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Evaluating a novel cervical orthosis to support neck weakness in patients with amyotrophic lateral sclerosis/motor neurone disease

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Abstract

Objective

Current practice and guidelines recommend the use of neck orthoses for people with amyotrophic lateral sclerosis (ALS) to compensate for neck weakness and to provide surrogate neck control. However, available options are frequently described by patients as restrictive and unsuitable and there was a need for a new device that addressed the needs of people with ALS.

Methods

This project utilised a co-design process to develop a new neck orthosis that was more flexible yet supportive. Following development of a prototype device, an extended evaluation phase of work was undertaken. The evaluation methods included a questionnaire and interviews with patients and carers.

Results

Twenty six patients were recruited to the study, with 20 of these completing all phases of data collection. Participants described the impact of neck weakness on their life and limitations of existing supports. Evaluation of the new orthosis indicated positive views regarding the range of movement, support provided, flexibility of use, and elements such as the appearance and comfort. Feedback enabled modification of design to achieve better optimal fit.

Conclusions

The results of this evaluation highlight the value of this alternative option for people with ALS, and potentially other patient groups who require a neck orthosis.

Key words: amyotrophic lateral sclerosis; cervical orthosis; co-design; head drop; motor neurone disease; neck orthosis; neck support; neck weakness; collar; user centred design

Background

Severe weakness of the neck extensor muscles has been described in various neuromuscular disorders including myasthenia gravis, spinal muscular atrophy and amyotrophic lateral sclerosis (ALS) / motor neurone disease (1). People with ALS who exhibit neck weakness can find it difficult to ambulate, and may suffer pain due to the flexed neck position (2). Current practice and guidelines for care of patients with ALS recommend the use of neck orthoses to compensate for patient neck weakness and to provide surrogate neck control (3 4). A review of orthoses available, described discomfort and restriction for patients, and concluded that there was a need for a new device that fully addressed the needs of people with ALS (5).

Methods

D*esign of the orthosis*

The initial stages of this project utilised a co-design process with users, health care professionals and designers to develop a new prototype neck orthosis. Details of the design process are reported elsewhere (6). The orthosis (called the Sheffield Support Snood [SSS]) consists of a snood-like base made of stretchable fabric which follows the contours of the neck, upper thorax and skull. The outer surface of the snood is covered in the “loop” of a hook and loop material which then allows various support structures (covered in Velcro – the “hook”) to be attached to the SSS (see Figure 1 and Figure 2). The whole surface is loop material, allowing supports to be placed in any position. This enables the degree of support to be varied when needed, either during specific tasks in a day, or as support requirements change with disease progression.

Insert Figure 1 and Figure 2 around here

Following production and CE marking of the device, an evaluation phase was undertaken over a 6 month period using a mixed method approach. This evaluation aimed to examine the perceptions and experiences of patients with ALS and their carers of the SSS compared to other neck collars that they had tried, and to underpin further refinement of the device.

Participants

Patients were recruited consecutively if they met the following eligibility criteria: i) A diagnosis of ALS/MND; ii) Symptoms of neck weakness with an MRC muscle score of 4 or less in at least one neck muscle; iii) Patients who had previously tried or were at the stage of starting to trial a neck orthosis; and iv) Aged over 16 and willing and able to participate in the study. Potential participants were predominantly recruited from neuromuscular clinics run at the host site, together with individuals referred to the study from clinics at six other specialist centres around England. Patients were excluded if they had other co-morbidity that would have affected their ability to benefit from a neck support; if they had no carer present who would be able to fit or remove the SSS; or if they had clinically overt dementia. Participants were unable to complete the study if, during the fitting appointment if was found that a suitable fit for the SSS could not be achieved. As the study was primarily explorative a formal sample size was not calculated. However, a target sample of 20 patients was planned in order to achieve saturation of the qualitative data (7).

Qualitative data

Qualitative interview data were collected either in the clinical research unit at the local hospital, or at the patients' home at two time points: before fitting of the snood; and again one month later. Where initial interviews were carried out at the hospital site, they took place one to two hours prior to fitting of the SSS. In-home initial interviews were carried out one to three days prior to fitting. The fitting of the SSS was carried out by two members of the team (a physiotherapist experienced in working this patient group, and a product designer). A carer was requested to be present at the fitting appointment.

Data collection was carried out by a researcher experienced in interviewing people with ALS. Ethical approval for the study was granted by the local NHS Research Ethics Committee, and ethical and humane principles of research were followed at all times. At the first session the researcher took written informed consent for participation and publication, administered the questionnaire, and carried out the semi-structured interviews. Patients communicated either verbally or via communication aids/writing, with carers present in most cases during the interview. Interviews typically lasted 45 minutes to one hour.

Quantitative data

A neck support questionnaire was developed to collect quantitative data in the evaluation. The questionnaire comprises an 11 item tool assessing neck support use, comfort and satisfaction. It uses a seven point Likert rating scale for each item of “strongly agree” to “strongly disagree” (see Appendix 1). The semi-structured interviews were based on a topic guide that had been developed based on previous expertise of the research team (see Appendix 2).

Quantitative data were analysed using R version 3.1.2. Descriptive statistics (means, medians, modes, percentiles were calculated) and the Wilcoxon test was used to explore any differences between patient ratings of their existing collar and the SSS. Qualitative interviews were recorded and transcribed. The interview transcripts were read line-by-line with labels (or codes) assigned to views and perceptions voiced by participants (7). Data within each of these codes were then re-examined to identify and bring together similar themes and subthemes across the set of interviews. Systematic coding and retrieval of data was supported by Atlas Ti software.

Results

Twenty six patients were recruited to the study, with 20 of these completing all phases of data collection. Three potential participants could not be successfully fitted with the SSS, and therefore were unable to complete the study. In addition, three participants died in the

interval between fitting and follow up. The final sample comprised 12 females and 14 males, with nine rating their neck weakness as severe and 17 rating their neck weakness as mild-moderate at the time of initial fitting. The data provide insights regarding the impact of neck weakness on patient quality of life and limitations of existing neck supports, and provide an evaluation of the new orthosis.

i) Impact of neck weakness

Participants described a number of ways in which neck weakness adversely impacted on their lives. These included: problems with eating and saliva for example, *"I'm having to try and get him a bit upright by sort of gently pushing on his head, that causes a problem with meal times" Participant (P) 3;* the effect on social interaction due to a low eye level, *"If anybody talks or wants to talk to her she can't see who it is" P7,* the challenge in getting around, "s*he won't go out a lot because she can't support her head" P7;* and discomfort and neck pain, "*it's looking down that brings on my neck pain" P2.*

ii) Limitations of existing neck supports

Patients had tried or were using a range of existing devices including; a foam collar (seven individuals; the Head Master (four patients), Beanie collar (two participants) and a single individual reported using either the Hereford, Oxford, Vista Aspen, Miami, Stro II, or Traction Fixer. Two patients used travel neck cushions. Daily usage of supports varied from none (six patients) to 1-4 hours, and one person who used a Beanie collar for much longer (10 hours).

During the interviews participants described their views of collars that they were using or had tried in the past. Their comments were grouped into five main themes: difficulty in fitting, for example, *"they're not easy to fit on your own" P5*; lack of physical support, *"I didn't find that it gave me any support" P4*; being overly restrictive, *"it really was like body armour from medieval times" P1*; feeling uncomfortable, "*that put too much pressure on her collar bone*

and her chin was slipping off the chin guard" P18; or being unsuitable, "*It is as if I have to explain I haven't just suffered a car crash" P11.*

iii) Evaluation of the orthosis

The questionnaire evaluation comprised 11 questions, with responses recorded on the Likert scale: strongly agree, agree, somewhat agree, neither agree or disagree, disagree somewhat, disagree, and strongly disagree. We obtained evaluations of a previously worn collar from 24 participants, and an evaluation of the SSS from 20 participants. Nineteen participants evaluated both a previously worn collar and the SSS. There is some missing data for questions that related to eating and drinking (questions 2, 3, 4) since some participants had either not tried their collar during eating and drinking, or had a gastrostomy and were not fed orally. See Table 1.

Insert Table 1 around here

We compared participant ratings of their existing collar to their ratings of the SSS. A descriptive summary of these data in the form of stacked bar charts is provided in Figure 3. Qualitative and quantitative data are outlined in detail in the following sections.

Insert Figure 3 around here

Level of support and range of movement

The participants rated the level of support provided by the SSS positively (mean of 2.15 "agree"), with the most common response being "strongly agree". The lower and upper quartiles of 1 & 3 indicate consistency in the positive opinion regarding the support provided. There was however, no statistical difference between rating of the support provided by the SSS versus previous collars. The interview data provide examples of positive opinion regarding the level of support with the SSS, *"it stops the head tilting forward. It's worked absolutely perfect" P20.* Four patients drew attention to the reduction in their neck pain as a result of the improved support, *"with the collar on I get no pain at all" P16.* Also, three

participants commented on how the feeling of warmth relieved pain or discomfort, *"I like the warmth that it provides" P5.*

A key area of positive ratings was in terms of the range of movement offered by the SSS with mean of 2.6, median and mode of 2 "agree" that the SSS allowed an acceptable range of movement. This compared very favourably with other collars which had average rating of 3.88 to 5. There was a significant difference in evaluation of range of movement between the SSS and other collars (N=19 p=0.04). During the interviews participants described this positive aspect, *"it's not as restrictive or as tight as the wire one" P3.*

Appearance

The appearance of the SSS was rated more positively than previously worn collars (p=0.005, N=19)), and the interview data confirmed these positive views, *"it looks like an item of clothing" P20.* However, despite this there was a degree of ambivalence amongst participants concerning the appearance of the SSS. The median rating was 3.5 and the modal rating was 4 "neither agree nor disagree".

Fitting the collar

All the collars scored poorly for the ability to fit without assistance, with a mean of 6.4 (SSS) versus 5.83 (previously worn collars), and a median and mode of 7 "strongly disagree" for both groups. Individual participant views confirmed that fitting the SSS without assistance was not possible for most, for example," *I can't fasten it at the back because my hands won't go up there" P15*. Fitting was predominantly carried out by a carer, with all but two participants reporting that that the SSS was not difficult for a carer to fit, *"she pops it on and job done" P2*, although practice could be required, "*it's got easier as we've gone along" P12.*

Breathing, eating and swallowing

Overall the SSS did not have an adverse impact on breathing, eating or swallowing (means of 3.2, 3.8, 3.4 and 3.4 for questions 1 to 4 respectively). The interquartile ranges for these questions were large suggesting considerable variation in participant experiences. Interview data confirmed this variation in perceptions, for example these contrasting views: *"I have eaten in it and it's not a problem" P10* versus *"can't possibly eat with it on" P11.* A feeling of tightness and pressing on the neck was an obstacle to use of the SSS for two participants, *"it makes my swallow harder as it presses on my Adam's apple" P2.*

Perspiration

Ratings of perspiration experienced when wearing the SSS were most frequently either "agree somewhat" or "neither agree nor disagree". Here again there is evidence of individual disparity in rating (interquartile range 2.75 to 5) suggesting variation in experience. No significant difference between the SSS and previously worn collars was detected.

Usage

We explored whether reported usage of the SSS was significantly different to that of other collars. While there were individual reports of increased usage of the SSS, there was no significant difference in the number of hours the SSS was used during the one month data collection period, versus the number of hours the previously worn collar was typically used. Sixteen of the 20 patients reported that they intended to continue using the SSS (most using it exclusively, a few combining with use of other collars). Four participants had found that it was not suitable for their needs, and did not intend to use it in the future. For two of these, the fit was described as being too tight and restricted swallowing or speaking. The other two participants also had issues with the fit, reporting that was too high on their neck. Three reported that they would prefer to use their previous supports, the other participant had found none of those available to be satisfactory.

An important positive aspect for participants who intended to continue use of the SSS, was the flexibility that the new support offered. This flexibility was described in terms of firstly, the ability to adjust the level of support according to individual need or at different points in the day, *"I love the idea that you can adjust it in lots of different ways" P3*. Secondly, participants valued flexibility in terms of changing the level of support as the disease progressed *"If my neck gets any worse then we might have to put a little bit more support into it" P7.*

Overall satisfaction

Overall participant rating of satisfaction with the SSS was on the boundary of agree and neither agree nor disagree (mean 3.65) which compared favourably with mean rating of 4.88 for the previously worn collars. The majority of satisfaction ratings for the SSS were positive with an interquartile range from 1 ("strongly agree") to 4 ("neither agree nor disagree"), whereas satisfaction for previously worn collars tended to be considerably lower with an interquartile range from 4 to 6 ("disagree"). Patients were significantly more satisfied with the SSS (N=19 p=0.01). There was no significant difference in regard to frustration experienced with the SSS versus previously worn collars.

Discussion

This study explored patient and carer views and experiences of using a newly-developed neck orthosis, the Sheffield Support Snood. Participants were positive regarding the level of support offered by the SSS compared to other devices that had been tried. There was also positive feedback regarding the range of movement that was possible. Other areas of perceived advantage for the SSS described in the interviews were: the flexibility of use offered, the reduction of pain/discomfort, and the feeling of warmth provided. Sixteen of the 20 patients intended to continue to use the SSS. The study sample included patients at earlier and later stages of disease progression, with use of the SSS described positively by those with both less and more severe neck weakness.

Limitations of this study are the small sample size and predominance of patients from a single specialist clinic. The observation period for use of the new support was also relatively short, and we recognise that there was only a single study arm. While our sample is in line with other qualitative studies which typically consist of around 10 to 30 participants, the limited quantitative data means that the statistical analysis should be treated with caution.

While the SSS was rated significantly more positively than other supports, there was considerable variation in views and experiences across the sample. Areas of more mixed ratings included the ease of eating and drinking wearing the SSS (with some patients reporting that it aided eating, whereas in contrast others found that a feeling of tightness made use during mealtimes not possible), and the fit (with some participants finding it too high up on their neck). The SSS was also perceived (as with most other collars) as being difficult to fit, with almost all patients requiring assistance. Carers did not report that there were any key issues in fitting the SSS, although some initial training and practice could be required. As a result of the evaluation phase further slight modifications were made to the design, in order to address feedback regarding sizing issues and also pressure around the neck.

The project illustrates the value of including patients and their family carers in the design process, with use and performance improvements over current neck orthoses and a sense of ownership among research participants, who have since acted as advocates for the product (6). The considerable variability in views and experiences of technology in patients with ALS has been previously reported (8). The current study highlights the importance of having a range of devices available, to enable patients to select those that are suitable and acceptable to them at different stages of the disease. Choice has been described as a central emphasis of care in ALS (9).

While the new orthosis was designed for patients with ALS, there is potential for usage in other neurological conditions causing neck fatigue. Unlike other collars, as well as being

adjustable for increasing need, it offers the facility to reduce the amount of support provided over time, and thereby could also be used in conditions where patients regain function.

Conclusions

The results of this evaluation highlight the significant impact of neck weakness on the everyday lives of people with ALS, and the value of an orthosis which better addresses the requirements of this group of patients. The client-focused development process that was used during the design of the SSS, provided valuable insights and feedback to underpin the production of an orthosis that was suitable for patient needs. While developed specifically for people with ALS, there is potential for the SSS to be suitable for a wider range of patients requiring cervical orthosis.

Disclosure of interests

This project was funded by the National Institute for Health Research (Innovation for Invention Programme) project number II-ES-0511-21003. The views and opinions expressed therein are those of the authors and do not necessarily reflect those of the i4i programme, NIHR, NHS or the Department of Health.

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We would like to thank the patients and their carers who volunteered to try the new orthosis, and who gave up their valuable time to provide us with their assessments and detailed feedback.

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Table 1 Neck collar questionnaire responses

	Previous collar Mean rating† (Median) Lower & upper quartiles	**SSS** Mean rating† (Median) Lower & upper quartiles	**Wilcoxon Signed rank test for paired samples**
1. This collar causes no restriction to my natural breathing	N=24 2.79 (2) 1 & 5	N=20 3.2 (3) 1 & 5	N=19 p=0.849
2. I experience no additional difficulties eating a meal due to wearing this collar	N=24 4.29 (5) 2 & 6	N=15 3.8 (3) 2 & 5.5	N=10 p=0.393
3. I experience no additional difficulties drinking due to wearing this collar	N=17 4.06 (4) 2 & 6	N=16 3.44 (2.5) 1.75 & 6	N=12 p=0.319
4. This collar causes no restriction to my natural swallowing	N=19 2.89 (2) 1 & 4.5	N=18 3.44 (2.5) 2 & 5	N=14 p=0.630
5. I feel that this collar offers support	N=24 3.29 (3) 2 & 4.25	N=20 2.15 (2) 1 & 3	N=19 p=0.098
6. I experience no perspiration around my head, neck, shoulders or neck as a result of wearing this collar	N=24 3.42 (3) 2 & 5	N=20 3.75 (3) 2.75 & 5	N=19 p=0.409
7. I find this collar visually attractive	N=24 5.17 (5) 4 & 6	N=20 3.3 (3.5) 2 & 4	N=19 *p=0.005
8. I have an acceptable range of head movement wearing this collar	N=24 3.88 (4.5) 2 & 5	N=20 2.6 (2) 2 & 3	N=19 *p=0.040
9. I find this collar very easy to fit on my own	N=24 5.83 (7) 5 & 7	N=20 6.4 (7) 6 & 7	N=19 p=0.088
10. I feel no frustration at all whilst wearing this collar	N=24 4.42 (5) 3 & 6	N=20 3.65 (3.5) 2 & 5	N=19 p=0.065
11. I am extremely satisfied with this collar	N=24 4.88 (5) 4 & 6	N=20 3.15 (3) 1.75 & 4.25	N=19 *p=0.010

† Rating derived from seven point Likert scale: 1=strongly agree, 2=agree, 3=agree somewhat, 4=neither agree nor disagree, 5=disagree somewhat, 6=disagree, 7=strongly disagree.

* significant at 0.05 level

Appendix 1. Neck support questionnaire

Date:

Subject ID:

Are you still using the Support Snood?: Yes ☐ No ☐

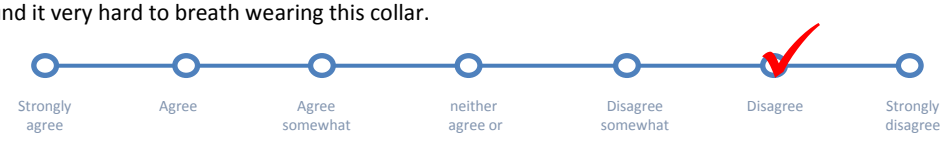
If you have stopped using the Support Snood please explain why?

Please state how long you wear/wore the Support Snood in a typical 24 hr period:

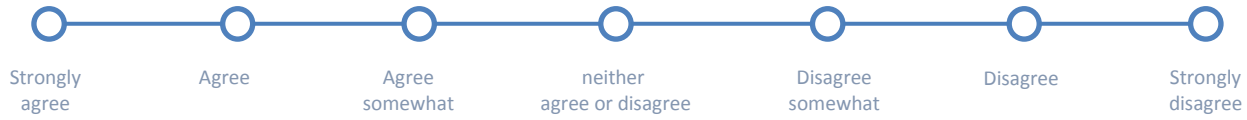
Do you prefer the Support Snood to other collars you have used? Yes ☐ No ☐
If yes please explain why.

Please read the following statements and place a mark on the circle you feel best describes your feelings regarding the Support Snood , as shown in the example below.

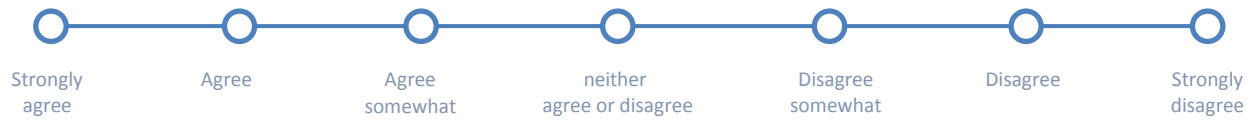
I found it very hard to breath wearing this collar.



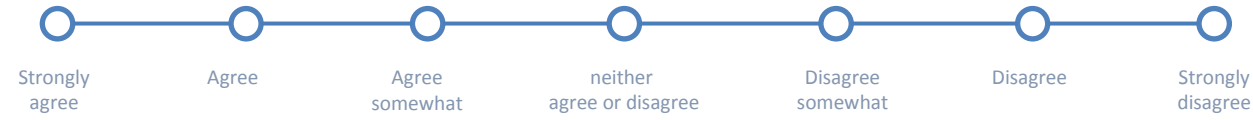
This collar causes no restriction to my natural breathing.



I experience no additional difficulties eating a meal due to wearing this collar.

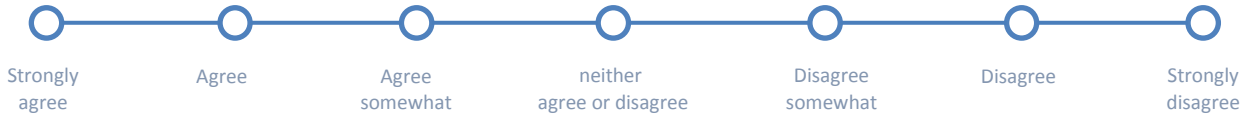


I experience no additional problems drinking due to wearing this collar.



ALS neck collar

I feel that this collar offers support.



I experience no perspiration around my head, shoulders or neck as a result of wearing this collar.

I find this collar visually attractive.

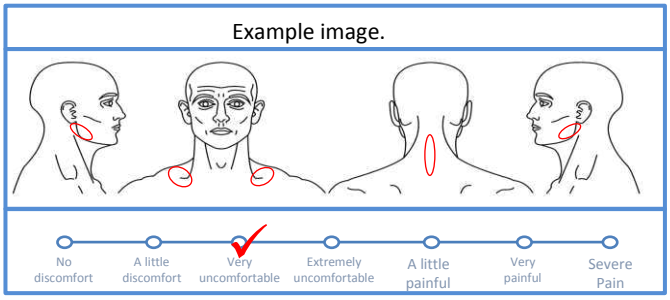
I have an acceptable range of head movement wearing this collar.

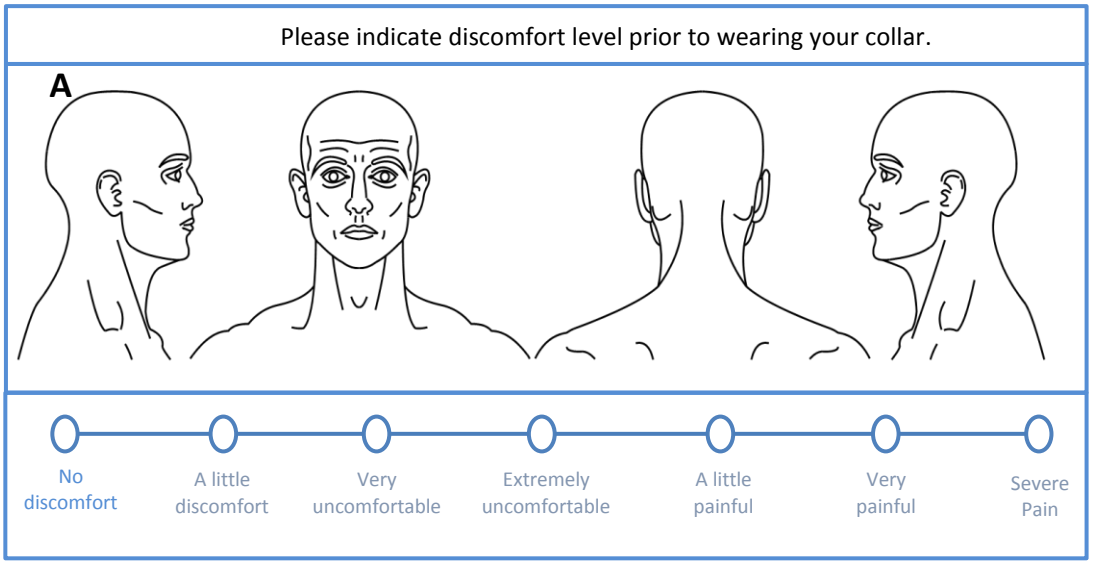
I find this collar very easy to fit on my own.

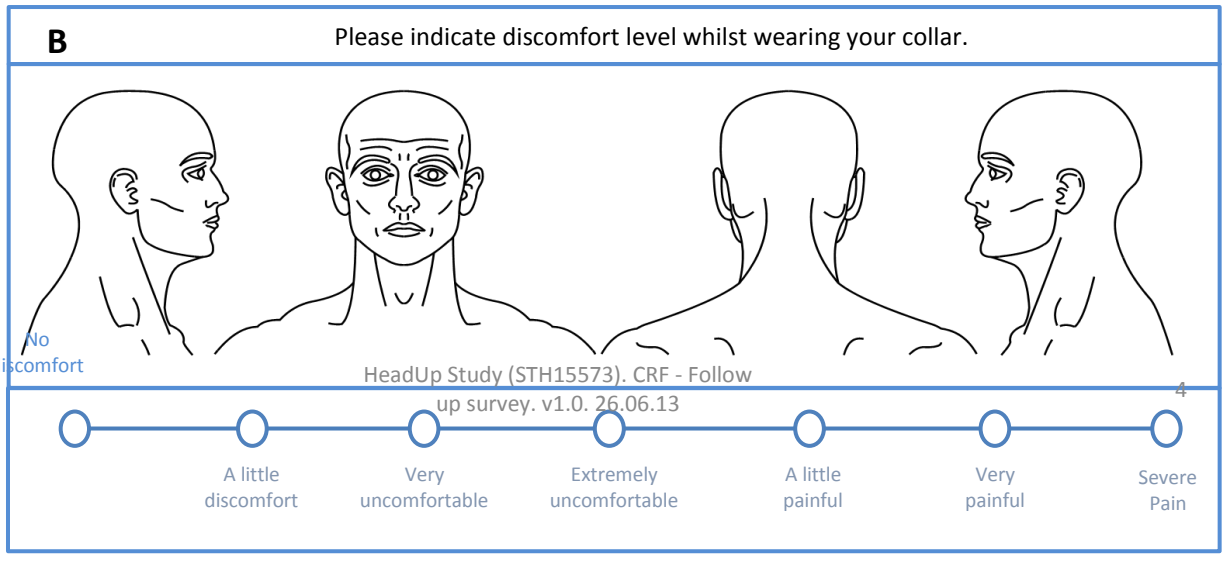
I feel no frustration at all whilst wearing this collar.

I am extremely satisfied with this collar.

Please could you circle on the images below the areas you feel discomfort and indicate on the scale the degree of discomfort experienced:
A. Prior to wearing the Support Snood .
B. Whilst wearing the Support Snood .







Appendix 2 Interview topic guide

1. Information and Consent

2. Effect of neck weakness

Level of weakness – how would you describe it? Duration of difficulties, extent of difficulties impact on daily living, socio-emotional aspects, partner/family.

3. Baseline interview - currently used collars

Knowledge of/experience of neck supports available, sources of information, own perceptions, reported perceptions of others. Describe your experience of using a neck collar/s. What were the successful things about the collar/s. What were the unsuccessful things about the collar/s. What did you want the collar/s to help with? Describe putting on and taking off the collar/s.

4. Follow up interview - Support Snood

Discussion of initial perceptions/expectations of new support and information received. Exploration of pattern of use of the new collar, factors underpinning pattern of use, decision-making process regarding usage. Discussion of any particular positive and/or negative aspects of the collar. Views regarding comfort level, level of support, any pain/discomfort, ease of fitting/removal, ease of adjustment, appearance. Exploration of any impact of the collar on daily living (include communication, eating, washing, dressing, travelling, leisure activities), and any impact on social and emotional aspects of life including impact on partner/family.