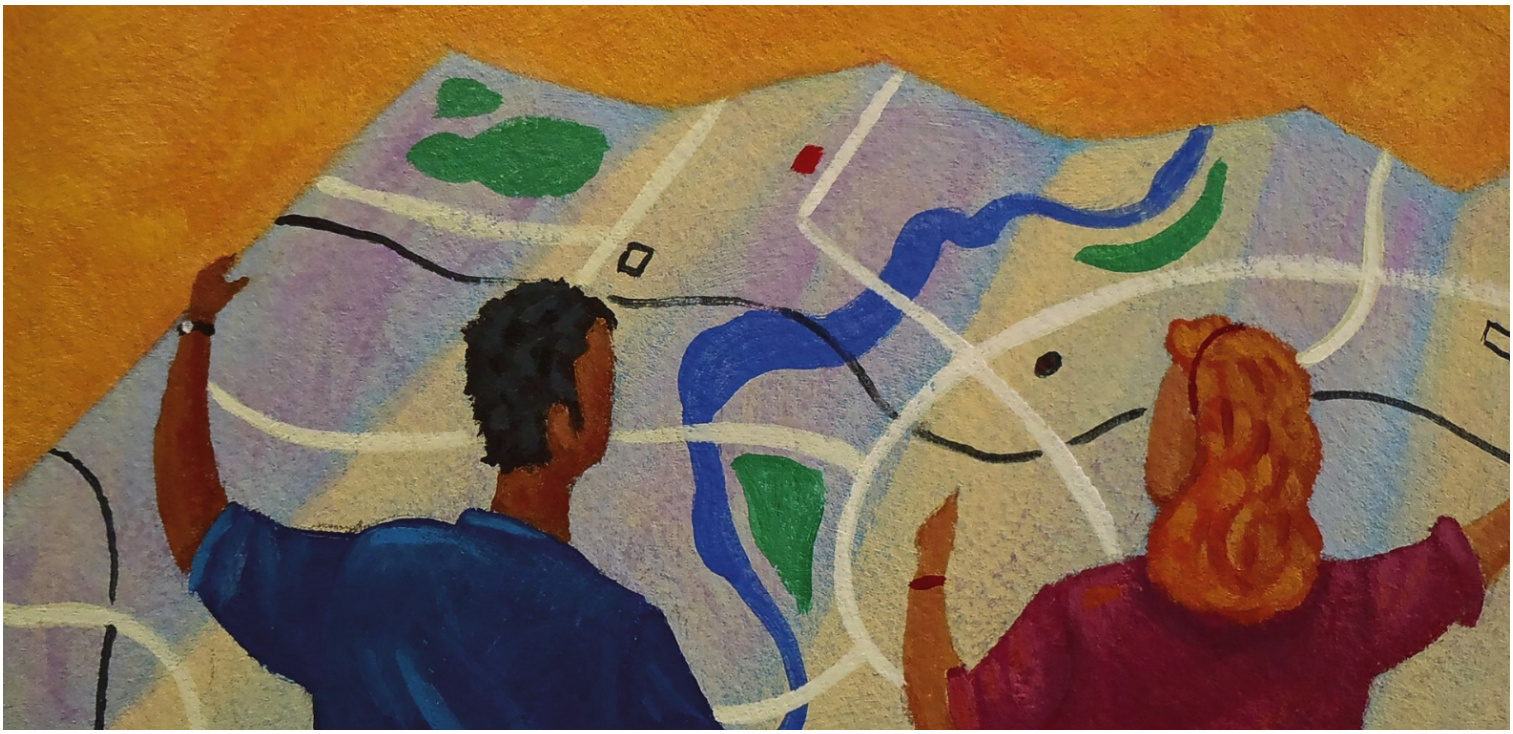
BREATHING DIFFICULTIES: HAVING A TRACHEOTOMY

From the Massachusetts General Hospital
ALS Parenting At a Challenging Time (ALS PACT Program)



Massachusetts General Hospital
Founding Member, Mass General Brigham

Healey & AMG Center
Sean M. Healey & AMG Center for ALS
at Massachusetts General Hospital

Parenting with ALS:
Guidance for Supporting your Children

From the Massachusetts General Hospital
ALS Parenting at a Challenging Time (PACT) Program

1. Supporting Children's emotional well-being: child development and resilience
2. Communication with children about a parent's ALS diagnosis
3. Communication about pseudo-bulbar affect (PBA) and frontotemporal dementia (FTD)
4. Genetic subtype of ALS
5. Changes in Motor Skills and Mobility
6. Bulbar symptoms: Speech, swallow, salivation, and eating
7. Breathing difficulties: Using a BiPAP machine
8. **Breathing difficulties: Having a Tracheotomy**
9. Including children in caring for a parent
10. Talking to children about an imminent loss
11. Supporting children through the end of someone's life
12. Legacy leaving
13. Bereavement
14. Seeking professional mental health support for your child

The entire series is available in both English and Spanish and can be downloaded at no cost from https://www.massgeneral.org/als-pact

**Parenting with ALS: Guidance for Supporting your Children" is intended for educational purposes. It is not medical treatment or advice. If you have questions about your or a loved one's medical condition, please contact a medical provider.*

**Resources noted in the "Parenting with ALS: Guidance for Supporting your Children" are intended solely for illustrative or informational purposes. It does not constitute an endorsement or recommendation of any products, services, websites, or organizations mentioned herein. Readers are encouraged to exercise their own judgment and discretion to evaluate the information.*

Breathing Difficulties: Having a Tracheotomy

Many patients never opt for a tracheotomy procedure, but when a BiPAP machine no longer adequately manages respiratory insufficiency, the choice to move to mechanical ventilation may be offered.

This handout offers:

- an overview of types of breathing difficulties
- a brief description of an endotracheal tube and tracheostomy and how they work
- a description of tracheostomy care, given that children may be asked to help with this
- guidance for talking about a tracheotomy with children of different ages

Physical changes and changes in daily functioning

Breathing difficulties can be a complication of ALS very quickly or only after a long period of time, depending on the person's rate and pattern of progression of ALS. It is rare that ALS begins with breathing difficulties. In most cases, the symptoms begin elsewhere in the body and spread to include the muscles that help with breathing.

Breathing difficulties might include:

- Feeling short of breath doing activities that used to be easy, for example, walking, daily activities like eating, bathing, or getting dressed, and eventually even when sitting down.

- Feeling short of breath while lying down- it may feel more comfortable to sleep with pillows, or sleep sitting up.

"Airway protection" is the process of keeping food and saliva out of the airways. This important part of breathing can be affected early in people with bulbar onset ALS (See handout entitled, "Bulbar symptoms: Speech, swallow, salivation, and eating" for more information about this subtype of ALS, found here: https://www.massgeneral.org/als-pact).

Breathing difficulties from problems with "airway protection" can include:

- Coughing when eating or swallowing saliva

- Difficulty clearing secretions or saliva from the mouth or throat

Equipment and Interventions

Mechanical ventilation: Endotracheal tube and tracheostomy tube

At some point, a BiPAP machine may no longer be enough to maintain a healthy balance of oxygen and carbon dioxide (See the handout, "Breathing difficulties: Using a BiPAP machine" for information about how to prepare children for its use). When carbon dioxide builds up because of difficulty exhaling, a person might feel very fatigued or sleepy. At this point, a person with ALS might shift to using mechanical ventilation.

In an emergency situation, a temporary breathing tube can be inserted through the mouth and into the windpipe and hooked to a ventilator to help someone breathe temporarily. This is called an endotracheal tube. It is temporary and can only remain in for about 10-14 days at the most. Prior to 14 days, the endotracheal tube can be removed, or a tracheostomy (or "trach") can be placed for more permanent ventilation.

A tracheostomy tube is placed through the neck, into the airway below the vocal cords, and also below where the esophagus (which connects to the stomach) and airway (which connects to the lungs) separate. This tube is attached to a breathing machine ("ventilator" or "respirator") that provides oxygen and also removes carbon dioxide from a person's body. In general, when people with ALS have a tracheostomy, they need to rely on a ventilator to help them breathe. A few people can spend short periods of time off the ventilator, if they have a tracheostomy placed when their breathing muscles are still strong enough. As the symptoms progress, they will need to rely on the ventilator all the time.

It is uncommon for people with ALS to talk and eat with the tube in place. As ALS symptoms progress, people lose the ability to speak and swallow. As a result, when people with ALS have a tracheostomy procedure, they have a feeding tube placed, as well. And, they make a plan for communication using means other than speech–for example, writing, typing, or using an eye-gaze machine to type words onto a screen (See the handout called "Bulbar symptoms: Speech, swallow, salivation, and eating" for more information about alternatives).

Because home nurses are not usually paid for by insurance, families usually care for person with ALS, and for the tracheostomy site after this surgery. The "stoma," where the tube enters the neck, has to be cleaned and kept dry to prevent infection. Because ALS makes it difficult for people to cough, and having the tracheostomy creates more secretions and mucous in the airway, "suctioning" is used to remove these secretions. "Humidification" helps to thin the secretions and make them easier to remove.

The Experience of Children and Families

The new equipment to help with breathing will be very obvious and some children will be openly curious about it, while others maybe more wary or anxious. Children will notice that a parent's voice sounds different after a tracheostomy, or that the parent is communicating nonverbally instead of speaking.

Strategies for parents with age-specific guidance

Some of the challenges that you may face in helping your children adjust to these changes include explaining the purpose of new equipment to children of different ages, helping children think about how to talk about these changes with peers, and making decisions about whether to ask children to help with any aspects of breathing care.

Knowing in advance about changes like new equipment is easier for children than being surprised, so if it's possible, try to talk with your children before the tracheostomy is placed. Explain why the equipment is needed, how it will help, how it will change the parent's and family's routine, and whether the child will be asked to help out in new ways.

Talking about a Tracheotomy

Toddlers/early preschoolers (2-4 years):

Dad needs some extra help to breathe because he is sick (or, because of ALS). This machine will help Dad breathe. It has a long name: "ventilator." This tube connects the machine to his body. It makes loud noises, but that means it is working. You can look at it with your eyes, but it is not for touching with your hands or body. What does this machine sound like to you?

Preschoolers/early school-age (4-6 years):

You know that Dad has been having some trouble with breathing in and out, because he is sick (or, because of ALS). He will be starting to use a new kind of breathing machine soon, called a "ventilator."

He will go to the hospital and his doctor will put a tube into his neck. That tube will connect to the breathing machine. Here is a picture of what someone looks like with the tube. He will be able to talk and eat with the tube attached. You can look at this machine but it is not for touching. The doctors want the buttons/dials to stay this way. What are you wondering about? You might have questions when Dad gets home and we will talk about this again.

Teens and young adults (13+ years):

You know that Dad has been sick [whatever words you are using to talk about ALS] and that he was using the BiPAP to help him breathe. It's become harder for him to breathe so his doctors suggested a new kind of breathing machine that's called a ventilator or respirator. This machine will be attached to him with a special tube, called a tracheostomy tube, that will go into his neck. The ventilator will move the air right down into his lungs and also help his body push the air out again, without his body having to do so much work. He will go to the hospital soon to have the tube placed. Then he'll be home. I will help take care of his neck where the tube goes in. (Depending on the needs of your family: That might be something you could help with.)

The doctors have shown us how to set the buttons/dials but we may need to make adjustments. We can show you what we're doing when that happens. We have some pictures if you'd like to see how this will look. What questions do you have right now about this? How are you feeling about this change?

Resources

- Tracheostomy teaching sheet: https://www.thoracic.org/patients/patient-resources/resources/mechanical-ventilation.pdf

- Symptom management for ALS, Later respiratory issues: a description of issues to consider in making decisions about using a ventilator, written by the MGH ALS Multi-disciplinary Clinic team: https://www.massgeneral.org/neurology/als/patient-education/symptom-management