



Caring for Children with
**Neuromuscular
Disorders**



American Family
Children's Hospital

uwhealth.org/kids

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OVERVIEW

This guide book has been designed for you, the family of a child who has a neuromuscular disorder (NMD). It is also for you to share with your local health care providers and others who will be providing care and support for your child in your local community. Some families find that learning about caring for their child with special health care needs can be overwhelming and even frightening. These feelings are to be expected.

This guide book will define how muscles help healthy lungs work. How NMD affects how healthy lungs work and what can be done to help your child's lungs work their best. How respiratory treatments move secretions to make breathing easier. We will also explore the role of good nutrition and how that impacts breathing. We will also look at nutritional supports and supplements.

Please keep in mind not all treatment options and techniques are used for every child. For many families, caring for your child at home requires a change in lifestyle and a big time commitment. Through experience, we have learned this can be overwhelming at times. We want you to know that you have options and the team will work with you to identify available support options.

Many families worry about the cost for medications, supplies, equipment and home nursing. Even with insurance, co-pays, deductibles and benefit caps, the cost of care for your child can be overwhelming. There are many financial aid programs for children with special health care needs. These programs vary depending on where you live. Equipment and services providers will be able to identify the costs of their services which will not be covered through insurance or Medicaid waiver programs. Team members are available to help answer questions about programs that may be available.

Some families find it helpful to talk with other families who are happy to share what their experiences have been and the choices they have made. We can connect you with another family if you would like to explore that option. Also included are resources located on the internet.

Using this guide book, the staff at American Family Children's Hospital will lead you through the day to day cares and help you to feel more confident safely caring for your child with NMD.

THANK YOU!

This booklet was created by Craig Becker and Rhonda Yngsdal-Krenz in collaboration with the University of Wisconsin Pediatric Pulmonary Center. As we partner with patients and families, it is our hope to work together on their life's journey. We want to especially thank the Poole and Kuester families for helping us with the creation of this booklet and adding their thoughts that come from the hearts of their children. Thanks to all the children for sharing your beautiful smiles. We would also like to give a special thanks to Dr. Mary Schroth who is passionate about her work with the neuromuscular population and works tirelessly to provide care and support to patients and families.





Figure A



Figure B

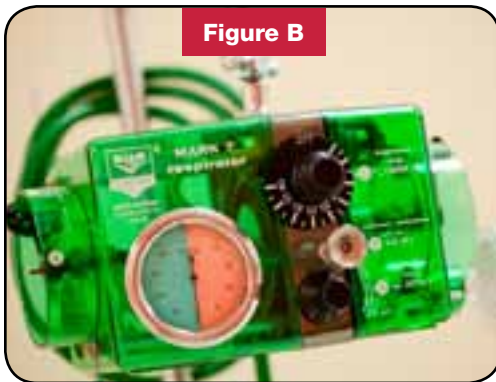


Figure C



Figure D



PROBLEMS WITH BREATHING

The Respiratory System plays an important role in maintaining health. This is especially true for children who have a Neuromuscular disorder (NMD).

Breathing Muscles

There are two important muscles for breathing. The diaphragm is the large muscle at the bottom of the rib cage. The diaphragm works by pulling the rib cage down so the lungs expand. The intercostal muscles are the muscles between the ribs. These muscles help to lift up and expand the shape of the rib cage. The diaphragm and intercostal muscles work together when you breath in (inhale). In healthy lungs both the top and the bottom of the ribcage expand when you inhale.

Lungs that expand normally help the rib cage to grow and maintain a normal chest wall shape. For lungs to work their best, they need a chest wall shape that allows lungs to fully inflate.

Different NMDs affect the muscles for breathing in different ways. However, the possible complications are the same.

Under-Developed Lungs and Chest

Children affected with NMD early in life are at risk for under developed lungs. In NMD's that affect children under the age of 2 years, the chest wall and the lungs may not fully develop because the muscles for breathing are not strong enough to take in big breaths. Big breaths help the lungs grow and expand the chest wall.

Weak Cough

Every day the nose and the lungs make mucus or phlegm, also called secretions. Secretions are moved out of the nose and lungs by coughing, clearing the throat or swallowing. During a cold or illness, more secretions are produced in the nose and lungs.

In children with NMD, secretions are more difficult to clear because of their respiratory muscle weakness. This respiratory muscle

weakness interferes with their ability to cough. If secretions are not cleared with coughing, they will collect in the small airways of the lungs and cause a blockage called plugging. Plugging of the small airways can make some of the air sacs collapse, this is called atelectasis. Plugging of the airways can also cause pneumonia.

Shallow Breathing

During normal breathing, we inhale or take in oxygen and exhale or let out carbon dioxide. During sleep, muscles relax, including the muscles we use for breathing.

Children with NMD have muscles that are weak. This puts them at risk for shallow breathing during sleep. Sometimes their breaths become so shallow that they do not take in enough oxygen and they do not exhale enough carbon dioxide. As a result, the oxygen level in their blood drifts down and their carbon dioxide level goes up. This is called hypoventilation.

FAMILY-CENTERED CARE

Your health care team is not only concerned about your child, but the entire family and will focus on family-centered care. You, your child, your family and your health care providers will work together as a team. Our desire is for each team member to participate in the decision making process, provide feedback and suggestions to come up with the best options for your child as well as for the entire family.

CARE OPTIONS

There are different care options available to help improve the function of the Respiratory System in children with NMD.

Expanding Under-developed Lungs

Because the lungs cannot inflate completely, the lungs may not fully develop. Under-developed lungs can lead to decreased lung function. Underdeveloped lungs can be expanded with daily breathing exercises. There are

three ways to exercise and expand the lungs.

Resuscitator Bag and Mask

A resuscitator bag (Figure A) and mask may be used to give a larger breath to your child when breathing in. The mask is placed over the nose and mouth and the bag is squeezed to give a larger breath to your child when breathing in. This is repeated for several breaths. Your care providers will teach you how, when and how often to do this.

Intermittent Positive Pressure Breathing (IPPB)

IPPB (Figure B) is a machine that gives a deeper breath when the child breathes in than they are able to inhale on their own. The breaths are given by a mask over the nose and mouth, or through a mouthpiece.

Cough (Assist) Machine

The Cough Machine (Figure I) can be set to deliver an inhale breath only by turning the time for the exhale to 0. Four sets of 5 inhale breaths can be given every day to help stretch the lungs and chest wall. The Cough Machine can also be used to strengthen a cough.

Airway Clearance Techniques

One of the ways to help remove secretions is to loosen them in the airways before coughing. The techniques available to loosen secretions include chest physical therapy (CPT), postural drainage (PD), intrapulmonary percussive ventilation (IPV), and Vest Therapy. No one technique has been proven to be better than any other; they all mobilize secretions. The best option for your child is the one that helps remove the most secretions and is well tolerated.

Chest Physical Therapy (CPT)

CPT (Figure C) means using your hands or palm cups to clap on different parts of the chest while positioning the child to help move secretions from that part of the lung. A mechanical percussor (Figure D) is a vibrating device that does the same thing

and is sometimes used in place of hands or palm cups.

Postural drainage (PD)

PD (Figure E) means positioning the child so the secretions in the lungs will drain with gravity. Typically the child will lay on their tummy, back or side with the lower half of the body positioned higher than the chest. This allows the secretions to flow with gravity from the smaller airways to the larger airways where secretions can be removed by coughing or suctioning.

Intermittent percussive ventilation (IPV)

IPV is a mechanical device (Figures F & G) that provides mini bursts of air into the lungs at a set rate and pressure using a mask or mouthpiece. Medication or saline is placed in the nebulizer cup to moisten the air going into the lungs. This is like performing CPT to the inside of the lungs. The machine sounds like a choo-choo train.

Vest Therapy

Vest therapy (Figure H) is a machine that delivers air at a set frequency or rate and pressure to the chest vest. The chest vest is worn by the child and the air vibrates the chest loosening the secretions in the lungs. The chest vest looks like a life jacket with hoses that connect to the machine.

Strengthen the Cough

After the secretions are loosened in the lungs, the secretions need to be removed by coughing. Providing a stronger cough can be met in two ways.

The Cough (Assist) Machine

This machine blows air into the child's lungs for a brief set time giving a bigger breath (Figure I). This is followed by the machine pulling the air out of the child's lungs for a set time. This is like taking in a deep breath and then coughing. The breaths are given using a mask over the nose and mouth, or through a mouthpiece.



Figure E



Figure F

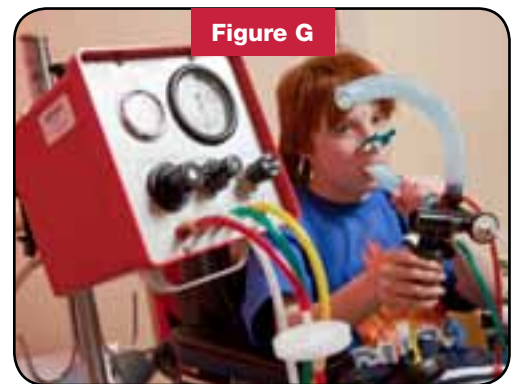


Figure G



Figure H

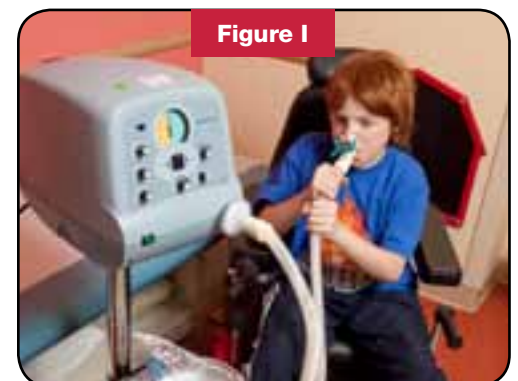


Figure I



Figure J

Manual cough assist

Manual cough assist requires someone else to assist with coughing. Manual cough assist means that someone uses their hands to apply pressure under the child's diaphragm during coughing. This will increase the force of the diaphragm to move air out of the child's lungs.



Figure K

Removing Secretions

Secretions that have been coughed up into the back of the throat or pool in the nose need to be removed. This can be accomplished by suctioning.

Bulb suction

Bulb suction (Figure J) is a balloon shaped rubber device with a long narrow tube on one end. The bulb is squeezed and the long narrow tube is gently placed into one nostril with the other nostril held shut. Once the bulb is released, secretions are pulled into the bulb.



Figure L

Suction machine

The suction machine (Figures K & L) generates a negative pressure that pulls the secretions out. The secretions are removed through a small tube that is gently placed into the child's nose and/or mouth when needed.

Pulse Oximeter

Sometimes it may be difficult to tell when your child is starting to have trouble breathing. A pulse oximeter (Figure M) may tell you there is a problem before your child shows signs of needing help. A pulse oximeter is a machine that tells how much oxygen is in the blood without drawing any blood.



Figure M

A soft Band-Aid like cover (probe or sensor) is placed on your child's finger or toe. This probe has a red light that shines onto the child's skin. It senses the amount of oxygen in the blood and digitally reads a number in percent or oxygen saturation. Normal oxygen saturation is 94% or above. If the oxygen saturation is less than 94%, the child probably needs to be coughed. The oximeter also reads the heart rate, which can assist with determining if the child is having trouble breathing.



SHALLOW BREATHING OPTIONS

Children with NMD often have low oxygen levels and high carbon dioxide levels especially when they sleep. There are two main options to deliver bigger breaths or ventilation during sleep and allow the respiratory muscles to rest.

Non-invasive Ventilation

Non-invasive ventilation (Figure N) means that nothing has to be placed inside of the body. These respiratory assist devices use a small mask that is placed over the nose or the nose and mouth. The mask is held in place by a strap or head gear. The mask is attached to a hose that connects to a bi-level positive airway pressure (BiPAP) assist device or a mechanical ventilator. These machines give more air into the lungs than the child can take on their own while sleeping. When the child breathes in, the machine gives a deeper breath by delivering air into the lungs up to the set inspiratory positive airway pressure (IPAP). When the child exhales, the pressure drops so that exhalation is easier. Some of the pressure is left in the lungs (expiratory positive airway pressure or EPAP) to help keep the lungs inflated and prevent the lungs from collapsing. This machine is set to assist the child's breathing. A breathing rate is also set so your child receives a certain number of breaths each minute. This helps to rest the muscles for breathing, especially during sleep.

These respiratory assist devices can also be used during the day to help with shortness of breath. A mouthpiece can replace the mask. Nose clips may be needed to help prevent air from leaking out of the nose.

A great deal of time may be needed to help your child become comfortable with using the respiratory assist device. Respiratory therapy and your home care equipment company will work with you and your child to become more comfortable.

Complications of Non-Invasive Ventilation

Some complications have been noted when using non-invasive ventilation.

Breakdown of facial skin

Sometimes your child's face may have red tender spots or areas where the skin is torn. These pressure points happen if the nasal mask is worn for long periods of time or is placed too tight on the face. There are ways to prevent pressure sores from happening. If your child is using the nasal mask continuously, release the mask from the child's face every two hours. Keep the skin free of oils and wash the nasal mask before wearing. In addition, skin protective coverings such as Duoderm, foam tape or elastogel can be used to redistribute the pressure over a larger area.

Flattening of facial features

Sometimes children who wear the nasal mask for more than 14 to 16 hours per day will develop flattening of the nasal bridge and upper lip and gums. If the child can tolerate different mask shapes or nasal pillows, this may prevent the changing of facial features. It is best to rotate the masks worn for respiratory assist devices.

Invasive Ventilation

Invasive ventilation means that something is placed inside the body. Invasive ventilation requires the placement of a tracheostomy tube along with breathing support from a mechanical ventilator.

Tracheostomy tube placement

A tracheotomy is a surgical procedure where a small hole (stoma) is made through the skin and into the trachea or wind pipe that allows a breathing tube called a tracheostomy tube (trach tube) to be placed (Figures O & P). An Ear Nose and Throat (ENT) doctor makes this opening during surgery. Breathing through a trach tube means that air moves directly into the trachea instead of through the nose and mouth. Because the trach tube enters the trachea below the voice box (larynx), it is important to understand that there are changes that occur after a child

receives a tracheostomy tube:

- Less able to talk or make noise.
- The nose cannot filter the air the child breathes.
- The upper airway cannot moisten (humidify) the air the child breathes.
- The upper airway cannot warm the air your child breathes.
- There is a higher risk of breathing things directly into the lungs. These things might include water, dust, powder, or aerosols.

Care is needed for the tracheostomy tube on a regular basis such as cleaning, changing of the tube and removing secretions or suctioning. The tracheostomy tube allows easy access for attachment to a ventilator.

Mechanical Ventilator

Mechanical ventilator (Figure Q) or respirator is a machine that can either assist or take over the child's breathing. This machine is more complex than BiPAP and can be set to deliver a specific size and number of breaths per minute. It also has more options for additional breathing support. Ventilators that are used in the home are smaller and more portable than hospital ventilators. Training in the hospital is required for families and care providers to learn about the ventilator and the trach tube to ensure safe use of these pieces of equipment in the home and community.

For more detailed information on tracheostomies and mechanical ventilation, please request the *Trach Training and Discharge Manual* from the Pediatric Pulmonary Center at American Family Children's Hospital.

Speech Therapy

A tracheostomy does not change or remove the voice box (larynx). The larynx contains the vocal cords. Most children with uncuffed trach tubes can make sounds with the trach tube in place. A child's ability to speak with the trach tube in place will depend on the amount of airflow around the trach tube. It also depends on whether the child's vocal



Figure N



Figure O



Figure P

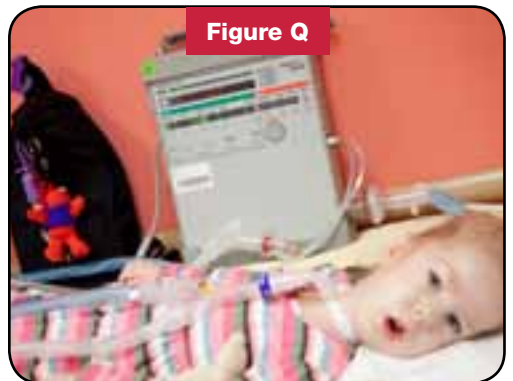


Figure Q



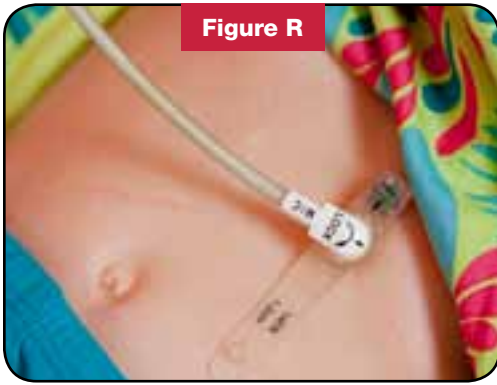


Figure R

cords are able to move. Most children have softer voices than usual when the trach tube is in place. The voice is softer because less air is able to flow around the trach tube and between the vocal cords. If your child has a cuffed trach tube he or she may not be able to speak because the cuff fits snug and does not allow the air to flow around the trach tube and between the vocal cords. The child's pulmonary or ENT doctor will be able to explain how well the child may be able to speak or vocalize.



Figure S

Something to keep in mind is that the non invasive and/or invasive ventilation interventions are not for everyone and not every family can do this for a variety of reasons. In addition, some children do not do well with the non-invasive breathing support or require the breathing support 24 hours per day. Requiring breathing support for 24 hours per day greatly impacts the dynamics of your home and the lives of those within the home. As the caregiver, you will make the best decision you can for your child. Your child's healthcare team will be there to assist and support you.



Figure T

NUTRITION

Maintaining good nutrition is key to slowing muscle breakdown. Eating small amounts of food frequently and keeping up with fluids will help balance the energy that is needed for breathing and muscle strength.

Weight issues

Being overweight or underweight can be an issue for children with NMD. Energy and calorie needs may be lower due to lower muscle mass and decreased movement.

Overweight

Being overweight can occur easily as children are not moving around as much and are less active. Calorie needs are typically lower due to the lower muscle mass and decreased movement. These needs can be 35-40% less than children without NMD.

Underweight

Being underweight can be a result of a lack of appetite, difficulty chewing and swallowing food or because more calories are being used for breathing than they are able to eat.

Supplements

Often children with NMD require special diets and supplements that help ensure the child is receiving the proper nutrients to slow down muscle loss and prevent constipation and dehydration.

NMD reduces your child's ability to move about freely, causing them to be more at risk for decreased bone density, osteoporosis (weak and brittle bones), fractures (bone breaks) and scoliosis (curving of the spine). Given the reduced bone density, it is important to encourage weight bearing as tolerated and to provide as much physical activity as possible. Taking vitamin D and calcium supplements also helps keep the bones stronger. It is also important to be sure there is adequate protein in the diet to help reduce the weakening of the muscles.

Chewing and Swallowing

Chewing and swallowing food can be difficult due to the weak muscle tone. For children too weak to chew or swallow all their calories by mouth, a tube can be placed through the skin and into the stomach. This tube can be used to give a nutritionally complete liquid formula directly into the stomach to give more calories.

Gastrostomy Tube (G-tube)

For children who are too weak to eat or swallow, a gastrostomy tube or G-tube (Figure R) can be surgically placed. A G-tube is a small feeding tube placed through the skin and the stomach wall, and directly into the stomach. The tube allows the child to get liquid nutrition and medicines. The G-tube can also be vented to let extra air or fluid out of the stomach.



Figure U



Figure V

Gastro-esophageal Reflux Disease (GERD)

Gastro-esophageal Reflux Disease (GERD) is the movement of food and acid from the stomach back up into the esophagus or stomach tube. GERD causes people to spit up or vomit food or liquids. This is also known as acid reflux. GERD is a common problem for children with NMD. Because they have weakened muscle tone, it is easier for food and liquid to come back up. If the child has difficulty swallowing, the refluxed food can go into the lungs causing irritation and/or an infection in the lungs. GERD can be treated with medications. However, for weaker children with NMD, it may be safest for them to have a surgical procedure called a Nissen fundoplication; often referred to as Nissen.

With a Nissen, the surgeon wraps a portion of the child's stomach around the bottom of the esophagus creating a one-way valve that allows food to go into the stomach, but makes it difficult for food to go back up the esophagus.

Constipation

Constipation can be caused from decreased muscle strength making it harder to physically pass the stool. Also if there is not enough water in the diet, stools can be harder and even more difficult to pass. A diet lower in fat may be helpful. In addition, there are medications that may be used.

MUSCULOSKELETAL HEALTH

Children with NMD are at risk for developing joint contractures (joints that cannot fully straighten or bend) and loss of range of motion. The joint capsule, tendons, muscles and ligaments can all lose their flexibility if they are not able to actively move through their entire range. When joint contractures happen they can make daily living tasks, such as dressing, bathing, and transfers more difficult. If a muscle has some joint contractibility, contractures will make movement more difficult for the weak muscle.

To help avoid contractures, daily stretching activities are necessary. Your physical or occupational therapist will work with you and your child to learn how to safely move a joint through its entire range of motion. Incorrect positioning during stretching can cause pain so it is important to use good techniques. Bracing and splinting are also recommended to provide a more prolonged stretch (Figures S & T). Good positioning is important to help prevent contractures. To help fit range of motion exercises into an already busy day try to break up activities throughout the day. Using songs or games will also make stretching more enjoyable and feel like play.

Play is an important way for all children to gain cognitive skills, interact with peers and enjoy themselves. Creative adaptations allow children with NMD to maximize their active movement for play. Switches and assistive devices let children with very limited active motion to independently operate toys and access computers. An occupational therapist can help to adapt games and other toys to let children with limited strength participate. Sensory play with different textures such as sand, fleece, beads, water, or gooey mixtures let children explore their environment with their hands. Toys with lights, sounds, or vibration also provide multi-sensory input. Some communities have recreation departments offering adapted baseball, soccer or basketball to individuals who use wheelchairs. In the school system, adaptive physical education teachers, occupational therapists, or physical therapists can help adapt recreational activities so children with neuromuscular disorders can play with their peers.

Adaptive equipment will be important to help provide support necessary for performing daily tasks such as toileting, bathing, and safe transfer. Different types of equipment are available to meet the various needs. Due to the progressive nature of NMD and growth of the child, equipment needs change over time. Occupational and physical therapists are

good resources for helping to determine what equipment is most appropriate. Home care equipment providers will help match adaptive equipment to the needs of your child and family. They can also help to determine how equipment can be funded.

Physical Therapy

Children with NMD have significant muscle weakness and do not develop strength with age. As the child grows, a greater demand is placed on the already limited muscle strength. With an imbalance of muscle strength and the inability to overcome gravity, children often have jug-handle positions of their arms and frog positioning of the legs.

A Physical Therapist can assist your child with the following:

- Strengthening and range of motion
- Maintain skin elasticity
- Maintain muscle length and strength
- Maintain joint integrity
- Improve circulation (blood and lymphatic)
- Improve body awareness/sensory input
- Provide opportunities to use muscles in gravity eliminated positions like suspension systems and in bath tub/pool

Splints

Splints help hold joints and muscles in position to decrease pain and contracture development (Figure U). Examples include:

- Night splints for positioning (resting hand splints or knee splints)
- Day splints for function (neoprene hand splints, ankle, foot orthotics [AFO])
- Weight considerations of splints (especially during day)

Equipment

(Figure V)

- Wheelchairs: power or manual
- Stander
- Bathroom and shower chair
- Car seat
- Additional sitting positions: tumbleform, positional chairs

TREATMENT AND INTERVENTION PLAN AT A GLANCE

Managing care at home is both time consuming and complex. When you and your child are at home, it is sometimes difficult to know when to call the physician and how to intervene. It is important for you to feel supported at home when there are changes in your child's baseline condition. We have created a tool that you can use to assess how well your child's lungs are working and when to communicate with the medical team. When you are in doubt, call the pulmonary center and describe symptoms and interventions that have been taken. In order to help assess your child, we have created a list of things to consider. Points are given for each of the things on the list. When the points are added up, you will have a number that allows you to see how often airway clearance treatments are needed,

the use of BiPAP and what action needs to be taken. The number also relates to a care zone that ranges like a stop light from green to red. The yellow and red zones alert you to changes in your child's baseline and the steps that need to be taken to monitor your child more closely.

- The green zone 0-4; your child is doing well.
- The yellow zone 5-11; your child is not doing as well as usual but is still able to do most daily activities.
- The red zone 12-16; your child is sick and you need to contact the pulmonary center.

The following two pages provide a quick reference tool to assess your child and guide you with the needed intervention.

Respiratory Treatment and Intervention Plan

This is a tool to help you decide how often treatments are needed and when to contact your healthcare team for intervention and support.

Points	0	1	2	3	Total Points
Respiratory Rate The number of breaths in a minute Normal:	Normal	5 over normal	10 over normal	Greater than 10 over normal	
Heart Rate The number of times the heart beats in a minute. Normal:	Normal	10 over normal	20 over normal	Greater than 20 over normal	
Measured Oxygen Saturation	Greater than 94%	91%-94% and improves after cough	91%-94% and does not improve after cough	Less than 90%	
Oxygen (if applicable)	No oxygen	Using less than 1 lpm	Using 1 to 3 lpm	Using more than 3 lpm	
Breath Sounds	Clear	Noisy in the throat but clears after a cough	Noisy in the chest but clears after a cough	Noisy and does not clear after a cough	
Color	Normal	Pale	Pale and blue tinged lips	Blue tinged lips and face	
Work of Breathing	Normal	The sides of the nostrils are moving in and out with each breath (flaring)	The skin around the ribs are moving in and out with each breath (retractions)	A grunting noise is heard with each breath (grunting)	
Secretions with Cough Machine	None	Small amount	Moderate amount	Large amount	
Mental State	Alert	Sleepy	Sleepy but can be woken up	Listless and sleepy	
Cough	None	Rare	Few	Many	

Frequency Guidelines

	Total Points	BiPAP	Treatments
Green	0-4 points	Usual Use	Do therapy as your doctor prescribed in the home
Yellow	5-8 points	Usual Use	Increase therapy so your child receives two extra therapies a day and watch for cold like symptoms
Yellow	9-11 points	Usual Use plus two extra hours during the day and at night	Call your Doctor and report the change. Increase therapy so your child receives four therapies during the day
Red	12-15 points	On all the time with 2 hour breaks twice a day	Call your Doctor and report the change. Plan for a hospitalization. Increase therapy so your child receives therapy every four hours
Red	≥ 16 points	On all the time	Call your Doctor and report the change. Plan for an emergency room visit.

PROTOCOL FOR MANAGEMENT OF AIRWAY SECRETIONS DURING A COLD

Children with NMD can have a very hard time clearing mucus from the lower airway during a cold. More frequent use of non-invasive support, the cough machine and breathing treatments are required. Follow these airway clearance steps in the order shown below:

- Every four hours and as needed during a cold:
 - Airway secretion mobilization for 10-20 minutes.
 - Cough machine, four sets of five breaths to remove the loosened secretions followed by suctioning secretions from the mouth.
 - Postural drainage for 15-30 minutes, which means lying with his bottom higher than his chest so that gravity helps to move the secretions out of the lungs.
 - Cough machine, four sets of five breaths to help remove the remaining drained secretions followed by suctioning secretions from the mouth.
- Use the cough machine every time your child sounds rattley or has trouble coughing out secretions. You cannot use this machine too much. If your child has difficulty getting secretions out from the back of the throat, also suction the mouth and nose.
- If your child is on non invasive support or a ventilator, use the machine while sleeping including naps during the day. Because your child is weaker during a cold, he may also need to use breathing support while awake.
- Use a pulse oximeter to check your child's oxygen saturations as needed when well and more frequently when sick. If the oxygen saturation is less than 94%, use the cough machine to clear secretions and recheck the oximetry. If the oxygen saturation is 92% or less and the cough machine is not increasing the oxygen saturation, place your child on BiPAP, do airway clearance followed by the cough machine, drainage and cough machine and contact the pulmonary center.

If your child's oxygen saturation does not remain greater than 90% while on BiPAP without oxygen, your child may need to be in the hospital for more intense therapy. Follow your emergency plan for contacting the pulmonary center or call 911.

Nutrition Intervention Plan

If your child is not tolerating feedings or vomiting, this can lead to dehydration. This can be a very serious complication. The pulmonary center should be contacted immediately.

DURING A HOSPITALIZATION

During each admission, your child is evaluated by the health care team in order to tailor a treatment plan that will meet your child's needs. Protocols are used as a guide. Therapy will be given with modifications when necessary. Hospitalizations can be very stressful and can create an extra burden on you as the care providers. We encourage you to let the medical team take charge of your child's care plan with your insights and suggestions. We hope this will be a time when you can be your child's parent and not have to balance the roles of both parent and care provider.

We understand and respect that you are the expert on your child; however, when your child is hospitalized, it is important to remember that what works at home to move your child through a crisis may not work in the hospital. The hospital may use different methods than you use at home. For example, medications may interfere with optimal airway clearance or the need

for sleep is balanced with the need for frequent airway clearance. The hospital is surrounded with medical professionals that will work with you to create a treatment plan.

Intubation

During an acute illness, another decision may be whether you prefer to have your child intubated (placement of a breathing tube) should your child become very sick. One of the things you and your doctor should consider is whether the illness and difficulty breathing is a sudden change due to a cold and may be reversible, or whether it seems to be a result of your child becoming gradually weaker overall.

The child will be sedated during the procedure to place the breathing tube (endotracheal tube or ET tube). The breathing tube will pass between the child's vocal cords so the child will not be able to talk while the breathing tube is in place. The breathing tube will be attached to a breathing machine or ventilator that will help the child breathe more comfortably.

The Respiratory Therapists will be following guidelines to help assist your child with daily therapy. To help prevent getting pneumonia while on the ventilator, your child's head will initially be raised up and will not be placed in the drainage position. The cough machine will be used to help remove secretions. Other devices may be used to help loosen the secretions in the lungs. Aggressive airway clearance will provide the greatest chance for success in having the ventilator and ET tube removed. This process is called extubation.

Extubation

Extubation is having the ET tube removed. The removal of the ET tube will occur once the medical team feels the child is strong enough to do more of the breathing on his own. Once the child is extubated, breathing support by nose mask (BiPAP) will be initiated and aggressive airway clearance will continue.

If your child does not tolerate breathing support by nose mask despite the best efforts of the healthcare team another difficult decision has to be made. The decision at this point is whether to:

- Re-intubate and wait to see if the child will become stronger and try again to extubate.
- Consider tracheostomy tube placement.
- Consider providing no breathing support and use comfort care measures only.

There are no right answers and as a parent, you will make the best decision that you can for your child. This decision is best made prior to an illness or a stressful situation.

SUPPORT OPTIONS

Families will need to determine which respiratory and medical interventions are best for their child and family. Families may or may not choose interventions based on their personal beliefs and definition of quality of life. You may choose some interventions for your child and may not want others. You may set some criteria as to what interventions you are willing to try and for how long. You may choose quality of life with no interventions beyond comfort care.

Comfort Care

The healthcare team can help you to explore options of pain free (palliative) care and hospice.

Palliative Care

Palliative care will treat you and your child's physical, emotional and spiritual needs. There are three main goals to ensure the best possible quality of life:

- Reduce pain and symptoms caused by NMD and/or therapy.
- Support you and your child during this transition and loss.
- Help your family provide support to your child and each other.

Hospice Care

Hospice care can also assist during end of life care. You and your child's wishes will be met. The goal will be to keep your child free of pain and comfortable.

There is no correct answer and the decisions are your family's personal choice. The option that works best for you and your family today may not be what is best in the future. As your child's disease progresses and care needs change, we are happy to help you think through the options available to you and your family. When patients and families choose the options that work best for them, we firmly believe that they are acting in their child's best interest and we will support those decisions.

HOME

When you bring your child home from the clinic or hospital, you are not alone! You will still have contact with the pulmonary team, the home care equipment provider, the home nursing agency, your child's local doctor or pediatrician, and others. All of these people are available to answer questions and to help you to care for your child at home. There will be clinic appointments with the pulmonary team to monitor your child's progress and overall health. Your child will see his local doctor or pediatrician for regular visits. Your home equipment provider will make routine home visits to check your child's home care equipment and supplies and to answer any questions you may have. Most parents find it helpful to keep business cards and phone numbers for their health care team in one place. Keeping a notebook or binder with information and phone numbers is a great way to stay organized and keep all of your child's resources and supports in one central place.

FINANCIAL PROGRAMS FOR EQUIPMENT AND SUPPLIES

Many families worry about the cost of home care equipment, supplies, and home nursing. Even with insurance, the co-pays, deductibles and benefit caps can quickly add up. Before your child goes home we will work with you to arrange financial support. There are many financial aid programs for children with special health care needs. Some of these programs start paying where your insurance leaves off. Your social worker will help you to explore the financial programs available to you.

To qualify for these programs, it will need to be determined that your child is not able to participate in usual activities because of a physical condition that is expected to last at least 12 months. Children with NMD can often qualify because they need special care that is usually provided only in a hospital setting.

The following is a short description of some of the programs used by many families of children with NMD's who live in Wisconsin.

Medical Assistance (Medicaid)

Medicaid is for families with a limited income and assets. It provides insurance coverage for children with special health care needs.

Katie Beckett

Katie Beckett program is a medical assistance waiver program that does not look at family income and assets to determine who is eligible. Coverage is determined by the medical needs of your child. It will help cover costs for home care equipment, supplies and services.

Supplemental Security Income (SSI)

SSI is for children who are determined to be disabled and their family income falls below a certain level. Your home and one automobile are not considered in determining your eligibility. To qualify, combined family savings, stocks and

retirement accounts cannot exceed a specified amount.

There are other financial programs that may be available to you and your family. Feel free to talk with your social worker to learn more about the programs and how to apply.

RESEARCH

Gallant efforts are being made towards finding a cure for NMD. As a hopeful health care team, we too will do our part in working towards a cure. You and your child may be asked to be a part of a research project. This decision will be up to you and your child and will not impact the care you receive.

WEBSITES

There are many websites that will help you on your journey. Here are just a few that you may find helpful:

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

This website provides an overview of the different types of Neuromuscular Disorders.

<http://www.mda.org/>

This national website is dedicated to fighting over 40 diseases that affect people of all ages.

<http://www.fsma.org>

Families of Spinal Muscular Atrophy—a comprehensive, on-line information and resource center. The website provides information on the annual conference, research & clinical trials, family experiences, legislative action, news letters and more.

www.curesma.org

Linked with the Families of Spinal Muscular Atrophy site, this is a fundraising site to help find a cure.

<http://jcn.sagepub.com/cgi/content/abstract/22/8/1027>

Consensus Statement for Standard of Care in Spinal Muscular Atrophy, Journal of Child Neurology.

<http://www.fsma.org/FSMACommunity/MedicalIssues/Respiratory/>

Breathing Basics: Respiratory Care for Children with SMA.

<http://www.fsma.org/FSMACommunity/NewlyDiagnosed/>
Caring Choices: For Parents of Infants Newly Diagnosed with SMA Type I.

MEETING YOUR HEALTH CARE TEAM

There will be a team of health care workers involved in your child's care. Please remember, you and your child are important members of our team. We want to work with you to provide the best care for your child. The healthcare team includes:

- **You, the parents or caregivers.**
- **Your child.**
- **Other family members** such as siblings, grandparents, aunts, uncles and friends.
- **Pediatric pulmonary doctors** will manage the care for your child's lung issues.
- **Pediatric orthopedic doctors** will assess for scoliosis and when to do surgery and evaluate whether other surgeries may be beneficial for muscles and joints to work optimally.
- **Orthotics** fit braces to support the back and spine and to help ankles stay in position.
- **Pediatric rehabilitation** will assess mobility and function for daily living skills and toiletry needs and help determine ways to maximize function.
- **Vocational rehabilitation** will assess school concerns and the schools ability to provide services to the child.
- **Physical and occupational therapy** help with maintaining flexibility of the joints, assessing bracing and equipment needs, and optimizing mobility.
- **Wheelchair provider** helps with devices such as specialized seating systems, wheelchairs and scooters.
- **Advanced practice nurses** have advanced education and expertise in a specialty area, such as pediatric pulmonary medicine.
- **Health psychology** has expertise in helping children and families cope with chronic illness.
- **Home care specialists** or **Durable Medical Equipment (DME) provider** will show you how to use home care equipment before leaving the hospital. They will also visit your home to check on your child's equipment.
- **Research coordinator** will assist families with education and research participation.
- **Social workers** help families with emotional, physical and economic concerns related to your child's illness, treatment, hospitalization, discharge and return home. They can refer you to financial and social service resources, and coordinate with agencies near your home.
- **Staff nurses** provide your child's daily care while in the hospital.
- **Respiratory Therapists** evaluate and treat your child's breathing problems.
- **Registered Dietitians/Nutritionists** evaluate your child's diet and maintain a proper weight through special diets and nutritional supplements.
- **Pharmacists** prepare and deliver medications and explain how to use the medications.
- **Your child's primary care doctor** or **pediatrician** will see your child for routine child care and well child visits.
- **Home nursing agency** will provide home nursing if needed.
- **Spiritual care** such as ministers, chaplains, priests, rabbis and other clergy who can offer support and healing.
- **Child life specialists** help children and families adjust to the hospital or stressful situations or procedures through therapeutic play.
- **Other community resources** such as school staff, local utilities and EMS (emergency medical services) will work with you and the team to make sure your child is safe in your community.





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