Recommendations and precautions

Immunisations should be kept up-to-date including the influenza and pneumococcal vaccine.

For children under the age of two, who have had recurrent chest infections and are at risk of intubation and ventilation, it is advisable to have the RSV vaccination.

Pregnancy in SMA Type 2 will 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

Bone density is poor owing to non-ambulation. Vitamin D supplementation is recommended, and calcium as indicated.

Feeding difficulties and care (continued)

Where weight is not gained with dietary modifications or with an unsafe swallow, feeding supplementation is recommended. This can be either with the insertion of a naso-gastric tube or a more permanent feeding tube into the stomach (gastrostomy). If reflux is present, a nissen fundoplication is indicated.

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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Feeding difficulties and care

- 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 be reviewed by a dietician. Simple dietary changes may be enough to help weight gain.
- Gastroesophageal reflux can occur in SMA. 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.

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Respiratory

Individuals often have weak respiratory muscles, which can cause nocturnal hypoventilation (under-breathing at night) and make it difficult to cough effectively. This can make them more vulnerable to respiratory infections.

Signs of nocturnal hypoventilation include morning headaches, frequent turning at night, fatigue during the day, poor concentration and chest infections.

Treatment for this is non-invasive ventilation, involving 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.

Spinal muscular atrophy (SMA) Type 2

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

In general, SMA Type 2 affects a person’s physical abilities, such as moving, walking and breathing, but does not affect their mental development. Although SMA Type 2 may shorten life-expectancy, improvements in care standards mean that most people with the condition live long, fulfilling and productive lives.

People living with SMA Type 2 are usually able to sit, but not stand or walk unaided. They may also have the following symptoms: breathing problems, weakness in their arms and legs, twitching of the muscles in the arms, legs or tongue and scoliosis.

Some people with SMA Type 2 have the following symptoms: breathing problems, weakness in their arms and legs, twitching of the muscles in the arms, legs or tongue and scoliosis.

In some cases, contracture of the hands, feet, chest, and joints develops as muscles atrophy. Many children with SMA Type 2 develop scoliosis as they grow.

Anaesthetic precautions

As there is a likelihood of respiratory muscle weakness, individuals with SMA Type 2 undergoing surgery should have a pre-operative evaluation including lung function tests, sleep study and cough assessment.

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 an individual 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, 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.

Chest infections – general advice

Low threshold for the use of antibiotics is recommended.

Intensive physiotherapy should be carried out in conjunction with cough augmentation techniques including cough machines, with oxygen saturations of 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 taken to avoid the risk of raised carbon dioxide (waste gas) levels with oxygen therapy.

Sticky secretions can be helped with saline nebs although suction may be needed if excess secretions are produced.

Excess oral secretions may be a problem and can be treated with medication, such as glycopyrronium bromide to reduce hypersalivation.

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