



Anaesthetic precautions (continued)

- ▶ Where respiratory muscle weakness is present, familiarisation with ventilatory support is warranted prior to procedure in case it is required post-operatively.
- ▶ After having a general anaesthetic, individuals must be weaned from invasive ventilator support (intubation) to non-invasive support. If an individual was able to breathe by themselves prior to surgery, the aim would be to try to wean them back to their pre-op baseline.

Recommendations and precautions

- ▶ Immunisations should be kept up-to-date including the influenza and pneumococcal vaccine.
- ▶ Pregnancy in SMA Type 3 may cause breathlessness. It is recommended to have a full respiratory assessment prior to conceiving.
- ▶ Routine cardiac checks are advised in any individual who complains of chest pain or discomfort.

Fractures/traumas

- ▶ As bone density may be poor in SMA Type 3, vitamin D supplementation is recommended. Further investigations for low bone density are required if there have been two or more fractures.

Feeding difficulties and care

Feeding supplementation may occur when individuals start to experience weight loss and/or an unsafe swallow.

- ▶ A meal time of longer than 30 minutes is indicative of feeding issues and should warrant evaluation by a speech and language therapist.
- ▶ Weight loss should also be reviewed by a dietician. Simple dietary changes may be enough to help weight gain.

For consensus care guidelines agreed by doctors and patient groups across the world, visit: www.treat-nmd.eu/care/sma/care-standards



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Muscular Dystrophy UK
 Fighting muscle-wasting conditions

Alert card

▶ Spinal muscular atrophy Type 3

Name _____

Date of birth _____ NHS number _____

If presenting at an emergency department, contact the neurology/neuromuscular team and respiratory team at:

_____ as soon as possible on:

Activate your alert card today to receive your vital care plan:

Email info@muscular dystrophyuk.org or call our Freephone helpline 0800 652 6352



Highly commended



The Information Standard Certified Member

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Spinal muscular atrophy (SMA) Type 3

SMA is a genetic condition affecting the part of the nervous system that controls voluntary muscle movement.

In general, SMA affects a person's physical abilities but does not affect their mental development.

SMA Type 3 is the mildest form of childhood SMA.

Most children with SMA Type 3 are able to stand unaided and walk, although many find walking or getting up from a sitting position difficult. Over time, the muscles will become weaker, resulting in some losing the ability to walk when they get older.

Breathing and swallowing difficulties are very rare and SMA Type 3 does not usually affect life-expectancy.

Respiratory

In SMA Type 3, individuals sometimes have weak respiratory muscles, which can cause nocturnal hypoventilation (under-breathing at night) later in life.

Signs of nocturnal hypoventilation include morning headaches, frequent turning at night, fatigue during the day, poor concentration and chest infections. This should be investigated further at a specialist centre. If required, treatment for this is non-invasive ventilation, which involves a small breathing machine attached to a mask.

The aim of this treatment is to make breathing more comfortable, improve daytime sleepiness and poor concentration and correct oxygen and carbon dioxide (waste gas) levels which are altered as a result of the weak breathing muscles.

In SMA Type 3, weak respiratory muscles can make it difficult to cough effectively and there may be vulnerability to respiratory infections.

Intubation and ventilation are indicated in the presence of an acute reversible event unless there is an advance directive stating otherwise.

Chest infections – general advice

- ▶ Low threshold for the use of antibiotics during chest infections is recommended.
- ▶ During a chest infection, it is recommended that intensive physiotherapy be carried out in conjunction with cough augmentation techniques including cough machines. This should occur when oxygen saturations are less than 95 percent on room air.
- ▶ Oxygen therapy may be needed during hospitalisation for a chest infection. If ventilator support is being used, then oxygen therapy should be combined with the ventilator. Care should be used to avoid the risk of raised carbon dioxide (waste gas) levels with oxygen therapy.

Anaesthetic precautions

- ▶ As there is a likelihood of respiratory muscle weakness, individuals with SMA Type 3 undergoing surgery should have a pre-operative evaluation including lung function tests, sleep study and cough assessment.

[continued overleaf ▶](#)