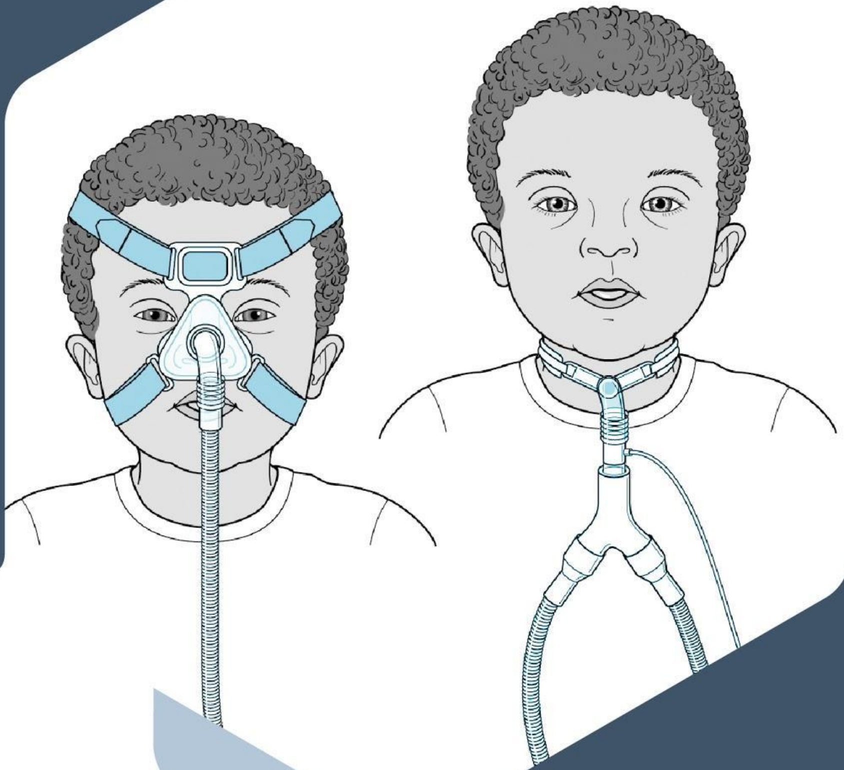

When a child may use a ventilator for a long time

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This booklet is for families of children who may use a breathing machine (ventilator or **vent**) to help them for a long time. It gives you info about this medical problem and the treatment options. It also talks about how to make decisions about these things if needed.

Topics Covered

This info comes from parents of children with this problem and medical experts. It is for families with children with many different reasons for this problem. So you should ask your medical team (doctors, nurses, respiratory therapists, and other providers) any specific questions that you have about your child. The medical team will probably have other important things to talk about with you—not just what is in this booklet. If your child can understand what is happening to him or her, you and the medical team should talk about all of this with your child. This should be done in a way he or she can understand.

What is chronic respiratory failure?

- *A medical problem that prevents a person from breathing on his or her own without help.*
- *Often a vent is needed to help with breathing.*
- *There are many causes of this problem, some of which can get better.*
- *There is a range of how bad it can be.*
- *When a person does not use a vent, serious problems can happen.*

When a child cannot breathe on his or her own and there is no short term reason for this (like an infection that can be treated), the child has “chronic respiratory failure.” This means the child has a problem getting rid of carbon dioxide and sometimes cannot get enough oxygen. “Chronic” means it will likely last a long time, usually months, years, or sometimes the child’s entire life.

There can be many reasons for this problem. These include:
- abnormal or damaged lungs
- abnormal or damaged airways (the tubes that air travels through to the lungs)
- abnormal heart
- weak breathing muscles
- abnormal chest wall
- abnormal or damaged brain or nerves that control breathing.

A child can have more than one reason for this problem. Your medical team should be able to explain your child's reason(s).

A child's breathing usually can be helped with a ventilator (**vent**) and other machines and treatments (see below). However, a vent cannot cure the problem. Some children can get better because the reason for the breathing problem gets better. If the medical team thinks your child's breathing can get better, they will say so. Even if it can get better, it is hard to guess how long it will take or how much better it will get.

There is a range of how bad this breathing problem can be. Some children use a vent to breathe for them (life-support). Some only use a vent for extra help. Some children only use the vent when asleep; others use it all day. Some use a vent more than usual when they have a cold or are sick.

For many children, the breathing problem gets better or stays the same. Some children’s breathing gets worse over time, depending on the reason for their breathing problem and if the child gets other illnesses. So a child can start out only using the vent during sleep but over time need it more and more until he or she uses it all day and at night.

If a vent is not used for a child who needs it, serious problems can happen. These can include: difficult breathing, discomfort with breathing, poor weight gain, less energy, problems with school work, headache, worse or more lung illnesses, heart failure, and death.

How can a vent help my child?
What can a vent do and not do?

- *A vent is a machine that pushes air into the lungs to help breathing.*
- *A child gets the help from the vent through a trach, nose prongs, or mask.*
- *Using a vent can have several benefits for a child.*
- *There are limits and risks with using a vent.*

A vent is a high-tech machine that breathes for or helps a person's breathing by pushing air into the lungs. Vents used outside the hospital have batteries that allow a child to travel outside the home. Every few hours the vent needs to be charged. Some children get help from a vent through a tracheostomy or **trach** (a tube that goes through the front of the neck into the windpipe) (see Figure 1).

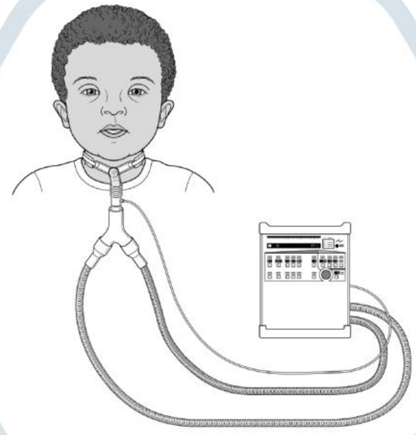

Figure 1.
Trach and vent

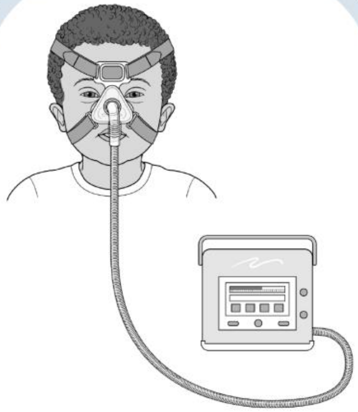

Figure 2.
NIV

Others get help through nasal prongs (short tubes that fit just inside the nose) or a mask that fits over the nose or nose/mouth; this is called noninvasive ventilation (**NIV**) (see Figure 2). Which way your child should get help from a vent depends on many factors that can be explained by the medical team. Vents can provide more or less help. The medical team choose vent settings based on what your child needs.

Using a vent at home can have health benefits, depending on the child. For some children, a vent breathes for them and provides life-support. For other children, benefits can include:

- Making breathing more comfortable
- Help with growth and weight gain in a young child and more energy in an older child
- Living outside the hospital and traveling outside the home
- Making it easier for the child to join in play and school activities
- Fewer lung illnesses or infections or helping the child better handle lung illnesses so that he or she does not have to go to the hospital

However, a vent cannot do some things:

- A vent cannot cure any medical problem
- Even with the help of the vent, many children still get serious lung illnesses. Vents sometimes cannot give enough help during a serious illness, meaning the child may have to go the hospital.

If your child uses a vent for life-support, you must be ready to handle vent emergencies. For example, if your child becomes separated from the vent, he or she must be put back on quickly or he or she could die. This can happen when the vent tubing detaches from the trach, the trach comes out, or the mask/prongs are not on the face like they should be. This is why a child who uses a vent for life-support is watched all the time.

Vents have alarms to warn of some problems and emergencies. So alarms cannot be ignored. Many children will have other monitors (like an oxygen monitor) to provide extra safety.

To be safe, you must know how to help your child's breathing by hand, using "low-tech" equipment. Also, many children who use a vent at home cannot clear their lungs and windpipe of mucus. Mucus can build up and can suddenly block air getting into the lungs, which can cause death. You must know how to help clear and suction out your child's mucus.

Vents rarely break. Families have a back-up vent when their child uses a vent for life-support.

There are other risks depending on if a child uses a trach and vent or NIV that are talked about in those sections below.

What other equipment might my child use?

- *Other equipment is needed to care for a child who uses a vent at home.*
- *Most children use a G-tube for feeding.*

To care for a child who uses a vent at home, other medical equipment and supplies are needed. This usually includes a self-inflating bag and mask, suction machine, heated humidifier, and oxygen monitor (pulse ox) (see Figure 3). Some children also need oxygen, "cough assist machine", chest vest, carbon dioxide monitor, and wheelchair. Some children already used some of this equipment before using a vent.

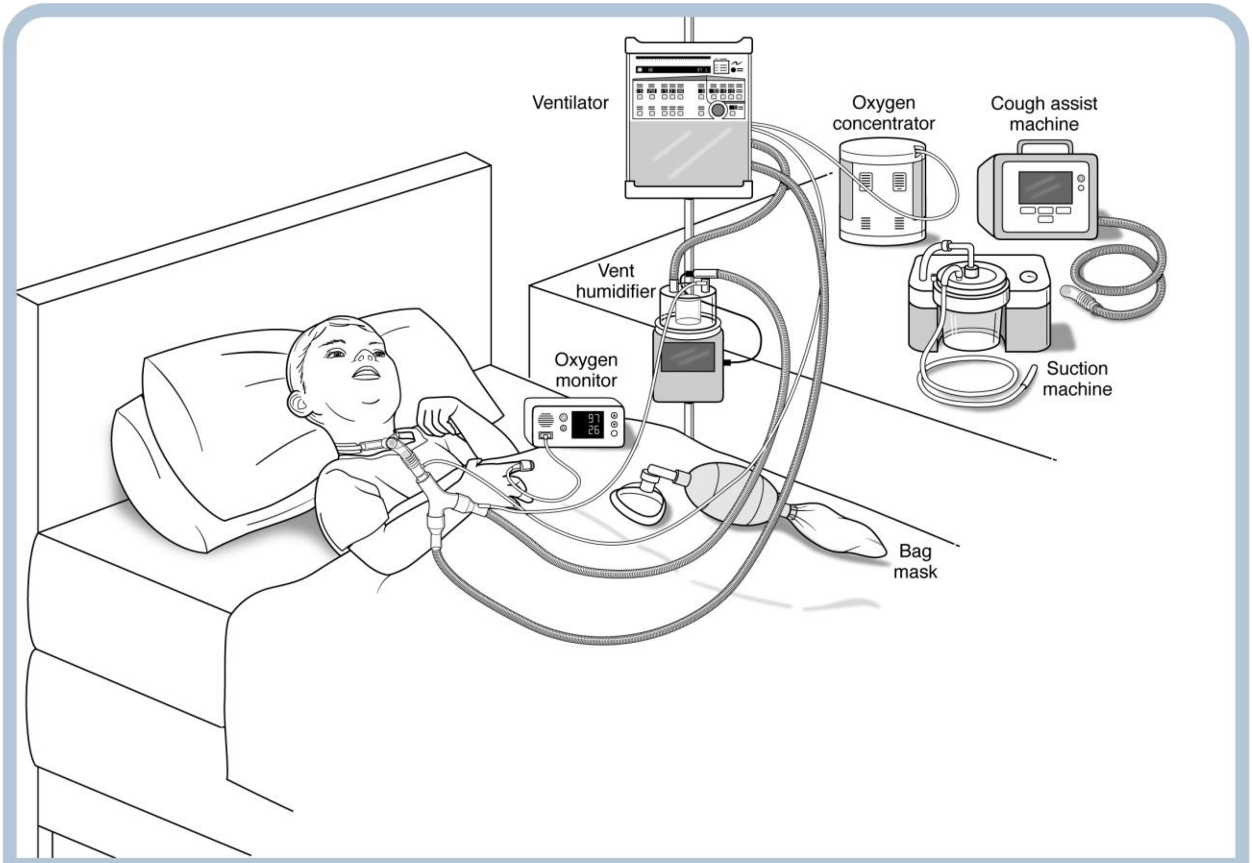

Figure 3. Equipment commonly used at home for child who uses a vent

Most children who need a lot of help from the vent or use the vent most of the day cannot safely eat or drink by mouth. They need a feeding tube to be fed safely. The most common type of feeding tube is a gastrostomy tube (**G-tube**). A G-tube is a plastic tube that goes through the skin of the stomach area into the stomach. A surgery is needed to put in a G-tube.

The medical team can give more detail about what equipment your child needs. You will be trained on how to use all equipment before (or for some things shortly after) going home.

For a child in the hospital, how long does it take to get home with a vent?

- *There are many steps before your child can go home using a vent, most importantly learning to care for your child on your own.*
- *These steps can take weeks or longer.*
- *Some children will go to a transitional care or rehab hospital before going home.*

If your child is started on a vent in the hospital, there are many steps before your child can safely leave the hospital. These steps include:

- Making sure your child is healthy enough to leave the hospital.
- Being trained to care for your child, to use your child's equipment, and to handle emergencies.
- Showing that you can care for your child without help from the medical team.
- Having your home inspected to make sure it is safe for your child.
- Arranging home nursing and delivery of home equipment and supplies.

These steps often take many weeks. When your child is leaving with a trach and vent, these steps take longer than when a child leaves using NIV.

In some hospitals, your child is sent to a rehabilitation (rehab) facility for some of these steps after your child is healthy enough to leave the hospital and before going home. Some children need to go to a rehab hospital for more rehab (like physical therapy) before going home. Sometimes, if a family cannot care for their child at home, the child will stay in a transitional or rehab hospital until the child no longer needs the vent. Rarely, when it is not possible for a family to care for their child at home, the child will live in a chronic care home after leaving the hospital or rehab. Families can visit their child regularly in a chronic care home.

How does using a vent at home affect the child and family?

These sections give more info about how using a vent at home may affect your child and family. It is not possible to cover every possible thing. So you should ask questions.

Some hospitals try to have a meeting between you and an “expert” family who already have a child using a vent at home. This meeting gives you a chance to hear about what their life is like and ask them questions. It can be hard to find an expert family with a child that is the same age and has the same medical problem as your child. This meeting can be face to face or over the phone. These meetings are less common when NIV may be started.

How does using a vent at home affect the child?

- *It is hard to know exactly how using a vent for a long time will affect your child.*
- *Many children who use a vent have a good quality of life.*
- *Using a vent can cause social and emotional problems for some children.*

Using a vent affects each child differently. Sometimes the child’s other medical problems affect the child more than using a vent.

Some of the benefits of using a vent are talked about above. Overall, studies on the quality of life of people who use a vent for a long time have mixed results. In other words, quality of life is good for some people and poor for others. Or quality of life is good in some ways and poor in others. Some older children and adults who use a vent say they have a better quality of life than others think they do. Some older children and adults say the vent is a good thing that helps them breathe and keeps them alive. These people make using a vent a “normal” part of their enjoyable lives. But many children who use a vent just cannot tell us about their experience with it because they are too young or cannot communicate.

When a child has a serious or life-limiting illness (one that cannot be cured and will shorten life), many families want to know how long will their child live with the help of a vent. Often the medical team cannot guess how long because a vent only helps a person’s breathing and cannot cure or treat other problems. Also, children who use a vent for life-support or are very fragile can die suddenly. Many of these deaths are due to sudden illness, but some are due to accidents (like the vent separating from the child, or not being able to clear mucus from the windpipe or trach).

Using a vent can lead to social and emotional problems for a child, depending on the child’s age and mental ability (attention, thinking, memory). A child can feel ashamed and lonely because the vent or trach makes him or her different (how he or she looks, what he or she can do, who he or she can play with) from other children. A child can feel upset about losing privacy because he or she needs others for everything from simple things (toileting, dressing) to treatments (suctioning). Their medical problems and these challenges can make them feel sad or depressed.

How does having a child who uses a vent affect the family?

- *Having a child who uses a vent is difficult and stressful for a family.*
- *It takes a huge amount of time and attention to care for a child who uses a vent.*
- *Families can feel separated from others and have money troubles.*

Having a child who uses a vent is difficult and stressful. This stress can cause problems for many parts of a family's life. Some families can have these challenges even if their child does not use a vent at home because of their child's other medical problems.

There are many reasons why it is difficult and stressful:

- Most of all, caring for a child who uses a vent is a lot of work. Most children must be watched 24 hours a day by an awake, focused adult, who can handle emergencies. They need many treatments and medicines every day. Getting used to having a child who uses a vent is very hard when the child first goes home with a vent.

- Families say they feel like both a parent and a nurse and that their home can feel like a hospital.

- Families can feel alone and separated from the community. They can have less contact with other people. Other people may not know how to act or relate with a family with a child with a serious medical problem.

- A child who uses a vent needs lots of care and support outside the hospital. But community resources can seem disorganized and not enough. It can be annoying and take a lot of time to plan and get the care a child needs. For example, there is often not enough home nurses to meet the needs of families. There are even less resources for adults who use a vent at home. So families can have an even harder time when their child becomes an adult.

- When a child is medically fragile or has a problem that will shorten his or her life, families can worry about their child's future health. When a child becomes suddenly ill and/or is in the hospital, a family's stress goes up and their routine gets messed up.

- Many families have money troubles. Even with health insurance, they usually have to use their own money to pay for medical expenses and supplies. Plus, one or more family members usually has to stop working or work less to care for their child.

The demands and stress of caring for a child who uses a vent can be hard for families in other ways:

- Travel can be very hard, because of all the equipment, supplies, and the treatments that the child needs.
- Family members often do not sleep well or long enough. This can make them feel tired during the day.
- Family members can feel scared, angry, powerless, guilty, and sad. Family members can have mental health problems (like depression) and problems with their relationships.
- Brothers and sisters of the child who uses a vent can have their own stresses and difficulties. For example, they can feel jealous or left out because the family spends so much time caring for the child who uses a vent.

If a family member worries about these challenges, he or she should talk about them with the medical team, especially the social worker. Social workers may be able to help find resources or services that can provide more help to the family.

Even with all these demands and difficulties, families usually do not say they would make a different decision. Many say their child makes their family life better. Families often get help from extended family, friends, and online groups of other families with children with similar problems.

Families who have a child who uses a vent often say "new" families will get used to things. They suggest staying strong and thinking about the good things both for themselves and their child.

How do I care for a child on a vent at home?

- *Families will be trained to care for their child who uses a vent.*
- *Most children get home nursing to help with their care.*

You will be trained to care for your child and use all his or her equipment. Not all hospitals teach the same things or in the same way. Most families will be trained to handle emergencies. This includes ways to try to restart a heart and give breaths with low-tech equipment (CPR or cardiopulmonary resuscitation).

When possible, 2 adult family members are fully trained, especially when the vent is life-support for their child or it is dangerous if their child becomes separated from the vent or cannot clear mucus without help.

At first, families feel overwhelmed by the amount they have to learn and by how their child's care is their responsibility. Before long, you will be an expert at caring for your child.

Families usually have home nursing to help at home. Home nursing can mean different types of paid home health professionals. Types include nurses, licensed practical nurses, aides, visiting nurse service workers, and others.

There are several things to know about home nursing:

- Your child will "qualify" for a certain number of hours per week of home nursing. The number of hours is decided by your insurance company. The number of hours your child qualifies for may be less than you hope for. Families and home nursing agencies try to "fill" those hours. There are often not enough home nurses to fill all the hours, plus nurses can call out sick. You have to care for your child when there is no home nurse. The number of hours your child qualifies for may increase or decrease over time depending on your child's needs.

- Home nurses have a range of skills and experience. Some have more experience than others caring for a child who uses a vent. Sometimes home nurses are trained differently from how families are trained, and families have to show the nurse how they like their child to be cared for. There can be high turnover (many changes) among home nurses. Sometimes, families find a nurse who is very capable, devoted, and who becomes like family.

- Some families ask other family members or friends (usually non-medical people) to help care for their child. Some families use their own money to pay for additional home nurses.

- Having nurses or non-family members in the home means you lose some privacy.

You need a plan for what to do in case there an emergency in your area (a disaster plan), like no electricity in your home for a long time. This plan should include things like:

- knowing how long batteries last in each piece of equipment
- knowing where you can go with your child that is safe and has electricity (like a friend's or family's home, hotel, or public safe place in another town)
- a list of emergency phone numbers
- a "go" bag with supplies, medicines, and formula for at least 3 days

What is long-term invasive ventilation?

- *Long-term invasive ventilation is help from a vent through a trach.*

This section is about breathing support from a vent through a trach. We suggest you read this section, even if the medical team thinks your child may only need NIV. This is because NIV cannot support some children over time, meaning their families and medical team might one day have to decide whether to have a trach placed.

What is a trach and how can it help my child?

- *A trach is a short curved tube that goes into a hole made through the neck to the windpipe.*
- *Often a child will have a different short term breathing tube in the windpipe before a trach.*
- *Having a trach is a big deal, but it can help some children and can have many benefits.*

A trach is a short curved tube, usually plastic or silicone, that goes into a hole made through the neck to the windpipe (trachea). A trach allows easier breathing with a vent, allows suctioning of mucus, or allows air to enter below a blocked nose, mouth, or upper airway. Having a trach is not painful, but people say they can feel that it is there.

Suctioning through a trach is uncomfortable because it makes a person cough and feel short of breath for a short time. But suctioning is needed to clear mucus and makes breathing more comfortable after it's done.

The type, size, and shape of the tube are chosen by the medical team based mostly on the child's age and size. A child might have different trach tubes as the child grows. Trach tubes can be cuffed or uncuffed. A cuff is a soft balloon around the outside end of the tube. The balloon can be blown up to make a seal between it and the windpipe. This seal helps the vent blow air into the person's lungs better by stopping too much air from escaping around the trach tube. For some, the cuff does not always have to be blown up. Sometimes the cuff is blown up when the child is asleep but not when awake. The decision to use a cuffed or uncuffed trach tube is made by the medical team. Sometimes, when a child with an uncuffed tube is sick or has surgery, the medical team will put a cuffed tube in for a short time.

The doctors who do the surgery to put the trach in will give you more details about the trach surgery, the risks of the surgery, and the follow-up that is needed.

Recovery after the surgery happens in the ICU. The child is kept asleep (sedated) and mostly still for about 7 days to give time for the connection (stoma) between the neck skin and windpipe to heal. It is dangerous if the trach tube comes out too soon after the surgery because it can be hard to put the tube back in. After 7 or so days, if the child is doing well, the goal usually becomes getting the child off the sedating medicines, teaching, and making plans to leave the ICU.

Once the stoma heals, you can learn how to change the trach tube, which will become easy for you.

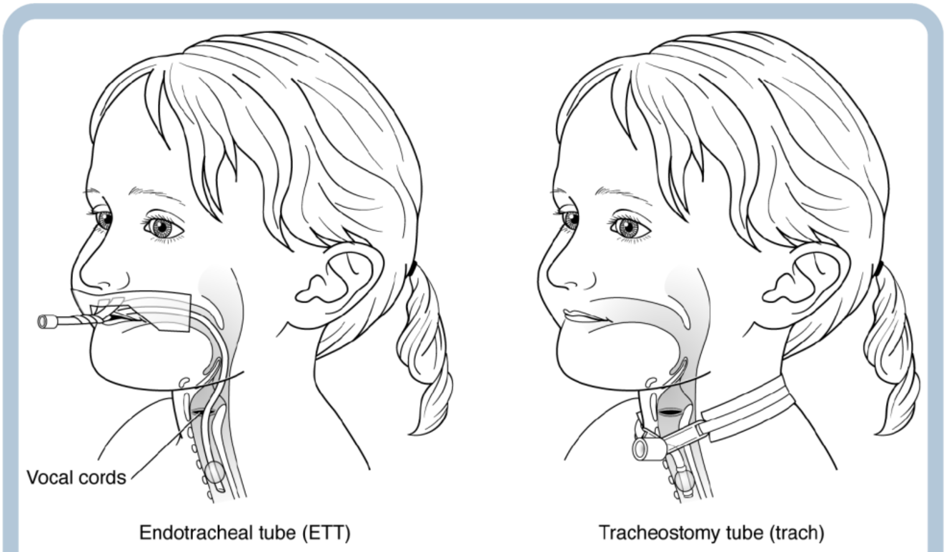

Figure 4. Short-term and trach tubes

Before a trach, many children get help from a vent through an endotracheal tube (**ETT**, see Figure 4). This tube goes from the mouth through the vocal cords and into the windpipe.

Having an ETT for too long leads to problems. It is painful; most patients require lots of pain and sedation medicines to be comfortable with it. These medicines have side effects like sleepiness, confusion, and stomach and bowel problems, like constipation and nausea. Having an ETT also means that the child cannot speak or eat by mouth, cannot move or be moved too much, and cannot leave the ICU. This limits what the child can do, like playing and doing rehab. If this short term breathing tube comes out, it is an emergency and needs to be put back in. This emergency procedure has its own risks. Since an ETT goes through the vocal cords and windpipe, it can cause them long-term damage and problems. Compared to a trach, an ETT may cause more infection and bleeding. How long an ETT can be used safely depends on many things and can be discussed with the medical team.

Having a trach is a big deal but it is *not* a bad thing or a failure. Adult patients who have had both an ETT and trach say that the trach is more comfortable. In addition, the trach allows patients to stop taking pain and sedation medicines. The trach allows safer and better breathing support, better suctioning, and greater freedom. Getting a trach earlier rather than later can mean less time spent in the hospital. Some people who used NIV 24 hours a day or needed lots of suctioning have said getting the trach made them feel safer and more comfortable. Some families have said a trach made caring for their loved one easier. So a trach can improve quality of life in certain cases.

What do you need to know about a trach?

- *If your child has a trach, you must be trained to care for it because it has risks.*
- *A trach can cause long-term problems.*

A trach requires lots of attention. If your child gets a trach, you will receive training on how to care for it and your child. You must show that you can care for your child before your child goes home.

Beyond the general risks of using a vent at home (see above), there can be risks and problems with having a trach.

Risks include:

- *Mucus plug*. Mucus can block the trach tube or the windpipe below the trach tube, making it hard or impossible for air to get into the lungs. These plugs can build up over time or occur suddenly. Suctioning or changing the trach tube can be life-saving. There can be more and thicker mucus when a child has an illness, like a cold.

- *Child becomes separated from vent*. The vent tubing can separate from the trach tube by accident. When this happens, the child is no longer getting breathing help from the vent. If a vent is life-support for a child, not fixing this can lead to death.

- *Trach coming out of child*. Trach tubes are held in place by soft ties or holders that go around the neck. Even with these, trach tubes can still come out. When this happens, the child is no longer getting breathing help from the vent. Not putting the trach tube back in quickly can lead to some children dying. Trach tubes can be put in the wrong way and not be in the windpipe but look like they are in from the outside.

Long-term problems include:

- *Speech problems*. For a young child who is learning to speak, the trach can delay his or her ability to speak, sometimes forever. Speech is not possible when a trach cuff is blown up. Many children who need a trach and vent have difficulty speaking for other reasons.

Some older children can speak if they have strong enough lungs, and sometimes a child can have a special speaking valve placed on the outside of his or her trach tube. This valve helps to force air through the vocal cords to speak and can have other benefits. A “speaking” valve should only be used as directed by a speech therapist and ear, nose, and throat (ENT) or lung doctor.

- *Swallowing problems*. Many children with a trach have a hard time swallowing, usually because of their medical problems. A child with a trach who does not use a vent all day can be tested by the “speech and swallow team” to see if he or she can take food or drink by mouth. Eating or drinking when a child is not ready can cause food or drink to go into the lungs (aspiration). This can happen without signs of problems like coughing. Food or drink going into the lungs can cause severe short-term and long-term lung illnesses. A cuff on the trach does not stop aspiration.

Rarely, families and the medical team decide to allow a child at risk for food or drink going into his or her lungs to eat or drink by mouth because the joy of eating and drinking is more important.

- *Infection*. A trach is a man-made object inside the body, so it has some bacteria on it. Normally, this bacteria does not cause a problem and does not need to be treated. But trachs can increase a child’s risk of infection. This is because it makes it easier for germs to get in the windpipe and lungs. The most common kinds of infections are 1) an infection of the windpipe (tracheitis); 2) an infection of the lungs (pneumonia); and 3) an infection of the skin near where the trach enters the body or where the trach ties are (cellulitis). Skin infections from fungus (like yeast) are common because wetness can become trapped under the trach or ties. If these infections are from bacteria or fungus, they can be treated with antibiotics. Some infections can be treated at home; some have to be treated in the hospital. Many children who use a vent are at higher risk of infections because of their other medical problems.

- *Granulomas*. A granuloma (see Figure 5) is new tissue that grows in the skin where the trach tube goes into the neck or inside the windpipe near the trach tube. It usually happens because of rubbing by the trach tube or damage from infections. They can be big enough to make it difficult to put the trach tube in during tube changes or block air going in and out of the lungs. Granulomas are not painful but they can bleed. They sometimes require medical treatment or surgery.

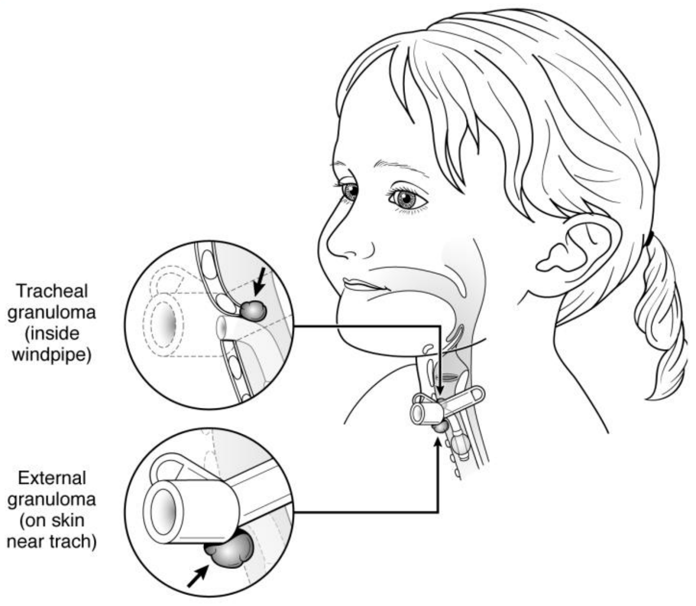

Figure 5. Common granulomas

- *Bleeding*. Minor bleeding can sometimes occur with suctioning, coughing, cough assist treatments, infection, or damage due to dryness or rubbing of the trach tube. Severe bleeding or bleeding that does not stop can sometimes be a sign of an open sore or the trach tube breaking into an artery. A child with severe bleeding or bleeding that does not stop needs urgent medical attention.

- *Sores*. Too much pressure on the skin from the trach tube or trach ties can cause sores. It is important to check the child’s skin often and to adjust the trach tube and ties to make sure that there is not too much pressure on the skin.

- Other less common problems can happen with trachs. These usually result from rubbing or pressure from the tube on the windpipe. Families must be careful not to blow up the balloon on a cuffed tube too much to not damage the windpipe.

At some hospitals, well children with trachs have their windpipe examined in the operating room every so often to make sure things are okay.

Can my child stop using the vent and get the trach out?

- *Some children can safely stop using the vent, but this can take months or years. A smaller number can have the trach removed.*

Depending on your child's reason for his or her breathing problem and his and her other medical problems, it may be possible for your child to stop using the vent. The medical team will tell you if they think there is a chance your child could stop.

Usually, children who have premature or damaged lungs have the best chance to stop using the vent. Even these children take months or years for their lungs to improve enough to stop using the vent.

Children with weak breathing muscles or abnormal/damaged brain or nerves often do not improve enough to stop using the vent.

There are a lot of steps before a child can safely stop using the vent. A lung doctor or other expert must direct these steps (called weaning). Weaning needs to happen before the medical team will consider taking out the trach tube (called decannulation). The steps can be explained in more detail by the medical team, if and when it becomes possible.

Even if a child gets better to the point where the child can stop using a vent, the child's breathing and lungs are usually not normal. The child remains at higher risk for lung illness and infection than other children do. Sometimes a child stops using the vent but has to go back to using it. Sometimes a child's trach is taken out but the child needs to use NIV or oxygen. A small number of children who stop using the vent and have their trach tube taken out have breathing problems again. This is usually because their breathing and lungs were not ready or because of a new illness. In these cases, families may have to decide whether or not to have a trach put in and use a vent again.

Children who used to have trachs can sometimes have problems with their windpipe later on.

What is long-term noninvasive ventilation (NIV)?

- *Long-term NIV is breathing help from a vent through a mask or nasal prongs.*
- *Some children can use NIV; some cannot or should not use it.*
- *NIV may not give enough help when a child is sick or needs a surgery.*
- *NIV can cause long-term problems.*

This section is about breathing help from a vent through a mask or nasal prongs (see Figure 6). Just like using trach and vent, NIV can have many benefits for a child. NIV is usually started in the hospital. Sometimes it is started at home.

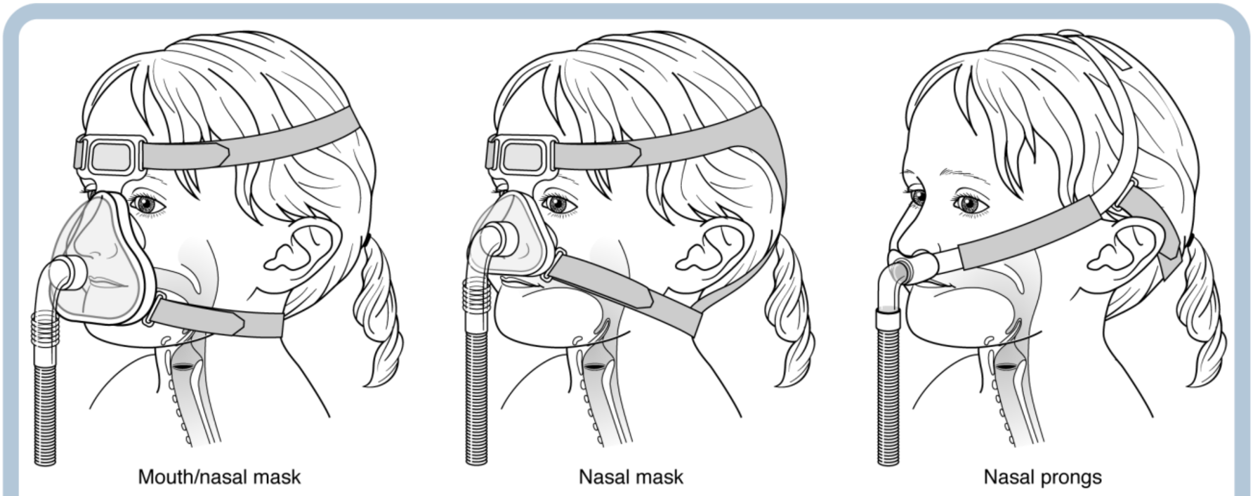

Figure 6. Different types of NIV

NIV is also called bilevel positive airway pressure (BPAP or BiPAP). Some experts also say continuous positive airway pressure (CPAP) is NIV. CPAP provides less help than BPAP.

NIV is usually <u>not</u> used:
- When a child has an abnormal airway (mouth, windpipe, or upper part of the lungs) that does not easily allow air into the lungs.
- When a child does not have a good gag reflex and/or cannot cough out mucus. These are needed to protect the lungs.
- When the amount of vent support that is used (the air pushed in) is too much for NIV to easily provide or makes the child uncomfortable.
- When a child uses a vent for breathing help for most of the day for many months or longer, some experts think that a trach is safer.
- When a child is very young, some experts think that a trach is safer.
- When a child just will not keep the mask or prongs on or the mask or prongs do not fit the child (see below).

If your child uses NIV and becomes sick or is having a procedure or surgery, NIV may not give enough help. In these cases, your child may need to be intubated (a short-term breathing tube [ETT] is placed through the mouth down into the windpipe). For some children, it can be take longer and be harder to remove the ETT safely and get back to NIV.

A child can have problems from using NIV. These include:

- *Skin damage*. Because the mask or prongs press on the skin, there can be injury to the skin. This is more likely to happen the longer the mask/prongs are used, if they do not fit well, or if they are strapped too tightly on the face. Your medical team can give suggestions on how to spot and decrease skin damage.

- *Abnormal growth of skull and face*. Over years of using a mask, the pressure can cause the upper jaw, cheekbones, and/or eye sockets to not grow as much as the rest of the face (called mid-face hypoplasia). This can make a child's eyes seem big and look like they are coming out farther than normal and make the upper teeth not line up with the lower teeth (an under bite). This is more common in children who start using NIV when young.

- *Stomach pain*. Because NIV is pushing air into the back of the mouth before going down the windpipe, some of the air goes into the esophagus and stomach. This can cause stomach pain and a feeling of being full (bloated). The higher the pressures from the vent, the more likely this can happen. Over time, this usually becomes less of a problem.

- *Not using NIV.* Some children do not use NIV as much as suggested by the medical team because they fight wearing the mask or prongs. This is more likely to happen with small children and children with an abnormal brain—in other words, children who cannot understand why they use the vent but can try to take it off. Usually families figure out tricks to get their child to keep the mask/prongs on or the child gets more used to it. It is rare, but sometimes it is just not possible to get a child to wear the mask/prongs. Teenagers also sometimes refuse to use NIV as a way of trying to take control of their lives, but this not good for their health. Over time, not using NIV can lead to the problems talked about above (discomfort with breathing, less energy, worse lung illnesses or more lung illnesses, heart failure, and death).

- Other problems can include eye pain, dry mouth and nose, and feeling trapped in a small place.

Some older children who use NIV all day can use a mouthpiece when awake and a mask/prongs when asleep. Mouthpiece ventilation (see Figure 7) means the person puts his or her lips around a mouthpiece to get vent support. The mouthpiece is put near the face so a person can reach it just by moving his or her head. The person does this from time to time when he or she feels the need for breathing help. A child must be physically and mentally able to use the mouthpiece for it to be a good option.

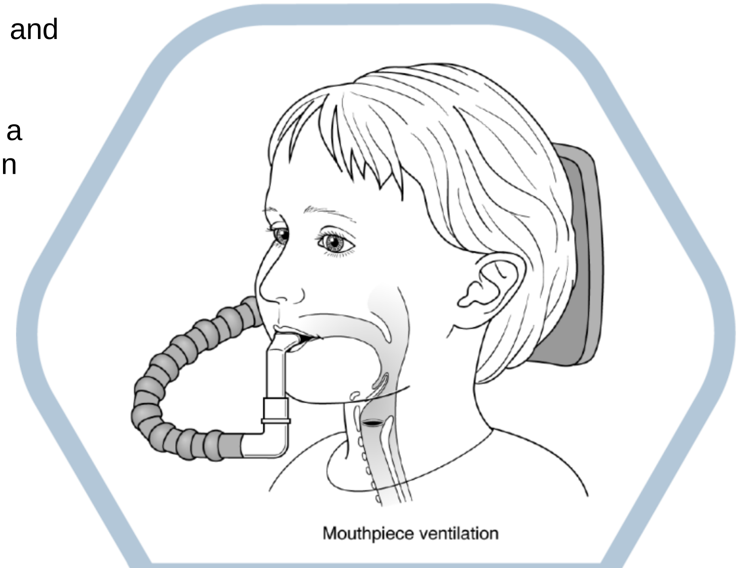

Figure 7.
Mouthpiece ventilation

Can my child stop using long-term NIV?

- *Some children can stop using NIV, but this can take months or years.*

Depending on your child's reason for his or her breathing problem and other medical problems, it may be possible for your child to stop using NIV. The medical team will tell you if they think there is a chance your child could stop.

Usually, children who have premature or damaged lungs have a better chance to stop using NIV. Even these children take months or years for their lungs to improve enough to stop using the vent.

Children with weak breathing muscles or abnormal/damaged brain or nerves often do not improve enough to stop using the vent.

There are a lot of steps before a child can safely stop using the vent. A lung doctor or other expert must direct these steps (called weaning).

Are there any other options?

- *In some cases, a family and medical team can choose not to have a child use a vent if it will not make the child better.*
- *Not using the vent usually means the child will die from chronic respiratory failure.*
- *This does <u>not</u> mean the family and medical team are "giving up." Instead, they are focusing on a goal they believe is most important, like avoiding more suffering.*

When a child has chronic respiratory failure, there are usually no options to support breathing other than using a vent for the long term. If there are other options, the medical team will explain them. Like any medical treatment, a trach cannot be placed or your child cannot be sent home from the hospital using NIV unless you agree to it. Therefore, a decision to use or not use a vent has to be made.

When a child has a medical problem that will shorten his or her life or the child is really suffering from a problem that cannot be cured, a family and medical team do not have to use a vent. Instead of helping the child's breathing with a vent, a family and medical team choose to focus on a different goal. This may be helping the child feel comfortable or not putting him or her through medical procedures. Not using a vent usually means the child will die or have other serious medical problems from chronic respiratory failure. When a child will die can vary from minutes to years, depending on the child. In these cases, <u>not using a vent is a very loving decision and does not mean the family and medical team are "giving up."</u>

Sometimes the choices are not always "yes" or "no" to the vent. Sometimes a family and medical team will decide to try NIV but also decide not to have a trach in the future, if NIV is not helpful. Or sometimes NIV is used to try to make a child's breathing more comfortable, and not to extend his or her life.

<u>If you and the medical team choose not to use the vent, the medical team will continue to care for your child and your family</u>. They will do everything they can to keep your child comfortable, reach the goal that you and the team decided for your child, explain what is likely to happen, answer any questions, and help your family. This choice and the sadness that follows a child's death can be very hard. Families who make this choice say it was the right decision, because they made it with love and the goal of not letting their child suffer. Support groups and counseling can help with grief and sadness.

Even after a child is already using a vent at home, a family and medical team can decide to stop using the vent if it is not helping. Not using the vent can lead to the child's death or other medical problems (talked about above). Also, just because a family and medical team chose to use a vent for their child, it does not mean they have to choose other procedures or treatments in other situations.

In many of these situations, Palliative Care experts are part of the medical team. These providers can help treat pain, aid decision-making, and support in other ways.

How to make a decision about using a vent for a long time

- *Most families and medical teams take a shared approach to making medical decisions.*
- *Decisions should not be rushed or very delayed.*
- *Families should ask any question.*
- *The child should be told everything and take part in decisions, if possible.*

When making hard medical decisions, most families and medical teams prefer a shared approach. This means the medical team gives you info about your child's problem, the possible ways to care for your child (including focusing on comfort), and what those options might mean for your child and family. You share your hopes, goals, and values with the medical team. You should also share your worries. The medical team helps to make sure your hopes and goals can happen and explains which option(s) is more likely to get to your goal. The medical team will try to lessen the worries that they can. You and medical team then decide together what is the best option for your child and family, based on your values and realistic goals.

Some families prefer a different approach to decision making. They may want the medical team to take a bigger or smaller role in deciding. You and medical team should talk about how your family likes to make hard decisions.

People take in info and make decisions at different speeds. No one should feel rushed to make a decision. You should tell the medical team if you feel rushed or if you need things explained again or in your primary language. But not making a decision for too long can cause problems for the child; the medical team should say if they think waiting longer is a problem and why.

Here are some suggestions on how decisions might or should happen.

Ask any question
You should ask any question and ask for any info you need. The medical team will try to answer all your questions, though sometimes they will not have all the answers. It can be hard to answer questions like:

- Will my child get better? How much better will my child get?
- How long will my child live if he or she uses a vent? How long will my child live without using a vent?

The medical team will likely ask you questions to make sure you understand what you are being told. They ask to learn more about what you are thinking and what is important to you. If you feel like you are getting different info or confusing messages from different members of the medical team, you should say so.

Many families go to the internet for info. Like other things on the internet, posts about medical problems, using a vent, trachs, and other things talked about in this booklet are not always true or may not apply to your child or family's situation. It is important for you to share what you read online with the medical team so that the team can help explain what is true and related.

Even with all the available info, families can have questions or thoughts like:

- Is using a vent for the long term the right thing to do?
- Is having a trach placed the right thing to do?
- Will using a vent improve my child's quality of life?
- Is using a vent too much for my child or my family?

- Is not using a vent giving up on my child?
- None of these options is good.

It is okay to have these sorts of questions, and the medical team will not judge you for any question or thought. The team may even ask if you have these sorts of questions or if you wonder if your child is suffering. They ask because they should not guess what you think or they too have these questions. Sometimes the medical team assumes you want your child to use a vent. If this is wrong, you should speak up about what you are thinking.

Families who have faced these sorts of decisions say they thought about how much their child is suffering and about their child's quality of life with and without the medical procedure. Families should think about the "whole" child and situation and not just focus on getting the child out of the hospital. Thinking through these hard questions and decision is often not as simple as weighing pros and cons.

The child's opinion matters

If your child is old enough and can understand what is going on, you should think about telling your child as much as possible and have him or her take part in decisions. Telling your child things at his or her level can make things less scary for him or her. These are not easy things to talk about with a child. The medical team can help talk about these things in a way your child can understand. If your child cannot be a part of the discussion but you know what he or she would want, that should be talked about.

Getting other opinions

Besides asking questions, some families want the opinions of the medical team, another medical person who knows their child, other family members, a religious minister, or other person(s) to help decide. Sometimes people will give an opinion on what is the best option, even if not asked. Opinions can be helpful, but you should not feel pressured by anyone into any decision.

Differences of opinion

People can have different opinions on what is the best option. When this happens, it is important for everyone to continue to talk about what they are thinking and why. It is rare, but when people really cannot agree, an Ethics or Palliative Care expert is usually asked to help reach a shared decision.

Other Resources

Here are a few related resources that some families and medical providers have found helpful when making hard decisions.

Courageous Parents Network
(https://courageousparentsnetwork.org/)
Has talks and tools from parents of sick children and pediatric medical providers (like https://courageousparentsnetwork.org/videos/two-families-two-paths/)

Caring Decisions: A handbook for parents facing end-of-life decisions for their child
(https://www.rch.org.au/caringdecisions/about_us/About_Us/)
An online handbook to help families who are facing decisions about life support for their seriously ill child.

Making critical care decisions for your baby
(https://www.togetherforshortlives.org.uk/resource/making-critical-care-decisions-baby/)
A leaflet for families who have a child with a life-limiting or life-threatening illness.

Ottawa family decision guide
(https://decisionaid.ohri.ca/docs/das/OFDG.pdf)
A general resource to people identify their decision making needs, plan next steps, track their progress, and share their views about the decision.

Acknowledgements

We would like to thank all the families and providers who helped to make this booklet possible. Funding for this booklet came from the Eunice Kennedy Shriver National Institute of Child Health & Human Development (K23 HD082361).

Updated 9/23/2020

Glossary

Aspiration – When anything but air gets into the lungs. Aspiration can happen suddenly and cause signs like coughing or happen little by little without any signs. Aspiration can cause short-term illnesses and long-term damage.

BPAP or **BiPAP** (bilevel positive airway pressure) **–** Usually a kind of NIV. With BPAP, more air is pushed in when breathing in than when breathing out. A BPAP machine can be used with a trach.

Cellulitis – A skin infection.

Chronic respiratory failure – A long-term medical problem with breathing that has many different causes. It can lead to other medical problems and death if a vent is not used help the person's breathing.

CPAP (continuous positive airway pressure) **–** Usually a kind of NIV. With CPAP, the same amount of air is pushed while breathing in and out. A CPAP machine can be used with a trach.

Cuff - A soft balloon around the outside of the far end of some trachs and ETTs, which can be blown up or let down.

Decannulation – A planned removal of a trach when the person no longer needs it. There are many steps that must happen before decannulation. When decannulations are unplanned (on accident), it is often an emergency and the trach must be put back in right away.

ETT (endotracheal tube) **–** A plastic tube that goes from the mouth through the vocal cords and into the windpipe and is connected to a vent or other equipment to help with breathing.

Granuloma – Abnormal tissue that grows because of rubbing or damage from infections. Granulomas usually grow in the skin where the trach tube enters the neck or inside the windpipe near the trach tube.

G-tube (gastrostomy tube) **–** A plastic tube that is used for feeding. The tube goes through the skin of the stomach area into the stomach. A surgery is needed to put in a G-tube.

Home nursing – Paid health professionals who come to the home to provide care. There are different types of nurses with a range of skills and experience.

ICU (intensive care unit) – An area of a hospital where the sickest or most complex patients get care for short-term illnesses or after surgeries.

Invasive ventilation – Help with breathing from a vent through an ETT or trach.

Life-limiting illness – A medical problem that cannot be cured and will cause the person to have a shorter life than a person without the problem.

Life-support – Doing or giving something that keeps a person alive.

Long-term ventilation – Using a vent outside the hospital and for an extended time, usually months, years, or sometimes the rest of a person's life.

Medically fragile – When a person needs long-term medication, treatments, or machines and constant care from others.

Medical team – The group of professionals who provide medical care, includes doctors, nurses, therapists, social workers, nutritionists, and other providers.

Mouthpiece ventilation – A type of NIV. The person puts his or her lips around a mouthpiece to get vent support when he or she feels the need for it.

Mucus – Fluid made by different parts of the body to keep things moist and trap outside material. Mucus can be thin or thick. When thick, it can build up and suddenly block air flow into the lungs, so a person must be able to get it out of the windpipe.

Nasal prongs – Short plastic tubes that fit just inside the nose to give oxygen or NIV.

NIV (noninvasive ventilation) – Help with breathing from a vent through nasal prongs, a mask, or a mouthpiece.

Pneumonia – An infection of the lungs.

Quality of life – How much a person is satisfied with his or her life. Many things make up quality of life, including health, comfort, ability to do things, and relationships.

Sedation medicine – A drug to make a person calm or sleep.

Speaking valve – A small device that can be put on the outside of a trach to help some people with a trach to make sounds with their voice.

Stoma – An opening into the body made by a surgery.

Suctioning – Removing mucus or anything else from the nose, mouth, ETT, or trach. Suctioning can help avoid plugs that block air getting into the lungs and help prevent infections.

Trach (tracheostomy) – A short curved tube that goes through a hole in the front of the neck into the windpipe. A person breaths through the trach, either with nothing attached to it or with it connected to a vent or other equipment to help with breathing. A surgery is needed to put in a trach.

Trachea (windpipe) – The part of the body in the neck and chest for air to travel to the lungs.

Tracheitis – An infection of the trachea (windpipe).

Vent (ventilator) – A machine that pushes air into a person's lungs to help his or her breathing.

Vocal cords – The part of the body at the top of the windpipe used for making a voice and sounds and help protect the lungs from aspiration.

Weaning – Decreasing the amount of help a vent gives a person in planned steps.