



Recommendations and precautions

- ▶ Immunisations should be kept up-to-date.
- ▶ As children with SMA Type 1 are at high risk of respiratory complications, it is advisable to have the RSV vaccination.

Fractures/trauma

- ▶ Bone density is poor owing to non-ambulation. Vitamin D supplementation is recommended, and calcium as indicated.
- ▶ Children are at risk after relatively low-impact injury. If more than two fractures have occurred, investigations for low bone density should be arranged at a specialist centre managing SMA.
- ▶ If the child has a suspected fracture: because of poor bone density, the radiologist at A&E should use a low threshold for X-ray.

Feeding difficulties and care

Feeding supplementation occurs when children start to experience weight loss and/or an unsafe swallow.

- ▶ Gastroesophageal reflux can occur in SMA. In children, symptoms may be subtle (weight loss, poor feeding, crying after feed or when lying down and coughing). In others it may be severe (stopping breathing with a change in skin colour – blue). Anti-reflux medication should be prescribed and investigated further if required.
- ▶ Feeding supplementation occurs with the insertion of a nasal gastric tube.
- ▶ In some cases, gastrostomy insertion with a nissen fundoplication is indicated to prevent the feed passing from the stomach into the oesophagus (gastroesophageal reflux).



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 1

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



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Spinal muscular atrophy (SMA) Type 1

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, such as moving, walking and breathing, but not their mental development.

Babies and children with SMA Type 1 have severe muscle weakness, which can result in problems moving, eating, breathing and swallowing. These symptoms arise at birth or during the first few months of life.

The muscles of babies with SMA Type 1 are weak, which makes their limbs limp and floppy. They are usually unable to raise their head or sit without support.

Respiratory

- ▶ Breathing problems caused by weakness in the baby's chest muscles are inevitable.
- ▶ Signs of weakening respiratory muscles include poor sleep quality, sucking in of the chest wall muscles, and a change in lip or skin colour.

- ▶ In some individuals, assisted ventilation is indicated, such as non-invasive ventilation. This 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.
- ▶ Assisted coughing helps to clear sputum and mucus plugs and suction and physiotherapy techniques can help this. In some cases a cough machine is indicated.
- ▶ Excess oral secretions can be treated with medication such as glycopyrronium bromide to reduce hypersalivation.
- ▶ Breathlessness and sticky secretions can be helped with saline nebs although suction may be required if excess secretions are produced.

Chest infections

- ▶ Low threshold for the use of antibiotics during chest infections is recommended.

- ▶ Oxygen therapy is often needed during hospitalisation for a chest infection or to palliate symptoms. If ventilator support is being used, then oxygen therapy should be combined with the ventilator. Care should be taken to avoid the risk of raised carbon dioxide (waste gas) levels with oxygen therapy.

Anaesthetic precautions

- ▶ In early stages of SMA, muscle cells develop certain abnormalities which can lead to dangerous reactions to muscle-relaxing drugs often used during surgery.
- ▶ When a child with SMA must undergo surgery (for example, to correct scoliosis or to insert a gastrostomy) special precautions need to be taken. Ideally the surgery should occur in a specialist centre with staff experienced in managing these individuals.
- ▶ After having a general anaesthetic, children must be weaned from invasive ventilator support (intubation) to non-invasive support. If a child was able to breathe by themselves prior to surgery, the aim would be to try to wean them back to their pre-op baseline.

