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

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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 Coordinating Centre (EPPI-Centre) core key wording strategy version 0.9.4¹⁸ 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.

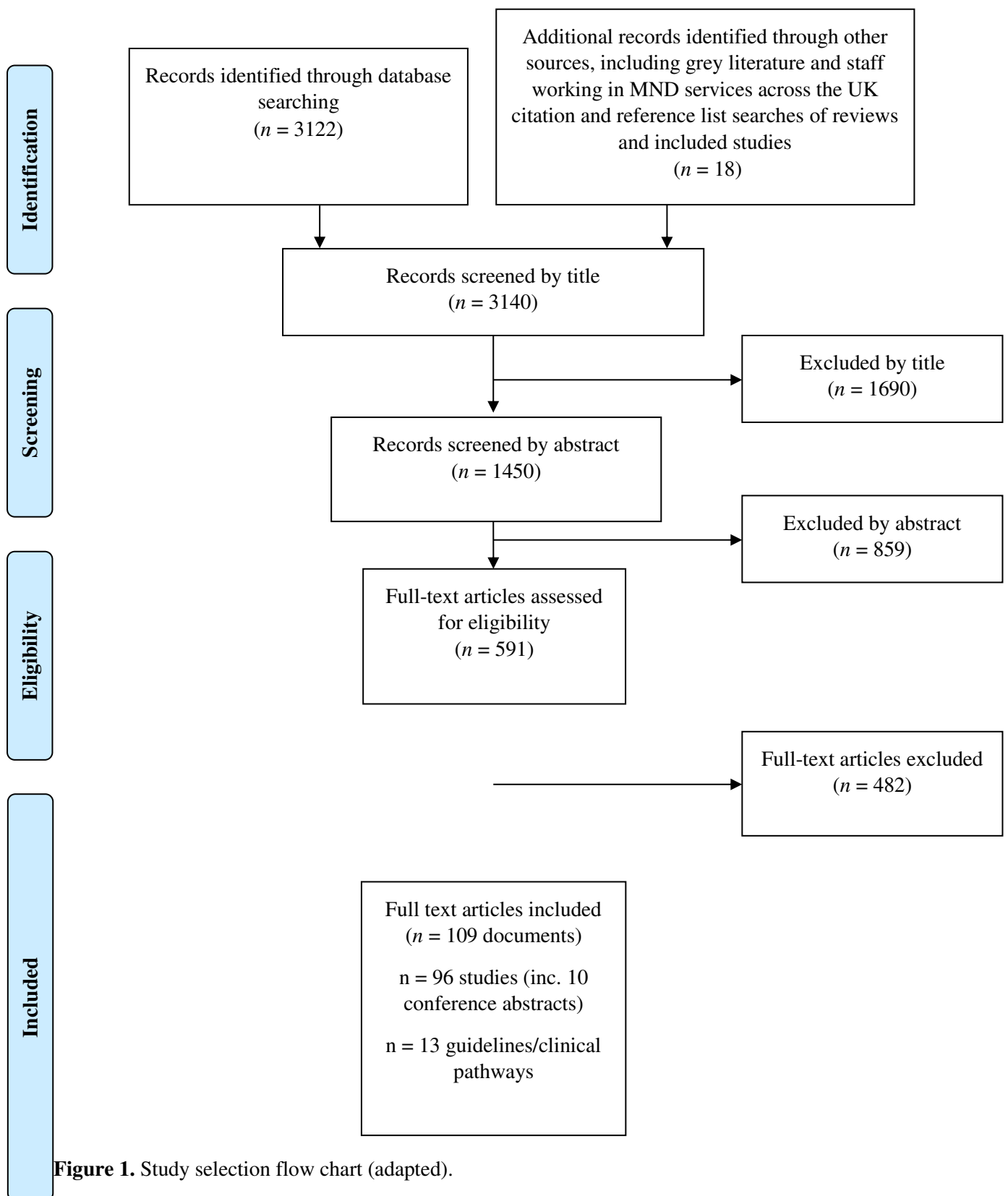


Figure 1. Study selection flow chart (adapted).

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 |
| Georgouloupoulou 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 |
| Stasinou 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 dietitians²⁵ 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 guideline¹²⁹ 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 guideline¹²⁰ 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 toolkit¹³¹ 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/m² 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/m² and ii) patients >70 years old with BMI < 21 kg/m². 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 toolkit¹³¹ 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/m², weight stabilisation for BMI of 25-30 kg/m² and weight loss for BMI > 35 kg/m². 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 guideline¹²⁹ recommended evaluation of a patient's diet, hydration, nutritional intake and fluid intake. The British Medical Journal Best Practice (BMJ BP) ALS guideline¹²³ 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} dietitian⁷⁴ or an experienced phoniatrix,⁸¹ otolaryngologist⁹² 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, videostroboscopy⁹² or flexible endoscopic evaluation of swallowing⁹² 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 MND¹²⁹ 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 guideline¹²⁰ 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 guideline¹²⁷ 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 difficulties¹²⁵ 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 MND¹³² 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 manoeuvre¹⁰² 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 tongue¹²⁶ and provision of a "swallow reminder" to support swallowing.¹²⁵ The MNDA patient information leaflet¹²⁵ 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 guideline¹²⁷ 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 MDT⁸¹ 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 guideline¹²⁹ 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 resource¹²⁷ 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 guideline¹²⁷ and swallowing difficulties leaflet¹²⁵ 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 MND¹²⁹ advises that the need for eating and drinking aids and altered utensils to assist with eating be considered. The MNDA eating and drinking guideline¹²⁷ 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 leaflet¹²⁵ 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 Centre⁷⁹ 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 study¹⁰³ 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 guideline¹²⁰ 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 guideline⁶⁵ 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 weakness⁸⁹ 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 guideline¹²³ 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 patients⁸⁷ 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}

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Several guidelines and related publications also provided additional information on other aspects of gastrostomy. The MNDA eating and drinking¹²⁷ and RCN¹³² guidelines, Bede *et al.*¹²² and Burgos *et al.*¹²⁴ all recommended the consideration of gastrostomy where required. The NICE MND guideline summary¹³⁰ 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 toolkit¹³¹ 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 feeding¹²⁸ 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 tube¹²⁸ and advice on gastrostomy options may be provided by the nutrition nurse.¹²⁸

The MNDA eating and drinking resource¹²⁷ and MNDA enteral feeding patient information leaflet¹²⁸ 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 guideline¹²³ and The MNDA enteral feeding tube patient information leaflet¹²⁸ 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 leaflet¹²⁸ 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 guideline¹²⁰ 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 guidance¹²⁰ 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 guide⁵⁸ advised that the feeding tube be used to maintain hydration. Professionals involved in providing hydration advice included the dietitian^{121 128} SLT¹²⁵ 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 approach 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.

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