 2013[86]	Ireland	Journal	Review of case notes	260
ProGas Study Group 2015[87]	UK	Journal	Prospective cohort study	330
Rafique 2013[88]	UK	Conference abstract	Review of case notes	33
Rio 2010[89]	UK	Journal	Review of case notes	159
Rooney 2015[90]	Republic of Ireland	Journal	Prospective cohort study	511
Roubeau 2015[91]	France	Journal	Prospective cohort study	117
Rubin 2012[92]	USA	Journal	Case report	1
Ruffell 2013[93]	UK	Journal	Survey	177
Russ 2015[94]	USA	Journal	Review of case notes	21
Sakel 2010[95]	UK	Conference abstract	Evaluation of clinical pathway	30
Sancho 2010[96]	Spain	Journal	Prospective cohort study	30

Author Year	Country of Study	Type of Publication	Study Design/Data Type	Study Population (n)
Sarkar 2017[97]	UK	Journal	Review of case notes	14
Selkirk 2017[98]	USA	Journal	Retrospective data analysis	68
Sharma 2010[99]	USA	Journal	Case series	54
Sharpley 2013[100]	UK	Conference abstract	Prospective cohort study	54
Smith 2014[101]	UK	Conference abstract	Prospective cohort study	26
Spataro 2011[102]	Italy	Journal	Review of case notes	150
Stasinos 2016[103]	UK	Conference abstract	Review of case notes	7
Stavroulakis 2014[104]	UK	Journal	Qualitative study	10
Stavroulakis 2013[105]	UK	Journal	Qualitative study	10
Stavroulakis 2013[106]	UK	Journal	Survey	NR
Strijbos 2017[107]	Netherlands	Journal	Review of case notes	45
Swetz 2017[108]	USA	Journal	Retrospective data analysis	1974
Sznajder 2016[109]	Poland	Journal	Retrospective data analysis	48
Thompson 2017[110]	UK	Journal	Prospective cohort study	107
Tomasello 2012[111]	Italy	Journal	Review of case notes	44
Tsou 2012[112]	USA	Journal	Review of case notes	5759
Van Der Steen 2009[113]	Netherlands	Journal	Cross-sectional study	122
Vanis 2012[114]	Bosnia and Herzegovina	Journal	Review of case notes	359
Verschueren 2009[115]	France	Journal	Prospective cohort study	35

Author Year	Country of Study	Type of Publication	Study Design/Data Type	Study Population (n)
Wight 2012[116]	UK	Conference abstract	Review of case notes	30
Wills 2009[117]	UK	Conference abstract	Review of case notes	53
Zamietra 2012[118]	USA	Journal	Review of case notes	15
Zhang 2012[119]	Australia	Journal	Review of case notes	86
Zhang 2013[15]	USA	Journal	Survey	148
UK Guidelines and related documents				
Andersen 2012[120]	Europe/UK	Journal/Guideline	Guideline	NA
Barber 2015[121]	UK	Journal/Guideline	Guideline/clinical pathway	NA
Bede 2011[122]	International/UK	Journal/Guideline	Guideline summary	NA
BMJ BP 2017[123]	UK	Guideline	Guideline	NA
Burgos 2018[124]	Europe/UK	Journal/Guideline	Guideline/recommendations	NA
MNDA- Swallowing difficulties 7A 2017[125]	UK	Patient information Leaflet	Advice	NA
MNDA - Living with MND 2015[126]	UK	Patient information Leaflet	Advice	NA
MNDA- Eating and drinking with MND 2017[127]	UK	Patient information Leaflet	Advice	NA
MNDA-Tube Feeding 7B 2015[128]	UK	Patient information Leaflet	Advice	NA
NCG 2016[129]	UK	Guideline	Guideline/clinical pathway	NA

Author Year	Country of Study	Type of Publication	Study Design/Data Type	Study Population (n)
Oliver 2017[130]	UK	Guideline/clinical pathway	Guideline summary	NA
PENG 2016[131]	UK	Guideline	Guideline	NA
RCN 2016[132]	UK	Guideline	Guideline	NA
BMJ BP, British Medical Journal Best Practice; MNDA, Motor Neurone Disease Association; MND, Motor Neurone Disease; PENG, Parenteral and Enteral Nutrition Group; RNC, Royal College of Nursing; NCG, NICE Clinical Guideline.				

Table 2. Study characteristics

Outcome and synthesis of results

Key findings relating to types of nutritional care from the most detailed and informative of included studies have been summarised descriptively and is presented separately by i) study data (journal articles and conference abstracts describing how nutritional care is delivered to pwALS/MND) and ii) local/national guidelines/clinical pathways and related documents (describing how nutritional care should be delivered to pwALS/MND).

Types of nutritional care delivered

Weight measurement and calculation of percentage weight loss

A total of 17 studies[25 29 32 33 58 59 69 71 72 74 82 84 89-91 102 119] described the use of weight measurement and/or calculation of percentage weight loss as part of the nutritional management of pwALS/MND. In most cases weight measurement was undertaken at a multidisciplinary ALS/MND clinic[29 90] or in patients' homes.[59] Weight was measured either at each visit,[29] at assessment, follow-up,[91 102] premorbid period, at diagnosis,[71] at admission,[82] or at admission for enteral feeding tube insertion.[89] Weight loss (> 10% baseline) was used to inform the consideration of enteral feeding.[90] Rio *et al*.[89]

advised that patients with MND be routinely weighed in order to identify weight loss and the provision of dietetic support. The professionals identified as being involved included a team of specialist neurologists, physiotherapists, occupational therapists, speech and language therapists (SLTs), dietitians, specialist ALS nurses,[90] clinicians trained in ALS care, specialist dietitians[25] and nutritionists.[49]

Six guidelines and related documents[120 121 124 127 129 131] recommended measurement of weight at diagnosis and follow-up. The NICE MND clinical guideline[129] advised that weight should be assessed at diagnosis and at multidisciplinary team (MDT) assessments, and support, advice and interventions provided as required.[129] It was recommended that the core MDT should include: neurologist, specialist nurse, dietitian, physiotherapist, occupational therapist, respiratory physiologist/related professional, SLT, and palliative care specialist.[129] The EFNS guideline[120] also advised that the MDT team should include professionals relevant to nutrition including gastroenterologist, occupational therapist, SLT and dietitian. Barber *et al*.[121] stated that weight monitoring was important and should involve a dietitian. The Motor Neurone Disease Association (MNDA) provided an in-depth guideline to inform patients and support professionals on nutritional issues.[127] This advises that patients should monitor their own weight at home or ask to be weighed regularly at health appointments. The Parenteral and Enteral Nutrition Group (PENG) dietetic outcomes toolkit[131] included an informative care study describing the home-based nutritional care of a patient with MND, in which weight was measured as part of nutritional care.

Calculation of body mass index (BMI)

Fourteen studies[25 27 49 58 59 71 72 82 84 89 91 102 109 119] described and used the calculation of body mass index (BMI) in the nutritional management of pwALS/MND. Both Rio *et al*.[14] and Pena *et al*.[82] used BMI of 18.5 kg/m^2 as a threshold for identification of malnutrition. Whilst in Roubeau *et al*.[91] malnutrition was classed as : i) patients < 70 years old with BMI < 18.5 mg/m^2 and ii) patients >70 years old with BMI < 21 kg/m^2. The time point when BMI was measured varied. In Abdelnour-Mallet *et al*.[25]

patients visited the ALS clinic every 3 months for neurological, nutritional and respiratory monitoring by a MDT. In Limousin *et al*.[71] BMI was determined at premorbid, time of diagnosis and final study examination. In Fasano *et al*.[49] BMI was measured at diagnosis, at the time percutaneous endoscopic gastrostomy (PEG) was first required and at three and six months post-PEG. In Allen *et al*.[27] BMI was described at a maximum of three months prior to gastrostomy. Whilst in Rio *et al*.[89] BMI calculations was used at the time of enteral feeding tube receipt. Details of professionals undertaking the task of measuring BMI was scarce.

Two guidelines related documents[124 131] provided recommendation on the use of BMI. Burgos et al.[124] advised that calculation of BMI be part of the complete nutritional assessment for pwALS at diagnosis and monitored over time to identify early malnutrition. The case study in the PENG toolkit[131] also reported the calculation of BMI.

Setting of nutritional goal(s)

One study of 148 providers reported on setting nutritional goals.[15] These included weight maintenance or weight gain regardless of ideal body weight. Although 8% of providers stated an aim for weight maintenance if patients had lost > 10% of their ideal body weight.

Two guidelines,[124 131] reported on setting nutritional goals. Burgos *et al*.[124] advised that weight gain be recommended for patients with baseline BMI < 25 kg/m^2, weight stabilisation for BMI of 25-30 kg/m^2 and weight loss for BMI > 35 kg/m^2. Nutritional goals for a patient with MND were set as part of their nutritional management in the PENG case study.[131]

Calculation of energy requirements

Calculation of energy requirement was reported in 4 studies,[15 59 89 119] and methods used varied. Rio *et al*.[89] described the calculation of energy requirements for pwMND at feeding tube insertion and calculated daily energy requirements using the Soulsby and Weeks recommendation of 105-126 kJ kg

(25 – 30 kcal/kg) current body weight recommended for adults. Whilst Jesus *et al*.[59] described the application of Bilnut ™ software (Nutrisoft, Tours, France) in the assessment of protein energy intakes and distribution of key nutrients and Zhang *et al*.[15] used a range of methods including Harris Benedict equation, Mifflin St Jeor equation and indirect calorimetry. Details of who and where the assessment was undertaken was limited.

Calculations of energy was reported in one guideline document by Burgos *et al*.[124] and recommended that energy requirements be calculated for non-ventilated ALS patients if indirect calorimetry was not available.

Screening tool used

Four studies[72 78 98 109] described the use of a nutritional screening tool. Lopez-Gomez *et al*.[72] reported that the Malnutrition Universal Screening Tool and the Subjective Global Assessment was applied in the measurement of nutritional risk. Whilst Sznajder *et al*.[109] and Nunes *et al*.[78] reported the use of Nutritional Risk Screening 2002 tool. No guidelines or related documents were identified.

Assessment of diet/nutritional intake

Four studies[40 59 84 90] described the assessment of diet/nutritional intake. Jesus *et al*.[59] used the duration of an average main meal and 3 day dietary recall. Plowman *et al*.[84] described calorie counting and in Rooney *et al*.[90] it was reported that adequate nutrition was assessed by the dietician at each visit and patients with developing difficulties in swallowing were discussed by the MDT.

Two guideline[123 129] documents described the assessment of diet and nutritional intake. The NICE MND guideline[129] recommended evaluation of a patient's diet, hydration, nutritional intake and fluid intake. The British Medical Journal Best Practice (BMJ BP) ALS guideline[123] advised that a dietitian should be involved in diet modification for patients with mild to moderate symptomatic dysphagia with mild weight loss (<10%).

Assessment of swallowing/dysphagia

Twelve studies[41 58-60 72 74 78 81 84 92 97 102] presented evidence on the assessment of swallowing/dysphagia in pwALS/MND. Evidence of formal swallowing assessment was undertaken or confirmed by a SLT,[41 60 74 97] dietitian[74] or an experienced phoniatrician,[81] otolaryngologist[92] or was referred to speech pathology for evaluation.[92] The assessment was performed either at an outpatient clinic,[60] a multidisciplinary ALS/MND clinic,[74] or a non-specialist neurology clinic. Several methods were used to assess swallowing and dysphagia including subjective discomfort on swallowing,[59] presence of cough associated with food intake,[59] recurrent lung infection,[59] Hillel ALS swallow severity score,[60] the DePippo test,[59] assessment of swallowing of solid food and liquids,[102] modified barium swallow study,[84 92] water swallow test,[102] the Eating Assessment Tool-10 (EAT-10),[72] flexible laryngoscopy,[92] spirometry assessments, videostroboscopy[92] or flexible endoscopic evaluation of swallowing[92] and flexible endoscopic evaluation of swallowing with sensory testing (FEESST).[92] Details on when the assessment was undertaken was limited, Onesti *et al.*[81] reported that patients were observed periodically every 3 to 6 months. Nunes *et al.*[78] and Sarkar *et al*.[97] reported that a swallowing evaluation was performed to assess the risk of aspiration prior to PEG.

Eight guideline documents[120 121 124 125 127 129 130 132] provided information on the assessment of swallowing/dysphagia. Both the NICE clinical guideline for MND[129] and Oliver *et al*.[130] advised that a swallowing assessment should be organised if there are issues with swallowing or swallowing difficulties were suspected. Barber *et al.*[121] recommended that occupational therapists refer patients for instrumental swallowing assessment where required. The EFNS guideline[120] states that patients should be referred to a dietitian when dysphagia develops. Burgos et al.[124] advised screening for dysphagia be undertaken part of clinical evaluation for all ALS patients at diagnosis and follow up, at approximately a 3 month frequency depending on need. The MNDA Eating and Drinking guideline[127] stated that patients should request GPs to refer them to a SLT for a swallowing assessment if they had concerns about coughing/choking. The MNDA patient information leaflet on swallowing difficulties[125] also described the role of the SLT in evaluating swallowing function/dysphagia and

described the possible requirement for additional tests. The Royal College of Nursing (RCN) guideline for MND[132] also recommended that the SLT undertake assessment of swallowing.

Provision of advice on swallowing/dysphagia

The delivery of advice on swallowing and dysphagia was reported in several included publications. Five studies[81 85 90 92 102] described the provision of advice on swallowing/dysphagia. This included advice on nutritional intervention including advice on the chin-tuck manoeuvre[102] and taking small serial sips to limit aspiration.[92] Rubin *et al*.[92] also suggested that during FEESST, positional manoeuvres and swallowing techniques could be tested to minimise the risk of aspiration and assist with the identification of the least restrictive diet. A range of professionals were included in provision of advice on swallowing and dysphagia. In Pols et al.[85] gastroenterologist provided advice, whilst in Rooney *et al*.[90] SLT within the multidisciplinary ALS/MND clinic team provided such advice and in Rubin *et al*.[92] a speech pathologist was involved in this care.

Five guidelines reported provision on advice on swallowing and dysphagia.[120 125-127 132] Majority of the reports recommended early referral to SLT for dysphagia management in MND and stated that this care should be provided by SLT and additional care provided by occupational therapists[127 132] and care should include provision of advice on dysphagia diet counselling and postural manoeuvres[120 126 132] e.g. chin tuck manoeuvre; how to manage coughing, gagging and choking while eating and drinking,[127] posture for eating, advice on biting cheeks and tongue[126] and provision of a "swallow reminder" to support swallowing.[125] The MNDA patient information leaflet[125] also recommends patients seek advice on swallowing from their GP, SLT and dietitian.

Food fortification/enrichment advice

No studies reported on food fortification or enrichment advice, but this was addressed in three guidelines.[121 124 127] Barber *et al.*[121] recommended that neurologists work with dietitians to provide this advice. For patients with muscular fatigue and long lasting meals, it is advised that they enrich meals

with deficient nutrients or energy.[124] The MNDA eating and drinking guideline[127] also recommends that dietitian should offer advice on vitamins, supplements and how to fortify and enrich meals drinks.

Advice on modification of food texture and Provision of thickeners

Four studies included information of provision of advice on modification of food texture.[40 59 81 102] Advice was provided by the MDT[81] and was described as being part of nutritional education for early dysphagia, with the provision of advice on fluids and food consistency.[102] Jesus *et al.*[59] and Onesti *et al.*[81] reported the use of thickener to support food and/or fluid intake in 10% and 56 % of patients, respectively.

Several guideline publications recommended advice be sought on how patients with swallowing difficulties should modify the texture of their food. The NICE MND clinical guideline[129] stated that modification of food and drink consistency and palatability was necessary. Burgos *et al.*[124] recommended fractionation of food in cases of fatigue and long lasting meals and also advised that dietetic counselling be used to modify texture of solids and liquids to support swallowing and reduce aspiration, which may be informed by the instrumental assessment of swallowing function. Barber *et al.*[121] recommended the gastroenterologist be involved in the provision of thickeners and the MNDA eating and drinking resource[127] advised that thickening agents were available on prescription.. Other professionals included the SLT[125 127] and dietitian.[132]

Provision of oral nutritional supplements

The use of oral nutritional supplements was reported in three studies[59 90 102] but no further details were provided. Similarly, small numbers of guideline publications (n=3)[124 125 127] described provision of oral nutritional supplements to pwALS/MND. Burgos *et al.*[124] recommend these be used if weight loss progresses. The MNDA eating and drinking guideline[127] and swallowing difficulties leaflet[125] both advise that these can be discussed with the dietitian, with some available from the chemist and some via the GP on prescription.

Provision of equipment or physical aids to support eating

No studies reported on provision of equipment or physical aids to support eating. However, three guideline documents[125 127 129] contributed information on how equipment or physical aids to support eating may be provided to pwALS. The NICE clinical guideline for MND[129] advises that the need for eating and drinking aids and altered utensils to assist with eating be considered. The MNDA eating and drinking guideline[127] recommends the occupational therapist be involved in discussion of the need for equipment or physical aids. These may include a perching stool to help with food preparation, adapted cutlery and crockery, insulated plates to help keep food warm during longer mealtimes, and arm supports. The MNDA swallowing difficulties patient information leaflet[125] also indicates this support be provided by the occupational therapist but also suggests the SLT may provide advice.

Additional pre-gastrostomy nutritional management

The conference abstract by Wight *et al.*[116] described the use of "formal nutritional review" in pre-gastrostomy but provided no further details on what this review constituted.

Enteral nutrition

A wide range of gastrostomy methods were identified including PEG, per-oral image-guided gastrostomy (PIG), radiologically inserted gastrostomy (RIG), surgical gastrostomy and parenteral nutrition. Indications for gastrostomy were marked weight loss,[80] BMI < 18.5;[36] difficulties with oral feeding/swallowing;[36 80] unsafe swallow, prolonged difficult meals, recurrent aspiration,[36] chest infection,[36] concerns from patients, family or professionals e.g. SLT and dietitian;[80] anticipation of future need, air swallowing, frequent coughing on attempts to swallow, non-invasive ventilation (NIV) mask causing eating difficulties, severe secretion difficulties, dysphasia,[80] problems opening the mouth to eat and difficulty in feeding due to limb weakness. Several studies[56 60 104 105] reported discussions of gastrostomy, the provision of information and advice on gastrostomy and non-oral feeding, including information pack on post-gastrostomy management.[104]

Percutaneous endoscopic gastrostomy (PEG)

A large number of literature (51 studies) reported on the use of PEG as enteral feeding. There was a clinical preference to refer for PEG those patients with good respiratory function who would be able to tolerate endoscopy. Chavada *et al.*[36] recommended that gastrostomy be considered at >10% of pre-morbid weight loss, whereas, the ProGas Study,[87] recommended an earlier threshold of 5% weight loss. Stavroulakis *et al.*[106] indicated that in addition to having good respiratory function, the method of gastrostomy should also be guided by service availability, anatomical contraindications, patient clinical condition, previously failed gastrostomy insertion, and patient management. Chavada *et al.*[36] and Oliver *et al.*[79] recommended that PEG was inserted if patients had a forced vital capacity (FVC) >50% predicted value and have a low associated risk whilst in Rio *et al.*[89] PEG tube insertion was recommended in patients with (FVC >60% predicted or sniff nasal inspiratory pressure > 40 cm water). The timing of insertion was stated to be a decision to be made by the clinician with the patient. Reports by Oliver *et al.*[79 80] described members of the MDT involving those working in specialist palliative care or neurology services; particularly a SLT and dietitian for PEG as well the patient's family and carer would be involved in discussions regarding the use of PEG. Chavada *et al.*[36] described PEG delivery in an inpatient hospital setting in an endoscopy unit by an experienced endoscopist and specialist nurse. Qualitative research based at one MND Care Centre[79] described the delivery of post-gastrostomy training for patients who had received PEG. Training following the procedure was delivered by a range of professionals including a dietitian, specialist nurse, and other ward nurses.

Gastrostomy training at home was also available from range of staff including MND care centre professionals, community nurses, GPs, dietitians, PEG nurses, and private company nurses supplying feeds. Thompson *et al.*[110] noted that among those professionals reported to be involved were a neurology/MND specialist dietician, endoscopy specialist nurse and ALS specialist nurse. Sakel *et al.*[95] described an evaluation of a multidisciplinary integrated care pathway for gastrostomy feeding of people with MND. The aim of the pathway and associated checklist was to coordinate care and improve communication between professionals. Criteria for placing patients on the updated care pathway included the risk of malnutrition or symptoms of dysphagia. It was reported that the

provision of written information on PEG feeding and eating and drinking was improved. For patients receiving PEG nutrition, the mean prescribed energy was 21.8 kcal/kg (1195 kcal/day).[89]

The use of PEG in patients having inadequate respiratory function requiring NIV during tube insertion or being already established on NIV was reported in 3 studies.[51 88 101] Smith *et al.*[101] reported that patients received respiratory function tests prior to the procedure and the involvement of a respiratory physician was described.

The use of PEG with gastropexy was reported in a single study[103] and was delivered by two experienced endoscopists with the involvement of an anaesthetist. Other studies reported the use of nasal unsedated seated percutaneous endoscopic gastrostomy (NuPEG)[44] and gastric pressure relief bags during enteral feeding to reduce the symptoms of bloating.[83]

Several guidelines and related resources described the provision of PEG to patients with ALS/MND. The EFNS guideline[120] suggests 10% weight loss be used as an indication for PEG/RIG. Barber *et al.*[121] stated the important of professionals working with a dietitian for discussions around enteral feeding options. The BMJ BP ALS guideline[65] recommended that PEG be offered for patients with moderate symptomatic dysphagia with significant weight loss (>10%) and before FVC <50% predicted value. Three local guidelines/checklists were also available that provided site-specific guidance on PEG procedures.

Per-oral image-guided gastrostomy (PIG)

The use of PIG is less widespread than PEG and reported in three studies.[36 87 104] PIG was preferred for use in frail patients, those with reduced respiratory function and those in which the risk of endoscopy was viewed as being too high. PIG was mainly performed in a radiology unit by an interventional radiologist.[36]

Limited information was provided in guidelines and related documents. The BMJ BP ALS guide,[123] recommended the use of PIG, as an alternative to PEG in patients with poorer respiratory function.

Radiologically inserted gastrostomy (RIG)

Twenty-two studies[26 27 34 44 52 53 60 62 68 70 73 79 85-87 89 90 96 100 106 117 119] described the use of RIG in the nutritional management of pwALS/MND. In common with PIG, RIG was preferred for frailer patients, with poorer respiratory function,[44 89 90] higher risk of endoscopy, weight loss of > 10% of baseline,[90] increasing dysphagia,[90] failure of PEG due to diaphragm muscle weakness[89] and the capacity to use NIV during the RIG procedure.[70 89] In Lewis *et al.*[70] the decision to offer enteral feeding was agreed by a MDT, including neurologists, clinical nurse specialist, specialist SLT, and specialist dietician. All RIG procedures were performed either in the radiological interventional suite and RIG tube insertion was undertaken mainly by experienced operators and was performed with the placement of a mushroom-cage gastrostomy tube. Twenty percent of insertions required respiratory support. In Rio *et al*.[89] patients were prescribed a mean of 27.1 kcal/kg (1521kcal/day). Datta *et al*.[44] reported that the RIG was used prior to the development of a newer PEG procedure in patients who were at risk of sedation-related complications, reduced respiratory function or inability to lie flat.

Two guidelines and related document provided details on RIG, Barber *et al.*[121] advised that professionals work with a dietitian in considering RIG. The BMJ BP ALS guideline[123] recommended RIG as an alternative to PEG in patients with more advanced respiratory difficulties. A local guideline advised that the nursing staff should liaise with i) the dietitian on the RIG feeding regime, ii) the nutrition team to arrange patient/family education on use of the RIG tube, and iii) with the district nurse to initiate RIG care in the community.

Surgical gastrostomy

Seven studies[32 48 87 99 106 109 112] described the use of surgical gastrostomy, however, this was used in minority of patients[87] and no guidelines were identified.

Gastrostomy – unspecified method

Six additional studies described the use of gastrostomy but did not specify the method used.[29 37 56 83 90 93]

Several guidelines and related publications also provided additional information on other aspects of gastrostomy. The MNDA eating and drinking[127] and RCN[132] guidelines, Bede *et al.*[122] and Burgos *et al.*[124] all recommended the consideration of gastrostomy where required. The NICE MND guideline summary[130] noted that the use of gastrostomy should be discussed early and regularly with patients and, if needed, this should be undertaken without delay. Barber *et al.*[121] emphasised the importance of providing information on gastrostomy to patients to allow them to make their informed decision on acceptance or refusal. Two local guidelines provided area-specific guidance on discussion and referral for gastrostomy procedures. The MND case study in the PENG dietetic outcomes toolkit[131] illustrated that the patient's husband was involved in enteral feeding support at home by administering feed, fluids and medications via the gastrostomy tube. The case study also demonstrated that support from district nurses for gastrostomy management was provided and the neurology/MND specialist dietitian can provide home-based gastrostomy feed modification and changes in management, training on tube management and provision of supplements for tube feeding. The MNDA patient information leaflet on enteral tube feeding[128] described that the dietitian should provide post-gastrostomy advice in the hospital to patients and carers on tube use and management. It was also stated that most hospitals typically provide patients and carers with a written information guide on gastrostomy care upon discharge. The specialist nurse may also give advice in the hospital to patients and carers on tube management. The dietitian should advice on the use of specially prepared gastrostomy liquid feed and fluids for use with the gastrostomy tube[128] and advice on gastrostomy options may be provided by the nutrition nurse.[128]

The MNDA eating and drinking resource[127] and MNDA enteral feeding patient information leaflet[128] advises that if a patient makes the decision not to accept a feeding tube, the dietitian and SLT will continue to provide support.

Use of gastrostomy supplemented by oral nutrition intake

Although no studies reported on use of gastrostomy supplemented by oral nutrition intake. The MNDA eating and drinking guideline,[127] the BMJ BP ALS guideline[123] and The MNDA enteral feeding tube patient information leaflet[128] recommended that patients consider continuing their oral food and/or fluid intake alongside their use of gastrostomy in order to support their calorie intake, hydration and to allow continued enjoyment of eating and drinking as long as it was tolerated. In addition the MNDA enteral feeding tube patient information leaflet[128] stated that the dietitian and SLT be involved in this support.

Parenteral nutrition

Parenteral nutrition was described in four studies.[25 50 55 115] The EFNS guideline[120] stated that home parenteral nutrition could be used for patients with advanced ALS.

Nasogastric feeding tube

Two studies,[89 109] reported on the use of nasogastric tube feeding (NGT). It was prescribed in a limited number of patients with a mean prescribed energy of 30.6 kcal/kg (1388 kcal/day) and was mainly being used in cases of end-stage MND or for patients who refused gastrostomy.[89]

Limited guidelines and related resources[120 128] presented information on the use of nasogastric feeding tubes. The MNDA advised that they should be for short term use (< 4 weeks).[128] The EFNS guidance[120] recommends the use of NGT feeding for short term use when PEG/RIG is not appropriate. One local clinical guideline stated that, if gastrostomy was not possible, alternative feeding options should be discussed with the patient, including the use of NGT or nasojejunal tube.

Hydration advice

Advice on hydration was reported in a single case report by Cleary *et al.*[40] where a hydration protocol "free-water" was used.

The importance of provision of advice on hydration was described in five guideline publications.[121 125 127 128 132] The MNDA eating and drinking guide[58] advised that the feeding tube be used to maintain hydration. Professionals involved in providing hydration advice included the dietitian[121 128] SLT [125] and nurse (specialism not defined).[132]

Discussion

In this mapping review we identified 109 documents including 13 UK guidelines/clinical pathways and other related documents that provided evidence on nutritional management in pwALS. The evidence provided information on multiple types of nutritional management and highlighted that a wide range of professionals provide input into aspects of nutritional care. However, no evidence on the commissioning of nutritional care to pwALS/MND was identified. National guidelines presented a high-level overview and gave general guidance/recommendations for nutritional care for the pre-gastrostomy and post-gastrostomy populations but provide little specific detail.

Healthcare settings in which nutritional care was delivered were often not clearly reported. Similarly, whilst job roles that provide nutritional support were reported, there was little information available on the number of professionals and team compositions that should be involved. Where details were available, nutritional care was often delivered by the MDT in the context of a clinic setting. Some limited evidence on home-based care was available. However, details on how services are delivered in community settings were sparse.

The consensus in the included evidence was that weight management plays a central role in nutritional management. Evidence typically indicated that this should be part of nutritional assessment and follow-up. Other included data suggested patients may input by monitoring their own weight or

requesting to be weighed at visits. Monitoring of percentage weight change may be useful in identifying malnourished patients. Weight change is also used to inform decisions on the potential need for artificial nutrition. In some studies and guidelines, 10% weight loss was used as a threshold, whilst in others a 5% threshold was recommended in order to initiate earlier discussion and receipt of gastrostomy where appropriate.

The majority of included studies related to the delivery of gastrostomy (indeed it can be noted that much of the evidence related to non-proactive care). PEG and RIG were typically the most frequently used methods. The method of gastrostomy selected was determined largely by service availability but also by patient respiratory function. Most studies and guidelines recommended that PEG be placed before FVC < 50% predicted. However, there was also evidence that some sites were able to administer PEG with NIV support.

Data on the use of parenteral nutrition and nasogastric/nasojejunal tubes were areas in which evidence was very limited. Where the use of nasogastric tubes was discussed, recommendations were that this care should be considered a short term option. Few studies described the use of a range of different screening tools but no pattern was observed in which tool is most typically used.

Consistent evidence of the role of the SLT in swallowing assessment and provision of advice on the management of swallowing difficulties was identified. The dietitian and physiotherapist may also be involved in this care. There is limited published evidence on the work undertaken by dietitians in supporting pwALS. Some evidence was reported on the role of occupational therapists providing practical support to aid eating and drinking through the provision of equipment and aids.

Strength and limitations

The strength of our mapping review lies in the systematic approached used in undertaking this review. In addition, comprehensive searching was undertaken to identify international studies and guidelines,

making the review generalisable to a wide population. Whilst this review identified a range of evidence on nutritional management of pwALS, there was limited literature on oral nutritional care in the early stages of the disease, the types of professional involved during each stage of the disease and the optimal time to undertake the various assessment. Despite the limitations of the evidence base, this mapping review highlights further research in the area of ALS nutritional care is needed. Further good-quality research is vital to help inform how nutritional management is undertaken, by whom and where improvements are needed in care management. It is crucial that the gaps in the evidence base needs are addressed.

Conclusion

The evidence identified in this mapping review has highlighted the requirement for further primary research providing specific details on how nutritional management of pwALS is structured and delivered across the healthcare systems. This review has identified that nutritional management involves a wide range of healthcare professionals, but that the specific detail of roles involved and their remit at local level is not well documented in the research evidence.

Statement of Authorship

M.E, M.C, S.W, T.S, V.H, C.M made substantial contribution to the conceptualisation of the research; M.E, E.C, D.B, G.H, V.H, C.M undertook data curation. M.E, E.C, S.W, T.S, V.H, C.M acquired the funding; M.E, M.C, T.S, V.H, CM made substantial contribution to the development of the methodology; M.E, C.M undertook formal analysis; M.E, M.C, C.M undertook the investigation; M.E, V.H, C.M supervised; M.E, D.B, G.H, CM were involved with project administration; M.E did visualization; M.E drafted the paper and all other authors critiqued the paper for important intellectual content.

Conflicts of interest

CM received grants from National Institute for Health Research (NIHR), during the conduct of the study; personal fees from Biogen, undertook consultancy work for Orphazyme and Orion, and received speaker fees from Merz, outside the submitted work. M.E, E.C, M.C, D.B, G.H, S.W, V.H and T.S declared no conflicts of interest.

Acknowledgements/Funding

We would like to acknowledge Rachel Archer for her support in undertaking this mapping review. This project is funded by the National Institute for Health Research (NIHR) Programme Grants for Applied Research Programme (Grant Reference Number RP-PG-1016-20006) and supported by the NIHR Sheffield Biomedical Research Centre. The views expressed are those of the authors and not necessarily those of the NIHR or the Department of Health and Social Care.

References

1. Logroscino G, Beghi E, Zoccolella S, et al. Incidence of amyotrophic lateral sclerosis in southern Italy: a population based study. *Journal of Neurology, Neurosurgery & Psychiatry* 2005;76(8):1094-98.

2. Zoccolella S, Beghi E, Palagano G, et al. Predictors of long survival in amyotrophic lateral sclerosis: a population-based study. *Journal of the neurological sciences* 2008;268(1-2):28-32.

3. Hobson EV, McDermott CJ. Supportive and symptomatic management of amyotrophic lateral sclerosis. *Nature reviews Neurology* 2016;12(9):526-38. doi: https://dx.doi.org/10.1038/nrneurol.2016.111

4. McDermott CJ, Shaw PJ. Diagnosis and management of motor neurone disease. *British Medical Journal* 2008;336(7645):658-62. doi: 10.1136/bmj.39493.511759.BE

5. Desport JC, Preux PM, Truong TC, et al. Nutritional status is a prognostic factor for survival in ALS patients. *Neurology* 1999;53(5):1059-63.

6. Jawaid A, Murthy SB, Wilson AM, et al. A decrease in body mass index is associated with faster progression of motor symptoms and shorter survival in ALS. *Amyotrophic Lateral Sclerosis* 2010;11(6):542-48. doi: http://dx.doi.org/10.3109/17482968.2010.482592

7. Marin B, Desport JC, Kajeu P, et al. Alteration of nutritional status at diagnosis is a prognostic factor for survival of amyotrophic lateral sclerosis patients. *Journal of neurology, neurosurgery, and psychiatry* 2011;82(6):628-34. doi: https://dx.doi.org/10.1136/jnnp.2010.211474

8. NICE. Nutrition support for adults: Oral nutrition support, enteral tube feeding and parenteral nutrition. 2006. https://www.nice.org.uk/guidance/cg32.

9. EFNS Task Force on Diagnosis and Management of Amyotrophic Lateral Sclerosis, Andersen PM, Abrahams S, et al. EFNS guidelines on the clinical management of amyotrophic lateral sclerosis (MALS) -revised report of an EFNS task force. *European journal of neurology* 2012;19(3):360-75. doi: https://dx.doi.org/10.1111/j.1468-1331.2011.03501.x

10. Miller RG, Jackson CE, Kasarskis EJ, et al. Practice Parameter update: The care of the patient with amyotrophic lateral sclerosis: Multidisciplinary care, symptom management, and cognitive/behavioral impairment (an evidence-based review) Report of the Quality Standards Subcommittee of the American Academy of Neurology. *Neurology* 2009;73(15):1227-33. doi: 10.1212/WNL.0b013e3181bc01a4

11. Miller RG, Rosenberg JA, Gelinas DF, et al. Practice parameter: the care of the patient with amyotrophic lateral sclerosis (an evidence-based review: report of the Quality Standards Subcommittee of the American Academy of Neurology. *Neurology* 1999;52(7):1311-23.

12. Wills A-M, Hubbard J, Macklin EA, et al. Hypercaloric enteral nutrition in patients with amyotrophic lateral sclerosis: a randomised, double-blind, placebo-controlled phase 2 trial. *Lancet (London, England)* 2014;383(9934):2065-72. doi: https://dx.doi.org/10.1016/S0140-6736(14)60222-1

13. O'Brien MR. Healthcare professionals' knowledge of motor neurone disease. *British Journal of Nursing (Mark Allen Publishing)* 2004;13(18):1080-4.

14. Rio A, Cawadias E. Nutritional advice and treatment by dietitians to patients with amyotrophic lateral sclerosis/motor neurone disease: a survey of current practice in England, Wales, Northern Ireland and Canada. *Journal of human nutrition and dietetics : the official journal of the British Dietetic Association* 2007;20(1):3-13.

15. Zhang M, Hubbard J, Rudnicki SA, et al. Survey of current enteral nutrition practices in treatment of amyotrophic lateral sclerosis. *e-SPEN journal* 2013;8(1):e25-e28.

16. Korner S, Hendricks M, Kollewe K, et al. Weight loss, dysphagia and supplement intake in patients with amyotrophic lateral sclerosis (ALS): impact on quality of life and therapeutic options. *BMC neurology* 2013;13:84. doi: https://dx.doi.org/10.1186/1471-2377-13-84

17. National Institute for Clinical Excellence. Motor neurone disease: assessment and management. (NG42): NICE London 2016 available from: https://www.nice.org.uk/guidance/ng42

18. EPPI-Centre (2001) Core Keywording Strategy: Data Collection for a Register of Educational Research Version 0.9.4. London: EPPI-Centre, Social Science Research Unit. 2001 available from: EPPI Keyword strategy 0.9.4.doc (ioe.ac.uk)

19. James CM, Harper PS, Wiles CM. Motor neurone disease - A study of prevalence and disability. *Quarterly Journal of Medicine* 1994;87(11):693-99.

20. Oakley A, Gough D, Oliver S, et al. The politics of evidence and methodology: lessons from the EPPI-Centre. *Evidence and Policy* 2005;1(1):5-31.

21. Peersman G, EPPI-Centre. A descriptive mapping of health promotion studies in young people. 1996

22. EPPI-Centre. A systematic map and synthesis review of the effectiveness of personal development planning for improving student learning. 2003

23. James K, Randall N, Haddaway N. A methodology for systematic mapping in environmental sciences. *Environmental Evidence* 2016;5(7)

24. Miake-Lye I.M., Hempel S., Shanman R., et al. What is an evidence map? A systematic review of published evidence maps and their definitions, methods, and products. *Systematic Reviews* 2016;5:28.

25. Abdelnour-Mallet M, Verschueren A, Guy N, et al. Safety of home parenteral nutrition in patients with amyotrophic lateral sclerosis: a French national survey. *Amyotrophic lateral sclerosis : official*

publication of the World Federation of Neurology Research Group on Motor Neuron Diseases 2011;12(3):178-84. doi: https://dx.doi.org/10.3109/17482968.2010.531741

26. Ahmed O, Jilani D, Sheth S, et al. Radiologically Guided Placement of Mushroom-retained Gastrostomy Catheters: Long-term Outcomes of Use in 300 Patients at a Single Center. *Radiology* 2015;276(2):588-96. doi: 10.1148/radiol.15141327

27. Allen JA, Chen R, Ajroud-Driss S, et al. Gastrostomy tube placement by endoscopy versus radiologic methods in patients with ALS: a retrospective study of complications and outcome. *Amyotrophic lateral sclerosis & frontotemporal degeneration* 2013;14(4):308-14. doi: https://dx.doi.org/10.3109/21678421.2012.751613

28. Ammar T, Rio A, Ampong MA, et al. Replacement of mushroom cage gastrostomy tube using a modified technique to allow percutaneous replacement with an endoscopic tube in patients with amyotrophic lateral sclerosis. *Cardiovascular and interventional radiology* 2010;33(3):590-5. doi: https://dx.doi.org/10.1007/s00270-009-9763-8

29. Aridegbe T, Kandler R, Walters SJ, et al. The natural history of motor neuron disease: assessing the impact of specialist care. *Amyotrophic lateral sclerosis & frontotemporal degeneration* 2013;14(1):13-9. doi: https://dx.doi.org/10.3109/17482968.2012.690419

30. Astrow AB, Sood JR, Nolan MT, et al. Decision-making in patients with advanced cancer compared with amyotrophic lateral sclerosis. *Journal of medical ethics* 2008;34(9):664-8. doi: https://dx.doi.org/10.1136/jme.2007.022731

31. Atassi N, Cudkowicz ME, Schoenfeld DA. Advanced statistical methods to study the effects of gastric tube and non-invasive ventilation on functional decline and survival in amyotrophic lateral sclerosis. *Amyotrophic lateral sclerosis : official publication of the World Federation of Neurology Research Group on Motor Neuron Diseases* 2011;12(4):272-7. doi: https://dx.doi.org/10.3109/17482968.2011.577786

32. Bach JR, Gonzalez M, Sharma A, et al. Open gastrostomy for noninvasive ventilation users with neuromuscular disease. *American journal of physical medicine & rehabilitation* 2010;89(1):1-6. doi: https://dx.doi.org/10.1097/PHM.0b013e3181c55e2c

33. Beggs K, Choi M, Travlos A. Assessing and predicting successful tube placement outcomes in ALS patients. *Amyotrophic lateral sclerosis : official publication of the World Federation of Neurology Research Group on Motor Neuron Diseases* 2010;11(1-2):203-6. doi: https://dx.doi.org/10.3109/17482960903207377

34. Benstead T, Jackson-Tarlton C, Leddin D. Nutrition with Gastrostomy Feeding Tubes for Amyotrophic Lateral Sclerosis in Canada. *The Canadian journal of neurological sciences Le journal canadien des sciences neurologiques* 2016;43(6):796-800.

35. Boylan K, Levine T, Lomen-Hoerth C, et al. Prospective study of cost of care at multidisciplinary ALS centers adhering to American Academy of Neurology (AAN) ALS practice parameters. *Amyotrophic Lateral Sclerosis and Frontotemporal Degeneration* 2016;17(1-2):119-27. doi: http://dx.doi.org/10.3109/21678421.2015.1091478

36. Chavada G, El-Nayal A, Lee F, et al. Evaluation of two different methods for per-oral gastrostomy tube placement in patients with motor neuron disease (MND): PIG versus PEG procedures. *Amyotrophic lateral sclerosis : official publication of the World Federation of Neurology Research Group on Motor Neuron Diseases* 2010;11(6):531-6. doi: https://dx.doi.org/10.3109/17482968.2010.494306

37. Chhetri SK, Bradley BF, Majeed T, et al. Motor neurone disease in Lancashire and South Cumbria in North West England and an 8 year experience with enteral nutrition. *Journal of clinical neuroscience : official journal of the Neurosurgical Society of Australasia* 2016;24:47-51. doi: https://dx.doi.org/10.1016/j.jocn.2015.07.007

38. Chio A, Ilardi A, Cammarosano S, et al. Neurobehavioral dysfunction in ALS has a negative effect on outcome and use of PEG and NIV. *Neurology* 2012;78(14):1085-9. doi: https://dx.doi.org/10.1212/WNL.0b013e31824e8f53

39. Clavelou P, Blanquet M, Peyrol F, et al. Rates of progression of weight and forced vital capacity as relevant measurement to adapt Amyotrophic Lateral Sclerosis management for patient Result of a French multicentre cohort survey. *Journal of the Neurological Sciences* 2013;331(1-2):126-31. doi: 10.1016/j.jns.2013.06.002

40. Cleary S, Kizar S, Kalra S, et al. Using active rehabilitation to decrease the risk of pneumonia in end-of-life ALS and dementia care. *Canadian Nursing Home* 2008;19(2):4-10.

41. Cocks N, Ferreira H. What information do UK speech and language therapists use when making oral versus nonoral feeding recommendations for adults with oropharyngeal dysphagia? *Dysphagia* 2013;28(1):43-57. doi: https://dx.doi.org/10.1007/s00455-012-9411-3

42. Connolly S, Heslin C, Mays I, et al. Health and social care costs of managing amyotrophic lateral sclerosis (ALS): an Irish perspective. *Amyotrophic lateral sclerosis & frontotemporal degeneration* 2015;16(1-2):58-62. doi: https://dx.doi.org/10.3109/21678421.2014.957322

43. Czell D, Bauer M, Binek J, et al. Outcomes of percutaneous endoscopic gastrostomy tube insertion in respiratory impaired amyotrophic lateral sclerosis patients under noninvasive ventilation. *Respiratory care* 2013;58(5):838-44. doi: https://dx.doi.org/10.4187/respcare.02024

44. Datta A, Copsey H, Baumer D, et al. Changing practice from radiologically inserted gastrostomy (RIG) to nasal unsedated seated gastrostomy (NUPEG): Our experience. *Amyotrophic Lateral Sclerosis and Frontotemporal Degeneration* 2016;17(Supplement 1):290-91. doi: http://dx.doi.org/10.1080/21678421.2016.1232066/0025

45. de Bucourt M, Collettini F, Althoff C, et al. CT fluoroscopy-guided percutaneous gastrostomy with loop gastropexy and peel-away sheath trocar technique in 31 amyotrophic lateral sclerosis

patients. *Acta radiologica (Stockholm, Sweden : 1987)* 2012;53(3):285-91. doi: https://dx.doi.org/10.1258/ar.2011.110662

46. Dorst J, Dupuis L, Petri S, et al. Percutaneous endoscopic gastrostomy in amyotrophic lateral sclerosis: a prospective observational study. *Journal of neurology* 2015;262(4):849-58. doi: https://dx.doi.org/10.1007/s00415-015-7646-2

47. Evans KE, Leeds JS, Robson HE, et al. Neurodegeneration as a prognostic factor post gastrostomy insertion: To peg or not to peg? *Gut* 2010;59(Supplement 1):A50. doi: http://dx.doi.org/10.1136/gut.2009.209015q

48. Faria GR, Taveira-Gomes A. Open gastrostomy by mini-laparotomy: A comparative study. *International Journal of Surgery* 2011;9(3):263-66. doi: 10.1016/j.ijsu.2010.11.019

49. Fasano A, Fini N, Ferraro D, et al. Percutaneous endoscopic gastrostomy, body weight loss and survival in amyotrophic lateral sclerosis: a population-based registry study. *Amyotrophic lateral sclerosis & frontotemporal degeneration* 2017;18(3-4):233-42. doi: https://dx.doi.org/10.1080/21678421.2016.1270325

50. Fini N, Georgoulopoulou E, Vinceti M, et al. Noninvasive and invasive ventilation and enteral nutrition for ALS in Italy. *Muscle & nerve* 2014;50(4):508-16. doi: https://dx.doi.org/10.1002/mus.24187

51. Fitton F, Wood GJ, Weeraman S, et al. The use of non-invasive ventilation during percutaneous endoscopic gastrostomy insertion in patients with impaired respiratory ventilation. *American Journal of Respiratory and Critical Care Medicine* 2010;181(1 MeetingAbstracts)

52. Foley G, Timonen V, Hardiman O. Understanding psycho-social processes underpinning engagement with services in motor neurone disease: A qualitative study. *Palliative Medicine* 2014;28(4):318-25. doi: http://dx.doi.org/10.1177/0269216313512013

53. Foster A, Given M, Thornton E, et al. Removal of T-fasteners 2 days after gastrostomy is feasible. *Cardiovascular and interventional radiology* 2009;32(2):317-9. doi: https://dx.doi.org/10.1007/s00270-008-9473-7

54. Georgoulopoulou E, Fini N, Vinceti M, et al. The impact of clinical factors, riluzole and therapeutic interventions on ALS survival: a population based study in Modena, Italy. *Amyotrophic lateral sclerosis & frontotemporal degeneration* 2013;14(5-6):338-45. doi: https://dx.doi.org/10.3109/21678421.2013.763281

55. Graziani A, Martelli A, Quercia O, et al. Pulmonary scintigraphy as a method to investigate gastrobronchial communication in tracheostomized patients. *Respiratory medicine case reports* 2015;16:29-31. doi: https://dx.doi.org/10.1016/j.rmcr.2015.05.015

56. Greenaway LP, Martin NH, Lawrence V, et al. Accepting or declining non-invasive ventilation or gastrostomy in amyotrophic lateral sclerosis: patients' perspectives. *Journal of neurology* 2015;262(4):1002-13. doi: https://dx.doi.org/10.1007/s00415-015-7665-z

57. Gundogan K, Yurci A, Coskun R, et al. Outcomes of percutaneous endoscopic gastrostomy in hospitalized patients at a tertiary care center in Turkey. *European Journal of Clinical Nutrition* 2014;68(4):437-40. doi: 10.1038/ejcn.2014.11

58. Jackson-Tarlton CS, Benstead TJ, Doucette S, et al. Correlating factors in the recommendation of feeding tubes in the nutritional management of amyotrophic lateral sclerosis. *Amyotrophic lateral sclerosis & frontotemporal degeneration* 2016;17(7-8):515-21.

59. Jesus P, Massoulard A, Marin B, et al. First assessment at home of amyotrophic lateral sclerosis (ALS) patients by a nutrition network in the French region of Limousin. *Amyotrophic lateral sclerosis : official publication of the World Federation of Neurology Research Group on Motor Neuron Diseases* 2012;13(6):538-43. doi: https://dx.doi.org/10.3109/17482968.2012.701309

60. Johnson J, Leigh PN, Shaw CE, et al. Eating-derived pleasure in amyotrophic lateral sclerosis as a predictor of non-oral feeding. *Amyotrophic lateral sclerosis : official publication of the World Federation of Neurology Research Group on Motor Neuron Diseases* 2012;13(6):555-9. doi: https://dx.doi.org/10.3109/17482968.2012.704925

61. Kak M, Issa NP, Roos RP, et al. Gastrostomy tube placement is safe in advanced amyotrophic lateral sclerosis. *Neurological research* 2017;39(1):16-22.

62. Kalava A, Clendenen S, McKinney JM, et al. Bilateral thoracic paravertebral nerve blocks for placement of percutaneous radiologic gastrostomy in patients with amyotrophic lateral sclerosis: a case series. *Romanian journal of anaesthesia and intensive care* 2016;23(2):149-53. doi: https://dx.doi.org/10.21454/rjaic.7518/232.scl

63. Kara O, Kizilarslanoglu MC, Canbaz B, et al. Survival after Percutaneous Endoscopic Gastrostomy in Older Adults with Neurologic Disorders. *Nutrition in Clinical Practice* 2016;31(6):799-804. doi: http://dx.doi.org/10.1177/0884533616648132

64. Kawa C, Stewart J, Hilden K, et al. A retrospective study of nurse-assisted propofol sedation in patients with amyotrophic lateral sclerosis undergoing percutaneous endoscopic gastrostomy. *Nutrition in clinical practice : official publication of the American Society for Parenteral and Enteral Nutrition* 2012;27(4):540-4. doi: https://dx.doi.org/10.1177/0884533612443712

65. Kehyayan V, Korngut L, Jette N, et al. Profile of patients with amyotrophic lateral sclerosis across continuum of care. *The Canadian journal of neurological sciences Le journal canadien des sciences neurologiques* 2014;41(2):246-52.

66. Kirbis M, Koritnik B, Leonardis L, et al. Amyotrophic lateral sclerosis in Slovenia - Analysis of a patient cohort at the Ljubljana institute of clinical neurophysiology. *Zdravniski Vestnik* 2015;84(7):528-35.

67. Kirstein MM, Korner S, Schneider A, et al. Percutaneous endoscopic gastrostomy with and without jejunal extension in patients with amyotrophic lateral sclerosis. *European journal of gastroenterology & hepatology* 2018;30(3):257-62. doi: https://dx.doi.org/10.1097/MEG.0000000000001054

68. Kurien M, Andrews RE, Tattersall R, et al. Gastrostomies Preserve But Do Not Increase Quality of Life for Patients and Caregivers. *Clinical gastroenterology and hepatology : the official clinical practice journal of the American Gastroenterological Association* 2017;15(7):1047-54. doi: https://dx.doi.org/10.1016/j.cgh.2016.10.032

69. Lavernhe S, Antoine J-C, Court-Fortune I, et al. Home care organization impacts patient management and survival in ALS. *Amyotrophic lateral sclerosis & frontotemporal degeneration* 2017;18(7-8):562-68. doi: https://dx.doi.org/10.1080/21678421.2017.1332076

70. Lewis D, Ampong M-A, Rio A, et al. Mushroom-cage gastrostomy tube placement in patients with amyotrophic lateral sclerosis: a 5-year experience in 104 patients in a single institution. *European radiology* 2009;19(7):1763-71. doi: https://dx.doi.org/10.1007/s00330-009-1307-8

71. Limousin N, Blasco H, Corcia P, et al. Malnutrition at the time of diagnosis is associated with a shorter disease duration in ALS. *Journal of the neurological sciences* 2010;297(1-2):36-9. doi: https://dx.doi.org/10.1016/j.jns.2010.06.028

72. Lopez-Gomez JJ, Torres-Torres B, Gomez-Hoyos E, et al. Influence of a multidisciplinary protocol on nutritional status at diagnosis in amyotrophic lateral sclerosis. *Nutrition (Burbank, Los Angeles County, Calif)* 2018;48:67-72. doi: https://dx.doi.org/10.1016/j.nut.2017.11.010

73. Lowe AS, Laasch HU, Stephenson S, et al. Multicentre survey of radiologically inserted gastrostomy feeding tube (RIG) in the UK. *Clinical Radiology* 2012;67:843-54.

74. Marin B, Beghi E, Vial C, et al. Evaluation of the application of the European guidelines for the diagnosis and clinical care of amyotrophic lateral sclerosis (ALS) patients in six French ALS centres. *European journal of neurology* 2016;23(4):787-95. doi: https://dx.doi.org/10.1111/ene.12941

75. Martin NH, Landau S, Janssen A, et al. Psychological as well as illness factors influence acceptance of non-invasive ventilation (NIV) and gastrostomy in amyotrophic lateral sclerosis (ALS): a prospective population study. *Amyotrophic lateral sclerosis & frontotemporal degeneration* 2014;15(5-6):376-87. doi: https://dx.doi.org/10.3109/21678421.2014.886700

76. Martin NH, Lawrence V, Murray J, et al. Decision Making About Gastrostomy and Noninvasive Ventilation in Amyotrophic Lateral Sclerosis. *Qualitative health research* 2016;26(10):1366-81. doi: https://dx.doi.org/10.1177/1049732315583661

77. Murphy M, Quinn S, Young J, et al. Increasing incidence of ALS in Canterbury, New Zealand: a 22-year study. *Neurology* 2008;71(23):1889-95. doi: https://dx.doi.org/10.1212/01.wnl.0000336653.65605.ac

78. Nunes G, Santos CA, Grunho M, et al. Enteral feeding through endoscopic gastrostomy in amyotrophic lateral sclerosis patients. *Nutricion hospitalaria* 2016;33(5):561. doi: https://dx.doi.org/10.20960/nh.561

79. Oliver D, Udoma M. Improving quality of life in patients with advanced motor neurone disease. *European Journal of Palliative Care* 2011;18(5):214-17.

80. Oliver D, Campbell C, Sykes N, et al. Decision-making for gastrostomy and ventilatory support for people with motor neurone disease: variations across UK hospices. *Journal of palliative care* 2011;27(3):198-201.

81. Onesti E, Schettino I, Gori MC, et al. Dysphagia in Amyotrophic Lateral Sclerosis: Impact on Patient Behavior, Diet Adaptation, and Riluzole Management. *Frontiers in neurology* 2017;8:94. doi: https://dx.doi.org/10.3389/fneur.2017.00094

82. Pena MJ, Ravasco P, Machado M, et al. What is the relevance of percutaneous endoscopic gastrostomy on the survival of patients with amyotrophic lateral sclerosis? *Amyotrophic lateral sclerosis : official publication of the World Federation of Neurology Research Group on Motor Neuron Diseases* 2012;13(6):550-4. doi: https://dx.doi.org/10.3109/17482968.2012.684215

83. Phippen A, Brennan E, Ealing J, et al. Farrell valve relieves bloating in gastrostomy patient. *BMJ supportive & palliative care* 2017;7(3):258-60. doi: https://dx.doi.org/10.1136/bmjspcare-2017-001375

84. Plowman EK, Tabor LC, Wymer J, et al. The evaluation of bulbar dysfunction in amyotrophic lateral sclerosis: survey of clinical practice patterns in the United States. *Amyotrophic lateral sclerosis & frontotemporal degeneration* 2017;18(5-6):351-57. doi: https://dx.doi.org/10.1080/21678421.2017.1313868

85. Pols J, Limburg S. A Matter of Taste? Quality of Life in Day-to-Day Living with ALS and a Feeding Tube. *Culture, medicine and psychiatry* 2016;40(3):361-82. doi: https://dx.doi.org/10.1007/s11013-015-9479-y

86. Power S, Kavanagh LN, Shields MC, et al. Insertion of Balloon Retained Gastrostomy Buttons: A 5-Year Retrospective Review of 260 Patients. *Cardiovascular and Interventional Radiology* 2013;36(2):484-91. doi: 10.1007/s00270-012-0456-3

87. ProGas Study G. Gastrostomy in patients with amyotrophic lateral sclerosis (ProGas): a prospective cohort study. *The Lancet Neurology* 2015;14(7):702-9. doi: https://dx.doi.org/10.1016/S1474-4422(15)00104-0

88. Rafique J, Luck P, Chaudhry N, et al. Are NIV supported PEG insertions (NSPI) in patients with neuromuscular degenerative disorders (NMD) safe and effective? *Thorax* 2013;68(SUPPL. 3):A155-A56. doi: http://dx.doi.org/10.1136/thoraxjnl-2013-204457.329

89. Rio A, Ellis C, Shaw C, et al. Nutritional factors associated with survival following enteral tube feeding in patients with motor neurone disease. *Journal of human nutrition and dietetics : the official journal of the British Dietetic Association* 2010;23(4):408-15. doi: https://dx.doi.org/10.1111/j.1365-277X.2010.01057.x

90. Rooney J, Byrne S, Heverin M, et al. A multidisciplinary clinic approach improves survival in ALS:A comparative study of ALS in Ireland and Northern Ireland. *Journal of Neurology, Neurosurgery and Psychiatry* 2015;86(5):496-501. doi: http://dx.doi.org/10.1136/jnnp-2014-309601

91. Roubeau V, Blasco H, Maillot F, et al. Nutritional assessment of amyotrophic lateral sclerosis in routine practice: value of weighing and bioelectrical impedance analysis. *Muscle & nerve* 2015;51(4):479-84. doi: https://dx.doi.org/10.1002/mus.24419

92. Rubin AD, Griffin GR, Hogikyan ND, et al. A new member of the multidisciplinary ALS team: The otolaryngologist. *Amyotrophic Lateral Sclerosis* 2012;13(2):229-32. doi: http://dx.doi.org/10.3109/17482968.2011.643898

93. Ruffell TO, Martin NH, Janssen A, et al. Healthcare professionals' views on the provision of gastrostomy and noninvasive ventilation to amyotrophic lateral sclerosis patients in England, Wales, and Northern Ireland. *Journal of palliative care* 2013;29(4):225-31.

94. Russ KB, Phillips MC, Wilcox CM, et al. Percutaneous endoscopic gastrostomy in amyotrophic lateral sclerosis. *The American journal of the medical sciences* 2015;350(2):95-7. doi: https://dx.doi.org/10.1097/MAJ.0000000000000517

95. Sakel M, Sansom W, Lamming J, et al. Completed audit loop for a multidisciplinary care pathway for the gastrostomy feeding of people with motor neurone disease. *Proceedings of the Nutrition Society* 2010;69(OCE7):E520-E20. doi: 10.1017/s0029665110004544

96. Sancho J, Servera E, Chiner E, et al. Noninvasive respiratory muscle aids during PEG placement in ALS patients with severe ventilatory impairment. *Journal of the neurological sciences* 2010;297(1-2):55-9. doi: https://dx.doi.org/10.1016/j.jns.2010.06.022

97. Sarkar P, Cole A, Scolding NJ, et al. Percutaneous Endoscopic Gastrostomy Tube Insertion in Neurodegenerative Disease: A Retrospective Study and Literature Review. *Clinical endoscopy* 2017;50(3):270-78. doi: https://dx.doi.org/10.5946/ce.2016.106

98. Selkirk SM, Washington MO, McClellan F, et al. Delivering tertiary centre specialty care to ALS patients via telemedicine: a retrospective cohort analysis. *Amyotrophic Lateral Sclerosis and Frontotemporal Degeneration* 2017;18(5-6):324-32. doi: http://dx.doi.org/10.1080/21678421.2017.1313867

99. Sharma A, Bach JR, Swan KG. Open Gastrostomy under Local Anesthesia for Patients with Neuromuscular Disorders. *American Surgeon* 2010;76(4):369-71.

100. Sharpley D, Angus R, Parker R. Ventilation, nutrition and survival in patients with motor neurone disease. *European Respiratory Journal* 2013;42(SUPPL. 57)

101. Smith MR, Matsou A, Nathani N, et al. Non-invasive ventilation during percutaneous endoscopic gastrostomy insertion in motor neurone disease patients - A safe and effective multi-disciplinary approach. *Gut* 2014;63(SUPPL. 1):A17-A18. doi: http://dx.doi.org/10.1136/gutjnl-2014-307263.36

102. Spataro R, Ficano L, Piccoli F, et al. Percutaneous endoscopic gastrostomy in amyotrophic lateral sclerosis: effect on survival. *Journal of the neurological sciences* 2011;304(1-2):44-8. doi: https://dx.doi.org/10.1016/j.jns.2011.02.016

103. Stasinos I, Lim M, Dent H, et al. Anaesthetist assisted percutaneous endoscopic gastrostomy with gastropexy; a safe and efficient procedure. *United European Gastroenterology Journal* 2016;4(5 Supplement 1):A716. doi: http://dx.doi.org/10.1177/2050640616663689

104. Stavroulakis T, Baird WO, Baxter SK, et al. The impact of gastrostomy in motor neurone disease: challenges and benefits from a patient and carer perspective. *BMJ supportive & palliative care* 2014;0:1-8. doi: https://dx.doi.org/10.1136/bmjspcare-2013-000609

105. Stavroulakis T, Baird WO, Baxter SK, et al. Factors influencing decision-making in relation to timing of gastrostomy insertion in patients with motor neurone disease. *BMJ supportive & palliative care* 2013;0(1):1-7. doi: https://dx.doi.org/10.1136/bmjspcare-2013-000497

106. Stavroulakis T, Walsh T, Shaw PJ, et al. Gastrostomy use in motor neurone disease (MND): a review, meta-analysis and survey of current practice. *Amyotrophic lateral sclerosis & frontotemporal degeneration* 2013;14(2):96-104. doi: https://dx.doi.org/10.3109/17482968.2012.723722

107. Strijbos D, Hofstede J, Keszthelyi D, et al. Percutaneous endoscopic gastrostomy under conscious sedation in patients with amyotrophic lateral sclerosis is safe: an observational study. *European journal of gastroenterology & hepatology* 2017;29(11):1303-08. doi: https://dx.doi.org/10.1097/MEG.0000000000000959

108. Swetz KM, Peterson SM, Sangaralingham LR, et al. Feeding Tubes and Health Care Service Utilization in Amyotrophic Lateral Sclerosis: Benefits and Limits to a Retrospective, Multicenter Study Using Big Data. *Inquiry : a journal of medical care organization, provision and financing* 2017;54:46958017732424. doi: https://dx.doi.org/10.1177/0046958017732424

109. Sznajder J, S Lefarska-Wasilewska M, Klek S. The influence of the initial state of nutrition on the lifespan of patients with amyotrophic lateral sclerosis (ALS) during home enteral nutrition. *La influencia del estado inicial de la nutricion en la esperanza de vida de pacientes con esclerosis lateral amiotrofica (ALS) durante la nutricion enteral en casa* 2016;33(1):3-7. doi: https://dx.doi.org/10.20960/nh.v33i1.7

110. Thompson AG, Blackwell V, Marsden R, et al. A risk stratifying tool to facilitate safe late-stage percutaneous endoscopic gastrostomy in ALS. *Amyotrophic lateral sclerosis & frontotemporal degeneration* 2017;18(3-4):243-48. doi: https://dx.doi.org/10.1080/21678421.2016.1274330

111. Tomasello G, Bellavia M, Damiano G, et al. Enteral nutrition: Our experience with percutaneous endoscopic gastrostomy (PEG) and revision of literature. *Progress in Nutrition* 2012;14(2):141-43.

112. Tsou AY, Karlawish J, McCluskey L, et al. Predictors of emergent feeding tubes and tracheostomies in amyotrophic lateral sclerosis (ALS). *Amyotrophic lateral sclerosis : official publication of the World Federation of Neurology Research Group on Motor Neuron Diseases* 2012;13(3):318-25. doi: https://dx.doi.org/10.3109/17482968.2012.662987

113. Van Der Steen I, Van Den Berg JP, Buskens E, et al. The costs of amyotrophic lateral sclerosis, according to type of care. *Amyotrophic Lateral Sclerosis* 2009;10(1):27-34. doi: http://dx.doi.org/10.1080/17482960802103131

114. Vanis N, Saray A, Gornjakovic S, et al. Percutaneous endoscopic gastrostomy (PEG): Retrospective analysis of a 7-year clinical experience. *Acta Informatica Medica* 2012;20(4):235-37. doi: http://dx.doi.org/10.5455/aim.2012.20.235-237

115. Verschueren A, Monnier A, Attarian S, et al. Enteral and parenteral nutrition in the later stages of ALS: an observational study. *Amyotrophic lateral sclerosis : official publication of the World Federation of Neurology Research Group on Motor Neuron Diseases* 2009;10(1):42-6. doi: https://dx.doi.org/10.1080/17482960802267480

116. Wight AG, Bennett J, Ward K, et al. Improving the patient journey for patients referred for niv in motor neurone disease: Early impact of national guidance. *American Journal of Respiratory and Critical Care Medicine* 2012;185(MeetingAbstracts)

117. Wills A, How A, Squires N, et al. Nutritional intervention in patients with motor neurone disease - 5 years' experience from a regional care and research centre. *Journal of Neurology, Neurosurgery and Psychiatry* 2009;80(4):465. doi: http://dx.doi.org/10.1136/jnnp.2008.167387

118. Zamietra K, Lehman EB, Felgoise SH, et al. Non-invasive ventilation and gastrostomy may not impact overall quality of life in patients with ALS. *Amyotrophic lateral sclerosis : official publication of the World Federation of Neurology Research Group on Motor Neuron Diseases* 2012;13(1):55-8. doi: https://dx.doi.org/10.3109/17482968.2011.641570

119. Zhang L, Sanders L, Fraser RJL. Nutritional support teams increase percutaneous endoscopic gastrostomy uptake in motor neuron disease. *World journal of gastroenterology* 2012;18(44):6461-p.66. doi: https://dx.doi.org/10.3748/wjg.v18.i44.6461

120. Andersen PM, Abrahams S, Borasio GD, et al. EFNS guidelines on the Clinical Management of Amyotrophic Lateral Sclerosis (MALS) - revised report of an EFNS task force. *European Journal of Neurology* 2012;19(3):360-75. doi: http://dx.doi.org/10.1111/j.1468-1331.2011.03501.x

121. Barber C. Management of bulbar symptoms in motor neurone disease: a community speech and language therapist perspective. *British Journal of Neuroscience Nursing* 2015;11(1):41-46.

122. Bede P, Oliver D, Stodart J, et al. Palliative care in amyotrophic lateral sclerosis: A review of current international guidelines and initiatives. *Journal of Neurology, Neurosurgery and Psychiatry* 2011;82(4):413-18. doi: http://dx.doi.org/10.1136/jnnp.2010.232637

123. British Medical Journal Best Practice. Amyotrophic lateral sclerosis. The right clinical information, right where it's needed. 2017

124. Burgos R, Breton I, Cereda E, et al. ESPEN guideline clinical nutrition in neurology. *Clinical nutrition (Edinburgh, Scotland)* 2018;37(1):354-96. doi: https://dx.doi.org/10.1016/j.clnu.2017.09.003

125. Motor Neurone Disease Association. Swallowing difficulties 7A 2017. Available from: https://www.mndassociation.org/app/uploads/2017/05/07a-swallowing-difficulties.pdf

126. Motor Neurone Disease Association. Living with motor neurone disease 2015. Available from: https://www.mndassociation.org/app/uploads/2015/07/Living-with-MND-STANDARD-PDF-Oct-20-1.pdf

127. Motor Neurone Disease Association. Eating and drinking with motor neurone disease 2017. Available from: https://www.mndassociation.org/app/uploads/2017/05/Eating-and-drinking-with-MND-final-web-PDF-2017.pdf

128. Motor Neurone Disease Association. Tube feeding 7B 2015. Available from: https://www.mndassociation.org/app/uploads/2015/03/07B-Tube-feeding.pdf.

129. NICE guideline. Motor neurone disease: assessment and management 2016. Available from: https://www.nice.org.uk/guidance/ng42/resources/motor-neurone-disease-assessment-and-management-pdf-1837449470149

130. Oliver D, Radunovic A, Allen A, et al. The development of the UK National Institute of Health and Care Excellence evidence-based clinical guidelines on motor neurone disease. *Amyotrophic Lateral Sclerosis and Frontotemporal Degeneration* 2017;18(5-6):313-23. doi: http://dx.doi.org/10.1080/21678421.2017.1304558

131. PENG The Parenteral and Enteral Nutrition Group. The dietetic outcomes toolkit 2016. Available from: https://www.peng.org.uk/pdfs/publications/dietetic-outcomes-toolkit.pdf

132. RCN. New guidance for nurses treating patients with MND 2016. Available from: http://mnd.rcnlearning.org.uk/.