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The nutritional management of people living with Amyotrophic Lateral Sclerosis (ALS): A national survey of dietitians

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Abstract

Background

People living with Amyotrophic Lateral Sclerosis (ALS) face many challenges to taking adequate nutrition. Growing evidence links weight loss with negative prognostic outcomes. We aimed to explore the practice of dietitians in the UK with regards the nutritional management of ALS.

Methods

A national online survey was disseminated via professional groups, social media, and newsletters to UK healthcare professionals, between September and November 2018. The

survey examined the nutritional management of ALS. Dietitian responses are reported in this paper.

Results

In total, 130 dietitians responded to the survey. Two thirds reported that ALS comprised less than a 20% of their total patient caseload. Fourty two percent reported that nutritional screening took place in their organisation. Half of dietitians reported that patients were referred for dietetic assessment at 'about the right time' although 44% reported referrals were made too late. The majority (83%) of dietitians used resting energy expenditure predictive equations not validated in ALS. When setting weight goals, dietitians reported most frequently recommending weight maintenance if the patients BMI was 18.5-25kg/m^2 (72%), 25-30kg/m^2 (98%), and over 30kg/m^2 (79%). In addition, 43% reported that people with ALS were not weighed frequently enough.

Conclusions

While the importance of early nutritional assessment is recognised, the timeliness of dietetic input and on-going monitoring of nutritional status in ALS care might not currently be ideal. Dietitians report using energy requirement predictive equations and setting weight goals that may not promote positive outcomes. Further research is required to understand the optimal nutritional management of ALS.

1. Introduction

Amyotrophic Lateral Sclerosis (ALS; known as Motor Neuron Disease, or MND, in the UK) is a progressive neurological condition that causes the degeneration of motor neurons and the progressive loss of muscle mass, with a prevalence of 5-7 per 100,000 [(1)]. In addition to dysphagia, other consequences of having ALS present challenges to taking adequate nutrition, including weakening grip strength, fatigue, and poor appetite [(2)]. Reduced nutritional intake, in conjunction with the presence of disease related hyper-metabolism, contributes to a prevalence of malnutrition estimated to be between 15-55% in people with ALS [(3,4,5)]. Malnutrition is a negative independent prognostic indicator of survival, highlighting the importance of identifying and treating nutritional issues in ALS [(6, 7)]. Evidence in support of the effectiveness of nutritional interventions to correct malnutrition in ALS is scant, although some studies have demonstrated that oral nutrition support can prevent further weight loss [(8,9)]. Post hoc analysis of randomised controlled trial data, found that people with fast progressing

ALS who added a high fat supplement to their daily diet, had improved survival [10]. Therefore, improving nutritional status with a high calorie diet could improve nutritional and prognostic outcomes in ALS.

There is a lack of ALS-specific guidance supporting healthcare professionals with the nutritional management of ALS. Dietetic texts and guidance, cover the nutritional management of ALS, but are not supported by a strong evidence base [11,12]. ALS guidelines recognise the high prevalence of nutritional compromise in ALS and the need to consider gastrostomy placement, but do not include specific recommendations about how malnutrition can be effectively identified, assessed and treated in clinical practice [13,14,15]. The successful management of disease related malnutrition requires a systematic approach to identification, cause analysis, intervention and monitoring [16]. In the UK, the Dietetic Process [17] has been adopted to inform the dietetic management of individuals requiring nutritional intervention. This cyclical process - involving nutritional assessment, identifying a nutritional diagnosis, nutritional monitoring and on-going evaluation - guides dietitians through the stages required to deliver and monitor effective dietetic interventions, including the management of malnutrition.

There is a paucity of research on the optimal approaches to identifying and treating the nutritional issues in ALS, with much of the literature focusing on post-gastrostomy care [18,19]. Dietitians are experts, qualified in the nutritional management of people at risk of disease related malnutrition. This makes dietitians' views and practice important when trying to understand the current nutritional management of ALS. Previous surveys of the nutritional management of ALS have described a variation in practice used to assess nutritional intake, status and requirements [20,21]. Rio and colleagues survey of 23 dietitians, found a range of nutritional assessments and dietetic interventions used in ALS [20]. The aim of this survey was to explore UK dietetic practice, in light of recent evidence demonstrating the raised energy requirements and potential benefits of optimising nutritional status in ALS.

Methods

The design of this cross sectional survey was informed by findings of a mapping review of the literature on the structure and input of nutritional management services for ALS and stakeholder workshops with healthcare professionals in the UK [22,23]. The survey questions were developed through discussions between the study team and modified in response to the

feedback from multidisciplinary team members (including 5 dietitians) who piloted the survey.

The survey was developed and hosted using the Qualtrics platform. The survey link was distributed via professional networks, UK MND care centres, and third-party organisations (e.g. MND Association) between September and November 2018. It was also publicised via relevant professional group social media, websites and newsletters. A snowball sampling technique was then used to optimise distribution of the survey, with participants asked to forward the link to the survey to relevant colleagues. Any healthcare professional in the UK who had experience of supporting the nutritional management in ALS was eligible to take part.

The survey included 109 questions, including 38 questions directed only at dietitians through conditional branching, aimed at identifying dietitians' views and practice with regards the nutritional management of ALS. Through conditional branching the additional questions related to a) why, how and when a person with ALS may be referred to a dietitian; b) nutritional assessment of people with ALS; c) nutritional interventions used in practice; d) monitoring of nutritional status. The full list of survey questions can be found in the Supplementary Information 1. The responses of only dietitians were analysed and reported in this paper, to explore their approach to the nutritional management of ALS. The survey took approximately 30 minutes to complete.

Data were analysed and summarised descriptively via SPSS®.

Ethical approval was granted by the Research Ethics Committee of the School of Health and Related Research at the University of Sheffield (ref: 018781), and governance approval for this study was granted by the Health Research Authority (ref: 18/HRA/2340). A consent form was included at the beginning of the online survey, which was required to be completed before participants could proceed.

2. Results

Of the 281 health professionals who responded to the survey, only the responses from the 130 (46%) dietitians were analysed and reported in this paper. Not all respondents answered every question in the survey. To make it clear about how many responded to each question, the percentage (%) and number of individual responses (n) to each question option and the total number of participants answering each question (N) will be presented (%; n/N). The full data

set, summarising the responses of dietitians to each question and the full list of options dietitians had to choose from when answering specific questions are presented in Supplementary Information 2.

On average, dietitians responding to the survey had been qualified for 12.7 years (N=129; SD 9.1) and supported people with ALS for 6.8 years (N=129; SD 5.6). The majority of dietitians reported they were currently providing dietetic care in ALS (87%; n=113/130) or had done in the past (12%; n=16/130), with only 1 respondent reporting having never provided care in ALS. Caseloads varied, with 66% (n=74/113) of dietitians being referred between 0-10 new people with ALS per year and 58% (n=65/113) having between 0-10 people with ALS on their current caseload. Most dietitians (69%; n=89/130) reported that people with ALS made up 0-20% of their total caseload. The vast majority of dietitians (97%; n=126/130) agreed that supporting the nutritional needs of people with ALS is either very or extremely important. Supplementary Information 3 summarises the characteristics and experience of the dietitians responding to the survey.

Dietetic process results

Identification of nutritional need

Under half of the dietitians (42%; n=54/129) reported that nutritional screening in ALS takes place in their organisation. Of the 54 dietitians reporting that nutritional screening takes place, 67% (n=36/54) reported a screening tool was used, with the Malnutrition Universal Screening Tool (MUST [24]) (83%; n=30/36) being the most frequently cited. Half of dietitians (50%; n=27/54) reported that screening takes place on a hospital ward and 44% (n=24/54) during an ALS clinic. Over half (56%; n=30/54) reported that screening takes place when there were indications that nutrition may be a concern, 52% (n=28/54) at diagnosis and 52% (n=28/54) during in-patient admissions.

Dietitians most frequently reported (43%; n=56/130) that their local healthcare teams were 'moderately successful' in identifying nutritional issues in people with ALS. The factors most frequently reported to result in a referral to the dietitian included an unsafe swallow assessment by a Speech and Language Therapist (85%; n=110/129); identified weight loss (84%; n=108/129); patient/carer reported weight loss (81%; n=105/129); and patient/carer reported poor dietary intake (78%; n=101/129). Dietitians reported receiving referrals from a variety of sources including from a doctor (91%; n=118/130), Speech and Language Therapist (89%; n=115/130); and a nurse (85%; n=111/130). Dietitians most frequently

reported (44%; n=57/129) patients being referred for dietetic assessment when nutritional problems were identified in ALS clinic. Only 19% (n=25/129) of dietitians reported that people with ALS were referred at the time of diagnosis. With regards to the timing of dietetic referral, 50% (n=64/129) reported that people with ALS were referred at 'about the right time' although 44% (n=57/129) reported that referral for dietetic advice was made too late.

Assessment

Assessing nutritional status

Following referral to a dietitian, 40% (n=51/129) of respondents reported that people with ALS were nutritionally stable but starting to experience nutritional problems (e.g., weight loss or reduced food intake); with 13% (n=17/129) reporting people with ALS presented with <10% weight loss and 23% (n=30/129) with >10% weight loss in the previous 6 months. Twenty-two percent (n=28/129) of dietitians reported discussing gastrostomy tube placement in the first consultation with people with ALS. Body weight (92%; n=119/130), Body Mass Index (BMI; 92%; n=119/130) and percentage weight loss over the previous three to six months (82%; n=106/130) were the most cited measures of nutritional status. Under a third (31%; n=40/130) of dietitians calculated mid-upper arm muscle circumference. Over half of dietitians (59%; n=75/128) did not calculate an ideal body weight (IBW) for in ALS. Where IBW was calculated, BMI was the most frequently reported calculation method (94%; n=50/53).

Estimating oral nutritional intake

With regards to the assessment of nutritional intake, 89% (n=114/128) of dietitians reported using a diet history (a retrospective record of usual dietary intake) to record patients' dietary intake. Very few dietitians (5%; n=6/128) reported asking people with ALS to record some form of diet diary. Nearly all (99%; n=127/128) dietitians estimated the nutritional content of dietary intake using their knowledge of the nutritional content of foods, with just under a quarter (23%; n=28/128) also using printed or electronic dietary analysis resources to make these calculations. Energy (95%; n=121/127), protein (94%; n=121/128) and fluid (97%; n=122/126) intakes were estimated at most or every dietetic review. Intake of other nutrients including fat, carbohydrate, vitamins, minerals and fibre was estimated less frequently.

Calculating nutritional requirements

The majority (83%; n=106/128) of dietitians reported that they use the Henry equation [25] to estimate a resting energy expenditure (REE) in ALS. Although 62% (n=86/128) of dietitians, in some or every ALS case, would add a stress factor to the estimated REE to predict total daily energy expenditure (TDEE), 28% (36/128) reported never adding a stress factor. Of those dietitians adding stress factors, the most frequently reported percentage stress factor was between six and ten percent (45%; n=39/86), with the hypermetabolic effect of ALS being the most frequently reported rationale for adding this (80%; n=68/85). Just over half of dietitians (55%; n=47/85) added a stress factor because of the metabolic effect caused by being in respiratory failure and 41% (n=35/85) because the patient is on non-invasive ventilation (NIV). A large majority of dietitians (94%; n=120/128) used the PENG guideline to estimate protein requirement i.e. converted from Nitrogen g/kg/day [26,27].

Planning nutrition and dietetic intervention

Just over half of dietitians (56%; n= 72/128) reported that their aim was to maintain weight regardless of premorbid weight or IBW, whereas 38% (n=49/128) would aim to achieve and maintain IBW. Table 1 describes the weight goals that dietitians would set, dependent on the BMI of the person with ALS. The majority of dietitians would aim for weight gain for BMI<18.5kg/m^2 (91%; n=115/130); and weight maintenance if the BMI 18.5-25kg/m^2 (72%; n=92/130), BMI 25-30kg/m^2 (98%; n=125/130) or BMI >30kg/m^2 (79%; n=100/127). Twenty-one percent (n=27/127) would aim for weight loss if BMI>30kg/m^2.

[Insert: Table 1. The weight goals dietitians would set dependent on the BMI (kg/m2) of a person with ALS. see separate file with table

Implementing nutrition and dietetic intervention

Dietitians most frequently (49%; n=63/130) reported that they believed their local healthcare team was 'moderately successful' at implementing nutritional management plans to address nutritional issues in ALS. Just over half (55%; n=72/130) reported fortifying diet and fluids (the 'food first approach') was only 'moderately effective' for in ALS; with 23% (n=30/130) feeling it was 'not very' or 'slightly' effective. A minority (22%; n=28/130) reported that the food first approach was 'very' or 'extremely' effective in meeting the nutritional requirements of people recently diagnosed with ALS. Just over half of dietitians (51%; n= 65/128) reported they would sometimes recommend oral nutritional supplements during their initial contact with people with ALS.

Monitoring and review

Nutritional monitoring

Similar to findings relating to implementing nutritional management plans, just over half of dietitians (51%; n=66/129) reported their local healthcare teams to be 'moderately effective' at nutritional monitoring in ALS. When asked about the effectiveness of their own nutritional monitoring, dietitians most frequently reported this was only 'moderately effective' (45%; n=57/127); compared with 11% (n=14/127) who felt they were 'not at all' or 'slightly effective' and 44% (n=56/127) reporting they were 'very' or 'extremely successful'.

Just over half of dietitians (54%; n=68/126) reported that they weighed people with ALS every three or more months with others weighing more frequently than this. Fourty-three percent (n=55/126) reported people with ALS were weighed too infrequently' with the majority (94%; n=121/129) reporting they should be weighed at least monthly. Sixty-two percent (n=79/127) were able to weigh patients who were unable to attend clinic and 82% (n=104/127) could weigh patients who were unable to stand on weighing scales. Dietitians reported having access to a range of weighing scales including step on scales (79%; n=102/130); wheelchair scales (65%; n=85/130); and hoist scales (45%; n=58/130).

Location, frequency and duration of nutritional monitoring

Dietitians most frequently reported that they routinely follow-up people with ALS that are receiving oral nutrition support every two to three months (43%; n=55/128). Around a third of dietitians (34%; n=43/128) reported spending 30 to 40 minutes with patients during a follow-up consultation, and the majority (76%; n=97/128) stated they review patients in their own homes.

3. Discussion

This is the largest published survey focusing on the nutritional management of ALS by UK dietitians, with 130 dietitians responding. However, ALS only accounted for a small proportion of the dietitians' caseloads with very few specialising in ALS alone. This is an important issue, as gaining ALS specific clinical experience will take longer to accumulate while dietitians are not in roles with a greater focus on ALS care.

Identification of nutritional need

Elia [24] recommends routine use of screening tools to identify patients at risk of malnutrition. The systematic screening for malnutrition risk allows for timely nutrition support interventions and onward referral for specialist advice [28]. The number of dietitians reporting that screening takes place in ALS (42%; n=54/129), is lower than the 99% of hospitals reporting in a national survey that they have a nutrition screening policy [29]. This may explain why people with ALS were reported to be often referred late for dietetic assessment. Dysphagia and other consequences of ALS present a challenge to meeting nutritional requirements and place people with ALS at high risk of malnutrition [5]. Although ALS guidance recommends assessing for nutritional problems, there is not an explicit recommendation for the use of screening tools such as MUST [15,24]. A study adopting MUST to screen for nutritional risk in a prospective cohort design found that over 90% of people with ALS presented with a high risk of malnutrition [28]. Malnutrition screening tools, sensitive to ALS-specific risk factors such as dysphagia, deteriorating hand-grip strength and sialorrhea, would facilitate timely nutritional assessment and intervention, allowing prioritisation of those who require a dietetic referral.

Assessment

National guidance on the management of disease related malnutrition recommends the assessment of nutritional status, nutritional intake and nutritional requirements [30]. In this study, dietitians most commonly reported using weight, BMI and percentage weight change as a measure of nutritional status. Only a third of dietitians used surrogate measures of fat-free mass such as mid-upper arm muscle circumference. A survey of 23 dietitians working in ALS centres reported similar results, but only 9% used mid-arm anthropometry compared with 31% in the present study [20]. Weight loss in ALS has been found to correlate with losses in both fat-free mass and fat mass [2]. Further research is required to identify the optimal nutritional assessment methods in ALS, to allow the evaluation of the efficacy of nutritional interventions' in attenuating the loss of muscle mass. Nutritional interventions that are found to slow the loss of fat-free mass have the potential to improve function and quality of life in ALS. Routine monitoring of fat-free mass would allow for the effectiveness of nutritional interventions to be monitored and modification of nutritional management plans.

Consistent with a previous survey, dietitians used diet histories to record the dietary intake in ALS [20]. A dietitian's knowledge of the nutritional content of food and drink was the most common method used to assess the nutritional content of dietary intake. Only a quarter of

dietitians used dietary analysis resources to estimate the nutritional content of the diet taken. The subjective nature of estimating dietary intake and analysing these records could result in underestimating the nutritional intake of people with ALS, and may lead to individuals not meeting their energy requirements [31].

The estimation of a person's energy requirement is an important step when devising initial dietetic plans and can be estimated using predictive equations [25,32,33,34]. In the present study 82% of dietitians reported using a resting energy expenditure (REE) predictive equation that is based on measurements validated with groups of healthy individuals [25]. Using these predictive equations to estimate the energy requirements of an individual with ALS is flawed as they do not account for the impact of the disease on an individual's REE. Indeed, there is growing evidence that people with ALS are characterised by a raised REE with nearly 50% being hypermetabolic [35,36] with REE estimated to be 10% greater than that of healthy individuals [32]. Predictive equations have been found to underestimate REE in 58% of ALS cases, compared to measured REE [37]. The use of predictive equations that are likely to underestimate REE in ALS could result in setting dietetic calorie goals that do not meet energy requirements, and contribute to the deteriorating nutritional status often observed in ALS [5]. The survey was disseminated prior to the release of the most recent edition of national dietetic guidance, which many UK dietitians refer to when estimating energy requirements [12]. This may explain why the Henry equation was still the most frequently cited predictive equation used, rather than the ALS specific energy predictive equations included in the new edition [12]. Routinely using predictions of REE or TDEE validated in ALS, should allow for the setting of dietetic goals that more accurately meet the needs of people with ALS [32,33,34,37].

Planning the nutrition and dietetic intervention

A thorough dietetic assessment informs the design of nutritional intervention plans that aim to improve clinical, nutritional and quality of life outcomes. The success of nutritional interventions can be measured against the dietetic goals set. Increased weight loss in ALS is associated with a shorter prognosis [38], while higher BMIs are positively associated with longer survival, with BMI 30-35 kg/m^2 predicting the greatest survival benefit even after adjusting for measures of disease severity [39].

By contrast, the majority of dietitians responding to our survey would aim for weight maintenance for people with ALS if they had a BMI>18.5kg/^{2}. Therefore, it appears that

dietitians may be continuing to advocate weight maintenance, despite the current evidence suggesting that weight gain predicts better outcomes in ALS. While there is a need for further prospective research to corroborate the relationship between weight gain and positive outcomes in ALS, dietitians may currently be missing an opportunity to improve the disease course of ALS by not setting weight gain as a dietetic goal.

Implementing nutrition and dietetic intervention

Following the identification of malnutrition risk, nutrition support interventions are recommended to address the risk factors and prevent further rapid deterioration in nutritional status [30]. Increasing the calorie density of the food and drink, has been recommended as the first line oral nutrition support intervention, often referred to as the 'food first' approach [40]. A systematic review found that optimising the energy content of normal diet taken can result in increased calorie intake and weight [41]. However, 23% (n=30) of dietitians reported that the food first approach was 'not very' or 'slightly' effective in people recently diagnosed with ALS. This may be related to patients being referred to them when they are starting to experience nutritional issues, and therefore require oral nutritional supplements following initial assessment.

Oral nutritional supplements have been found to be an effective method of stabilising weight in ALS [5,8]. Individuals with ALS supplementing their oral diet with whey protein significantly increased their BMI and were able to preserve lean body mass in another study [9]. Further research is required to evaluate the effectiveness of 'food first' approaches earlier in the disease course, as well as, the timely use of oral nutritional supplements.

Monitor and review

Close nutritional monitoring is required in the majority of people with ALS, in light of the often rapid and variable progression of the disease [4]. Dietitians responding to the survey reported infrequent nutritional monitoring and dietetic review in ALS. Nearly all agreed that people with ALS should be weighed monthly. Though weight is the most accessible measure of nutritional status in clinical practice, many people with ALS become unable to safely stand up. Most dietitians had access to stand-on weighing scales but fewer had the ability to weigh non-ambulatory patients. Individuals with ALS who are non-ambulatory are often at a later stage of disease and at higher risk of malnutrition, therefore require close nutritional monitoring. The majority (88%) of dietitians responding to the survey reported asking people with ALS to weigh themselves. A system of self-screening may be a more effective way of

identifying nutritional issues early in ALS [42]. However, this would only be effective for as long as the patient can weigh themselves. Inadequate nutritional monitoring and review may contribute to the weight loss observed in ALS, although the efficacy of such nutritional care pathways remain to be evaluated.

Strengths and limitations

While the sample in our survey included well over 100 dietitians with experience of working in ALS our recruitment may be biased in favour of those who with greater interest or experience of the condition . In particular, one possible reason for this may be the adoption of a snowball sampling approach via ALS and dietetic networks. Future studies should explore more systematic and cross-sectional sampling techniques which may increase the diversity of the sample.

As this survey only focused on the dietetic practice in the UK the results may not reflect dietetic practice in other countries, in contrast to previous investigations [20]. The validity of the findings reported here is strengthened by the majority of respondents having experience of the nutritional management of ALS.

4. Conclusions

The results from the present survey shed light on dietitians' nutritional management of ALS in the UK. While the importance of nutritional management in ALS is recognised, the findings suggest that timely identification of malnutrition risk and initiation of nutrition support interventions might not be ideal. The survey reports varied approaches to assessing energy requirements and setting dietetic goals that may not be in line with current evidence. More work is required to meet the specific nutritional needs of people with ALS. Future approaches could include access to dietitians soon after diagnosis and closer nutritional monitoring of people with ALS.

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Transparency Declaration

The lead author affirms that this manuscript is an honest, accurate, and transparent account of the study being reported. The reporting of this work is compliant with STROBE guidelines. The lead author affirms that no important aspects of the study have been omitted and there were no discrepancies from the study as planned.

References

1. McDermott CJ, Shaw PJ (2008). Diagnosis and management of motor neurone disease. *BMJ* **336,** 658-62.
2. Ngo S, Van Eijk R, Chachay V *et al* (2019). Loss of appetite is associated with a loss of weight and fat mass in patients with amyotrophic lateral sclerosis. *Amyotroph Lateral Scler Frontotemporal Degener* **0**, 1-9.
3. Dupuis L, Oudart H, Rene F *et al* (2004). Evidence for defective energy homeostasis in amyotrophic lateral sclerosis: benefit of a high-energy diet in a transgenic mouse model. *Proc Natl Acad Sci* USA **101**, 11159-64.
4. Genton L, Viatte V, Janssens J *et al* (2011). Nutritional state, energy intakes and energy expenditure of amyotrophic lateral sclerosis (ALS) patients. *Clin Nutr* **30**, 553-559.
5. Körner S, Hendricks M, Kollewe K *et al* (2013). Weight loss, dysphagia and supplement intake in patients with amyotrophic lateral sclerosis (ALS): Impact on quality of life and therapeutic options. *BMC Neurol* **13**, 84.
6. Desport JC, Preux PM, Truong TC *et al* (1999). Nutritional status is a prognostic factor for survival in ALS patients. *Neurology* **53**, 1059-63.

7. Rio A, Ellis C, Shaw C, et al. (2010). Nutritional factors associated with survival following enteral tube feeding in patients with motor neurone disease. *J Hum Nutr Diet* **23,** 408-415
8. Dorst J, Cypionka J, Ludolph AC (2013). High-caloric food supplements in the treatment of amyotrophic lateral sclerosis: a prospective interventional study. *Amyotroph Lateral Scler Frontotemporal Degener* **14**, 533–6.
9. Silva L, Mourão L, Silva A, *et al* (2010). Effect of nutritional supplementation with milk whey proteins in amyotrophic lateral sclerosis patients. *Arq Neuropsiquiatr* 2010 **68**, 263-268.
10. Ludolph A, Dorst J, Dreyhaupt J *et al* (2019). Effect of High-Caloric Nutrition on Survival in Amyotrophic Lateral Sclerosis. *Ann Neurol* **87**, 206-216.
11. Cawadias E, Rio A 2014, Neurological disease, Motor neurone disease in: Manual of Dietetic Practice. pp. 555-562 [Gandy, J editors]. John Wiley & Sons, Incorporated, Hoboken. 2014.
12. British Dietetic Association (BDA) (2018). Parenteral and Enteral Nutrition Specialist Group of the BDA (PENG). A pocket guide to clinical nutrition - Adult requirements section. 5th Edition. 3.11a-3.11b.
13. Miller R, Jackson C, Kasarskis E *et al* (2009). Practice Parameter update: The care of the patient with amyotrophic lateral sclerosis: Multidisciplinary care, symptom management, and cognitive/behavioural impairment (an evidence-based review). *Neurology* **73**, 1227-1233.
14. Andersen P, Abrahams S, Borasio G *et al* (2012). EFNS guidelines on the Clinical Management of Amyotrophic Lateral Sclerosis (MALS) - revised report of an EFNS task force. *Eur J Neurol* **19**,360–375.
15. NICE. Clinical guideline NG42 (2019). Motor neurone disease: assessment and management. https://www.nice.org.uk/guidance/NG42 (accessed July 2020).
16. Stratton R, Smith T, Gabe S (2018). Managing malnutrition to improve lives and save money. www.bapen.org.uk (accessed July 2020)
17. British Dietetic Association (2016). Model and Process for Nutrition and Dietetic Practice. https://www.bda.uk.com/professional/practice/processmodeldieteticpractice_2016_v (accessed November 2019)

18. Wills A, Hubbard J, Macklin E *et al* (2014). Hypercaloric enteral nutrition in patients with amyotrophic lateral sclerosis: a randomised, double-blind, placebo controlled phase 2 trial. *Lancet* **383**, 2065-72.
19. ProGas group. Gastrostomy in patients with amyotrophic lateral sclerosis (ProGas): a prospective cohort study. *Lancet Neurol* **14**, 702–709.
20. Rio A, Cawadias E (2007). 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. *J Hum Nutr Diet* **20**, 3-13.
21. Zhang M, Hubbard J, Rudnicki S *et al* (2013). Survey of current enteral nutrition practices in treatment of amyotrophic lateral sclerosis. *ESPEN J* **8**, e25-e28.
22. Zarotti N, Coates E, McGeachan A *et al* (2019). Health care professionals' views on psychological factors affecting nutritional behaviour in people with motor neuron disease: A thematic analysis. *Br J Health Psychol* **24**:953-969.
23. Halliday V, Zarotti N, Coates L *et al* (2021). Delivery of nutritional management services to people with amyotrophic lateral sclerosis (ALS). *Amyotroph Lateral Scler Frontotemporal Degener* Online ahead of print 1-10
24. Elia M (2003). The 'MUST' Report. Nutritional screening of adults: a multidisciplinary responsibility. Development and use of the 'Malnutrition Universal Screening Tool' ('MUST') for adults. https://www.bapen.org.uk/pdfs/must/must-report.pdf (accessed July 2019)
25. Henry C (2005). Basal metabolic rate studies in humans: measurement and development of new equations. *Public Health Nutr* **8**,1133–1152.
26. British Dietetic Association (BDA) (2011). Parenteral and Enteral Nutrition Specialist Group of the BDA (PENG). A pocket guide to clinical nutrition - Adult requirements section. 4th Edition. 3.13.
27. Elia M (1990). Artificial Nutrition Support. *Med Int* **82**, 3392-3396
28. López-Gómez J, Torres-Torres B, Gómez-Hoyos E *et al* (2018). Influence of a multidisciplinary protocol on nutritional status at diagnosis in amyotrophic lateral sclerosis. *Nutrition* **48**, 67-72.
29. Russell C, Elia M (2011). Nutrition screening survey in the UK and republic of Ireland in 2011. A report by the British Association for Parenteral and Enteral Nutrition (BAPEN). Hospitals, care homes and mental health units. Accessed from https://www.bapen.org.uk/pdfs/nsw/nsw-2011-report.pdf on 3/3/21.

30. NICE. Clinical Guideline CG32 (2017). Nutrition support for adults: oral nutrition support, enteral tube feeding and parenteral nutrition. https://www.nice.org.uk/guidance/cg32 (accessed July 2020)
31. Martin G, Tapsell L, Batterham M *et al* (2002). Relative bias in diet history measurements: a quality control technique for dietary intervention trials. *Public Health Nutr* **5**, 537–545.
32. Desport J, Preux P, Magy L *et al* (2001). Factors correlated with hypermetabolism in patients with amyotrophic lateral sclerosis. *Am J Clin Nutr* **74**, 328-334.
33. Vaisman N, Lusaus M, Nefussy B *et al* (2009). Do patients with amyotrophic lateral sclerosis (ALS) have increased energy needs? *J Neurol Sci* **279**, 26-29.
34. Kasarskis E, Mendiondo M, Matthews D *et al* (2014). Estimating daily energy expenditure in individuals with amyotrophic lateral sclerosis. *Am J Clin Nutr* **99**, 792–803.
35. Bouteloup C, Desport J, Clavelou A *et al* (2009). Hypermetabolism in ALS patients: an early and persistent phenomenon. *J Neurol* **256**, 1236-1242.
36. Steyn F, Ioannides Z, Van Eijk R *et al* (2018). Hypermetabolism in ALS is associated with greater functional decline and shorter survival. *J Neurol Neurosurg Psychiatry* **89**, 1016-1023.
37. Jesus P, Marin B, Fayemendy P *et al* (2018). Resting energy expenditure equations in amyotrophic lateral sclerosis, creation of an ALS-specific equation. *Clin Nutr* **38**, 1657-1665.
38. Shimizu T, Nakayama Y, Matsuda C *et al* (2019). Prognostic significance of body weight variation after diagnosis in ALS: a single-centre prospective cohort study. *J Neurol* **266**, 1412–1420.
39. Paganoni S, Deng J, Jaffa M *et al* (2011). Body mass index, not dyslipidemia, is an independent predictor of survival in amyotrophic lateral sclerosis. *Muscle Nerve* **44**, 20-4.
40. Burgos R, Breton I, Cereda E et al (2018) ESPEN guideline clinical nutrition in neurology. *Clin Nutr* **37**, 354-396.
41. Baldwin C, Weekes C (2012). Dietary counselling with or without oral nutritional supplements in the management of malnourished patients: A systematic review and meta-analysis of randomised controlled trials. *J Hum Nutr Diet* **25**, 411-426.
42. BAPEN (2018). Self-screening tool. https://www.malnutritionselfscreening.org/self-screening (accessed July 2020)

Supplementary information

Additional supporting information may be found online in the Supporting Information section at the end of the article.

Supplementary Information 1. Survey pro-forma.

Supplementary Information 2. Complete data set of survey responses from dietitians to the survey.

Supplementary Information 3. Characteristics and experience of dietitians participating in the survey.