

Respiratory Care Pathway for people living with Motor Neurone Disease in Dorset - Non-Invasive Ventilation

Abbreviations:	
NIV - Non-Invasive Ventilation	SNIP - Sniff Nasal Inspiratory Pressure
ADRT - Advance Decision to Refuse Treatment	TRC - The Respiratory Centre
MNDA - Motor Neurone Disease Association	UHS - University Hospitals Southampton
FVC - Forced Vital Capacity	ABG - Arterial Blood Gases

Assess: Motor Neurone Disease: assessment and management 2016 (NG42)1.14.7

Symptoms and signs:

Monitor the symptoms and signs listed to detect potential respiratory impairment. **[2010, amended 2016]**

Symptoms and signs of potential respiratory impairment:

Symptoms:

1. Breathlessness
2. Orthopnoea
3. Recurrent chest infections
4. Disturbed sleep
5. Non-refreshing sleep
6. Nightmares
7. Daytime sleepiness
8. Poor concentration and/or memory
9. Confusion
10. Hallucinations
11. Morning headaches
12. Fatigue
13. Poor appetite

Signs:

1. Increased respiratory rate
2. Shallow breathing
3. Weak cough - could be assessed by measuring peak cough flow
4. Weak sniff
5. Abdominal paradox (inward movement of the abdomen during inspiration)
6. Use of accessory muscles of respiration
7. Reduced chest expansion on maximal inspiration

Withdrawal Guidelines:

Withdrawal of Assisted Ventilation at the Request of a Patient with Motor Neurone disease:

Guidance for Professionals (Draft Consultation Document). 2015.

Association for Palliative Medicine of Great Britain and Ireland. Chair: Professor Christina Faull

Local Specialist Palliative Care advice for NIV withdrawal:

East Dorset: Consultant Dr Catherine Webb

t: 01202 705470

West Dorset: Consultant Dr Karen Steadman

t: 01305 215300

Local contacts:

The Respiratory Centre, University Hospital Southampton NHS Trust:

t: 02381 204325

Consultants : Dr Arjjana Sivaloganathan, Dr Jane Wilkinson

Physiotherapists: Fiona Cull, Dr Denise Gibson

t: 02380 777222 bleep 2080

uhs.homeventilationservice@nhs.net

UHS switchboard: for equipment only - **no clinical advice:**

02380 777222

'Those who qualify are asked to contact out of hours support via UHS switchboard and ask for the 'Respiratory Ventilator Service'. Switchboard takes the patients details and passes this onto the person on call who calls them back. This is not for clinical support - only for equipment support. Currently a telephone advice support only'.

Clinical advice from Respiratory Registrar UHS NHS Trust

t: 02380 777222

This pathway has been drawn up with the named service providers.
Revised by Dorset MND working party. June 2020. Review June 2022.

Respiratory Medicine:

East Dorset: Consultant - Dr Tim Shaw

t: 01202 303656

West Dorset: Consultant - Dr Will McConnell

t: 01305 225259

References:

BOURKE, S., BULLOCK, R.E., WILLIAMS, T.L., SHAW, P.J., and GIBSON, G.J., 2003. Noninvasive ventilation in ALS: Indications and effect on quality of life. *Neurology*. 61:171-177.

BOURKE, S., TOMLINSON, M., WILLIAMS, T., BULLOCK, R., SHAW, P., and GIBSON, G.J., 2006. Effects of non-invasive ventilation on survival and quality of life in patient with amyotrophic lateral sclerosis: a randomised controlled trial. *The Lancet Neurology*. 5:140-47.

FALLAT, R., JEWITT, B., Bass, M., Kamm, B., and NORRIS, F.H., 1979. Spirometry in Amyotrophic Lateral Sclerosis. *Arch Neurol*. Vol 36 Feb, 74-80.

LYALL, R.A., DONALDSON, N., POLKET, M.I., POLKEY, P.N., LEIGH, P.N., ND MOXHAM, J., 2001. Respiratory muscle strength and ventilatory failure in amyotrophic lateral sclerosis. *Brain* 124, p2000-2013.

MURPHY, P., JYALL, R., HART, N., POLKEY, M.I., 2010. Assessment of respiratory muscle strength in motor neurone disease: is asking enough? *European Respiratory Journal*. 35: p353-360.

NATIONAL INSTITUTE FOR HEALTH and CARE EXCELLENCE. 2016. *Motor neurone disease: assessment and management*. London. NG42.

SIIRALA, W., SAARESANTTA, T., VUORI, A., SALANTERA, S., OLKKOLA, K., AANTAA, R., 2012. Using respiratory rate and thoracic movement to assess respiratory insufficiency in amyotrophic lateral sclerosis: a preliminary study. *BMC Palliative Care*. 11: 26.

SIMONDS, A., ed. 2015. *European Respiratory Society Practical Handbook: Noninvasive ventilation*. Sheffield: The European Respiratory Society.

For further information for professionals or patients see:

<https://www.mndassociation.org/>

Information for patients and carers:

<https://www.mndassociation.org/>

Information sheets:

8A - Support for breathing problems Information sheet 8B - Ventilation for Motor Neurone Disease

8B - Ventilation in MND

8C - Withdrawal of ventilation in MND

8D - Trouble shooting for NIV

8E - Air travel and ventilation for MND

SITraN/University of Sheffield Information and videos about starting, living and later on with NIV:

www.mybreathing.mymnd.org.uk

Information for Healthcare Professionals:

<https://www.mndassociation.org/wp-content/uploads/information-sheet-p4a-mnd-just-in-case-kit.pdf>

<https://www.mndassociation.org/wp-content/uploads/information-sheet-p3-managing-saliva-problems.pdf>

This pathway has been drawn up with the named service providers.
Revised by Dorset MND working party. June 2020. Review June 2022.

<https://www.mndassociation.org/wp-content/uploads/information-sheet-p6-evaluation-and-management-of-respiratory-symptoms-in-mnd.pdf>

This pathway has been drawn up with the named service providers.
Revised by Dorset MND working party. June 2020. Review June 2022.