Anaesthetic precautions

- There is an increased risk of complications with general anaesthetic in SMA. If elective surgery is required, ensure preoperative assessment, including range of jaw opening, and communication between local and specialist respiratory teams. Where possible, surgery should occur in a specialist centre with staff experienced in managing SMA patients.
- People with SMA can have serious reactions to neuromuscular blocking drugs (muscle relaxants), including suxamethonium, which should be avoided whenever possible.
- After surgery, the individual may require transition from intubation to non-invasive ventilation. If able to breathe by themselves prior to surgery, aim to wean them back to their pre-op baseline.

Falls and fractures

- Minimise fall risks in all environments and use a seatbelt or harness when in a wheelchair. Exercise caution when lifting/moving individual to reduce risk of injury.
- Due to limited mobility, bone density can be poor. Vitamin D supplementation and adequate calcium intake should be ensured.
- Individuals are at higher risk of fractures even after low-impact injuries. Regular monitoring of bone health is expected, planned by specialist neuromuscular clinic.
- If at A&E with a suspected fracture, have a low threshold for X-ray due to poor bone density. It is essential for local team to contact specialist neuromuscular team regarding fracture treatment.

Swallowing and nutrition

- Individuals can experience difficulties with eating, drinking, and swallowing. Signs and symptoms of difficulty may include weight loss, poor appetite, choking, coughing, recurrent chest infections, respiratory secretions, and prolonged mealtimes. In these instances, early referral to specialist speech and language therapist and/or dietitian is important.
- Acid reflux (stomach acid flowing back into the oesophagus) can occur and increase the risk of aspiration (food entering the airway). These can lead to aspiration pneumonia which requires prompt antibiotic treatment.
- Maintaining adequate calorie intake may be difficult. A dietitian can advise on specialised diets, feeding techniques, and nutritional supplements.
- BMI should be interpreted with caution and not based on general population's growth.

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MUSCULAR DYSTROPHY UK

Alert card

Spinal muscular atrophy type 2

Name ..... Date of birth .....

NHS/CHI/H&C number .....

If presenting at A&E, contact the specialist team at:

.....

as soon as possible on: .....

For information and support, contact us on our helpline 0800 652 6352 or email info@musculardystrophyuk.org

Spinal muscular atrophy (SMA) type 2

SMA is a progressive genetic condition that causes severe muscle weakness and affects movement.

Symptoms of SMA type 2 show between 6 to 18 months old. Individuals are usually able to sit independently, and some can stand and walk with support. Others may not be able to do so. They may have breathing problems, weakness in the arms and legs, twitching or tremors in the tongue or hands, and scoliosis.

Pneumococcal and annual flu vaccinations should be kept up to date. COVID-19 vaccination should be given according to national guidelines. It is recommended that children under two who require ventilation (including BiPAP) receive RSV immunisation.

Treatments

- Disease-modifying drugs are available if eligibility criteria are met. Risdiplam (Evrysdi) and Nusinersen (Spinraza) are available for those with SMA types 1, 2, or 3. These drugs are most effective when given before muscle weakness onset. They can stop the progression of muscle weakness and may improve strength and motor function.
- Ask the individual if they have a treatment plan.

Respiratory function

- Weak respiratory muscles are common, which can cause nocturnal hypoventilation (shallow breathing at night). Signs include morning headaches, fatigue during the day, and poor concentration. Nocturnal non-invasive ventilation (NIV) may be required. Use of NIV should improve sleep quality, improve daytime sleepiness, and control abnormal oxygen and carbon dioxide levels.
- Weak respiratory muscles can make it difficult to cough effectively. Cough strength can be assessed using a cough peak flow test.

Respiratory infections and acute medical care

- Increased risk of pneumonia and respiratory tract infections. Low threshold for antibiotics and anticipatory care plan for acute care are recommended.
- Assess bronchial secretion/physiotherapy management and consider cough augmentation techniques such as assisted coughing, breath stacking with lung volume recruitment (LVR), and/or cough assist device to clear lower airway secretions. Dips in arterial oxygen saturation level are often due to retained secretions and can be improved by physiotherapy and cough augmentation. Nebulisers, medication, and suction may be required to manage secretions.
- NIV may be needed, or settings/duration of NIV use increased if individual is already receiving NIV. Oxygen therapy is sometimes required during hospitalisation but should be combined with the ventilator. Oxygen therapy (including high flow oxygen) should only be used on its own with close CO_2 monitoring to avoid the risk of raised CO_2 level.
- Swallowing difficulties (see section) can contribute to respiratory symptoms and chest infections.
- Due to low muscle mass, CK levels may be below the normal range. There may also be an increased risk of acidosis and low blood sugar.
- Care with dosing of paracetamol is important as individuals with SMA may have lower tolerance and require a smaller dose; it can be helpful to monitor liver function tests.
- Finding a vein can be difficult, and it may be necessary to use ultrasound to help identify a vein for cannulation and to use smaller-sized cannulas.