Paediatric Neurology

Use of Salbutamol in Neuromuscular conditions

This leaflet is aimed at parents and carers of children who have a neuromuscular condition and may benefit from the use of Salbutamol.

What is Salbutamol?
Salbutamol is a drug which acts mainly on certain receptors (called β-2 receptors present in the smooth muscle of blood vessels, the lungs and the intestines. Salbutamol has been shown to improve muscle strength and function in children and adults with certain neuromuscular conditions.

Which neuromuscular conditions can Salbutamol be used in?
Salbutamol has been shown to be beneficial in Spinal Muscular Atrophy, some types of Congenital Myasthenic disorders and a type of congenital myopathy called Central Core disease.

Use of Salbutamol in Spinal Muscular Atrophy (SMA)
Spinal muscular atrophy (SMA), results from deletion of Exons 7 and 8 in the Survival Motor Neurone (SMN1) gene. The severity of the SMA depends upon the number of copies of the ‘back-up’ gene called SMN2 which produces a less effective protein. Data from observational studies has shown that Salbutamol may improve motor ability in children with SMA by increasing the function of SMN2.

Salbutamol is currently recommended for treatment in SMA Type 2 and 3.

Salbutamol can be considered in children with SMA from the age of three years. In some children, it may be started earlier if clinically indicated; this will be discussed with you in detail.

Salbutamol in Congenital Myasthenic Syndromes/Central Core disease
Salbutamol has also been found to be effective in children with DOK7 related Congenital myasthenia and Central Core disease by improving muscle strength. The mechanism of action is different from SMA and is thought to be via up regulation of the Beta-2 receptors.
What tests are needed before starting Salbutamol?

- Blood pressure
- Urea and Electrolytes
- ECG (a test looking at the electrical activity and rhythm of the heart)

These can be carried out when your child attends the neuromuscular clinic at Addenbrooke’s Hospital or they can be carried out more locally to you, if it is more convenient. An assessment of a child’s strength and current ability using a specialised scoring system will be carried out by a physiotherapist before starting Salbutamol. This will be a baseline score to compare with any future assessments.

What are the possible side effects of Salbutamol?

Salbutamol can cause tremor in some children. Many children with SMA type 2 or 3 have a pre-existing hand tremor, which may get worse. This is thought to be related to increase in Potassium (a type of salt) levels in the blood.

If the tremor is significant and is affecting your child’s day to day activity, a dose adjustment may be needed. Other side effects may include muscle cramps, palpitations and headache. Salbutamol can decrease the potassium (salt) level in the blood and can also (rarely) lower your child’s blood pressure.

How is Salbutamol used in neuromuscular conditions?

Salbutamol is usually given orally for neuromuscular conditions, (it is given as an inhaled preparation if used to treat asthma).

A liquid or tablet preparation can be prescribed, depending on your child’s preference. Your neurologist will tell you how often your child should take it. This may increase over time depending on benefits and side effects.

Monitoring the effect of Salbutamol

- You may notice a difference in your child’s strength and ability to carry out activities. Some parents notice that their child does not seem to tire as quickly and some children may even find moving much easier.
- It is important that a physiotherapist assesses your child’s function around six to eight weeks after starting Salbutamol. This will give you and your neuromuscular team a clear indication of whether or not there has been an improvement.
- Your child will be seen again by the neuromuscular team around three months after starting Salbutamol. You will be asked questions by your neurologist to assess if there have been any adverse effects. Your child will also need a repeat blood pressure, ECG and blood test.
- If your child has tolerated the Salbutamol well and their tests are normal, your neurologist may increase it further. Your child will require repeat blood tests and an ECG every once in a while or if their dose of Salbutamol is increased.
**How long will my child be on Salbutamol?**

Your child can be on Salbutamol as long as they are tolerating it and it is not causing any adverse effects. There may come a point where your neurologist will look at whether the Salbutamol is still having any sustained benefit and whether to continue or discontinue it.

**What happens if my child becomes unwell and cannot take the medication?**

If this happens it is important you inform your neuromuscular team or the medical team at your local hospital. They will assess whether a blood test is needed to check the levels of potassium in your child’s blood and check that it is normal. Further advice about what to do if your child is unwell will be given by your neurologist, if your child is started on Salbutamol treatment.

**Contacts/Further information**

If you have any further questions or queries regarding Salbutamol and your child, please contact Jo Musson, paediatric neuromuscular nurse specialist on 01223 254976.

Alternatively, if you would like any general support or information on SMA, please refer to SMA Support UK at [http://www.smasupportuk.org.uk/](http://www.smasupportuk.org.uk/)

Or for the use of Salbutamol in Congenital Myasthenic Syndromes, please refer to MGA: MyAware at [http://www.myaware.org/congenital-myasthenia](http://www.myaware.org/congenital-myasthenia)

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We are now a smoke-free site: smoking will not be allowed anywhere on the hospital site.

For advice and support in quitting, contact your GP or the free NHS stop smoking helpline on 0800 169 0 169.

**Other formats:**

If you would like this information in another language, large print or audio, please ask the department where you are being treated, to contact the patient information team: patient.information@addenbrookes.nhs.uk.

Please note: We do not currently hold many leaflets in other languages; written translation requests are funded and agreed by the department who has authored the leaflet.

**Document history**

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