- Finding a vein can be difficult, and it may be necessary to use ultrasound to help identify a vein for cannulation and to use small-sized cannulas.

Feeding and swallowing

- A child's suck may be weak, and they may have difficulties chewing and swallowing. Signs include weight loss, poor appetite, choking, coughing, and prolonged mealtimes. Referral to speech and language therapy and a dietitian is necessary. If not consuming an adequate diet, dietetic team will consider nutritional supplements and vitamins.
- Feeding tube may be inserted to supplement oral food/fluid intake. Caregivers must be trained in use and care of tube.
- Acid reflux, where stomach acid flows back into the oesophagus, can occur. Symptoms include poor feeding, crying after meals, or coughing. Anti-reflux medication is recommended, with further investigation if necessary. Consider nissen fundoplication if medication is ineffective.
- Severe reflux can lead to aspiration pneumonia, where stomach contents enter the lungs, causing inflammation and respiratory issues. Seek urgent medical attention if struggling to breathe.
- Growth should not be based on general population and energy requirements should be calculated individually.

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, there should be a low threshold for X-ray due to poor bone density. It's essential for local team to contact specialist neuromuscular team regarding fracture treatment.

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MUSCULAR DYSTROPHY UK

Alert card

Spinal muscular atrophy type 1

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 1

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

Symptoms of SMA type 1 show within the first six months of life. Babies will have floppy limbs, be unable to raise their head or sit without support, and have difficulties moving, breathing, and swallowing. Many children with SMA will have normal cognitive development, but some may have cognitive impairments and developmental delays.

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. Zolgensma (Onasemnogene Abeparvovec) is available for SMA type 1. Risdiplam (Evrysdi) and Nusinersen (Spinraza) are available for SMA types 1, 2, or 3. These 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.

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 before surgery, aim to wean them back to their pre-op baseline.

Respiratory function

- Weak breathing muscles are common and may lead to nocturnal hypoventilation (shallow breathing at night) and a weak cough.
- Monitor respiratory function periodically; non-invasive ventilation (NIV) may be required. Use of NIV should improve sleep quality, reduce daytime sleepiness, and regulate oxygen and carbon dioxide levels.
- Check if the individual has an emergency care plan and follow advice. Signs of respiratory distress in children include fast or irregular breathing, flaring nostrils, wheezing, grunting, and changes in lip or skin colour.

Respiratory infections and acute medical care

- Increased risk of pneumonia and respiratory infections. Low threshold for the use of antibiotics is recommended. Anticipatory care plan for acute care is recommended.
- Oxygen therapy is often needed during hospitalisation or to ease symptoms. If using ventilator support, combine oxygen therapy with the ventilator. If giving oxygen to a child not on a ventilator, there is risk of suppressing their hypoxic drive to breathe, leading to CO_2 retention. This is dangerous and monitoring is necessary.
- Assess secretion management and consider cough augmentation techniques like assisted coughing, breath stacking with a LVR bag, and/or a cough assist device to clear lower airway secretions. Nebulisers, medication, and suction may be necessary for excess secretions.