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PSYCHOLOGICAL AS WELL AS ILLNESS FACTORS INFLUENCE ACCEPTANCE OF NON-INVASIVE VENTILATION (NIV) AND GASTROSTOMY IN AMYOTROPHIC LATERAL SCLEROSIS (ALS): A PROSPECTIVE POPULATION STUDY

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RUNNING TITLE: Predictors of decision-making in ALS

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Abstract

Objective: To identify factors associated with acceptance of non-invasive ventilation (NIV) and gastrostomy in an exploratory population-based study.

Methods: 78 people with ALS at least 6 months post-diagnosis, and 50 caregivers, were recruited from the South East ALS Register. Baseline physical, cognitive and psychological measures were obtained. Three-monthly follow-ups monitored whether patients had accepted or refused NIV or gastrostomy. Following an intervention decision, post-decision interviews repeated baseline measures and included further intervention-specific questionnaires.

Results: Thirty-two people with ALS made at least one intervention decision and of these 10 decided about both NIV and gastrostomy. While illness factors predicted those needing to make an intervention decision, cognitive and education status, and level of executive dysfunction were associated with decision-making and acceptance or refusal of interventions. Patients' understanding of their illness, their early approach to considering interventions and carer-related factors were also associated with treatment decisions.

Conclusions: Our findings highlight the complexity of decision-making and provide a platform for designing further studies. Cognitive and psychosocial factors may assume a greater role in palliative care decisions for people with ALS than has been explicitly recognised. Future work must clarify how to ensure patients are not inadvertently being denied suitable interventions.

INTRODUCTION

At late stages of Amyotrophic Lateral Sclerosis (ALS), symptoms of sleep-disordered breathing and latterly dyspnoea are managed using non-invasive ventilation (NIV) and dysphagia by gastrostomy.[1-3] NIV improves survival and quality of life[4] without increasing caregiver burden,[5] and gastrostomy may also prolong survival.[1] Thus understanding the factors influencing uptake of these interventions is important.

Prospective studies have estimated NIV and gastrostomy uptake at between 7-36%.[6 7] Preliminary data on gastrostomy use in Scotland indicate an uptake of 11%.[8] Little has been reported about uptake in England and Wales. In the UK, NIV referral rates are variable[9 10] and this is likely to apply to other countries.

Demographic, psychological and disease-related factors have been shown to affect decision-making about NIV and gastrostomy by people with ALS.[8 11-14] Early positive views towards gastrostomy and NIV are predictive of uptake when needed.[15 16] People who derive less pleasure from oral intake or are unable to feed themselves independently are more likely to accept gastrostomy.[17] This evidence on intervention uptake has mostly been derived from non-UK retrospective studies, most of which are clinic-based and therefore prone to bias. We therefore conducted a UK population-based prospective study investigating factors influencing decision-making about gastrostomy and NIV in ALS patients and caregivers. From a medical perspective, we predicted that significant weight loss, dysphagia and reduction in pleasure in eating would lead to uptake of gastrostomy, and that severe respiratory symptoms and sleep disruption would lead to uptake of NIV, with greater illness severity being evident in decision-makers. To improve interpretation of the findings and generate hypotheses for future research, we also explored cognitive and behavioural factors influencing decision-making, and investigated the specific beliefs patients had about their illness.

MATERIALS AND METHODS

Participants

Participants with a diagnosis of possible, laboratory-supported probable, probable or definite ALS[18] were recruited from the South East ALS Register (SEALS),[19] a population register covering the South-east region of England. Patients were 6-60 months post-diagnosis and had not been referred for either NIV or gastrostomy. We considered that patients might require time to adjust to the diagnosis and gain information, so defined eligibility at 6 months post-diagnosis. Caregivers, where present, were invited to participate in the study. Ethics approval was granted by the Joint South London and Maudsley and the Institute of Psychiatry Research Ethics Committee (11/H0807/1). Written informed consent was obtained from all participants consistent with the Declaration of Helsinki.

Procedure

Participants were assessed at study baseline on physical, cognitive, psychological and health service use measures (Table 1). They were also interviewed about interventions they might be offered in the future. Responses regarding NIV and gastrostomy were coded and classified for analysis as being either ‘active’ (e.g., actively seeking information and keen to find out more, or had considered information but no decision made at that point) or ‘passive’ (e.g., no mention of the intervention or aware of it but reluctant to think about it).

Table 1 near here

Participants were followed-up at three-monthly intervals to monitor actual decision-making about NIV and gastrostomy and measure health service use (data to be reported elsewhere).

We defined a decision as identification of a clinical need for an intervention and a clear decision made by the patient to accept or refuse it. This was confirmed by direct contact with the patient/caregiver, the healthcare professionals (HCPs) involved, or from clinic letters. Patients were considered decision-makers if they made at least one intervention decision during the course of the study. Patients who made more than one decision (either about different interventions or changed their mind over a specific intervention) were categorised as accepters or refuses based on the first decision made.

When an intervention decision was made, patients and their caregivers were invited to participate in a post-decision assessment. For those agreeing to an intervention, the assessment occurred after gastrostomy placement or NIV trial. For those refusing an intervention, follow-up sessions were arranged as soon as possible after decision confirmation.

Measures

Baseline measures completed by/about the person with ALS and, where available a caregiver, are listed in Table 1.

Post-decision, in addition to repeating baseline patient and caregiver measures, where feasible, we administered the Beliefs about Medicine Questionnaire (BMQ),[34] modified to

measure patients' beliefs about NIV or gastrostomy (as applicable), including the necessity for the intervention and their concerns about it.

Statistical analysis

Only a small group of people made a decision regarding NIV and gastrostomy so the statistical analysis is largely descriptive. We quantified the association between various variables and decision outcomes and ranked the associations to provide an ordering of hypotheses for future study. Associations between continuous or categorical baseline or post-decision variables and binary decision outcomes were quantified by standardized odds ratios measuring the change in odds of the outcome per one standard deviation change in the continuous predictor, or when changing the group, of a binary predictor. Odds ratios were generated by fitting logistic regression models. To rank the importance of the various associations, p-values from a test of independence were used. (*NB: These multiple p-values cannot be interpreted as formal significance tests*.)

RESULTS

Participants

We invited a total of 178 patients identified between August 2008 - December 2011from the SEALS register to participate (see Figure 1 for study flowchart). Of these, 81 consented to participate (three were later excluded from the analysis after discovering they had made an intervention decision between invitation and baseline); of the remainder, 26 were no longer eligible (most had made an intervention decision), six died, six could not be contacted, two were not followed-up following advice from HCPs, and 57 declined participation (see Figure

1). Seventy-eight people with ALS (44% of those initially identified) were included in the final analysis.

Figure 1 near here

Sample characteristics at baseline

The sample's baseline age was 62.5 years (SD 11.8). Most participants were men (49/78) and had sporadic (65/78) and non-bulbar onset (65/78) ALS. Most were in a relationship (62/78), had no dependents (61/78) and were not employed (65/78). At baseline assessment, participants were on average 7.8 months post-diagnosis. The group had a mean WTAR-predicted premorbid IQ of 103.2 (SD 11.0), 12.4 (SD 3.4) years of education, and a slightly elevated mean post-illness onset Apathy score (FrSBe). Otherwise, there was an absence of marked psychological, cognitive or behavioural dysfunction (Table 2). When asked about interventions that might be offered in the future, 63% (49/78) of participants provided a 'passive' (versus 'active') response about gastrostomy and 71% (55/78) about NIV.

Table 2 near here

Intervention decisions made (See also Figure 2)

Participants were followed up for on average 17.2 (SD 11.8) months until death or study end. Forty-two participants died during the study, 18 of these without making a decision. Of those who died, only 2 of the 24 (8.3%) who made a decision had lived alone, compared with 6 of the 18 (33.3%) who made no decision.

In total, 32 people (41%) made at least one intervention decision. Of these, 10 decided about both interventions. Nineteen people accepted and two refused NIV. Fifteen accepted and six refused gastrostomy. Two people who refused a gastrostomy subsequently agreed to one within the study timeframe. The majority of first decisions for the nine participants with bulbar onset who made decisions concerned gastrostomy (8/9 (89%) bulbar-onset decision-makers); for those decision-makers with non-bulbar onset (23), 52% of first decisions (i.e. 12/23) concerned NIV. Mean time from diagnosis to first decision was 21.1 months (SD 15.0 months). In patients who died during the course of the study, NIV decisions were taken closer to end-of-life (mean 2.7 months (SD 2.2) prior to death) than gastrostomy decisions (mean 6.1 months (SD 4.2)) as found elsewhere[3].

Figure 2 near here

Decision-makers

Associations between baseline variables and making a decision during the course of the study were quantified by means of odds ratios. Table 3 shows the 10 most important baseline variables (ranked according to p-value) in predicting whether a decision was made. We had predicted that significant weight loss, dysphagia and reduction in pleasure in eating would lead to uptake of gastrostomy, and that severe respiratory symptoms and sleep disruption would lead to uptake of NIV. Given our limited post NIV decision refusal data, we considered the two interventions together but in the overall decision-making process we found that being more unwell at study baseline (poorer speech and swallow, lower BMI, lower ALSFRS-R) and having poorer prognostic indicators (bulbar onset) were more likely to be associated with making a decision within the study's timeframe. Importantly, some non-illness variables were also associated with decision-making. Having a higher predicted premorbid IQ, more years of education and a more 'active' approach to gastrostomy and NIV at baseline (i.e. actively seeking information and keen to find out more, or had considered

information but had made no decision at baseline compared to those with a more passive approach who made no mention of the intervention or who were reluctant to think about it) were more likely to be associated with making a decision during the study.

Table 3 near here

Factors best differentiating between intervention accepters and refusers

Of the 32 participants who made ≥1 intervention decision, 21 agreed to complete a post-decision session (Figure 2). Of the remaining 11, six died before a session could occur, three were too unwell, and two could not be contacted. Sessions occurred a median of 10.0 months (range 3.3-34.0) following study baseline and 2.2 months (range 0.5-9.0) after the decision had been made.

Variables that best differentiated between participants who accepted or refused an intervention were identified by calculating odds ratios to evaluate the association between baseline or post-decision variables and 'refusal status'. The 10 variables most strongly associated with refusal were ranked according to p-values (Table 4). The most highly associated baseline variable was employment status: being employed at study entry was associated with subsequent refusal of an intervention. Those who felt they understood their illness less well, who appeared to have a more 'active' approach to interventions and those with fewer depressive symptoms at baseline were more likely subsequently to refuse an intervention.

Table 4 near here

Family-rated executive function/behaviour at baseline and closer to the decision (rated at post-decision) may be a relevant predictor of intervention refusal. Those with greater FrSBe informant-rated Executive Dysfunction and Disinhibition pre-illness were more likely to refuse. Additionally, at baseline and post-decision, being less religious was more likely to predict intervention refusal.

Post-decision those deriving more pleasure in eating were more likely to have refused an intervention. Pleasure in eating may be related to swallowing ability, which was also ranked highly. In addition, those who reported more concerns about gastrostomy and who felt it was less necessary were more likely to have refused.

Caregivers

A total of 50/65 available caregivers (17 men, 33 women), with mean age 57.6 (sd 12.2) years and a mean of 13.7 (sd 3.3) years of education completed measures at baseline; 14/17 repeated measures at post-decision (Table 5). Most caregivers were spouses or partners (45/50) of study patients, but also included a granddaughter, friend and son. Twenty caregivers were working. Odds ratios indicated that reporting better general psychological well-being and lower caregiver strain at baseline and post-decision were more likely to be associated with caring for intervention-refusing patients. At baseline, the most important variable was palliative care outcome, with a better palliative care outcome score being associated with a greater likelihood of intervention refusal by the person with ALS. By post-decision, however, refusers were more likely to be patients with a worse caregiver-rated palliative care outcome, perhaps resulting from refusing the intervention.

Table 5 near here

DISCUSSION

This is the first prospective UK population-based study of the complex factors that influence how people with ALS make decisions about NIV and gastrostomy. Twenty-seven percent of our sample made a decision about each intervention during the study. In terms of intervention uptake, 19% accepted gastrostomy, broadly consistent with previous reports (e.g.[6 7 16]). NIV use, at 24%, was slightly higher than previously reported,[9] possibly reflecting an increase in UK NIV referrals[10] or recently published NIV guidelines for use in ALS.[2]

This was an exploratory study to provide insights into the predictable complexity of these processes and thus lay the basis for future analyses with more participants and greater power to dissect factors implicated in this study. We found support for the expected importance of some illness-related physical factors in predicting the need for or uptake of interventions. However, the demonstration of the likely relevance of a range of psychological factors argues against the uncritical application of simplistic solutions (e.g., uncritical care pathway algorithms) for HCPs involved in advising people with ALS on choices relating to NIV and gastrostomy.

Predictors of decision-making

Most factors predicting decision-making were illness variables, with those more unwell at baseline more likely to make a decision about an intervention during the study. However, those with a higher predicted IQ, more years of education and a more active attitude towards interventions were also more likely to be decision-makers. Such patients may be more assertive in their consultations with HCPs or more active in their information-seeking about services and available interventions. Of concern, and important for future studies, those with lower IQ, fewer years of education and a more passive approach to interventions may be

disadvantaged in having less opportunity to make informed decisions. Future research might usefully explore whether systematic approaches to decision-making via the use, for example, of decision aids (e.g. [35-37]) might enable professionals working with people with ALS to be more systematic in their approach with patients for whom decisions may become necessary, in order to reduce the potential bias in which patients find themselves making intervention-related decisions.

Accepting vs. refusing interventions

Baseline vital capacity and subsequent dysphagia and loss of pleasure in eating were associated with uptake of gastrostomy, although weight loss (BMI) was not strongly associated with decision status (Table 4). Potentially linked to these illness variables is the finding that baseline employment status predicted refusal. These findings may all relate to the participants' perception of the physical need for the intervention, are consistent with previous studies,[38] and highlight the importance of considering timing when offering the intervention. We were unable to evaluate whether severe respiratory symptoms and sleep disruption would lead to uptake of NIV, due to the small number of NIV refusers providing data for analysis.

Although FrSBe-rated Apathy was the only clinically elevated baseline psychological-symptom score, our results suggested greater family-rated pre-illness onset Executive Dysfunction and Disinhibition, and post-onset Executive Dysfunction scores were more strongly associated with intervention refusal. Such individuals may be less able to weigh up information, leading to a more concrete view of the intervention. Signs of executive dysfunction/disinhibition in the patient may also influence HCPs' discussion of the intervention, possibly providing fewer details or emphasising difficulties with compliance. Elsewhere, an examination of epilepsy treatments[396] found that clinicians' presentation of options varied, subtly communicating their preferred option. Thus HCPs may communicate

their preferences to the patient, and may differ in the options presented depending on patient characteristics. The potential for inequitable provision of healthcare as an explanation for poorer prognosis in ALS patients with executive dysfunction has been raised elsewhere.[25]

Other psychological characteristics were also associated with intervention acceptance or refusal. Patients with fewer depressive symptoms at baseline were more likely subsequently to refuse an intervention, suggesting that refusing an intervention was not simply related to the presence of earlier depressive symptoms being associated with a longer term wish not to prolong life by accepting an intervention. Indeed, refusers were those with a more active attitude to interventions, suggesting a more complex decision-making process than one that was simply mood related. Prior to needing either intervention, refusers were more likely to express a desire to seek out information, to be actively thinking about the intervention, or to have already decided what they would do if they needed the intervention in future. Refusers may, therefore, have had a more active approach to their disease management, perhaps reducing their tendency to comply unquestioningly with HCPs' recommendations. Our finding that participants claiming to have no religious belief and to never attend religious services were more likely to refuse an intervention was consistent with previous findings whereby those opting to use NIV were more religious.[14]

In addition, those who felt they understood their illness less well were more likely to refuse an intervention, suggesting it is important for people to feel they understand the illness (and related interventions) for them to accept an intervention. Those less concerned about and believing they had more control over their illness were also more likely to refuse. Furthermore, those who viewed gastrostomy as being less necessary and those with more concerns about gastrostomy were more likely to refuse interventions.

Caregiver variables

Previous studies have not identified caregiver factors in decision-making.[7 40] Our finding that caregivers with better general psychological well-being and lower strain at baseline and at the time of a decision were somewhat more likely to care for patients who refused an intervention, suggests that caregiver characteristics require consideration. We have previously also shown that the number of other caregiver dependents is predictive of caregivers' psychological well-being[41] and future research relating to intervention decisions might usefully investigate caregivers' wider social context. The presence of caregivers may itself be important since, of those dying during the study, 91.67% of those making a decision had lived with a caregiver, compared with 66.6% who died without making a decision. Our findings in relation to the carer-rated PCOS focused on total PCOS scores; future research, however, could examine whether components of the PCOS (e.g. carer ratings of items relating to patients' other symptoms, anxiety over the illness on the part of the patient or their family, reduced feelings of well-being` or the lack of dealing with practical affairs) contributed most to PCOS-related findings.

Limitations

Potential limitations of this study may affect the interpretation of our findings. Follow-up data were restricted because not all decision-makers participated in a post-decision interview. No follow-up data were available on those refusing NIV, since NIV decisions were taken closer to death than gastrostomy-related decisions, with those refusing likely to be more unwell. During recruitment, NICE guidelines for NIV were published; although we cannot determine

whether this influenced the rates of uptake of NIV by our sample, the majority of those dying without making a treatment decision did so prior to mid-2010, so the NIV guidelines may have changed the practice of offering NIV during the study. It is also possible those completing a post-decision session were not representative of all decision-makers.

Analysis was based on grouping decisions about NIV and gastrostomy together. It is possible that decision-making about gastrostomy may be very different from decision-making about NIV. For example, gastrostomy could be perceived as a more physically-invasive procedure and less reversible than a decision to trial NIV would be.

Conclusions

This study provides a unique, prospective report on the actual decision-making of a population-based cohort of ALS patients and their caregivers in the UK. Our findings suggest that, rather than simply being limited to illness variables, IQ, education, employment status, religiosity, executive dysfunction and disinhibition, perceptions or beliefs about illness and interventions, and attitude to interventions or disease management from early in the disease may influence choices around NIV and gastrostomy. Our results therefore i) provide a potential broad framework to enhance understanding of the complexity of factors to be taken into account by HCPs advising people with ALS on choices relating to NIV and gastrostomy; ii) warn against simplistic algorithms to guide HCPs in assessing patient choices; and iii) argue for a ‘whole person’ biopsychosocial/spiritual understanding of patients’ decision-making, especially at the level of the individual patient with ALS.

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Table 1. Baseline measures completed by/about the person with ALS and their caregiver.

People with ALS	Baseline	Measure
	Illness variables	ALS Functional Rating Scale- Revised (ALSFRS-R)[20] ALS Severity Scale, speech and swallowing subscales.[21] Single item, visual analogue scale to measure pleasure in eating (rating out of 10).[17] Vital capacity (% predicted). Height and weight (to calculate Body Mass Index - BMI).
	Cognitive / behavioural measures	Wechsler Test of Adult Reading (WTAR)[22] to estimate premorbid IQ. Addenbrooke's Cognitive Examination Revised (ACE-R)[23] to screen for early cognitive change. Frontal Systems Behavior Scale (FrSBe, Family-rated),[24] used in other studies of ALS (see [25]) to assess everyday behavioural changes in the person with ALS in terms of apathy, executive dysfunction and disinhibition.
	Psychological Measures	Purpose in Life Scale (PILS)[26] to assess patients' views concerning the purpose they saw in their lives, attitudes to death and their freedom to make choices. Beck Depression Inventory-FastScreen (BDI)[27]: to screen for major depressive disorders and designed to measure psychological/non-somatic symptoms only. Brief Illness Perceptions Questionnaire (B-IPQ),[28] a brief measure of illness perceptions in those who are very ill or elderly. Idler religiosity questionnaire[29]: a measure of public and private religiousness. Palliative Care Outcome Scale (POS)[30]: a 10-item scale which addressed issues relevant to palliative care.
Carer	Health status	General Physical Health Rating (GPHR)[31]: a self-rating from 0 to 4 of the caregiver's own physical health.
	Psychological status	Caregiver Strain Index (CSI)[32]: to measure strain experienced by informal caregivers. General Health Questionnaire (GHQ)[33]: to assess caregivers' general psychological well-being
	Proxy rating of palliative care	Caregiver version of Palliative Care Outcome Scale (PCOS)[30]: to rate the problems for the patient and caregiver from the caregiver's perspective.

Table 2: Characteristics of patients at baseline and post-decision

	Baseline measures for whole sample (max N =78)		Baseline measures for subsequent decision-makers (max N=21)		Measures obtained post-decision (max N=21)	
	N	Descriptive	N	Descriptive	N	Descriptive
ALSFRS-R total (/48) # Mean (SD)	78	35.1 (7.5)	21	34.4 (7.7)	21	22.1 (8.0)
ALS Severity Scale – Speech (1-10) # Median (min-max)	78	8.80 (2-10)	21	7.0 (2-10)	21	6.0 (1-10)
ALS Severity Scale – Swallow (1-10) # Median (min-max)	78	10.0 (6-10)	21	8.0 (6-10)	21	5.0 (2-10)
Body Mass Index (BMI) Median (min-max)	76	24.4 (15-40)	21	23.5 (15-33)	20	21.7 (12-40)
Vital capacity (% predicted) Mean (SD)	68	73.8 (22.6)	18	74.1 (19.9)	12	52.8 (16.0)
Pleasure in eating (/10) # Median (min-max)	78	9.0 (0.5-10)	21	7.5 (0.5-10)	17/17	6.0 (0-10)
ACE-R total (/100) # Mean (SD)	53	90.7 (9.2)	14	91.9 (9.2)	3	86.7 (6.8)
ACE-R verbal fluency (/14) # Mean (SD)	71	10.9 (2.6)	20	11.6 (2.5)	6	10.5 (3.7)
ACE-R language (/26) # Mean (SD)	60	24.5 (2.2)	17	25.3 (1.2)	3	25.7 (0.6)
ACE-R memory (/26) # Mean (SD)	74	23.1 (3.5)	20	23.6 (3.6)	8	21.4 (2.9)
ACE-R visual-spatial (/16) # Mean (SD)	59	14.9 (1.8)	15	14.7 (2.4)	5	15.4 (0.6)
ACE-R attention-orientation (/18) # Mean (SD)	76	17.4 (1.4)	20	17.6 (0.7)	8	17.4 (0.9)
FrSBe Apathy pre-illness* Median (min-max)	48	48.0 (32-87)	17	49.0 (37-67)	13	53.0 (35-73)
FrSBe Disinhibition pre-illness * Median (min-	48	48.5 (34-103)	17	45.0 (37-66)	13	45.0 (39-76)

max)						
FrSBe Executive dysfunction pre-illness* Mean (SD)	47	50.1 (10.1)	16	48.6 (6.9)	13	49.5 (10.5)
FrSBe Apathy post-illness* Mean (SD)	48	65.7 (16.6)	17	65.8 (18.3)	13	78.5 (14.5)
FrSBe Disinhibition post-illness* Median (min-max)	48	54.0 (34-99)	17	51.0 (38-66)	13	53.0 (39-86)
FrSBe Executive dysfunction post-illness* Mean (SD)	47	53.7 (11.4)	16	53.1 (8.8)	13	53.4 (8.9)
Purpose in Life Scale (20-140) # Mean (SD)	74	101.1 (16.8)	21	107.8 (15.9)	21	94.9 (17.9)
Beck Depression Inventory (/21)* Median (min-max)	74	3.0 (0-15)	21	2.0 (0-14)	21	3.5 (0-13)
Palliative Care Outcome Scale (/40)* Mean (SD)	74	10.5 (6.5)	21	10.1 (6.4)	20	11.5 (5.5)
BIPQ: Effect on life (/10)* Mean (SD)	73	7.0 (2.8)	20	6.7 (3.2)	21	8.1 (2.8)
BIPQ: Timeline (/10)* Mean (SD)	73	9.3 (1.7)	20	8.8 (2.2)	21	8.9 (2.8)
BIPQ: Control over illness (/10)* Median (min-max)	73	8.0 (0-10)	20	7.5 (0-10)	20	9.5 (4-10)
BIPQ: Help from treatment (/10)* Mean (SD)	72	6.7 (2.9)	20	6.4 (2.8)	20	6.9 (2.7)
BIPQ: Symptoms (/10)* Mean (SD)	72	6.4 (2.5)	20	6.2 (2.4)	20	7.6 (2.5)
BIPQ: Concern about illness(/10)* Mean (SD)	73	7.1 (3.1)	20	6.5 (3.4)	20	7.5 (3.3)
BIPQ: Understand illness (/10)* Median (min-max)	74	2.0 (0-8)	21	2.0 (0-8)	21	1.0 (0-10)
BIPQ: Emotion effects (/10)* Mean (SD)	74	6.3 (2.8)	21	5.9 (2.7)	21	6.7 (2.7)
Idler: Has a religion:	51/2		12/9		14/	

Yes/No	5				7	
Idler: Attend religious service: ≥1x Yr/Never	38/36		11/10		12/9	
Idler: Strength/comfort: Yes/No	44/32		11/10		10/11	
BMQ-Gastrostomy: Necessity (5-25)* Mean (SD)	N/A		N/A		18	17.3 (4.4)
BMQ-Gastrostomy: Concerns (5-25)* Mean (SD)	N/A		N/A		18	13.6 (4.1)
BMQ-Gastrostomy: Differential (-20-20) Median (min-max)	N/A		N/A		18	3.5 (-7-13)
BMQ-NIV: Necessity (5-25)* Mean (SD)	N/A		N/A		6	16.7 (4.3)
BMQ-NIV: Concerns (5-25)* Mean (SD)	N/A		N/A		6	14.0 (4.2)
BMQ-NIV: Differential (-20-20) Median (min-max)	N/A		N/A		6	4.0 (-6-8)
BMQ-General: Harm (4-20)* Mean (SD)	N/A		N/A		21	8.6 (2.6)
BMQ-General: Overuse (3-15)* Mean (SD)	N/A		N/A		21	8.8 (2.9)

ALSFRS-R=ALS Functional Rating Scale Revised; ACE-R= Addenbrooke's Cognitive Examination-Revised; FrSBe=Frontal Systems Behavior Scale Family-rated (T scores reported: 60-64=borderline impairment; 65+=clinically elevated); Purpose in Life Scale (<92 Lack of purpose; 92-112 Indefinite; >112 Definite purpose); BIPQ=Brief Illness Perceptions Questionnaire; BMQ-Gastrostomy/NIV/General=Belief in Medicines Questionnaire regarding gastrostomy/NIV/medicines in general; *High scores indicate more problems/ greater dysfunction or stronger agreement with statement; #High scores indicate fewer problems/less dysfunction.

Table 3: Relationship between variables at baseline patient data with decision-making status – Odds ratios ranked according to p-value

Predictor measure	Made at least one decision n=32			Made no decision N=46			OR	95% CI	P value	Adjusted OR*
	N	Mean (sd)	Median (min-max)	N	Mean (sd)	Median (min-max)				
ALS-SS Speech (1-10)	32	8.06 (2.15)	9.0 (2-10)	46	9.26 (1.32)	10.00 (4-10)	0.48	0.28-0.82	0.007	
ALS-SS Swallow (1-10)	32	8.42 (1.48)	8.25 (6-10)	46	9.24 (1.12)	10 (6-10)	0.53	0.32-0.86	0.010	
WTAR-FSIQ	28	107.29 (9.61)	109.0 (81-117)	43	100.51 (11.12)	103.0 (75-117)	2.07	1.15-3.73	0.015	2.42
BL attitude to gastrostomy (Passive /active)	15 / 17			34 / 12			0.31	0.12-0.81	0.017	0.43
BL attitude to NIV (Passive / Active)	18 / 14			37 / 9			0.31	0.11-0.86	0.024	0.34
Years of education	32	13.53 (4.13)	13.00 (3-23)	46	11.67 (2.66)	11.00 (6-20)	1.80	1.08-2.98	0.024	
Region of onset (bulbar/other)	9/ 23			4/4 2			0.24	0.07-0.88	0.031	
BMI	32	23.80 (4.03)	23.40 (14.9-33.0)	44	25.74 (4.70)	24.60 (17.4-40.2)	0.62	0.37-1.04	0.069	
ACE-R attention-orientation (/18)	30	16.97 (1.99)	18.0 (9-18)	46	17.61 (0.75)	18.00 (15-18)	0.58	0.32-1.07	0.081	
ALSFRS-R total (/48)	32	33.38 (7.50)	35.50 (18-44)	46	36.35 (7.38)	37.50 (13-47)	0.67	0.42-1.07	0.090	

ALS-SS=ALS Severity Scale; WTAR-FSIQ=Full-Scale IQ predicted by Wechsler Test of Adult Reading; BL=baseline; BMI=Body Mass Index; ACE-R=Addenbrooke's Cognitive Examination-Revised; ALSFRS-R=ALS Functional Rating Scale. *Adjusted for illness variables (ALS-SS Swallow, ALSFRS-R, BMI) Adjusted odds ratios were calculated to evaluate the association between WTAR-IQ or baseline attitude to NIV and gastrostomy and making a decision, after controlling for the effects of illness variables (BMI, ALSSS Swallow, ALSFRS-R). Holding the effect of illness variables constant increased the predictive effect of WTAR-IQ, but decreased slightly the predictive effects of baseline attitude to gastrostomy and NIV.

Table 4: Relationship between variables at baseline and post-decision and refusal status – Odds ratios ranked according to p-value

Baseline measures									
Predictor measure	Accepted intervention n=24			Refused intervention N=8			OR	95% CI	P value
	N	Mean (sd)	Median (Min-max)	N	Mean (sd)	Median (Min-max)			
Employed y/n	2/ 22			4/4			0.09	0.01-0.67	0.02
BIPQ: Understand illness (/10)	23	1.87 (2.56)	0 (0-8)	7	4.14 (2.61)	5.00 (1-8)	2.27	0.95-5.41	0.07
Vital capacity (% predicted)	20	75.19 (19.56)	80.17 (38.8-108.3)	7	58.54 (18.20)	56.37 (30.4-82.0)	0.36	0.12-1.10	0.07
FrSBe: Executive Dysfunction pre-illness	16	48.44 (8.64)	48.50 (37-70)	5	57.40 (13.45)	53.00 (45-79)	2.39	0.79-7.20	0.12
FrSBe: Disinhibition pre-illness	17	46.71 (8.17)	45.00 (36-66)	5	56.20 (15.99)	48.00 (45-83)	3.27	0.71-14.97	0.13
FrSBe: Executive dysfunction post-illness	16	52.19 (11.13)	52.50 (37-79)	5	61.80 (11.63)	64.00 (50-79)	2.41	0.75-7.48	0.13
Idler: Attend religious service (≥ 1x Yr / never)	14 / 9			2/5			3.89	0.62-24.52	0.15
BL Attitude to treatment: Passive/ active*	13 / 11			2/6			0.28	0.05-1.69	0.17
ACE-R memory (/26)	22	21.73 (5.33)	24.00 (9-26)	7	25.00 (1.41)	25.00 (22-26)	2.73	0.62-12.02	0.19
BDI (/21)	24	4.17 (3.86)	4.00 (0-14)	7	2.00 (2.58)	1.00 (0-7)	0.44	0.13-1.48	0.19

Post decision Max N=21									
Predictor measure	Accepted intervention			Refused intervention			OR	95% CI	P value
	N	Mean (sd)	Median (Min-max)	N	Mean (sd)	Median (Min-max)			
Idler: Define as having a religion y/n	13/3			1/4			17.33	1.39-216.60	0.03
BMQ-specific (Gast): Necessity (5-25)	13	19.08 (2.56)	20.00 (14-23)	5	12.80 (5.22)	13.00 (7-21)	0.13	0.02-0.86	0.03
BMQ-specific (Gast): Concerns (/10)	13	12.38 (3.86)	11.00 (7-20)	5	16.60 (3.21)	18.00 (11-19)	3.68	0.90-15.01	0.07
Idler: Attend religious service (≥ 1x Yr / never)	11/5			1/4			8.80	0.77-100.26	0.08
FrSBe: Executive dysfunction pre-illness	10	46.20 (7.22)	46.00 (36-60)	3	60.33 (13.87)	64.00 (45-72)	5.02	0.81-31.10	0.08
Pleasure in eating (/10)	12	4.58 (3.43)	5.00 (0-10)	5	8.00 (1.87)	9.00 (6-10)	3.14	0.84-11.65	0.09
BIPQ: Control over illness (/10)	15	8.87 (1.46)	10.00 (6-10)	5	7.40 (2.79)	8.00 (4-10)	0.26	0.04-1.64	0.15
BIPQ: Concern about illness (/10)	15	8.07 (2.84)	10.00 (1-10)	5	5.60 (4.03)	7.00 (0-10)	0.48	0.18-1.32	0.16
FrSBe: Disinhibition pre-illness	10	47.70 (11.50)	43.50 (39-76)	3	59.00 (3.61)	60.00 (55-62)	4.20	0.56-31.25	0.16
ALS-SS Swallow (/10)	16	4.69 (2.58)	3.00 (2-10)	5	6.60 (2.30)	7.00 (4-10)	1.50	0.85-2.64	0.16

BIPQ=Brief Illness Perceptions Questionnaire; FrSBe=Frontal Systems Behavior Scale; ACE-R memory=Addenbrooke's Cognitive Examination-Revised memory subscale; BDI=Beck Depression Inventory-FastScreen; BMQ-specific (Gastrostomy): Necessity=Belief in Medicines Questionnaire, necessity of gastrostomy; BMQ-specific (Gastrostomy): Concerns=Belief in Medicines Questionnaire,

concerns about gastrostomy; BIPQ=Brief Illness Perceptions Questionnaire; ALS-SS=ALS Severity Scale; *Baseline attitude to intervention that first decision was about.

Table 5: Caregiver data at baseline and post-decision, including odds ratios for refusal status of patients

Baseline N=50									
	Accepted intervention			Refused intervention			OR	95% CI	P value
Predictor measure*	N	Mean (sd)	Median (min-max)	N	Mean (sd)	Median (min-max)			
1. PCOS (/40)	17	13.29 (6.84)	14.00 (2-25)	5	8.50 (6.76)	6.00 (3-18)	0.5	0.2-1.4	0.2
2. GHQ (/36)	13	14.20 (4.90)	13.00 (8-23)	3	12.67 (7.10)	14.00 (5-19)	0.7	0.2-2.9	0.6
3. CSI (/13)	17	6.24 (3.51)	6.00 (1-13)	5	5.60 (4.72)	4.00 (1-12)	0.8	0.3-2.3	0.7
4. GPHR (0-4)	17	1.06 (0.75)	1.00 (0-2)	5	1.00 (1.00)	1.00 (0-2)	0.9	0.3-2.3	0.9
Post decision N=14									
Predictor measure*	Accepted intervention			Refused intervention			OR	95% CI	P value
	N	Mean (sd)	Median (min-max)	N	Mean (sd)	Median (min-max)			
GPHR (0-4)	11	1.45 (0.82)	2.00 (0-2)	3	0.33 (0.58)	0.00 (0-1)	0.2	0.0-1.3	0.1
CSI (/13)	11	8.73 (2.69)	10.00 (5-13)	3	5.33 (3.06)	6.00 (2-8)	0.2	0.0-1.7	0.1
GHQ (/36)	9	17.00 (3.71)	18.00 (10-22)	2	13.00 (7.07)	13.00 (8-18)	0.3	0.0 - 2.3	0.3
PCOS (/40)	11	15.62 (5.72)	16.00 (4-24)	3	17.19 (4.24)	15.56 (14-22)	1.5	0.3-7.9	0.6

PCOS: Palliative Care Outcome Scale; GHQ: General Health Questionnaire; CSI: Caregiver Strain Index; GPHR: General Physical Health Rating; *High scores indicate greater strain, poorer health or well-being, or more problems.

Figure 1: Flowchart showing recruitment pathway

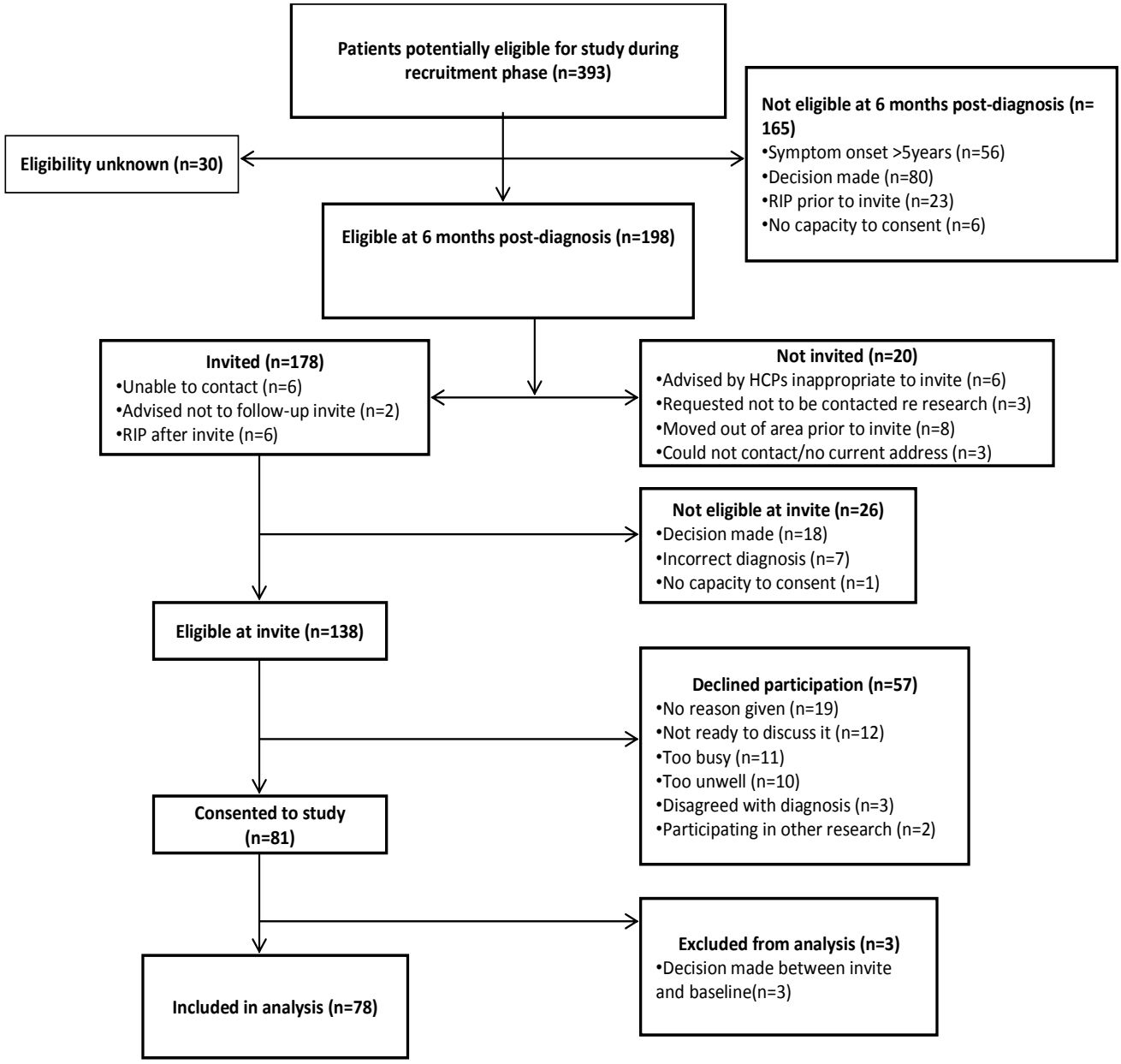
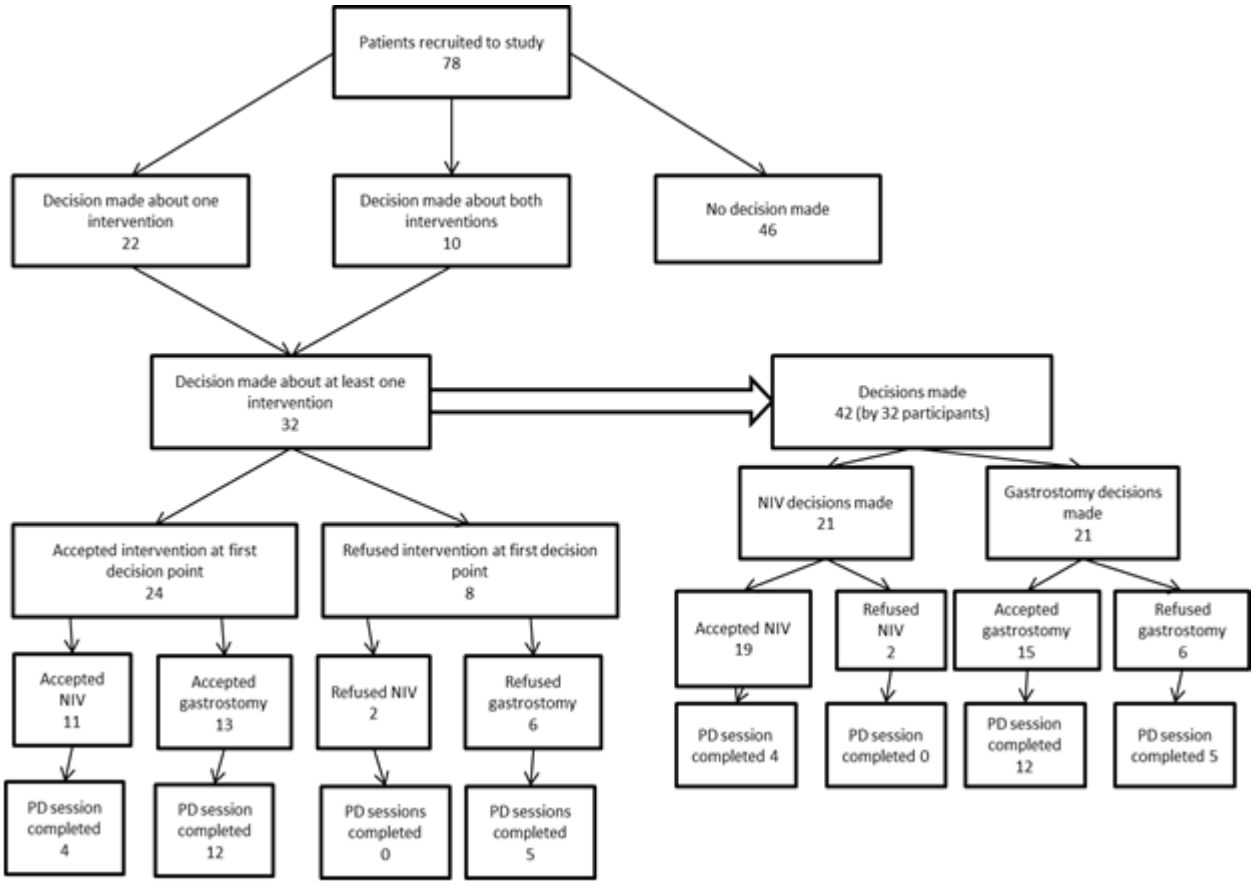


Figure 2: Flow diagram showing decisions made by participants during the study



First decision made (could have been about NIV or gastrostomy)

All decisions made