



How to choose respiratory equipment for children with neurodisability including NMD

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This is a general guide for healthcare professionals outlining commonly used respiratory equipment for children with neurodisability including Neuromuscular Disease (NMD). Respiratory equipment suitability and requirements will vary because there are many different types of neurodisability and NMD. Individuals will present differently, and their needs may change over time. It is important to understand devices that can be recommended; however, regional variability may result in some techniques and devices not being available in every country.

In the table below are some general principles and suggested tips to help you decide on the most appropriate equipment for your patient along with when and why it may be useful. Not every person with neurodisability or NMD will require this equipment, regardless of specific diagnosis, as the onset and severity of respiratory complications vary between individuals. The patient's respiratory equipment needs should be discussed with a respiratory healthcare provider who specialises in NMD or neurodisability. They can ensure the equipment is suitable for your patient and guide you in the correct use and programming of the appropriate equipment.

TYPE OF EQUIPMENT	WHY AND WHEN IT IS RECOMMENDED	SOME SIGNS AND SYMPTOMS YOU MAY HAVE THAT SUGGEST YOUR PATIENT MAY NEED THIS EQUIPMENT
<p>Lung volume recruitment (LVR) bag</p> <p><i>For example</i></p>  <p>©Breas</p>	<p>The LVR bag is recommended when vital capacity drops towards 50-60% predicted. This occurs often in conjunction with impaired mobility. This is because a lack of general mobility and respiratory muscle weakness makes it more difficult to take a deep breath – an essential component for an effective cough required to clear airway secretions.</p> <p>At other times, when the patient is unwell with a respiratory illness, airway secretions, fatigue or pain can make it difficult to take deep breaths to facilitate a strong enough cough to clear secretions from the large airways. This is because a respiratory illness leads to a decrease in respiratory muscle strength and fatigue¹.</p> <p>Respiratory physiotherapists may recommend using LVR bag or a resuscitation bag (bag valve, mask (BVM)). These devices help to get air behind secretions and provide a big inspiratory breath to enable an effective cough. They can be used to augment volume by providing a single breath or by stacking multiple breaths. It is usually used followed by manually assisted cough.</p> <p>Studies are ongoing to determine if there is any benefit to using this daily when well to help maintain some lung function over time^{2,3}.</p>	<p>When your patient gets a cough or cold:</p> <ul style="list-style-type: none"> • The cough is weaker and ineffective leading to difficulty clearing secretions • If they are getting tired and weak when they are unwell and find it difficult to take a deep enough breath to cough and clear secretions effectively • If they are taking a longer time than expected to get over a cough/cold with secretions that are lingering
<p>Non-Invasive Ventilation (NIV)</p> <p><i>For example Breas Vivo 3</i></p>  <p>©Breas</p>	<p>Lung function tests (including spirometry) and sleep studies help to assess the ability to maintain normal ventilation. Often the first changes to breathing occur during sleep⁴. The Respiratory Team will advise your patient about the timing and need for non-invasive ventilation (NIV) to support ventilation during sleep.</p> <p>Sometimes respiratory physiotherapists may wish to use NIV to support the patient to assist in clearing airway secretions when they are unwell with a respiratory illness.</p>	<p>If your patient:</p> <ul style="list-style-type: none"> • Wakes unrefreshed • Has morning headaches • Takes a long time to feel “awake” in the morning • Feels more tired, sleepy or falls asleep throughout the day • Wakes more frequently at night • Has frequent respiratory illnesses <p>Your patient may have sleep disordered breathing (SDB) and should be assessed by their specialist team.</p> <p>If a respiratory illness causes your patient to be tired and lethargic urgent specialist review is warranted. NIV may be used for a short period of time to provide rest to the breathing muscles and for support during chest (airway clearance) physiotherapy.</p>

Mechanical Insufflation-Exsufflation (MI-E)

For example Clearway 2



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MI-E is often recommended when lung function (spirometry) especially cough peak flow (CPF) (also known as PCF – peak cough flow) is less than 50% predicted or lower than 160L/min in children older than 12 years old⁵.

MI-E is also often recommended clinically for some patients in the presence of respiratory illness that results in airway secretions, fatigue and/or pain that makes it difficult to take a deep breath in and have a strong enough cough to clear secretions from the large airways⁶.

Sometimes if bones are very brittle (osteoporosis or osteopenia), it may not be advisable to use a manually assisted cough with hands on the chest wall.

Consideration of a short-term loan of MI-E device is advisable initially if available.

If the use of MI-E is required frequent times per year or daily to maintain lung health, long term provision should be considered.

If your patient:

- Gets frequent respiratory tract infections
- Does not get over coughs and colds as quickly or easily
- Has difficulty coughing and clearing airway secretions
- Has tried using LVR bag or BMV with manually assisted cough but, is still unable to cough and clear effectively

High Frequency Chest Wall Oscillation/Compression (HFCWO/C)

For example (during an acute illness) HillRom Therapy Vest



©HillRom

The HFCWO/C occasionally can be useful if your patient has frequent or regular respiratory secretions or their specialist team has highlighted there are persistent lung changes on chest X-ray or CT scan (e.g. chronic suppurative lung disease or bronchiectasis).

The HFCWO/C can be very helpful in clearing lower lung secretions. If your patient is already using techniques such as percussion and/or vibrations or other manual techniques to help secretions move and is clearing well these other effective techniques may be sufficient. This may influence if the HFCWO/C is necessary.

Use of HFCWO in the acute setting must be used in conjunction with NIV to reduce risk of respiratory compromise.

Frequent or prolonged respiratory illnesses with airway secretions that are not easily cleared even with use of a MI-E machine and other manual techniques and uses some form of assisted ventilation (e.g. NIV – see above).

The patient may benefit from a review of their airway clearance with your respiratory physiotherapist who specialises in NMD and/or Neurodisability. They may consider the HFCWO/C.

In our experience HFCWO/C, when appropriate, is often more useful in younger children.

Intrapulmonary Percussive Ventilation (IPV)

For example IPV[®]- 2C



©Percussionaire

IPV can sometimes be useful if your patient needs help to clear lower lung secretions during times of respiratory illness or if they have frequent or regular lung secretions due to persistent lung changes on chest X-ray or CT scan (e.g. chronic suppurative lung disease or bronchiectasis).

When patients require more help to breathe and need some form of ventilation (e.g. NIV or mouthpiece ventilation) IPV can provide ventilatory support with the addition of oscillations to aid airway clearance⁷.

There are other techniques that can be used to help clear lung secretions. Therefore, it is recommended that this is discussed with the specialist team.

If your patient:

- Has frequent or regular airway secretions that are difficult to clear even with the use of a MI-E machine as secretions are more in the peripheral airways
- Or frequent or prolonged respiratory illnesses with airway secretions that are not easily cleared even with use of a MI-E machine and manual techniques
- Is more dependent on NIV
- Requires increased ventilatory support when unwell with a respiratory illness
- Manual techniques are indicated to aid clearance of peripheral airway secretions but unable to be used due to osteoporosis/osteopaenia and/or pain

Your patient may benefit from a review of their airway clearance regime with their specialist team who may consider the IPV.

In our experience IPV is often more useful in older children/adults with spinal curves and/or brittle bones.

Mouthpiece Ventilation (MPV)

For example Mouthpiece Ventilation Mode



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MPV is a useful tool when your patient needs daytime non-invasive respiratory support, especially if you are looking to increase mask free time.

It is especially useful to facilitate speech, eating and drinking.

If skin breakdown has occurred on the face due to NIV mask, it can give the face relief from pressure during the waking hours.

MPV can be considered, if your patient:

- Is feeling more and more breathless throughout the day and finding it hard to speak in longer sentences.
- Is finding eating and drinking difficult due to breathlessness and fatigue and not new symptoms indicating impaired swallow (bulbar dysfunction).
- If either of these are noted it would warrant an urgent medical review.
- Needing to use NIV more and more during the day but does not want to wear a mask or needs to relieve pressure on their face

Home Suction unit

For example



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When swallowing is difficult, saliva and secretions can pool at the back of the throat. When your patient cannot clear these secretions easily, suctioning is a useful tool. Oral suction can clear secretions and prevent oral secretions from being aspirated into the lungs.

If your patient:

- Finds it hard to clear saliva or secretions from the back of their throat
- Finds themselves often coughing, spluttering, or choking on water or food.
- Has a “purring” sound when they breathe especially when they have a cough or cold
- Has speech that sounds “gurgly” or muffled because they cannot clear their throat easily

Let their specialist team know as they may have a poor swallow and if this is a new symptom, they may benefit from a Speech Pathology review including a swallowing assessment.

If your patient does have a poor swallow but can cough secretions to their mouth yet have difficulty to clear out of their mouth, they may benefit from a suction machine for home.

IF YOUR PATIENT ALREADY HAS RESPIRATORY EQUIPMENT

Patients may need to increase the frequency of use of their equipment and the settings may need to be modified to provide more support during a respiratory illness. Further advice can be obtained by contacting their respiratory team to discuss.

IF YOUR PATIENT DOES NOT HAVE RESPIRATORY EQUIPMENT

Your patient may not need respiratory equipment on a daily basis when well, however, when they become unwell with a respiratory illness, they may benefit from the short-term use of certain equipment until they recover. If you do not feel comfortable initiating this then contact their respiratory team for support. Their specialist team can determine if there are any ways you can access this equipment on loan or advise as to whether equipment should be provided long term. They can also assist in setting up the equipment on the most appropriate parameters.

References

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