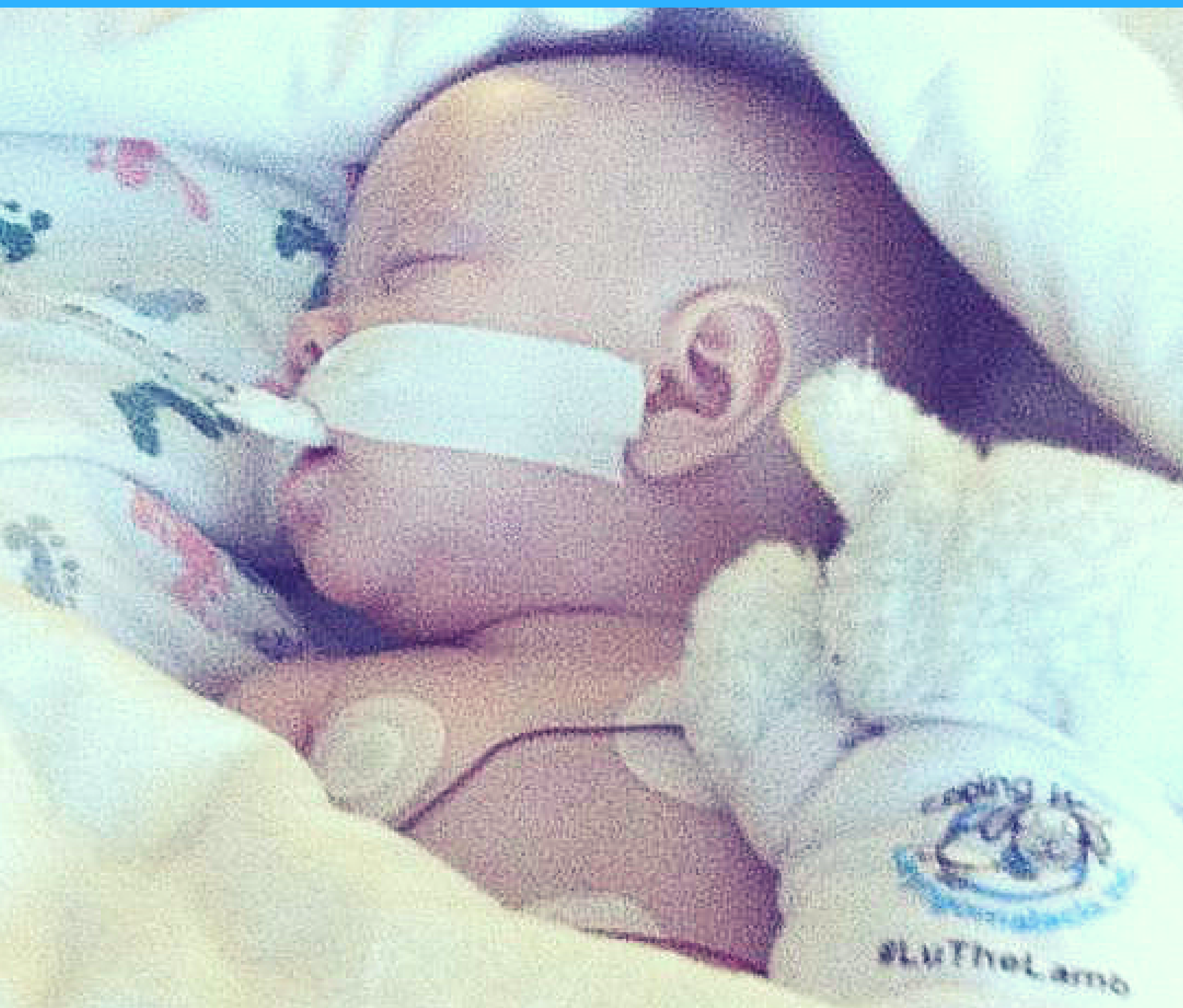
When It's More Than Noisy Breathing

A Guide On How To Cope With Airway Disorders



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Dr. Suzanne Forman D.O.

Otolaryngologist & Plastic Surgeon

A Message From Coping With LM's Founder

Since you are reading this book, I assume your baby has just been diagnosed with laryngomalacia, tracheomalacia or bronchomalacia.

You are probably feeling very overwhelmed, confused and scared. Not really sure how to even pronounce laryngo/tracheo/bronchomalacia let alone know where to begin. The doctor may have told you your baby has mild-moderate laryngo/tracheo/bronchomalacia and you are now in the "watch & wait" period. He may have ordered further testing such as a chest and neck X-ray, a sleep study or maybe even prescribed acid reflux medication. Your baby may be having difficulty feeding, every ounce is a victory. Nursing a baby with an airway disorder has been challenging and you are in need of advice. Maybe your baby has been turning blue and stops breathing. You are now headed into the hospital for surgery.

You are exhausted.

My hope is that you will find support, strength and education within the pages of this book. Knowledge is power and you are your baby's biggest advocate!

Stay strong, you will get through this.

Stephanie Hueston
CWL Founder & President
Lets chat: copingwithlm@yahoo.com

For Bappy,
thank you for showing me the good always follows the bad.

A heartfelt thank you to
Dr. Prasad John Thottam D.O., FAAP
and Dr. Suzanne Forman D.O. for donating
your time, knowledge and support.

And a very special thank you to all of our parents who
shared their stories, photos and hearts.
We will always walk this journey together.

Disclaimer

The information provided in this free resource is for advice only. It is not intended to diagnosis or treat any illness or disease. If you believe your child has an airway disorder please seek medical attention immediately. Do not change your child's current treatment plan before speaking to his/her doctor.

Coping With Laryngomalacia, Inc., Stephanie Hueston, Dr. Prasad John Thottam D.O., FAAP and Dr. Suzanne Forman D.O. are not held liable for unfavorable outcomes based on information provided in this free resource. As the parent, it is your responsibility to seek medical attention (for your child) when needed and to follow your child's treatment plan accordingly.

If your child stops breathing, call 911.

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Chapter 1.
All About Airway Disorders

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"Unfortunately, a lot of physicians are quick to say, "it's laryngomalacia, they will grow out of it." While true, LM can also cause failure to thrive, pectus excavatum, hypoxia, sleep apnea, feeding problems etc. The effect on parents and family watching their child struggle to breathe can be very difficult."

DR. PRASAD JOHN THOTTAM D.O., FAAP
MICHIGAN PEDIATRIC EAR, NOSE AND THROAT ASSOCIATES

What is Laryngomalacia?

Laryngomalacia is a disorder that affects the larynx (voice box) of infants. Laryngomalacia most often presents as noisy breathing or stridor as early as 2 weeks of age. Infants with laryngomalacia will have stridor and may also have difficulty in breathing, feeding and gaining weight, failure to thrive and respiratory distress. This can lead to poor quality of life and anxiety in parents (1).

The larynx of a child with laryngomalacia will demonstrate one or all of the following:

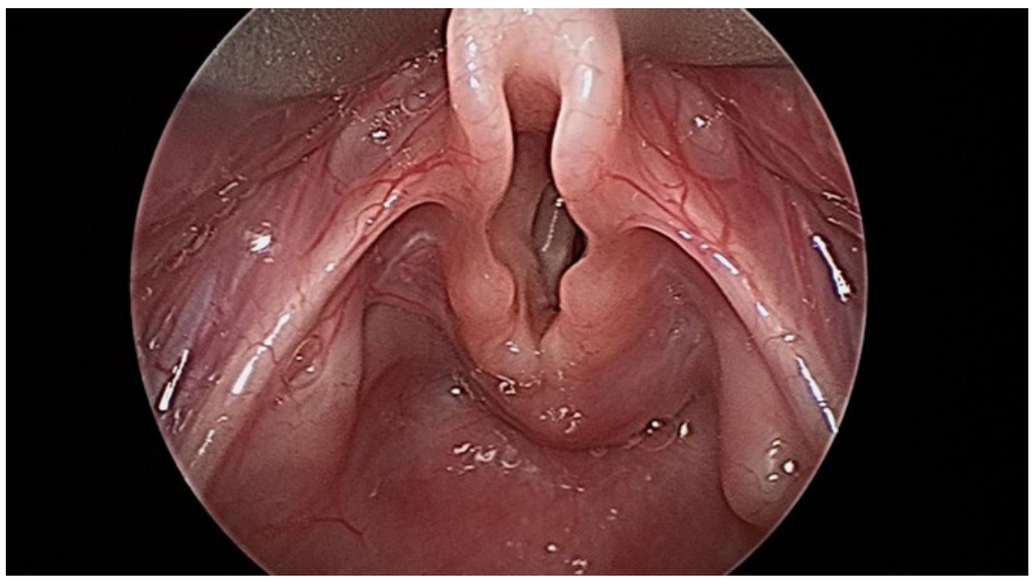
1.) excess tissue overlying the arytenoid cartilages (cartilage above the voicebox) with evidence of arytenoid cartilages falling into the airway (arytenoid prolapse).

2.) short aryepiglottic folds.

3.) omega shaped epiglottis and prolapse of the epiglottis into the airway.

This prolapse of airway structures during inspiration is what leads to airway distress in children with laryngomalacia. The noisy breathing in these children is caused by increased resistance of air flowing through the larynx due the tissue blocking the airway.

While we are still uncertain the exact cause of the disorder, the most common theory describes laryngomalacia as a delay in neurological function and tone of the laryngeal structures above the vocal cords (2). The natural course of this disease typically will progressively worsen up to age 4-8 months and resolve by 12 to 18 months. **Most children do not require surgical intervention.**



omega shaped larynx

What is Tracheomalacia?

Tracheomalacia is a condition where the tracheal cartilage (windpipe) rings soften, partially collapsing when a child is breathing. This collapse most often occurs during expiration. Tracheomalacia may be primary or secondary. Primary tracheomalacia is caused by anterior prolapse of the posterior tracheal wall while secondary tracheomalacia (rare) is caused by external compression. This external compression is most often caused from a vascular anomaly where an artery compresses and weakens the tracheal cartilage leading to collapse. Examples of vascular malformations that can cause this problem include aberrant right subclavian artery, pulmonary artery sling and vascular rings and slings. **In most cases this is managed without surgical intervention.**

What is Bronchomalacia?

Bronchomalacia is very similar to tracheomalacia however in bronchomalacia the cartilage softening occurs in the bronchi of the lungs. Bronchi are branches off of the trachea and also have cartilage that can become soft and collapse. When this collapse occurs in the bronchi it is called Bronchomalacia. Bronchomalacia can also be primary or secondary.

What is Pharygomalacia?

Pharyngomalacia is collapse of the walls of the pharynx. The pharynx is the area behind the nose and the mouth. It also surrounds the supraglottic structures. It can cause obstructive event but is mainly self limiting. Often it can be overcome with positive pressure or bypassing the collapse (nasal trumpet).

What is a supraglottoplasty?

A supraglottoplasty is a surgical technique in which the structures of the larynx are altered in order to prevent collapse of the airway and improve breathing. While the baby is asleep, the surgeon will use a laryngoscope to look into the mouth and evaluate the larynx. The surgeon will then alter the airway to decrease obstruction caused by laryngomalacia. Short aryepiglottic folds are cut in order to lengthen them and provide a more open airway (Figure 4 and Figure 5). Excess arytenoid tissue can be trimmed down so it no longer prolapses into the airway. A third technique that is performed is called an epiglottopexy. This technique attaches the epiglottis to the base of tongue to prevent the epiglottis from falling into the airway.

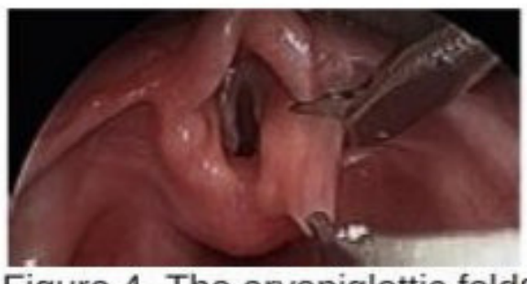


Figure 4- The aryepiglottic folds are being cut in a supraglottoplasty

Figure 5- Supraglottoplasty after bilateral incisions of the aryepiglottic folds

When is surgery needed?

Most of the time laryngomalacia will resolve at 12 to 18 months of age without surgical intervention. **Surgical intervention is warranted if a baby has severe laryngomalacia with recurrent apneic events, difficulty with feeding, failure to thrive and recurrent cyanosis and respiratory distress** (2).

Will my baby need more than one supraglottoplasty?

In otherwise healthy babies, most of the time a single supraglottoplasty will improve the airway obstruction caused by laryngomalacia and no further surgery will be needed. **Supraglottoplasty has been shown to have over a 90% success rate in healthy children with laryngomalacia** (3). There are times however, when a second surgery may be needed if your baby is still having symptoms of severe laryngomalacia such as recurrent apneic events, difficulty with feeding or failure to thrive. Studies have shown that this is more common in neurologically delayed children. Up to 70% of children with neurological delay and laryngomalacia may require a revision supraglottoplasty.

When is tracheotomy needed?

Tracheostomy is needed if your baby is still having symptoms of severe laryngomalacia after maximal supraglottic surgical intervention. This is rare in otherwise healthy children and is more common in babies who have neurological delay if a supraglottoplasty or revision supraglottoplasty does not work. **Often in these situations other airway factors co-exist which make a tracheostomy necessary**.

How common is Laryngomalacia, Tracheomalacia and Bronchomalacia?

Laryngomalacia is very common. **It is the most common cause of stridor in children and effects 35-75% of infants presenting with stridor** (5). Tracheomalacia and bronchomalacia are less common. Estimated incidence of tracheobronchomalacia is estimated to be around 1 in 2,100 (6).

What are the treatments for Laryngomalacia and Tracheobronchialmalacia?

First line treatment for laryngomalacia includes positioning as well as treatment for gastric reflux. Typically, parents will be instructed to position their child on their stomach when they are having difficulty breathing. It is important however to always be with the baby when they are on their stomach. Never leave a baby alone while they are lying on their stomach. Eating is typically easier for babies with laryngomalacia if they are sitting up. Babies with laryngomalacia will often be prescribed medication to decrease gastric reflux. **If these conservative measures do not help or if the baby has severe laryngomalacia surgery may be necessary**.

Tracheobronchialmalacia is most often treated conservatively with humidified air, chest physical therapy and slow, careful feedings. If babies with tracheobronchial malacia are in significant distress, positive pressure airways such as a CPAP may be helpful. There are surgical treatments for tracheobronchial malacia as well though they are rarely needed except for extreme circumstances. Surgical treatment may involve placing a stent in the trachea or bronchi to keep it from collapsing during breathing. As well, if tracheobronchialmalacia is caused by vascular anomalies surgery may be done to fix the artery that is causing obstruction. These surgeries are often done by pediatric cardiothoracic surgeons. **Sometimes tracheostomy may be needed for severe tracheobronchialmalacia to give time for the airway to develop and strengthen**.

What is the prognosis?
(for mild/moderate and severe cases)

The natural course of laryngomalacia will present as early as 2 weeks of age and worsen progressively up to age 4-8 months. **Symptoms typically resolve without surgical intervention by 12 to 18 months of age**. Laryngomalacia can be broken down into mild moderate and severe cases.

Mild laryngomalacia is associated with non-severe intermittent stridor and sporadic feeding difficulties.

Moderate laryngomalacia is associated with dyspnea and consistent feeding difficulties. Severe laryngomalacia is associated with recurrent cyanosis, apneic events and difficulty with feeding that is associated with aspiration and failure to thrive (2).

Patients with mild and moderate laryngomalacia are able to be treated with positioning and anti-reflux medications. Severe cases may need a surgical supraglottoplasty in order to decrease symptoms and allow the baby to breathe without difficulty and gain weight appropriately. **For severe cases, most babies without neurological disease will do well with minimal complications.**

What is the Watch & Wait Method of treatment?
When will symptoms improve?

Mild and moderate laryngomalacia can be treated with antacid medications and watchful waiting. As discussed above, **the natural course of laryngomalacia will worsen until age 4-8 months and then will begin to improve, resolving by age 12 to 18 months** (5).

More than one child with Laryngomalacia?

While laryngomalacia itself is not hereditary, laryngomalacia is more common in infants with neurological disorders. Some neurological diseases may be hereditary. Infants with seizure disorders, hypotonia, developmental delay, cerebral palsy, microcephaly and Chiari malformation are at an increased risk of having laryngomalacia. Laryngomalacia may also be seen in infants with CHARGE association and Pierre Robin sequence as well as 22q11 microdeletion syndrome (DiGeorge syndrome). (5)

Are there any prenatal tests to determine the baby has LM, TM or BM?

There are not currently any prenatal tests to determine if a baby has laryngomalacia, tracheomalacia or bronchomalacia.

Is there a link between Polyhydramnios and having a baby with Laryngomalacia or Tracheobronchialmalacia?

Studies have shown that babies with congenital tracheoesophageal fistula and esophageal atresia do have an increased risk of other upper airway abnormalities (7). Tracheomalacia is the most commonly seen disorder with TE fistula and esophageal atresia however laryngomalacia occurs as well (6). Babies with congenital tracheoesophageal fistula and esophageal atresia will often present with polyhydramnios **so there may be a link between polyhydramnios and laryngomalacia / tracheobronchialmalacia.**

What is the cause of Laryngomalacia and Tracheobronchialmalacia?

The exact etiology of laryngomalacia is not known. The underlying dysfunction is related to redundant mucosa, poor support of laryngeal cartilages and poor neurological tone. The most widely accepted theory of laryngomalacia describes laryngomalacia as a consequence of an immature neurological system (2).

Tracheobronchialmalacia may be primary or secondary. Secondary tracheobronchial malacia is caused by extrinsic compression on the trachea or bronchus most often by vascular abnormalities. The cause of primary tracheobronchial malacia is not known but may be related to other airway abnormalities (ie tracheoesophageal fistula/ esophageal atresia) or neurological immaturity (8)

Is there a relation to laryngomalacia and the MTHFR gene mutation?

There are not currently any studies linking Laryngomalacia to MTHFR gene. Defects in the MTHFR gene are however associated with neurological conditions such as anencephaly, homocystinuria and spina bifida. Babies with neurological conditions are at increased risk for laryngomalacia.

What is the difference between congenital and acquired laryngomalacia?

Laryngomalacia is almost always congenital. The natural progression of laryngomalacia will worsen over time until age 4-8 months.
Tracheomalacia may be primary or secondary.
Secondary tracheomalacia is often due to extrinsic compression of the airway, often by vascular structures.

Is a preemie at risk for developing Laryngomalacia?

To date there is there is no clear link between the risk of developing laryngomalacia in premature infants. Though it is theorized that some premature infants may have decreased neurological tone and thus **increased risk for laryngomalacia**.

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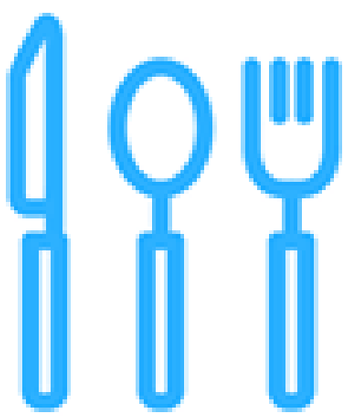
SYMPTOMS OF

LARYNGOMALACIA

STRIDOR, NOISY BREATHING

An audible wheeze when your baby breathes in. It is often worse when the baby is agitated, feeding, crying or sleeping on the back.



DIFFICULTY FEEDING. POOR WEIGHT GAIN, REFLUX, CHOKING WHILE FEEDING AND ASPIRATION.

RETRACTIONS AND CYANOSIS

Pulling in neck and chest with each breath and turning blue.

APNEA, THE STOPPAGE OF BREATHING.

Surgery is the treatment of choice if your baby is having significant blue spells, needs oxygen to breathe, has been diagnosed with Failure To Thrive or has heart and/or lung related conditions from not getting enough oxygen.

LEARN MORE AT:

CopingWithLM.org

Chapter 2.
Eight Common Misdiagnoses of Laryngomalacia

"Laryngomalacia is very common.
It is the most common cause of stridor in children and **effects 35-75% of infants presenting with stridor."**

DR. PRASAD JOHN THOTTAM D.O., FAAP
MICHIGAN PEDIATRIC EAR, NOSE AND THROAT ASSOCIATES

WHEN IT'S MORE THAN JUST SOME NOISY BREATHING

EIGHT COMMON MISDIAGNOSES OF LARYNGOMALACIA

COPINGWITHLM.ORG

newborn congestion	parent is told he/she is overreacting
uncontrolled reflux	some babies make noise when they breathe, they will outgrow it
baby swallowed amniotic fluid during delivery	food allergy
common cold	pyloric stenosis

Chapter 3.
The First Doctor's Appointment

"I found the What To Ask Your ENT document very helpful. Doctor appointments always felt rushed and I left with questions. This helped me show up better prepared and I also feel this document led to my son being cared for quicker and more thoroughly. When I would ask questions off the sheet, the doctor would follow through with examining that area and even found some things that she was not looking for to begin with."

NICOLE
MOM OF A LARYNGOMALACIA WARRIOR

Listen to your instinct. You know your baby better than any monitor or medical test. Let your voice be heard and if a doctor speaks over you, find a new doctor. Get to the best hospital. Bills can be paid later, your child needs to breathe now.

THE LARGEST DIRECTORY OF RECOMMENDED

Ear, Nose & Throat Doctors

only at CopingWithLM.org

Updated often, view our Find An ENT Directory **here**.

13 questions you should ask your child's Ear, Nose and Throat Doctor.

1. How severe is the Laryngomalacia? Do you suspect Tracheomalacia or Bronchomalacia?

2. During the scope, was GERD (reflux) present? How severe? What is the treatment?

3. If surgery is needed, how many surgeries (supraglottoplasty) have you (the ENT) performed? Will you perform a Bronchoscope during surgery? What are your thoughts about second and third surgeries?

4. If surgery is not needed, how long is the watch and wait period?

5. When do I need to call 911?

6. Do you suggest any of the following tests? If no, why not?

Sleep Study

Barium Swallow

Chest/neck X-ray

7. What are signs/symptoms my baby is aspirating his/her food?

8. Are there any home remedies I can do to help improve the symptoms?

9. When do you believe my baby’s symptoms will improve?

10. How can I get in touch with you (the ENT) after office hours?

11. Are any specialist (pulmonologist, cardiologist, gastro) follow ups needed?

12. What hospital are you (the ENT) affiliated with?

13. What is your Emergency Appointment Policy? Can my baby be seen the same day I call?

DON'T FORGET!

Get your child's medical portal log in information.

Ask that a summary of today's visit be sent to your child's pediatrician.

Get all prescriptions and double check dosage.

Ask about CPR training.

Go over safe sleep practices.

Chapter 3.

Caregiver Info Sheet

"Do you know CPR?"

DEBORAH
MOM TO A LARYNGOMALACIA WARRIOR

Doctor:

Phone number:

I have an airway disorder, your germs can hurt me!
Please wash your hands before touching mine!

I have (check all that apply):

Laryngomalacia
Tracheomalacia
Bronchomalacia

For sleep please put me on my:

Back
Side
Stomach

To help me breathe easy I use a / an:

Apnea / oxygen monitor
Oxygen
CPAP / BiPAP
Tracheotomy

To eat I use a:

Bottle
Breast / SNS
Feeding Tube
Honey Bear

Medication to be given:

Time:

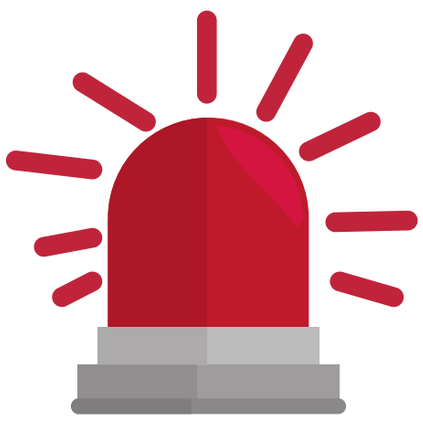
Route:

The nearest hospital is:

Phone number:

Special instructions from Mom and Dad:

If I stop breathing, call emergency services!



Chapter 5.
The Surviors Award

"If it weren't for this group and the moms who pushed me to trust my instincts, **I am pretty sure my son would have died in his sleep.** "

TRACEY
MOM TO A LARYNGOMALACIA WARRIOR

COPING WITH LARYNGOMALACIA, INC.

The Laryngomalacia Survivors Award

This award is presented to

you didn't outgrow it, you **survived** it!

Coping With Laryngomalacia, Inc.™

STEPHANIE HUESTON
CWL FOUNDER & PRESIDENT
CopingWithLM.org

Chapter 6.
Helping You Cope

Programs We Offer
&
How To Get Involved

"Perhaps the hardest part is learning how to cope with a journey you never thought you would have to walk."

STEPHANIE HUESTON
COPING WITH LM FOUNDER & PRESIDENT

HOW YOU CAN HELP A PARENT WHILE THEY ARE COPING WITH LARYNGOMALACIA

LEARN CPR

Take an infant and child CPR class at your local hospital. If the baby is on a monitor/oxygen, learn how to properly work the machines.

COOK DINNER

Drop off a hot, homemade meal.

GERM CONTROL

Wash your hands before asking to hold the baby. If you are sick, suggest postponing the visit until you are well.

CARE FOR SIBLINGS

Offer to take siblings to the park, out for ice cream or stay at the house and do a craft.

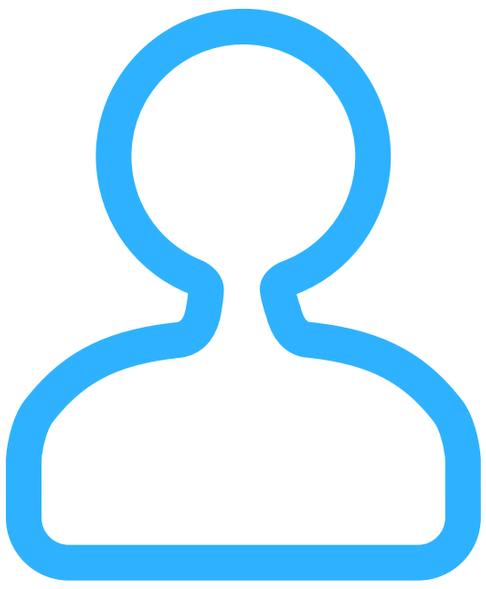
OFFER SUPPORT

Call and ask, "how are you?"

BRING PAJAMAS

Emergency hospital stays happen quickly. Parents run into the ambulance without many personal belongings. Stop by the hospital with comfy pajamas for Mom and Dad.

SPONSOR A CARE PACKAGE IN THEIR CHILD'S NAME AT COPINGWITHLM.ORG

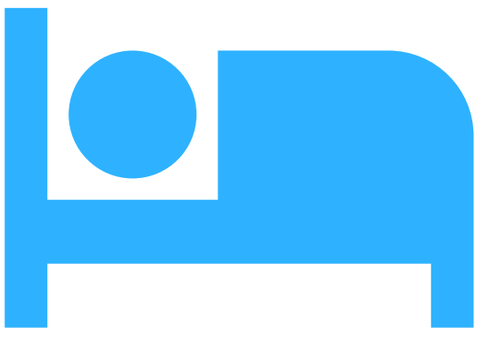


Parent Support Group on Facebook

With over 2,000 (and growing!) members worldwide, parents can connect directly to others who coping.

Care Package Program & Pint-Sized Powerhouse Grant Program

Thanks to our amazing and generous donors, we are able to fulfill our mission by offering programs that bring support and strength to our families.



Chibebe Snuggle Pod Program

Thanks to Chibebe's ongoing support, we are able to gift Snuggle Pods to our warriors within the USA (chibebe.com) and AUS (chibebe.com.au).

Malacia Meet-Ups

Meet local families who are coping!
We are always in need of hosts.
Want to help others? Contact us today!

Become An Awareness Leader

From blogging to speaking at your local hospital, there are many ways you can make a difference! Contact us to get started!

Give

Sponsor a care package, organize a Blanket & Book Drive, support our current fundraiser. Every dollar makes a difference!

Light blue words are hyper-linked.

Chapter 7.

Meet #LuTheLamb

Lu was diagnosed with LM as a little lamb. He was having a hard time breathing so Mama Sheep brought him to see the doctor. At three months old he had to go to the hospital and have surgery because his LM was so severe.

Ever since surgery, Lu has been breathing easy!

Lu now travels the world to provide his friends with support, love and strength as they cope with airway disorders.

You can follow on Lu on his journey by searching #LuTheLamb on Facebook, Instagram and Twitter!

"Although Vinny is too young to understand the significance of #LuTheLamb, Lu brought my husband and I so much comfort while in the hospital during the early days of diagnosis and supra #1. Every time I looked at Lu, I was reminded of how many people were fighting this fight alongside us. I thought of all the other families and children all over the world who had been in our shoes, and I felt strength and hope! Lu meant we weren't alone, even when the dark and quiet PICU told us otherwise."

-Brittany