RNM-01: Arterial blood gas analysis and pulmonary function tests comparison in Amyotrophic Lateral Sclerosis

Dr Maria Claudia Torrieri[1], Dr Umberto Manera[1], Dr Cristina Moglia[1,2], Dr Margherita Viglione[1], Dr Margherita Daviddi[1], Dr Enrico Matteoni[1], Dr Luca Solero[1], Dr Francesca Palumbo[1], Dr Rosario Vasta[1], Dr Antonio Canosa[1,2], Dr Fabrizio D'Ovidio[1], Dr Fulvia Ribolla[3], Dr Michela Bellocchia[3], Dr Luana Focaraccio[4], Dr Alessio Mattei[3], Dr Gabriele Mora[5], Prof Andrea Calvo[1,2], Prof Adriano Chiò[1,2]

[1]ALS Centre, "Rita Levi Montalcini" Department, University of Turin, Turin, Italy, [2]SC Neurologia 1U, AOU Città della Salute e della Scienza, Turin, Italy, [3]SC Pneumologia U, AOU Città della Salute e della Scienza , Turin, Italy, [4]Pneumology Unit, ASL Città di Torino, Turin, Italy, [5]ALS Centre, Istituti Clinici Scientifici Maugeri, IRCCS, Milan, Italy

Live Poster Session A, December 9, 2020, 5:10 PM - 5:50 PM

Background:
There is still lack of consensus among guidelines about the right timing for starting non-invasive mechanical ventilation (NIMV) in Amyotrophic Lateral Sclerosis (ALS) [1,2]. The importance of spirometry as reliable prognostic factor in ALS is widely recognized; conversely, few studies investigated the role of Arterial Blood Gas analysis (ABG).

Objective:
To investigate the role of ABG in predicting NIMV initiation in a cohort of ALS patients, determining the best cut-offs for Forced Vital Capacity (FVC%), carbon dioxide (pCO2), carbonate (HCO3-) and standard base excess (SBE) to suggest NIMV initiation and to predict survival.

Methods:
We included 488 ALS patients with concomitant ABG and spirometry, followed in the Turin ALS Centre, diagnosed from 2000 to 2015. Correlations between ABG and FVC were studied. ROC curves were computed to find the most sensitive cut-off for ABG/PFT parameters for starting NIMV within 3 months. Association between ABG/PFT values and survival was assessed, using Cox proportional hazards models, adjusted for sex, age, time interval disease onset-ABG/PFT, site of onset and respiratory symptoms. Kaplan-Meier curves were performed and compared with log-rank test.

Results:
ABG parameters showed a significant correlation with FVC% values (for pCO2 r= -0.350; for HCO3- r= -0.345; for SBE r= -0.325; p<0.001). FVC% declined significantly when pCO2 was >42 mmHg, HCO3- >26 mmol/L, or SBE >2 mmol/L. A single HCO3- elevation (>26 mmol/L) corresponded to reduced FVC% values. The most sensitive cut-offs for starting NIMV within 3 months were: 70% for FVC%, 42 mmHg for pCO2, 26 mmol/L for HCO3- and 2 mmol/L for SBE. Risk for death/tracheostomy increased significantly when pCO2 was >42 mmHg, HCO3- >26 mmol/L, SBE >2 mmol/L and FVC% <80%. An isolated HCO3- elevation was present in 25.6% of patients who showed the same frequency of respiratory symptoms of patients with normal ABGs (p=0.273 for dyspnea; p= 0.562 for orthopnea). Patients with an isolated HCO3- increase had a reduced survival than patients with normal ABGs (0.87 years, IQR 0.73-1.02 vs 1.39 years, IQR 1.23-1.55; p<0.001).

Discussion:
Current guidelines for NIMV initiation could lead to a delayed treatment of respiratory failure in ALS patients. Despite being a minimally invasive procedure, ABG turned out to be a sensitive and inexpensive tool for monitoring respiratory function in ALS. Unlike PFTs, ABG does not require patient collaboration and is not influenced by facial weakness. Moreover, it can detect nocturnal hypoventilation [3], an asymptomatic clinical condition which is frequently underestimated and associated with an increased risk of respiratory failure.

References:
1. Andersen PM, Abrahams S, Borasio GD et al. Eur J Neurol. 2012;19:360–375.
2. Miller RG, Jackson CE, Kasarskis EJ et al. Neurology. 2009 Oct 13;73(15):1218-26.
3. Boentert M, Glatz C, Helmle C et al. J NeurolNeurosurg Psychiatry. 2018 Apr;89(4):418-424.

RNM-02: Clinical practice recommendations for respiratory management in patients with ALS, prior to initiation of ventilatory support.

Jaap Bakers[1], Esther Kruitwagen[1], Leonard van den Berg[1], Anita Beelen[1]
[1]*UMC Utrecht academic hospital, Utrecht, Netherlands*

Live Poster Session A, December 9, 2020, 5:10 PM - 5:50 PM

Background:
Regular monitoring of respiratory function can help to detect respiratory weakness at an early stage, allowing prevention of complications and timely initiation of supportive treatment. Therefore, there is a need for easy to perform methods to monitor respiratory function, including cough effectiveness. Treatment methods that can be carried out at home with minimal instruction and supervision, can contribute to alleviate symptoms, and prevent avoidable complications.

Objective:
As part of the development of a clinical practice guideline for physiotherapy in ALS, we aimed to provide evidence-based recommendations for (1) early identification of reduced respiratory dysfunction and ineffective cough in ALS and (2) non-pharmacological management of respiratory dysfunction and ineffective cough.

Methods:
Reviews were undertaken to summarize the evidence on both research questions. A guideline development group drafted the recommendations from the evidence and additional factors that are considered relevant such as values and preferences of patients and carers, the expertise of the study group members, recommendations made in other relevant guidelines, costs and resource use. Draft recommendations were reviewed by external experts, including patients with ALS and their carers.

Results from literature:
1. Orthopnoea is an important indicator of respiratory dysfunction. Vital Capacity (VC) in sitting position is not sufficiently sensitive for early detection of respiratory dysfunction. There is evidence that Maximal Inspiratory Pressure (MIP), Sniff Nasal Inspiratory Pressure (SNIP), Peak Cough Flow (PCF) and the VC measured in supine position can early identify respiratory dysfunction. The PCF is a valid test for detection of ineffective cough.
2. There is evidence for the superiority of air stacking on improving cough strength than spontaneous coughing with manual support, even in bulbar patients. Inspiratory muscle strength training (IMT) has no effect on the deterioration of inspiratory muscle strength in people with ALS. There is a very poor level of evidence for a positive effect of expiratory muscle strength training.

Recommendations:
1. Orthopnea and other symptoms of respiratory weakness, supplemented with VC in supine position, MIP or SNIP are indicative for respiratory dysfunction. To detect ineffective cough, the PCF is the first test of choice.
2. In case of ineffective cough (PCF <270L/m), airway clearance techniques (ACT), especially air stacking, should be applied structurally, or earlier if there are symptoms of ineffective cough. In contrast to ACT, respiratory strength training is not recommended for routinely appliance. Consider IMT only in motivated patients, in whom diaphragm weakness is not evident or only mild.

Conclusions:
Symptoms of respiratory weakness in combination with relevant respiratory function tests can contribute to the early detection of respiratory dysfunction and ineffective cough in ALS. In the case of respiratory dysfunction and ineffective cough, several supportive, non-pharmacological interventions are available prior to initiation of ventilatory support.

RNM-03: Correlations between measures of ALS respiratory function: is there an alternative to FVC?

Dr Deirdre Murray[1], Dr. James Rooney[7], Prof. Adriano Chio[3], Prof Ammar Al-Chalabi[4], Prof Christopher McDermott[2], Prof. Phillip Van Damme[5], Professor Leonard Van Den Berg[6], Dr Dara Meldrum[1], Prof. Orla Hardiman[1]

[1]Academic Unit of Neurology, Trinity College Dublin, , Ireland, [2]Royal Hallamshire Hospital, Sheffield, United Kingdom, [3]CRESLA, Dipartimento di Neuroscienze , Turin, Italy, [4]King's College Hospital, London, United Kingdom, [5]KU Leuven, , Belgium, [6]UMC Utrecht, , The Netherlands, [7]University Hospital, LMU Munich, , Germany

Live Poster Session A, December 9, 2020, 5:10 PM - 5:50 PM

Background:
Respiratory function is a key indicator of decline in ALS. The REVEALS study is longitudinal study in six European sites, which aims to evaluate the decline in Forced Vital Capacity (FVC), Slow Vital Capacity (SVC), Peak Cough Flow (PCF) and Sniff Nasal Inspiratory Pressure (SNIP). The Covid-19 pandemic has severely curtailed respiratory assessments, which are aerosol generating procedures (AGPs), and alternatives are being sought. The aim of this interim analysis was to analyze the patterns and rates of decline in respiratory measures and to evaluate the potential for the lowest risk measure, SNIP to be utilized as a surrogate measure to the APGs.

Methods:
This was a prospective observational study. Patients attending one of six study sites with King's Stage 2 or 3 ALS completed baseline FVC/SVC/SNIP/PCF assessments and repeated assessments at 3 monthly intervals. Data were collected March 2018 to March 2020 after which a COVID-19 related study suspension was imposed. A descriptive analysis of participants stratified by cohort was performed. Correlations between outcomes were calculated. A Bayesian multiple outcomes random effects model was constructed to investigate rates of decline across measures with time from baseline and site of onset as fixed effects, in interaction with time from baseline, gender and cohort.

Results:
In total, 270 cases and 828 assessments were included (Mean age, 65.2±15.4yrs; 33%F; 60% Kings Stage 2: 81.1% Spinal onset). FVC and SVC were the most closely correlated outcomes (0.95). FVC had a correlation of 0.75 with PCF. SNIP showed the least correlation with other metrics 0.53 (FVC), 0.54 (SVC), 0.60 (PCF). Stratified by bulbar or spinal onset, correlation between FVC and SVC was 0.95 for both groups. In spinal onset patients PCF had a correlation of 0.76 and 0.77 with FVC and SVC respectively but was poorly correlated with SNIP (0.54). SNIP was also poorly correlated with each of the other metrics in spinal patients. In bulbar patients, the correlation between SNIP and PCF rose to 0.72 and PCF's correlation to FVC and SVC was reduced (0.66 and 0.70 respectively). All four measures significantly declined over time with SNIP in the bulbar onset group having the greatest rate of decline.

Discussion:
SNIP was not well correlated with FVC and SVC, probably because it examines a different aspect of respiratory function (max pressure rather than volume). Respiratory measures declined over time, but differentially according to site of onset. Rates of decline of SNIP and PCF showed the greatest ability to differentiate between bulbar and spinal onset ALS. FVC, SVC and PCF are APGs, currently curtailed and the results suggest that SNIP cannot be used as a surrogate for FVC and SVC, but is a complementary measure, declining linearly and differentiating spinal and bulbar onset patients.

RNM-04: Development of an online toolkit to support healthcare professionals to deliver an optimal non-invasive ventilation service to people living with motor neuron disease in the UK.

Miss Lucy Musson[1], Dr Theocharis Stavroulakis[1], Dr Susan Baxter[2], Professor Paul Norman[3], Mr David O'Brien[4], Dr Mark Elliott[5], Dr Stephen Bianchi[6], Dr Georgios Kaltsakas[7], Professor Christopher McDermott[1], Dr Esther Hobson[1]

[1]Sheffield Institute For Translational Neuroscience, University Of Sheffield, Sheffield, United Kingdom, [2]School of Health and Related Research, University of Sheffield, Sheffield, United Kingdom, [3]Department of Psychology, University of Sheffield, Sheffield, United Kingdom, [4]Medical School, University of Sheffield, Sheffield, United Kingdom, [5]Leeds Teaching Hospitals NHS Trust, Leeds, United Kingdom, [6]Sheffield Teaching Hospitals NHS Foundation Trust, Sheffield, United Kingdom, [7]Lane Fox Service, Guy's and St Thomas' NHS Foundation Trust, London, United Kingdom

Live Poster Session A, December 9, 2020, 5:10 PM - 5:50 PM

Background:
Non-invasive ventilation (NIV) is a complex intervention and its success relies on the intervention components and the way in which healthcare professionals (HCPs) deliver it. There is little evidence for how best to deliver NIV to people living with motor neuron disease (MND) (plwMND) to ensure optimal use. Therefore, there is a need to explore this and to develop resources to support staff.

Objectives:
To identify the best ways to provide NIV services to plwMND in the UK and to develop an evidence-based online toolkit to support HCPs to deliver an optimal NIV service to plwMND.

Methods:
Four work packages were carried out:
1) A systematic review identified factors associated with optimal NIV use.
2) Two UK cross-sectional surveys explored the NIV services provided by MND care centres, respiratory services and community teams.
3) Focus groups with HCPs and third-party stakeholders were carried out to contextualise the earlier findings and provided ideas of how to overcome obstacles preventing best clinical practice.
4) An online toolkit was created to display recommendations to support HCPs to deliver an optimal NIV service, involving user-centred testing.

Results:
There are many factors associated with optimal NIV use but services and individual practices vary. There is a strong desire for more training for HCPs. We synthesised our findings with current published guidance in the UK and translated these into evidence-based recommendations. We developed NIV4MND: a free online toolkit designed to support HCPs who deliver NIV to plwMND to develop and deliver best practice care. The toolkit covers each point in the patient care pathway from diagnosis to end of life and describes our recommendations, practical advice and the perspectives of other HCPs, patients and carers. The toolkit will be launched in December. It includes images, videos and podcasts and helps HCPs learn through reflective exercises. We used user-centred design methods to ensure that the toolkit is easily accessible and relevant to their learning needs.

Discussion and conclusions:
This free toolkit is available to anyone wishing to enhance their NIV service. We hope this will enable HCPs to support their patients so they get the most out of NIV and ultimately improve their quality of life and survival.

Acknowledgements:
A National Institute for Health Research Programme Grant for Research for Patient Benefit (PB-PG-1216-20041) funded this research.

RNM-05: Evaluation of remote respiratory testing implemented in response to the Covid-19 pandemic

Dr Deirdre Murray[1], Mr Simon Carty[1], Mr Colm Peelo[1], Ms Rachel Tattersall[1], Dr. Dara Meldrum[1], Prof Orla Hardiman[1]
[1]Academic Unit of Neurology, Trinity College Dublin, Dublin, Ireland, [2]Beaumont Hospital, Dublin, Ireland

Live Poster Session B, December 10, 2020, 5:10 PM - 5:50 PM

Background:
Regular monitoring of respiratory function is a key aspect of care in people with Amyotrophic Lateral Sclerosis (ALS). Forced or slow vital capacity (FVC/SVC) are widely used to guide timing of interventions such as non-invasive ventilation (NIV) and in clinical trials. The Covid-19 pandemic has severely curtailed FVC, SVC and Peak Cough Flow (PCF) testing, as aerosol generating procedures are considered to pose a risk of spread of Covid-19. Previously, remote monitoring showed promise in ALS/MND (1) and therefore a remote respiratory monitoring service was rapidly developed in our centre. Appropriate patients were sent a device (MIR Spirobank Essential) by post. Tests were completed via live video link, which allowed patients to be guided through the test technique and be observed. Test scores and graphs displayed on the device were shared with the clinician via the patients' webcam. After the first 5 months a service evaluation was conducted to guide future development.

Method:
The number of respiratory tests during April to September 2020 was compared with the same period in 2019. A survey of patients and carers was completed in order to evaluate their experience of completing the tests at home.

Results:
Between 21st April 2020 and 27th September 2020, 33 patients had remote FVC testing. Five patients were tested once, while 29 had serial assessments. A total of 105 FVC tests were completed. Additionally, 67 remote PCF tests and 17 in-clinic SNIPs were completed. During the same 5-month period in 2019, 125 individuals completed respiratory tests during routine clinic appointments (FVC= 30, SVC =48, SNIP = 150, PCF = 84). During Covid-19, five patients' FVC results aided decision making on gastrostomy insertion, nine aided NIV prescription and eight cough augmentation device prescription. Six patients and 10 carers completed the telephone survey. 94% reported feeling confident doing the tests at home, although 80% needed help from a carer. All considered the remote test to be 'more efficient' than in-clinic testing and 100% indicated that they would like to continue. The mean journey time to clinic was 100±58minutes. Only 12.5% indicated that they would prefer to complete the FVC test independently without the live-video support.

Discussion:
A combination of mini-spirometers and video consultation allowed respiratory monitoring to continue with a high degree of precision. In-clinic assessments were curtailed, but remote assessment of PCF and FVC, combined with subjective assessment facilitated proactive management. Patients and carers expressed satisfaction with the service, but expressed a preference for live-video support from clinicians. The feedback supports retention and expansion of the service. Reliability and validity compared with in-clinic tests requires evaluation.

1. Geronimo A, Simmons Z. Evaluation of remote pulmonary function testing in motor neuron disease. Amyotroph Lateral Scler Frontotemporal Degener. 2019;20(5-6):348-55.

RNM-06: Factors influencing the delivery of non-invasive ventilation to people living with motor neuron disease.

Miss Lucy Musson[1], Dr Esther Hobson[1], Dr Susan Baxter[2], Professor Paul Norman[3], Mr David O'Brien[4], Dr Mark Elliott[5], Dr Stephen Bianchi[6], Dr Georgios Kaltsakas[7], Professor Christopher McDermott[1], Dr Theocharis Stavroulakis[1]
[1]Sheffield Institute For Translational Neuroscience, University Of Sheffield, Sheffield, United Kingdom, [2]School of Health and Related Research, University of Sheffield, Sheffield, United Kingdom, [3]Department of Psychology, University of Sheffield, Sheffield, United Kingdom, [4]Medical School, University of Sheffield, Sheffield, United Kingdom, [5]Leeds Teaching Hospitals NHS Trust, Leeds, United Kingdom, [6]Sheffield Teaching Hospitals NHS Foundation Trust, Sheffield, United Kingdom, [7]Lane Fox Service, Guy's and St Thomas' NHS Foundation Trust, London, United Kingdom

Live Poster Session B, December 10, 2020, 5:10 PM - 5:50 PM

Background:
The success of non-invasive ventilation (NIV) for people living with motor neuron disease (MND) requires both good patient adherence and the delivery of effective ventilation. Good delivery of NIV requires healthcare professionals (HCPs) to work together with patients and their carers to identify and address factors that can impact upon its success throughout the care pathway (i.e., before starting NIV, initiation, ongoing monitoring and optimisation, withdrawal and end-of-life).

Objectives:
To explore factors that influence the delivery of NIV including ways of overcoming barriers to good clinical practice.

Methods:
A UK cross-sectional survey of HCPs involved in the respiratory management of people living with MND and a series of focus groups with HCPs and representatives from the MND Association were conducted to explore the perceived barriers to, and enablers of, good practice. Thematic analysis was used to analyse free-text responses in the survey as well as the focus group data. The themes of influencing factors were mapped onto the COM-B model (1) which proposes that behaviour is a function of a person's capability (e.g., skills, knowledge), opportunity (e.g., resources, support) and motivation (e.g., beliefs, intentions).

Results:
For the survey, 158 responses were included in the analysis. These were from 74 NHS trusts, 21 hospices and 2 social enterprises representing HCPs across all regions of the UK. Thirty HCPs and three representatives from the MND Association charity attended five focus groups.

The strongest factors influencing practice reflected the opportunity component of the COM-B model, with the main barriers related to aspects of the service (e.g., time constraints, staff, appointments, equipment, rooms and inpatient facilities), a lack of funding for the service, and a lack of training and guidelines. Other common factors influencing practice related to the capability component, especially the need to have knowledge about NIV, bulbar function, secretion management and guidelines. Factors related to the motivation component included perceived competence to deliver good practice, self-efficacy, beliefs about guidelines, and an accurate interpretation of the guidelines and how to apply them in their practice.

Discussion and conclusions:
The findings highlight a number of ways practice may need to change in order to deliver the best possible NIV service. Optimising the provision of NIV is likely to rely on having the requisite resources (opportunity), knowledge (capability) and beliefs about capabilities (motivation). There is a clear need for adequate funding and more training to support HCPs in their practice.

Acknowledgements:
A National Institute for Health Research Programme Grant for Research for Patient Benefit (PB-PG-1216-20041) funded this research.

References:
1) Michie S, van Stralen MM, West R. The behaviour change wheel: A new method for characterising and designing behaviour change interventions. Implementation Science. 2011;6:42.

RNM-07: Mouth occlusion pressure at 100ms (P0.1) in ALS: what is its significance?

Susana Pinto[1], Michael Swash[1,2], Mamede de Carvalho[1,3]
[1]Instituto De Medicina Molecular João Lobo Antunes, Faculdade De Medicina, Universidade De Lisboa, Lisbon, Portugal, [2]Barts and The London School of Medicine and Dentistry, Queen Mary University of London, London, United Kingdom, [3]Depart Neurosciences, Hospital de Santa Maria, Centro Hospitalar Lisboa Norte, , Portugal

Live Poster Session B, December 10, 2020, 5:10 PM - 5:50 PM

Introduction:
Airway pressure in the first 100ms of an occluded inspiration (P0.1) evaluates the respiratory center activity, increasing in the presence of respiratory muscle weakness. Its increased activity can compensate for respiratory muscles weakness in amyotrophic lateral sclerosis (ALS), but possible exhaustion over disease progression can reduce its benefit. We aimed to approach this subject.

Methods:
Consecutive ALS patients with P0.1 evaluated at first visit were included. Depending on P0.1 percentile, patients were divided in three groups: G1 (<25th percentile); G2 (25th–74th percentiles); G3 (75th percentile); two subgroups were further considered: SG0 (<10th percentile); SG1 (>90th percentile). Body mass index (BMI), functional ALS rating scale and its subscores, respiratory function tests, including forced vital capacity, maximal inspiratory (MIP) and expiratory pressures, percentage of P0.1 (%P0.1), blood gas analyses, phrenic nerve motor amplitude (MeanPhrenAmpl) were compared. P0.1/MIP and %P0.1 predictors were explored by linear and multinomial logistic regression analyses. SG1 patients were further studied and those with longitudinal assessment at 6 months (Wilcoxon test assessed differences between observations) were compared with those immediately adapted to non-invasive ventilation (NIV) with Mann-Whitney U test. $p<0.05$ was considered as significant.

Results:
From the 497 patients included, 124 were in G1 and G3 each, 249 in G2, 49 in SG0 and SG1 each. G1 included more men, with higher BMI ($p<0.001$). G3 had older women, with predominant bulbar phenotype ($p<0.001$). Lower respiratory function ($p<0.05$) was present in both groups. SG0 (%P0.1<51.73%, P0.1/MIP=1.48±1.02) had more spinal-onset men ($p<0.001$) with lower MeanPhrenAmpl ($p<0.004$). SG1 (P0.1>147.12, P0.1/MIP= 7.92±4.62) predominantly included older patients (p.0.033), women (p.0.012), with lower MeanPhrenAmpl (p.0.039). The 15 SG2 patients evaluated 6 months after (6±2.8months) presented a significant P0.1 decay, as seen with the other variables and increased arterial carbon dioxide tension (PCO2) and bicarbonate ion, but P0.1/MIP was >1. Those immediately adapted to NIV had significant lower functional status and significant arterial blood repercussions.

Discussion:
ALS patients with respiratory failure can show high or low P01 values, related to phenotype. A central respiratory drive exhaustion does not seem to preclude a respiratory failure in those patients with high P0.1 values. The role of spasticity and respiratory-metabolic-renal buffering system should be further addressed.

RNM-08: Practice variation, barriers, and support needs decision-making on gastrostomy in ALS

MSc Remko van Eenennaam[1,2,3], MD, PhD Willeke Kruithof[1,2], MD Esther Kruitwagen-van Reenen[1,2], MD, PhD Leonhard van den Berg[1,3], MD, PhD Johanna Visser-Meily[1,2], PhD Anita Beelen[1,2,3]
[1]ALS Centre Netherlands, Utrecht, the Netherlands, [2]Centre of Excellence for Rehabilitation Medicine, UMC Utrecht Brain Center, University Medical Center Utrecht, and De Hoogstraat Rehabilitation, Utrecht, the Netherlands, [3]Department of Neurology, UMC Utrecht Brain Center, University Medical Center Utrecht, Utrecht, the Netherlands

Live Poster Session B, December 10, 2020, 5:10 PM - 5:50 PM

Background:
Gastrostomy is recommended for patients with ALS experiencing weight loss or swallowing difficulties. However, a lack of evidence on effectiveness and optimal timing of indication may lead to practice variation.

Objectives:
To assess practice variation, barriers, and support needs in decision-making on gastrostomy amongst rehabilitation physicians of ALS care teams in the Netherlands.

Methods:
71 rehabilitation physicians of 38 ALS care teams in the Netherlands were invited to fill in an online survey on decision-making about gastrostomy. Survey items included multiple choice and dichotomous questions (yes/no), and open questions about 1) criteria for method of placement (PEG, PRG, or other), 2) timing of indication, 3) goals, and 4) decision-making, first introduction, and involvement other HCP's. Additionally, we asked about 5) barriers and 6) support needs.
Quantitative data questions were summarized. Responses to open questions were analysed for themes.

Results:
29 physicians responded (27 ALS care teams). 1) Selection criteria for method of placement (PEG vs PRG) included physician preference (69%), contraindication PEG (choice for PRG; 59%), local availability (31%), patient preference (24%), and less complications (choice for PEG; 17%). 2). All physicians used malnutrition/weight loss, dysphagia, and prolonged and effortful meals as indicators for gastrostomy, also frequently mentioned were recurring chest infections, insufficient intake of liquids), and reduced vital capacity. 3) Physician goals of gastrostomy included optimizing nutritional status (100%), ensuring safe food-intake (72%), and reducing effort of meals (59%). 4) Half of physicians introduces the topic of gastrostomy early after diagnosis, the other half later at indication. 5) 69% of physicians experiences barriers during the decision-making process, they most frequently mention patient readiness (45%), timing of indication (31%), and organizational barriers (21%). Physicians introducing the topic of gastrostomy later at indication more often reported experiencing barriers (93%) versus those introducing the topic early (43%; $p < .01$). 6) Support needs include a more evidence-based indication (35%) and better patient education (31%).

Discussion and conclusions:
Although physicians agreed on the most important indicators of gastrostomy, there was no consensus on cutoff values and as a result they reported struggling with the timing of indication. There is unwarranted practice variation in method of placement with physician preference and local availability being more frequently mentioned over patient preference. There is also unwarranted practice variation in the late introduction of the topic of gastrostomy, which might result from physicians struggling with unclarity over the timing and, especially, patient readiness.
More evidence on optimal timing of gastrostomy placement is needed. Until then, early and regular discussion of the topic gastrostomy and better patient education may promote patient readiness and support patient choice. We are conducting a qualitative study into decision-making process on gastrostomy from the perspectives of patients, caregivers, and their physicians.

RNM-09: Hyponatremia, Impending Sign of Respiratory Failure in Amyotrophic Lateral Sclerosis

Mr Wonjae Sung[1], Mr Sanggon Lee[1], Mr Jin Seok Park[1], Mr Ki-Wook Oh[1], Prof Seung Hyun Kim[1]
[1]Department of Neurology, College of Medicine, Hanyang University, Seoul, South Korea

Live Poster Session C, December 11, 2020, 12:05 PM - 12:50 PM

Background:
Amyotrophic lateral sclerosis (ALS) is a neurodegenerative disorder causing limb paralysis, dysphagia, dysarthria, and respiratory failure. As the disease progresses, many devastating complications occur. Among various problems, respiratory failure due to weakened breathing muscles is the main cause of death in ALS. Syndrome of inappropriate antidiuretic hormone(SIADH), caused by various conditions, is frequently observed in patients with respiratory failure. ALS patients having difficulty with breathing shows low level of sodium eventually diagnosed as SIADH. In many patients, SIADH was resolved by caring respiration problem and doing conservative treatment such as water deprivation. We report a case series of ALS patients showing SIADH, which is shown as the impending sign of respiratory failure.

Object:
We aimed to detect low levels of serum sodium in patients suffering from respiratory functional decline. We investigated whether this hyponatremia would be compatible with SIADH and reviewed the clinical course focusing on the treatment and respiration.

Methods:
Retrospective analysis of patients with low levels of serum sodium (serum sodium, <135 mmol per liter) was performed and presence of respiratory problem was evaluated in selected cases. Respiratory failure was detected by symptoms (eg, orthopnea, dyspnea) and arterial blood gas data (hypercapnia, hypoxia & desaturation). Electrolyte data (serum & urine sodium levels and osmolarity), hormonal data (thyroid and adrenal gland) and clinical information were checked to diagnose SIADH. Clinical courses treating hyponatremia to normal level were reviewed.

Results:
Nineteen ALS patients showing hyponatremia accompanied with respiratory difficulty were included. Among these patients, fourteen patients were diagnosed as SIADH. Except one patient, other thirteen patients were managed to resolve respiratory failure by non-invasive mask apply or tracheostomy. Combined with water restriction and dietary salt intake, all patients were recovered from SIADH after treatment.

Conclusion:
ALS patient having problems with breathing can manifest low levels of sodium which is compatible with SIADH. As this syndrome is known to be provoked by respiratory failure, many ALS patients can be targets since this disease frequently involves breathing muscles. Treating SIADH by solving respiratory failure with conventional treatment can make sodium level to normal. Physicians should always keep in mind that managing respiration is mandatory in short-breathed ALS patient showing hyponatremia.

RNM-10: Percutaneous endoscopic gastrostomy outcomes in patients with motor neurone disease: a retrospective cohort study of Western Sydney patients managed through a coordinated service in New South Wales, Australia

Dr Paulina Suo[1], Associate Professor Nicholas G. Burgess[1], Julie Labra[2], Dr Shea Morrison[2]
[1]Department of Gastroenterology and Hepatology, Westmead Hospital , Sydney, Australia, [2]St Joseph's Hospital, Sydney , Australia

Live Poster Session C, December 11, 2020, 12:05 PM - 12:50 PM

Background and Aim:
Dysphagia and malnutrition are prevalent in patients with motor neuron disease (MND). The American Association of Neurology guidelines suggest improved survival related to percutaneous endoscopic gastrostomy (PEG) placement, however, there is insufficient data to quantitate the survival advantage or guide optimal timing of insertion.
MND patients in Western Sydney, NSW, are managed through a coordinated service at St Joseph's (SJH) and Westmead (WMH) Hospitals. This study aims to examine the 30 day and 6 month mortality and adverse procedural events amongst MND patients who undergo PEG or radiologically inserted gastrostomy (RIG).

Methods:
Consecutive patients managed through the aforementioned service between March 2004 and June 2020 were included. Patient databases at SJH and WMH were used to collect demographic details including sex, median ages at diagnosis and gastrostomy insertion and deceased date. Disease characteristics included site of disease onset (bulbar/limb/cognitive), median ALSFRS-R score at time of gastrostomy insertion, predicted rate of decline calculated at initial visit ('slow', 'steady', or 'rapid') and markers of respiratory dysfunction (carbon dioxide retention, forced vital capacity, ≤50% vs >50%, or use of non-invasive ventilation at the time of gastrostomy insertion), location of PEG/RIG insertion (WMH vs other institution), BMI at insertion, riluzole use, disease duration between symptom onset and gastrostomy insertion (<20 months vs ≥20 months). The primary outcome was peri-procedural adverse events including sedation adverse events, unplanned intubation or NIV, pain, wound infection, peritonitis and perforation). Secondary outcomes were 30 day and 6 month mortality.

Results:
Of the 375 patients included, a total of 148 patients had been referred for PEG (n = 146) or RIG (n = 2). The median age at PEG insertion was 64 years and there were 70 female participants. Median survival post PEG placement was eight months. 46 patients (31%) experienced peri-procedural adverse events related to gastrostomy placement. 10 participants (6%) died within 30 days of gastrostomy placement and 52 participants (35%) died within 6 months of gastrostomy placement.
Peri-procedural adverse events were associated with PEG insertion at a non WMH site (p = 0.036) and baseline CO2 retention (p = 0.046). The DeltaFS score was strongly associated with peri-procedural adverse events (10% 'slow', 20.8% 'steady', 64.3% 'rapid', p=0.006)
There were no factors associated with 30 day mortality. Predictors of mortality at 6 months included limb site onset of disease (p = 0.017) and BMI <25 at time of gastrostomy insertion (p = 0.026).

Conclusion:
This study highlights that the predicted rate of decline calculated at initial visit (better outcome for slow progressors compared with rapid progressors), baseline nutritional state (BMI <25) and baseline CO2 retention as well as the site of insertion (WMH vs other institution) impacted on mortality and the rate of procedural adverse events.

RNM-11: Personal experiences of gastrostomy tube in people with Motor Neurone Disease – what does the evidence tell us? A systematic review and qualitative meta-synthesis of evidence (a meta-ethnography).

Dr Dominika Lisiecka[1], Dr Aine Kearns[2], Ms Fiona Bourke[2], Mr Ian Lawson[3], Ms Cath Muir
[1]Department of Nursing, Institute of Technology Tralee, , Ireland, [2]Department of Speech & Language Therapy, University of Limerick, , Ireland, [3]North Yorkshire Disability Forum, Whitby, England

Live Poster Session C, December 11, 2020, 12:05 PM - 12:50 PM

Introduction:
Placement of a gastrostomy tube is often recommended in MND to provide nutrition and hydration that is no longer safe or possible to receive orally. The lived experiences of a gastrostomy tube in adults with neurodegenerative disease such as MND are not well understood. The aim of this qualitative meta-synthesis was to review and synthetise the available evidence to inform clinical practice and identify research gaps.

Methods:
Meta-ethnographic synthesis of qualitative studies was conducted with systematic searching of eight databases from inception to May 2020. Qualitative studies reporting personal experiences of gastrostomy tubes in adults with neurodegenerative diseases (from decision making about insertion to living with a gastrostomy tube) were identified. To ensure the highest reporting standards recently published eMERGe meta-ethnography reporting guidance were followed. The registration with International Prospective Register of Systematic Reviews (Prospero) was completed (CRD42020180700). To allow for multi-perspectival interpretation of findings patient and public involvement was incorporated during the evidence synthesis and discussion, as two authors have personal experience of gastrostomy feeding and MND.

Results:
Of 2573 unique records identified, only eight fulfilled the review criteria. All studies recruited participants with Motor Neurone Disease; no other neurodegenerative diseases were represented. Decision making emerged as the most complex and predominant theme. In comparison, the everyday experiences of living with a gastrostomy tube were not investigated in depth. A "line of argument" synthesis illustrated how people decide for / against a gastrostomy tube and what formed the essence of the lived experience after gastrostomy tube insertion. The subjectively perceived swallowing ability was an important factor influencing decision making, unlike the objective severity of dysphagia.

Conclusion and recommendations:
This review showed that the lived experience of gastrostomy feeding in MND is complex and individualised. Health care professionals should provide a holistic approach across the trajectory of the illness, exploring perceptions and values of people with MND. This will allow them to provide appropriate individualised support throughout the entire experience of gastrostomy tube (during the decision-making time as well as and post insertion). The structure of health services should ensure that both health care professionals and patients are provided with sufficient time to build rapport, discuss management options including potential benefits and problems as well as examine personal values.
There is limited research on the experience of living with a gastrostomy tube in adults with neurodegenerative disease such as MND. Future research is indicated to explore the lived experiences of gastrostomy tube and inform clinical practice. These experiences should be investigated holistically rather than being divided into pre and post insertion.

RNM-12: The E-health Application To Modify ORal Energy intake and Measure Outcomes REmotely in ALS Clinical Trial (EAT MORE2)

Mansi Sharma[1], Jane Hubbard[1], Jamie Garry[1], Dr Anne-Marie Wills[1]
[1]*Massachusetts General Hospital, Boston , United States*

Live Poster Session C, December 11, 2020, 12:05 PM - 12:50 PM

Background:
Mobile health applications have successfully achieved self-monitoring of diet and physical activity, tailored feedback, and reminders for app use. However, there have been no mHealth apps designed for ALS nutrition, despite the importance of nutrition in ALS. In our previously-completed standard-of-care controlled, single center EAT MORE clinical trial, randomization of ALS patients to nutritional counseling supported by a mHealth application resulted in a consistent trend towards increased calorie intake, quality of life, and ALSFRS-R progression. Given the positive results from the EAT MORE study, we designed and launched a freely available mHealth ALS Nutrition app, E-health Application To Modify ORal Energy intake and Measure Outcomes REmotely in ALS (EAT MORE2), for people with ALS to manage their nutrition and promote weight gain remotely. People who download the app are given the option to use the app without participating in research or consent to share their data with the EAT MORE2 study team using an in-app waiver of consent.

Objective:
The EAT MORE2 clinical trial is designed as an entirely remote study with enrollment through online advertising and social media. The aim of this abstract is to provide an interim analysis of the first 6 months of enrollment and an assessment of remote recruitment strategies.

Methods:
The EAT MORE2 nutrition app was made available on iOS or Android mobile devices starting in March 2020 and enrollment was open to individuals diagnosed with ALS around the United States. App users were recruited using electronic advertisements by ALS clinics and registries, Instagram, Twitter and Facebook ads, and virtual presentations at ALS community groups.

App users who enrolled in the optional research study are asked to enter weekly weights (self-reported), calorie intake (through an eDiet diary) and disease progression monthly using the ALSFRS-R, global health survey, and active tasks: tapping test and sustained phonation.

Results:
As of September 23, 2020, a total of 125 people have downloaded the app and 42 participants have consented to the research study. The most effective advertising methods were emailed notifications by the MGH Healey Center and the CDC ALS registry. Of the 42 participants who enrolled in the research study, 41 self-identified as having a diagnosis of ALS (1 had PLS) and 85% completed the ALSFRS-R at baseline. The app shows approximately 50% adherence over time.

Conclusions:
Within six months, we exceeded our recruitment goal of 100 individual downloads over 12 months, however due to the opt-in nature of the study design, only 1/3 of users have agreed to participate in research. While we have not fully-enrolled, our app demonstrates the feasibility of entirely-remote recruitment and in-app consenting for mHealth research studies. This is particularly relevant in the setting of COVID-19.