

## RESPIRATORY CARE OF SMA AND CHOICES

### I. IDENTIFYING THE PROBLEM

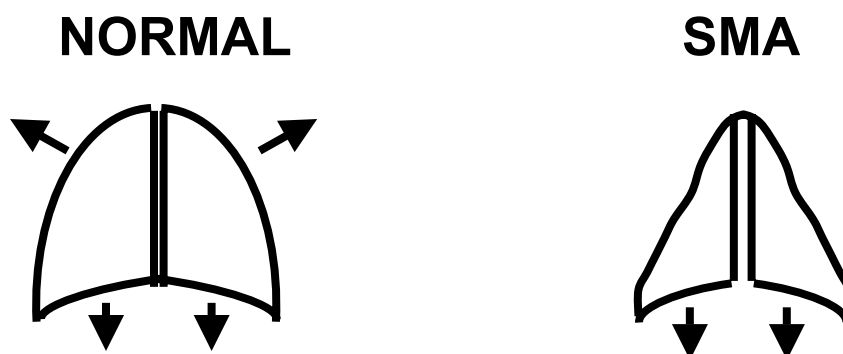
Respiratory care is critically important for individuals with spinal muscular atrophy and affects both their survival and quality of life. SMA affects the muscles for breathing as well as the muscles for moving the arms and legs.

#### NORMAL BREATHING MUSCLE FUNCTION

The muscles important for breathing include the intercostal muscles and the diaphragm. The intercostal muscles are the muscles between the ribs. These muscles help to lift up and expand the shape of the rib cage thereby helping the lungs to inflate. The diaphragm is the large muscle at the bottom of the rib cage. The diaphragm works by pulling the rib cage down in an almost opposite direction of the intercostal muscles, and also helps the chest wall and lungs to expand causing air to fill the lungs. What you see in someone without SMA is that the ribcage inflates with air at the top of the ribcage as well as at the bottom of the ribcage during inhalation.

#### SMA BREATHING MUSCLE FUNCTION

SMA causes very weak intercostals muscles, the muscles between the ribs. The strongest breathing muscle in SMA is the diaphragm. As a result, children with SMA type I and type II appear to breath with their stomach muscles. The belly or abdomen pooches out while they take a breath in. The top of the rib cage does not appear to move. In infants less than a year of age, the rib cage is very soft even though it consists of the ribs, which are bones and cartilage. Over time the rib cage may appear as though the bottom of the ribcage is pulled down. This gives the appearance of a bell shaped chest meaning that the top of the chest is narrow and the bottom of the chest is much wider. Sometimes the area of the sternum or the bone in the front that the ribs attach to becomes sunken. This is called pectus excavatum and happens because the intercostals muscles are not strong enough to help pull the top of the rib cage out against the diaphragm that is pulling down.



WHAT DOES THIS MEAN?

Individuals with SMA have respiratory muscles that are at a mechanical disadvantage. Because the lungs cannot inflate fully, the lungs may not fully develop. In addition, because the muscles are weak, the cough is very weak, and it is difficult to take deep enough breaths while sleeping to maintain normal oxygen levels and normal carbon dioxide levels.

## LUNG UNDERDEVELOPMENT

Lungs that are not fully developed may contribute to decreased lung function as the child grows. Normal lung expansion helps the rib cage to grow and maintain a normal chest wall shape. A normal chest wall shape provides the best mechanical advantage for the lungs to work optimally. One of our goals for the respiratory care of children with SMA is to provide maximum lung and chest wall development.

## WEAK COUGH

During viral respiratory infections, the nose and the lungs make a lot of secretions. In addition, everyone is weaker during a cold. If the secretions are not cleared from the lungs by coughing, the secretions will collect in the airways and cause plugging of the small breathing tubes of the lungs. If the secretions become infected this will result in pneumonia. Plugging of the small breathing tubes of the lungs results in collapse of some of the air sacs and this is called atelectasis. A consequence of atelectasis is decreased oxygen saturation, which means that there is not enough oxygen in the blood for the body to maintain normal functions. Oxygen saturation can be determined easily using a pulse oximeter.

## SHALLOW BREATHING DURING SLEEP

During sleep, the muscles relax including the muscles for breathing. During normal breathing, we breathe in oxygen and exhale carbon dioxide. When someone with muscle weakness falls asleep, their muscles for breathing also relax and their breaths become more shallow. Sometimes those breaths become so shallow that they do not inhale adequate oxygen and they also do not exhale enough carbon dioxide. This is called hypoventilation. As a result, their oxygen saturations during sleep may drop to less than 94% and their carbon dioxide level may increase.

## **II. WHAT CAN BE DONE**

### LUNG DEVELOPMENT

Lung underdevelopment can be overcome by giving insufflations or big breaths daily using any of the following: a resuscitator bag and mask, or a cough machine on the inspiratory setting only or an intermittent positive pressure breathing (IPPB) device. This exercises the muscles for breathing and expands the lungs to a larger volume than the person can do on their own.

1. Using the resuscitator bag and mask, the mask is placed over the nose and mouth and a breath is given to the person while they are inhaling. Several breaths are given over several minutes to help inflate the lungs.

2. A cough machine with an automatic cycle is a device that can be set to shift from inhalation mode to exhalation mode and then a pause before the next breath. The breaths are given by either a mask over the nose and mouth or a mouthpiece. Using the cough machine in the inhalation mode only, the lungs can be inflated to a set pressure. (Please see additional information below in Improve Cough.)
3. IPPB gives a breath in synchrony with an individual to a set pressure with a mouthpiece or a mask over the nose and mouth. A similar effect can be achieved using the cough machine.

Incentive spirometers typically given to adults after a surgery are not helpful to individuals with SMA because the spirometer inflates the lungs but only to the volume the person can do already. There is no extra expansion or benefit to using this device in individuals with SMA type I and II.

## IMPROVE COUGH/AIRWAY CLEARANCE

Coughing up secretions can be improved dramatically by using a cough machine or having someone else use their hand to assist with coughing. Manual cough assist means that another person uses their hand to apply pressure under the diaphragm in synchrony with the coughing person. This results in increased force of the diaphragm to move air out of the lungs. The cough machine inflates the lung to a set pressure for a set time following by sucking the air out of the lungs at a set pressure for a set time. This is very similar to what occurs during coughing in stronger people. When you cough, you inhale a big breath, close your throat, increase the pressure inside your chest, open your throat and then the air bursts out of the lungs. Typical settings for the cough machine are inhale pressure 30 to 40 cm of H<sub>2</sub>O for 1 second, exhale pressure -30 to -40 for 1 second and pause time 1 to 2 seconds. Four to 5 breaths are taken continually followed by suctioning the mouth to remove secretions coughed up and a rest period of 1 to 2 minutes. Four sets of 5 breaths is a typical treatment. The cough machine cannot be overused for the purpose of coughing and should be thought of as the way the person with SMA coughs. There is no other device that helps to clear the secretions out of the lungs. The cough machine is available through your home care durable medical equipment (DME) provider. The cough machines are known by brand as the In-exsufflator Cough Machine or the newer Cough Assist.

## AIRWAY SECRETION MOBILIZATION

When a person with SMA has a cold, the secretions build up and are difficult to cough out. One of the ways we help remove secretions is to loosen them in the airways before coughing. The techniques available to loosen secretions include manual or mechanical percussion, postural drainage, intrapulmonary percussive ventilation (IPV), and the Vest Airway Clearance system. No one technique has been proven to be better than any other; they all mobilize secretions.

1. Manual or mechanical percussion means using your hands or palm cups to clap on different parts of the chest while positioning the person to optimally drain that part of the lung. Mechanical percussion refers to using a vibrating device applied to the chest to loosen secretions.

2. Postural drainage means positioning the person so that the secretions in the lungs will drain with gravity. Typically the person will lay on their tummy, back or side with their bottom higher than their chest. This allows the secretions to move with gravity from the smaller airways to the larger airways.
3. IPV is a mechanical device that provides positive pressure by mask or mouthpiece into the airways at a set frequency and pressure that can be varied as tolerated. Medication or saline is nebulized with IPV. This is mechanical percussion from the inside of the lungs. The machine sounds like a choo-choo train.
4. The Vest Airway Clearance System is a mechanical device that delivers air at a set frequency and pressure to the chest through a vest that the individual wears. This device is expensive and has not been proven to be more effective than other modes of airway secretion mobilization.

## BREATHING SUPPORT OPTIONS

Individuals with SMA type I and many individuals with SMA type II have hypoventilation while they sleep at night. This means that they exchange carbon dioxide and oxygen at lower levels than is necessary for the body to maintain normal function. Their oxygen saturations drop and their carbon dioxide levels increase. To improve this problem, patients with SMA have to receive bigger breaths during sleep than they can do on their own. There are several options to accomplish better ventilation (bigger breaths) and oxygenation while sleeping including BiPAP, mechanical ventilator, and negative pressure ventilation. During illnesses the respiratory support device may need to be worn throughout the day also. There is no role for supplemental oxygen in patients with SMA until **ALL** other options have been exhausted.

1. BiPAP refers to bilevel or two level positive airway pressure. This is usually delivered through a mask over the nose. BiPAP provides a higher pressure and volume of air into the lungs during inhalation and inflates the lung greater than what the person can do without the BiPAP machine. During exhalation, the BiPAP pressure drops so that air can passively leave the lungs. The BiPAP machine can sense when the person is taking a breath and gives the breath in synchrony with the individual. A respiratory rate is also set so that the BiPAP will give breaths even when an individual with SMA is in a deep sleep and taking shallow breaths. The person can breath above the BiPAP rate and the BiPAP will deliver more breaths. CPAP (continuous positive airway pressure) should never be used in patients with SMA. Typical BiPAP settings in an individual with SMA include are inspiratory positive airway pressure (IPAP) 14-20, expiratory positive airway pressure (EPAP) 3-6, respiratory rate based on age of 14-30, inspiratory time based on age of 0.5 to 1.5 seconds, and respiratory rate and rise time based on comfort.
2. Mechanical ventilators or respirators come in a variety of models. The most portable model that is available is the LTV (laptop ventilator). This ventilator can be carried over the shoulder or placed on a wheelchair for transport. Portable external batteries are also available. Mechanical ventilators are more complex, but also allow for control of more variables. The ventilator can be set to deliver a specific size breath and a set number of

breaths per minute. Mechanical ventilation can be delivered with a nose mask, mouthpiece while awake, or through a tracheostomy tube.

3. Negative pressure ventilation refers to providing breaths into the lungs using a large chamber or tank that encircles the chest. The chamber is connected to a vacuum pump that takes the air out of the chamber and as a result the chest wall expands to bring air into the lungs. A Porta-Lung is an example of a negative pressure ventilator. It can be set to deliver a specific number of breaths per minute and a vacuum pressure.

### **III. PROTOCOL FOR MANAGEMENT OF AIRWAY SECRETIONS DURING A COLD**

During a cold, individuals with muscle weakness can have a very hard time clearing mucus from their lower airway. **You can help them by following these steps:**

1. **These steps should be done in the order shown below** every four hours and as needed during the cold.
  - a. Airway secretion mobilization for 10-20 minutes.
  - b. Cough machine, four sets of 5 breaths to remove the loosened secretions followed by suctioning secretions from the mouth.
  - c. Postural drainage for 15-30 minutes, which means laying with their bottom higher than their chest so that gravity helps to move the secretions out of the lungs.
  - d. Cough machine, four sets of 5 breaths to help remove the remaining drained secretions followed by suctioning secretions from the mouth.
2. 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 of the back of their throat, also suction their mouth.
3. If your child is on BiPAP or breathing support by nose mask, use the machine every time that they are asleep including naps during the day. Your child is weaker than usual during colds. They may also need to use the breathing support machine while they are awake.
4. Use a pulse oximeter to check your child's oxygen saturations as needed when well and more frequently when sick. Often you may not be able to tell when your child is having difficulty breathing and the pulse oximeter may tell you there is a problem sooner and that they need help coughing. 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 or equivalent and give airway clearance treatment and please contact your physician. If you cannot maintain oxygen saturation greater than 90% while on BiPAP and in room air, your child may need to be in the hospital for more intense therapy.

If your child is vomiting and not tolerating nutrition, they may become dehydrated. This can be a very serious complication in children with Spinal Muscular Atrophy. Please contact your physician.

## IV. CHOICES

**Non-invasive** respiratory care management means helping a child to breathe using a mask over the nose that is connected to a BiPAP machine or a mechanical ventilator. **Invasive** respiratory management refers to the use of a breathing tube that is placed in the mouth and passed between the vocal cords and into the lungs. **Invasive** respiratory management also includes the placement of a surgical hole in the neck to the large airway (trachea) for a tracheostomy tube. A tracheostomy tube bypasses the mouth and vocal cords and goes directly from the skin to the trachea. A respirator or ventilator is connected to the endotracheal tube or tracheostomy tube.

### SMA TYPE I

Children with SMA type I have a huge variation in muscle strength and their breathing support needs are difficult to predict. Almost all children with SMA type I benefit from supportive ventilation with BiPAP or a ventilator using a nasal mask while they sleep starting at an early age, 6 months for example. The breathing support needed while healthy ranges from no breathing support while sleeping to needing BiPAP while sleeping to needing breathing support 24 hours per day, 7 days per week with or without a tracheostomy tube. During colds all children require additional breathing support and may need to use their BiPAP or ventilator 24 hours per day until the cold goes away.

Parents of children with SMA type I have many very difficult decisions. First is whether you wish to pursue non-invasive breathing support per nose mask for your child. The methods and equipment although generally effective are also a tremendous amount of work, especially when the child has a cold. In addition, the child may not tolerate the nose mask breathing support.

Another decision is whether you desire to have your child intubated should they become very sick especially with a cold. Remember, everyone becomes weaker with a cold. 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 the child becoming gradually weaker over all. Using an aggressive airway clearance and extubation (removal of the breathing tube) protocol followed by placing the child on breathing support by nose mask provides the greatest chance for success. Some physicians are not aware of the protocol and tools that are available. However, when a child with SMA type I has a breathing tube placed one of the risks is whether the child will succeed in having the breathing tube removed and using breathing support by nose mask when the illness has resolved. If the child does not tolerate breathing support by nose mask despite the best efforts of the doctor another difficult decision has to be made. The decision at this point is whether to:

1. Wait and see if the child will become stronger and try again.
2. Consider transferring to a hospital that has a lot of experience with non-invasive respiratory care if your hospital does not.
3. Consider tracheostomy tube placement.
4. Consider letting your child go.

There are no right answers and as a parent, you will make the best decision that you can for your child.

Something to keep in mind is that the non-invasive methods for respiratory care of children with SMA are not for everyone and not everyone 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 24 hours per day breathing support by nose mask may be very difficult for the child as well as for the family. Again, you as the parent will make the best decision you can for your child.

## SMA Type II

Children with SMA type II should never require a tracheostomy, and in general are strong enough to be managed using non-invasive respiratory care techniques on a day to day basis. This means that when the child is healthy, they may use BiPAP or a ventilator with a nose mask or negative pressure ventilation while they sleep, and the Cough Assist device as needed. Some children with SMA type II do not require support for their breathing while they sleep. When a child with SMA type II develops a cold, they may need to use their nose mask breathing support more often and sometimes continuously. Remember that during a cold they are weaker than usual, but their strength improves after the cold has resolved. During a cold they need respiratory care treatments every 4 hours with airway secretion mobilization, cough machine use and postural drainage. The cough machine may be needed more often to clear the mucus. If oxygen saturation cannot be maintained at 92% or greater on BiPAP, they may need to be hospitalized and may need to have a breathing tube placed and attached to a ventilator. However, through aggressive use of airway secretion mobilization and airway clearance, the tube can be removed and the child placed on ventilation by nose mask and then weaned back to using the breathing support while they sleep only.

## SUMMARY

The respiratory care for children with SMA is critically important. The necessary tools include a cough machine, a method for airway secretion mobilization, a method for performing postural drainage, and a pulse oximeter to help monitor their oxygen saturations. In addition, a method for breathing support including a BiPAP machine or mechanical ventilator are also very important. For individuals with SMA type II, aggressive respiratory care using non-invasive techniques improves quality and quantity of life. For children with SMA type I, the non-invasive respiratory care techniques are beneficial and generally effective. Please keep in mind that not all techniques fit every child.