**Suppl. 1 Usual care at the study site provided for patients with NIV**

Once established on NIV, patients are followed-up by consultant respiratory physicians and NIV physiotherapists every 3 months at multi-disciplinary outpatient clinic, depending on the rate of progression of the condition. The NIV physiotherapists also offer a post-hoc clinic for NIV where required, and cough assist-therapy on an outpatient basis. Patients are provided with rescue antibiotics and the NIV physiotherapist have a contact number for any issues related to NIV and cough-assist therapy, available during office hours (i.e. Monday - Friday 9:00am – 4:30pm) with an automated telephone answering machine.

### **Suppl. 2 Phase 1: bespoke questions development**

#### Expert Panel 1 (initial development)

Questions were generated to identify nocturnal hypoventilation, respiratory insufficiency, acute respiratory events (e.g. chest infection), and NIV related issues. Their discussion was guided by: 1) the symptoms of hypoventilation as set out in the NICE guidelines; 2) questions used in current outpatient ventilation clinics; and 3) the results of a literature review exploring symptoms of hypoventilation described in MND (see Table Suppl.1). Whilst the self-report question set focuses on respiratory and NIV related aspects of MND, it also includes questions on adequacy of care (Q10) and swallowing (Q19 and 20) as these are commonly discussed within outpatient ventilation clinics, patients could also raise any other issue with the “anytime” message “button”.

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| Table Suppl.1: Broad symptom sets potentially indicating hypoventilation |
| 1. Anorexia
2. Anxiety
3. Concentration impaired
4. Decreased energy
5. Difficulty waking up in the morning
6. Dry or sore mouth
7. Early morning headache
8. Generalised fatigue
9. Hypersomnolence
10. Impaired cognition
11. Insomnia
12. Intermittent breathing
13. Memory problems
14. Nightmares
15. Ordinary activities an effort
16. Orthopnoea
17. Shortness of breath on exertion
18. Short sleep / frequent waking / fragmented sleep
19. Waking unrefreshed or tired
 |

A preliminary question set of 21 items was allocated for either a morning or evening slot depending on relevance. Each item was categorised; high priority notifications were generated for those relating to acute respiratory events, and triggered prompt intervention by the clinical team. Low priority notifications were generated for those indicative of general deterioration, that was not clearly acute. The question set also included two modified Borg dyspnoea scales [1], one each for morning and evening slot, and a sliding scale for both oxygen and NIV usage time.

#### Face validity assessment

The preliminary questions were tested for face validity, clarity and relevance. A weekly telephone follow-up by a non-clinical researcher identified any notification failure according to a predetermined protocol i.e. no notification was generated via the Careportal® despite for example NIV equipment failure or a significant worsening of respiratory function that resulted in unplanned healthcare utilisation. For the assessment, 40 patients were identified on the ventilation database and these patients were screened for a potential recruitment. 26 patients were identified as potential participants. Ten participants were perceived sufficient for the purpose, therefore the recruitment was carried out until we reached the target number of ten. We approached 12 patients of which two declined before consent and one died shortly after their consent, resulting in the total of nine participants for the face validity assessment.

Following the validation, five participants attended a focus group at the study site, and four participated in an individual interview at their home, as was their preference. The focus group had two facilitators; the researcher who had monitored the study data acted as the main facilitator, while the other was a physiotherapist involved in the care of the participants. A physician observer was also present to provide clarification if matters were left unclear in particular regarding their condition. The individual interviews were conducted by the main facilitator of the focus group.

The focus group and interviews were semi-structured during which each question was assessed. Based on the protocol, a question was deemed clear and understandable if at least 80% of the study participants agreed it as such. Nevertheless, questions that were perceived unclear by any participant were noted and participants were encouraged to make suggestions for modifications during the discussion and interviews. They were also asked whether the question set covered all the respiratory related symptoms of MND and NIV related issues they experience. All discussion was audio-recorded and listened later to ensure that the modified items reflected the discussion.

During the validity assessment, no notification failure was identified. 18 items (86%) were deemed clear and understandable by all participants and were retained. One item received 78% agreement on its clarity leading to re-wording and a modification of its response. Two items received 56% agreement, thus were deleted. An additional item was included to expand NIV related problems (e.g. mask leak).

#### Expert panel 2

The questions were further evaluated by the second expert panel. A representative from the telemonitoring company who issues Careportal® was also present to advise on formatting of questions. The same assessment criteria (i.e. 80% agreement) used for patient feedback sessions was employed for the panel.

10 items (50%) were deemed unclear therefore reworded and their response range was modified. One nested question was identified as a standalone question. Six items were added for missing symptoms and/or specifying problems. The panel also proposed the creation of a notification system for the evening Borg scale so that a worsening of the score by ≥2 points would trigger a high priority notification. The final question set (MND OptNIVent) was agreed on with an aim to optimise the care of MND patients, who were using NIV. A further representative from Docobo advised on the questions structure for display to the patients and in the display of alerts at the base station. Neither Docobo person influenced the panels’ discussion however the expertise in presentation was valuable.

**Reference**

1 Borg GA. Psychophysical bases of perceived exertion. Med Sci Sports Exerc 1982;14:377-381.

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| **Suppl. 3 MND OptNIVent question set****Morning question set** Nested question number in a bracket. High priority notifications are generated for those relating to acute respiratory events, triggering prompt intervention by the clinical team. Low priority notifications are generated for those indicative of general deterioration, but not clearly acute. (notification type indicated in CAPITAL) |
| 1 | In general, how well have you been sleeping during the past week?1. Well
2. Somewhat better
3. Same as usual
4. Somewhat worse (LOW)
5. My sleep has been disturbed (LOW)
 |
| 2 | In general, is your sleep refreshing?1. Yes
2. Yes, somewhat
3. No
 |
| 3 | In general, do you have headache when you wake up in the morning?1. Yes (HIGH)
2. Yes, occasionally (LOW)
3. No
 |
| 4 | How many pillows do you need behind your head to be comfortable (choose all that apply)?1. 2 or less
2. 3 or more (LOW)
3. I sleep semi-upright (LOW)
4. I can only sleep sitting up (LOW)
 |
| 5 | Please indicate which number on the scale, from 0 to 10, best describes the degree of breathlessness when you are in upright position. (Borg Scale here) |
| 6 | Have you used your ventilator in the last 24 hours?1. No, not in the last 24 hours (HIGH)
2. Night time only
3. Night time and some day time
4. Day time only
5. All the time (HIGH)
 |
| 7 | Tell us how many hours you have used the ventilator in the last 24 hours. (sliding scale here) |
| 8 | Are you having any problems with the ventilator (i.e. is there any issue with the machine or mask and the way it is working for you)?1. None whatsoever
2. Occasional problems
3. Occasional problems I would like you to address (LOW)
4. A lot of problems (HIGH)
5. It is not working at all (HIGH)

If patient answers 8c, 8d and 8e then Q9 will be asked  |
| (9) | What problems with the ventilator are you experiencing (please choose as many that apply)?1. Mask leak
2. Mask discomfort
3. Ventilator alarms
4. Others
 |
| 10 | How well does the personal care/help you receive where you live meet your needs at present?1. Very well
2. Reasonably well
3. Still need more support
4. My needs are not met (HIGH)
 |
| **Evening question set** |
| 11 | We would like to know if there has been a change in your breathing since last week. Is it:1. Better than the previous week
2. No change
3. More breathlessness than the previous week (HIGH)
 |
| 12 | Please indicate which number on the scale, from 0 to 10, best describe the degree of breathlessness you experienced during the day today. (Borg Scale here)If the patient’s answer is 2 point worse (i.e. increased number) than the previous week then HIGH alert is triggered |
| 13 | Are you able to cough on your own? 1. Yes
2. No (LOW)
3. I use a Cough Assist Machine/Lung Volume Recruitment Bag

If patient answers 13a then the Q14 will be asked  |
| (14) | Have you developed a new cough?1. Yes (LOW)
2. No
 |
| 15 | How effectively can you clear your chest (with or without equipment)? 1. I can clear my chest effectively
2. I am struggling to clear my chest (HIGH)
 |
| 16 | Have you had phlegm in the last week?1. I have not had phlegm
2. I have had some phlegm
3. I have had a lot more phlegm than normal (HIGH)

If patient answers 16b and 16c then Q17 will be asked |
| (17) | What colour is your phlegm/ sputum today? 1. It is clear
2. Yellow (HIGH)
3. Green (HIGH)
4. Other
 |
| 18 | Do you have difficulty with clearing the throat of saliva or secretions?1. Yes
2. No
 |
| 19 | Please describe your swallowing (please choose as many that apply).1. No problems
2. I am aided for my swallowing problems by a suction machine
3. I sometimes have difficulty with liquid
4. I sometimes have difficult with food
5. I have difficulty swallowing saliva
6. I am aided for my swallowing problems by a feeding tube

If patient answers 19b, 19c, 19d and 19e then Q20 will be asked |
| (20) | Have you had any swallowing problems in the last 24 hours (with or without suction machine)?1. No problems
2. Slight problems
3. Moderate problems (HIGH)
4. I have choking/swallowing problems that I need extra help with (HIGH)
 |
| 21 | Tell us how many hours you have used your oxygen, when not using the ventilator, in the last 24 hours? (Sliding scale here) |
| 22 | In general, how sleepy are you in the day?1. Not sleepy
2. Slightly sleepy
3. Very sleepy (LOW)
4. Hard to stay awake (HIGH)
 |
| 23 | Have you had any hallucinations or unusually vivid nightmares during the past week? By hallucination we mean an experience of seeing or hearing something not present.1. Yes (LOW)
2. No
 |
| 24 | In general, do you feel tired during the day?1. Yes
2. Yes, somewhat
3. No

If patient answers 24a or 24b then Q25 will be asked |
| (25) | What caused your tiredness during the past week (please choose as many that apply)? 1. A busy week
2. A mentally stressful week
3. My condition i.e. MND
4. Cold/ Chest infection
5. Not sleeping
6. Others
7. Unsure
 |
| 26 | How has your fatigue affected your daily activities during the past week? We use ‘fatigue’ to mean tiredness and/or lack of energy.1. My daily activities have not been affected
2. Slightly affected my daily activities
3. Moderately affected my daily activities (LOW)
4. Severely affected my daily activities (LOW)
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