Essential Information for Emergency Services when Assessing and Caring for a Child who has Spinal Muscular Atrophy

What is SMA

SMA is a rare, genetic, neuromuscular condition causing progressive muscle wasting (atrophy) and weakness leading to loss of movement. This may also affect strength, breathing and swallowing. There are different types of SMA and there can be a wide variation in the severity and impact in patients with the same type. SMA does not usually affect cognition.

This information sheet provides essential information for emergency services about:

- **Disease-modifying treatments**
- **Respiratory Care**
- **Nutrition Management**
- **Anaesthetic Precautions**
- **Fracture Management**

It's helpful to ask parents/carers which specialist neuromuscular centre oversees their child's care.

Disease-modifying treatments

These are now widely prescribed:
You will need to ask the parents / carers about their child's disease-modifying treatment.

- **Risdiplam / Evrysdi™** – daily, oral.
- **Nusinersen / Spinraza™** – 4-monthly, delivered via lumbar puncture.
- **Onasemnogene abeparvovec (Zolgensma™)** - this is a 'one-off' gene therapy; most children will require several months of oral steroids after their treatment which can in turn result in a risk of immunosuppression and adrenal suppression.

If on steroid treatment, check if there is an individual treatment plan and make sure that the advice in this is followed.

Respiratory Care

- This child may be known to a long-term ventilation team or respiratory medical or physiotherapy specialist.

- They may have an emergency care / respiratory ventilation / airway clearance plan.

Always check with the child's parents / carers and inform their neuromuscular and respiratory team of any A&E attendance.

- Be aware that, due to their muscle weakness or use of NIV, typical signs of respiratory distress may be masked for a child with SMA.

Always:

- **Take note of parental concerns and ensure rapid assessment by a senior member of the A&E team to determine whether the child is showing signs of respiratory compromise which include:**

  - Fatigue or altered level of consciousness.
  - Altered capillary blood gas.

- Consider whether ventilatory support is needed. Early support with non-invasive ventilation may be effective. In the majority of instances, BiPAP is preferred to CPAP - but needs to be delivered / coordinated by a specialist team with experience in the management of neuromuscular conditions.

- Note that high dose oxygen alone should not be given without blood gas monitoring in an unventilated child with SMA as it may suppress respiratory drive.

- If symptoms of respiratory distress continue or are severe, have a low threshold for intubation with urgent consultation with the child's specialist neuromuscular centre / respiratory team.

- Arrange chest X-ray and treatment with antibiotics.

- Monitor oxygen saturations and carbon dioxide clearance (capnography or blood gas).

- Review use of anticholinergics (e.g., oral glycopyrrolate or hyoscine), as these can increase the risk of mucous plugging and may need to be discontinued.

- Be aware that Respiratory Physiotherapy is a vital part of the management of SMA.

  - Ask parents/guardians for their child's most up to date respiratory physiotherapy programme and about their child's routine airway clearance programme.

  - If the parents are competent and confident, they can ask to assist with providing care themselves during their child's stay if clinically appropriate and on discussion with the child's physiotherapy/medical team.

Secretion clearance is critically important in the management of acute infection in SMA and early assessment by a respiratory physiotherapist is essential.

Respiratory physiotherapy techniques, nebulisers (saline, hypertonic saline, salbutamol, DNAse), antibiotics and suction may all be required to maintain airway patency.

Remember to review medical records which may contain patient-specific advice including care pathways.

Nutrition Management

- Remember to ask if this child has a Feeding Plan.
- As a result of low muscle mass, hypoglycaemia can occur after a short period of 'starvation'.
- Always check a BM / laboratory glucose level.
- Avoid prolonged fasting; have a low threshold for starting oral rehydration or intravenous fluids including dextrose if normal enteral feeding is not tolerated.
- Be aware that during a period of illness, gut motility may be reduced making a child at higher risk of vomiting or aspiration. The strain of coughing can also induce vomiting:
    - Consider a move to continuous or slower feeds.
    - Consider the use of a free-drainage bag on their gastrostomy or nasogastric tube if they have one to minimise their risk of vomiting.

Anaesthetic Precautions

SMA may increase the risk of complications with anaesthetics:

- Aspiration risk;
- Limited mouth opening;
- Non-reversible muscle relaxant agents should be avoided;
- Recovery of breathing may take longer, and children may need to be extubated to non-invasive ventilation;
- High dependency or intensive care facility should be routine post-surgery to allow for appropriate post-operative monitoring.

Always inform the child's Neuromuscular Centre of any planned procedure involving general anaesthetic.

Fracture Management

- **Children with SMA may have low bone density. Stress or low impact fractures may occur.**
- Have a low threshold for considering fracture in those complaining of bone pain or distress with movement or handling with a relevant history.
- An X-ray is recommended in all cases of suspected fracture.

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Version 1
Author: Clinical Experts with the SMA UK Information Production Team
Last updated: August 2023
Next full review due: August 2025

Published by:

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