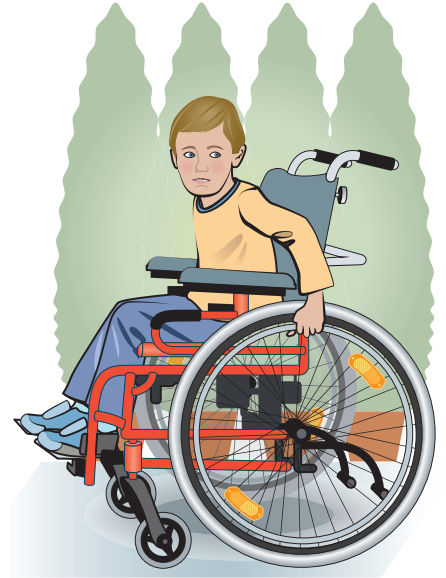


Treating Breathing Problems in Children with Neuromuscular Diseases

Mini-series 2 of 2

Neuromuscular diseases are diseases of the nervous system (nerves that control the body) and muscles. Some examples of neuromuscular diseases in children are: spinal muscular atrophy (also called Werdnig-Hoffman disease), Duchenne (doo-shen) muscular dystrophy, congenital muscular dystrophy and congenital myopathies. Most of these disorders are genetic or inherited (passed down in a family).



Neuromuscular conditions can cause breathing problems in several ways- directly and indirectly (because of related complications). To learn more about how neuromuscular diseases cause breathing problems see the ATS Patient Information Series “Breathing Problems in Children with Neuromuscular Disease, Part 1”.

This information sheet describes what can be done to treat common breathing problems seen in neuromuscular disease in children. Treatment depends on what specific disorder the child has and often is supportive with no possible cure. A specialist in neuromuscular disease such as a neurologist or geneticist can provide more information regarding treatment of the specific neuromuscular disease.

What can be done to treat breathing problems with neuromuscular disease?

Even in cases in which there is no cure for a neuromuscular disease, there are ways to help manage breathing problems that may improve your child’s health.

If the child has difficulty getting enough air and has low oxygen levels, there may be ways to help provide extra oxygen or other support for breathing. What options are possible for a child and how well they may work need to be discussed with a lung specialist. There are

children who need to use a machine (ventilator) to help them breathe. This could be done by a nasal or face mask or through a tracheostomy tube (a tube placed into the windpipe through the neck). Some children use this support only with sleep but those with severe weakness could require all the time. (see related *ATS Patient Information Series fact sheets, PAP in Children, Use of a Tracheostomy with a Child, and Sleep Problems with Neuromuscular Disease in Children*).

What can be done for scoliosis in neuromuscular disease?

Children with scoliosis from neuromuscular disease can have problems with lower lung function and airway clearance because the curve in the spine can effect the chest size and shape. If your child has scoliosis, he or she may benefit from use of a supportive chest jacket or surgery. Scoliosis surgery may be advised at times to help with other problems such as hip pain and sitting problems. It is important to consider how well your child may do with surgery and what breathing support may be needed for your child after surgery. An orthopedic surgeon can work together with your lung specialist to decide if scoliosis treatment may help lung function and how well your child may tolerate surgery.

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What can be done to prevent infections in neuromuscular disease?

Children with neuromuscular disease may have a weak cough that limits the ability to get mucus out of the lungs.

Airway clearance therapies such as a cough assist device and suctioning can help a child cough out mucus.

Good hand washing and avoiding ill contacts can help one avoid infection. Every child with neuromuscular disease should get a yearly influenza vaccine. Your child may also benefit from a booster pneumococcal pneumonia vaccine to help prevent infection from strains of *Streptococcus pneumoniae* bacteria. You can talk with your health care provider about whether your child needs a booster vaccine.

What can be done to protect the lungs if my child has swallowing problems with neuromuscular disease?

Children who cannot swallow well and are at risk of aspiration may require a feeding tube to provide nutrition. A speech pathologist can help evaluate your child's swallowing ability and if certain types of food or thickened liquids can be safely eaten.

How can you treat obstructive sleep apnea in neuromuscular disease?

Children with neuromuscular disease can be at higher risk for obstructive sleep apnea (OSA). Risk factors include

- Large Tonsils and/or Adenoids: Large tonsils and/or adenoids can block the airway.
- Obesity: Children who are very overweight are more likely to have sleep apnea.
- Problems with muscle tone: Children can have trouble breathing during sleep because the throat muscles relax and block the airway.

If your child has large tonsils and/or adenoids, he or she may benefit from surgery to remove them (called adenotonsillectomy). Symptoms of OSA should improve after surgery. However, because children with neuromuscular disease are at higher risk for OSA, your child will need to have another sleep study 2-3 months after surgery to be sure the sleep apnea is improved. If your child is overweight, talk to your health care provider about

a safe, effective weight control program. Children with neuromuscular disease and OSA often require support at night for breathing as was described earlier. (see related ATS Patient Information Series fact sheets, PAP in Children, Use of a Tracheostomy with a Child, and Sleep Problems with Neuromuscular Disease in Children).

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Rx Action Steps

- ✓ If your child has a neuromuscular disease, talk with your health care provider about symptoms to watch and how often tests should be done to check your child for breathing problems.
- ✓ Work with your child's health care provider to address conditions that can lead to more breathing problems such as difficulty swallowing or scoliosis.
- ✓ Discuss the risks and benefits of available therapy for breathing problems that your child develops.
- ✓ Wash your hands often and well when caring for your child. Use soap and water or alcohol-based hand sanitizer.
- ✓ Avoid having your child around people who are sick. Keep your child away from tobacco smoke.
- ✓ Have your child and caregivers get a yearly flu vaccine. Check to see if your child needs a booster pneumococcal pneumonia vaccine

Doctor's Office Telephone:

Other Resources

Muscular Dystrophy Association

<http://mda.org/disease>

The Cooperative International Neuromuscular Research Group

<http://www.cinrgresearch.org/aboutnd/diseases.cfm>

National Library of Medicine Medline Plus

<http://www.nlm.nih.gov/medlineplus/neuromusculardisorders.html>

Hull J, et al. British Thoracic Society guideline for respiratory management of children with neuromuscular weakness.

Thorax 2012;67:i1 e i40. Access at <https://www.brit-thoracic.org.uk/document-library/clinical-information/children-with-nmw/children-with-nmw-guideline/respiratory-management-of-children-with-neuromuscular-weakness-guideline/>

Interested in joining a patient-centered research network for sleep apnea? Visit www.MyApnea.org