

PedsCases Podcast Scripts

This is a text version of a podcast from PedsCases.com on “**Chronic Stridor in Children**” These podcasts are designed to give medical students an overview of key topics in pediatrics. The audio versions are accessible on iTunes or at www.pedscases.com/podcasts.

Chronic Stridor in Children

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Introduction

This podcast was written by both Hannah Kraicer-Melamed and Dr. Jonathan Rayment. I'm Hannah, a student at McMaster University and Dr. Rayment is a Pediatric Respiriologist at B.C. Children's Hospital.

Objectives

Today's podcast will discuss an approach to chronic stridor in children. Stridor is a common, distressing problem in children. We think about the common causes of acute stridor like croup, epiglottitis, or inhaled foreign bodies, but what happens when it doesn't go away? There are a number of important structural and functional causes of stridor that a physician caring for children needs to consider.

After listening to this podcast, the learner should be to:

1. Define stridor and differentiate it from wheeze
2. List the major causes of chronic stridor
3. Order appropriate investigation for a child with chronic stridor
4. Understand the management of the major causes of chronic stridor

If this is a newer topic for you, you might want to check out the PedsCases podcasts on Evaluation of Stridor and Croup, before listening to the rest of this podcast.

Case

Let's start with a case.

You are working in a community clinic, and your next appointment is Charlie, a 4-month-old whose parents are bringing them in for assessment of noisy breathing.

You enter the room and hear the following sound on inspiration: *insert noise of stridor*
<https://www.youtube.com/watch?v=oeoAze-CHng> .

What is your approach to this sound?

What is stridor

We should first start with an explanation of what stridor is.

Stridor is a harsh, high pitched noise, of one specific tone, that is produced by turbulent airflow moving through a narrowed large airway like the larynx or trachea. Stridor can often be heard without a stethoscope. The following is an example of stridor ***insert stridor sounds***.

<https://www.youtube.com/watch?v=oeoAze-CHng>

Stridor can be inspiratory, expiratory, or biphasic, but most commonly it is thought of as an inspiratory sound.

Wheeze is an expiratory sound primarily that is heard with a stethoscope within the chest walls due to the obstruction of small airways.

Stridor, on the other hand, is primarily an inspiratory noise that is usually audible without a stethoscope and due to obstruction of the large airways. Expiratory stridor and wheeze can be difficult to tell apart at first, however stridor tends to be much harsher in character and is best heard over the neck.

Some information on the location of the stridor can be inferred based on whether it is inspiratory or expiratory. We'll talk a little about that now.

The extrathoracic large airways are outside of the chest cavity and include the larynx and the upper trachea. The larynx, known as the voice box, is separated into three anatomical areas: the glottis, subglottis, and supraglottis. The glottis is the area that contains the vocal cords. The area above the vocal cords is called the supraglottis and the area below the vocal cords is called the subglottis, which will become relevant later on in this podcast. The intrathoracic large airways are inside the chest cavity and include the lower trachea and mainstem bronchi, both of which are supported by cartilaginous structures to maintain their stability. Narrowing of any of these structures can cause stridor.

The diameter of the airway normally changes during inspiration and expiration. However, when there is airway narrowing, due to a pathological process, this airway narrowing can result in turbulent air flow that can lead to stridor.

Inspiratory stridor is usually caused by narrowing of the extrathoracic large airways. On inspiration, the pressure inside the chest becomes lower than atmospheric pressure to allow air flow into the lungs. The extrathoracic airways are floppier than the intrathoracic airways, and during inspiration, the fast-moving air and pressure changes causes them to collapse a little bit. When pathological narrowing is present, this airway collapse during inspiration makes the passageway even smaller, leading to turbulent flow and inspiratory stridor. On expiration, the opposite occurs, and the pressures in the extrathoracic large airways are higher than atmospheric pressure, causing relative distention of the extrathoracic airways, which can, in most cases, relieve the narrowing and therefore relieve the stridor.

In contrast, expiratory stridor is caused by narrowing of the intrathoracic large airways. In this case, during inspiration, pressures are lower inside the thoracic cavity than in the extrathoracic airways (trachea, larynx) and atmosphere. Therefore, this pushes the intrathoracic airways

open, relieving the obstruction. On expiration, intrathoracic pressures are increased to push air out and this leads to compression of the intrathoracic structures, causing expiratory stridor.

Biphasic stridor is due to turbulent flow during both inspiration and expiration. This can be caused by a combination of intra- and extrathoracic pathologies or severe pathology in either compartment that cannot be overcome by the normal physiologic airway's distention associated with the respiratory cycle.

To summarize, on inspiration, this narrowing occurs outside of the thorax leading to inspiratory stridor. On expiration, this narrowing occurs inside the thorax and can lead to expiratory stridor or wheeze.

Case

Returning to the case of Charlie, the 4-month-old patient. You walk into the room and recognize the sound as stridor. You think to yourself that this is most likely due to obstruction of the extrathoracic upper airways because it is:

1. Inspiratory;
2. Heard without our stethoscope; and
3. Monotonic.

On immediate inspection, the baby is fussy, but in no respiratory distress. There is no nasal flaring, subcostal indrawing, tracheal tug, head bobbing, accessory muscle use or drooling. You are happy to note that the O₂ saturation is 99% on room air and the respiratory rate is 38 and the patient is afebrile. The patient is the 55th percentile for both height and weight. Seeing that the baby is not acutely unwell, we now have some time to speak with the family to figure out what is really going on here.

The family tell you that they started to hear the noise in the first week of Charlie's life. It was initially pretty quiet, however, in the last few weeks the noise has slowly gotten louder, especially when the baby is feeding or sleeping on their back. The parents say, "it's quite a loud noise for such a small human!". Charlie had an uneventful pregnancy and was born at term with no complications. They have otherwise been a healthy and happy baby is on no medications and their immunizations are up to date. The parents do not know of any relevant family history.

Given the time course, and the clinical picture, you recognize Charlie as having chronic stridor. **But**, if the time course had been short and the story fit we would be thinking about the acute etiologies of stridor like croup, anaphylaxis, foreign body aspiration, epiglottitis, and bacterial tracheitis, like has been discussed in the Acute Stridor PedsCases podcast.

Etiologies of chronic stridor

When thinking about the etiology of chronic stridor, it is helpful to use a structural approach. Stridor can either be caused by an intrinsic narrowing of the large airways, or due to an extrinsic compression. We can then work from the top of the airway down to the mainstem bronchi to ensure that we are not missing anything. The differential for chronic stridor is large, and we do not have time to cover every cause in this podcast, however we will attempt to address the major causes of chronic stridor in a clear and easy to follow differential.

Intrinsic narrowing

We will start with the most common group of reasons for stridor, intrinsic narrowing of the airway. Let's move from the top of the extrathoracic airways down into the chest cavity and the intrathoracic airways.

Naso- and oro-pharyngeal congenital abnormalities

We start with the nose and face. Nasal, pharyngeal, and craniofacial alterations that narrow the airway can lead to stridor. These include but are not limited to:

1. Choanal atresia or stenosis, which can be identified by trying to insert an NG tube into the patient's nares and identifying a blind end;
2. Intranasal masses, which can be identified with imaging or on direct visualization with flexible nasolaryngoscopy; and
3. Craniofacial abnormalities, like micrognathia, macroglossia, or Pierre Robin Sequence.

Therefore, ensuring the patency of the nasopharynx and the lack of masses in this area is important for narrowing down our differential. This assessment should be performed by a paediatric ear nose and throat surgeon.

Larynx

Next we move down into the larynx or throat.

Laryngomalacia

The most common cause of chronic inspiratory stridor in the pediatric population is laryngomalacia. Laryngomalacia is collapse of the larynx on inspiration. It is thought to be related to decreased tone in the structures of the larynx, edema, or redundant tissue. Laryngomalacia should be suspected in an infant with chronic stridor, made worse when supine and improved with prone positioning. The diagnosis can be confirmed by flexible nasolaryngoscopy, with findings such as short aryepiglottic folds, an omega shaped or prolapsed epiglottis, collapse of the supraglottis, or prolapse of the cartilaginous structures of the larynx. The symptoms peak at approximately 4-6 months of age, and spontaneously resolve as the child's airways grow by approximately 1 year of age. Once confirmed, treatment is geared to severity of presentation. In mild cases, watchful waiting is an appropriate strategy as most children will grow out of the stridor as they age. In severe cases, leading to failure to thrive or respiratory compromise, surgical repair with a supraglottoplasty can be considered. Management of any concomitant gastroesophageal reflux is also felt to be beneficial to reduce airway edema from acidic reflux.

Other laryngeal anomalies

In addition, congenital abnormalities like laryngeal webs can lead to narrowing of the airway by physical blockage of the larynx. This can be visualized with direct flexible nasolaryngoscopy.

Trachea

Moving down from the larynx, we reach the upper trachea and the glottis. We can first think about the vocal cords.

Vocal cord paralysis

Vocal cord paralysis can be either congenital or acquired and unilateral or bilateral. Congenital causes include neurologic defects like myelomeningocele, Chiari malformations, myasthenia gravis or hydrocephalus. Acquired causes are typically iatrogenic following surgery, or injury from intubation.

Unilateral vocal cord paralysis is more likely on the left side, which is thought to be related to the increased length of the recurrent laryngeal nerve. Unilateral vocal cord paralysis typically presents with weak cry, but not usually stridor, as the diameter of the airway can be largely maintained on inspiration.

Bilateral paralysis can lead to stridor in infants and children, due to decreased airway diameter and can often lead to respiratory compromise, aspiration, and breathing difficulties. More than half of children and infants with bilateral vocal cord paralysis require airway support. Diagnosis is usually made again via direct visualization of the vocal cords, with awake flexible nasolaryngoscopy. Management also depends on severity. Mild symptoms are amenable to watchful waiting until about 2-3 years as they may improve or resolve. After 2-3 years, it is unlikely to improve and surgical interventions, such as laryngeal reconstruction or insertion of a tracheostomy to protect the airway from aspiration are employed.

Subglottic stenosis

The second most common cause of chronic stridor in infants is subglottic stenosis. This is, as the name suggests, a narrowing of the area below the glottis, extending to the cricoid cartilage. Causes of subglottic stenosis can be characterized as congenital or acquired.

Congenital subglottic stenosis can be caused by abnormalities in the development, shape, or size of the structures below the glottis, including the cricoid cartilage or soft tissues of the larynx. This has been hypothesized to be related to failure to fully canalize the subglottic area. Congenital subglottic stenosis is associated with trisomy 21.

Acquired subglottic stenosis occurs most frequently from trauma and scarring due to endotracheal tube (ETT) intubation. The cricoid cartilage consists of a complete ring; therefore, this area is more likely to be damaged than anywhere else in the trachea. Risk factors for acquired subglottic stenosis are: increased time of intubation, low birth weight, infection of the airway, and gastroesophageal reflux disease or GERD. With transient intubation and inflammation, the process can be reversible, but with prolonged damage, fibrosis occurs, and the stenosis is usually permanent.

Diagnosis of subglottic stenosis is made based on history, physical examination, and can also include findings from chest X rays, but definitively requires direct visualization. This should be done by rigid or flexible bronchoscopy, with the patient breathing spontaneously (i.e. no positive airway pressure) in order to avoid artificial distention of the airway and masking of the stenosis. The severity of the stenosis can be graded based on the amount of occlusion.

The risk of subglottic stenosis can be reduced by decreasing the duration of intubation and minimizing the number of extubation attempts, if possible. Once subglottic stenosis occurs, watchful waiting is appropriate for mild presentations. However, if there is respiratory distress or compromise, surgical approaches such as tracheostomy and tracheal reconstruction are required.

Subglottic hemangioma

Infantile hemangiomas are benign vascular tumours that are the most common tumours in infants. They are commonly found on the skin, but can also be found in the liver, brain, lung, mediastinum, and importantly for our discussion in the large airways, classically in the subglottic region. The presence of subglottic hemangiomas is often, but not always, associated with cutaneous hemangiomas in the beard distribution (on the jawline of an infant, where a beard

would be in a man). Children with a beard hemangioma warrant urgent assessment by ENT if they develop any symptoms of stridor.

Subglottic hemangiomas follow the natural course of other infantile hemangiomas. They initially have a phase of rapid growth reaching their peak size at approximately 6 months of age. Following this, there is a plateau until 1 year of age and then a period of involution, usually about 3-5 years where they slowly disappear.

Given these timelines, a child with a subglottic hemangioma may initially be asymptomatic but will develop increasing stridor in the first 6 months of life as the hemangioma grows. If they get too large, they can cause severe respiratory compromise. Subglottic hemangiomas therefore require prompt treatment.

Diagnosis of subglottic hemangioma is made with direct visualization with flexible or rigid bronchoscopy. Once the diagnosis is made, patients are started on medical therapy, which consists of the β -blocker propranolol at 1mg/kg/day with escalation to 3mg/kg/day to prevent further proliferation and initiate involution. The mechanism is not currently well understood. This treatment normally lasts until the child is 8-12 months of age and is at the point in which normal involution would occur. Contraindications to propranolol treatment include heart conditions, hypoglycemia, and asthma.

Next, let's move farther down into the intrathoracic airways.

Tracheomalacia and bronchomalacia

Tracheomalacia and bronchomalacia are somewhat similar to laryngomalacia but occur in the intrathoracic large airways. Recall that narrowing of the intrathoracic large airways leads to an expiratory stridor. Tracheomalacia is abnormal collapse of the trachea and/or bronchi during expiration.

Tracheomalacia can either be primary, due to insufficient cartilaginous rings and support, or secondary, due to external compression of the trachea or compromise of the integrity of the trachea following surgery. In tracheomalacia and bronchomalacia, one normally appreciates a low-pitched monophonic stridor on expiration. Diagnosis is made with direct visualization via bronchoscopy. Treatment is based on severity, with respiratory compromise requiring tracheostomy and positive pressure support, while milder cases can allow for watchful waiting and conservative management as individuals may grow out of this.

*Clinical pearl: expiratory stridor can often be mistaken for wheeze; but administration of inhaled β -agonists (eg. Salbutamol) can actually exacerbate the stridor due to loss of bronchial smooth muscle tone. If you are ever treating an "asthmatic" child who paradoxically seems to worsen on salbutamol, consider tracheobronchomalacia.

Other trachea abnormalities

Other tracheal abnormalities such as tracheal stenosis and webs, can also lead to stridor and can be predicted based on CXR imaging.

Extrinsic compression

Now let's discuss things that compress the airway from the outside. Abnormalities of the great vessels, large airways, esophagus or other structures of the mediastinum can result in extrinsic large airways compression.

Vascular rings and slings are one of the most common **extrinsic** causes of stridor. These are malformations of the large thoracic blood vessels that lead to compression of the trachea and esophagus. The most common cause of a vascular ring is a double aortic arch but other abnormalities such as a pulmonary artery sling can also result in a similar presentation. Vascular rings can be diagnosed using MRI or CT with contrast.

The area surrounding the airway is also the embryologic origin of foregut development. Cysts in the foregut such as bronchogenic cysts, or esophageal duplication cysts can develop adjacent to the large airways and lead to extrinsic compression. These can sometimes be seen on plain film X-rays, but often cross-sectional imaging (such as CT scans) will be required to make a radiographic diagnosis.

In addition, external compression of the airway due to large mediastinal lymph nodes or mediastinal masses can cause stridor in patients with infectious or non-infectious conditions that lead to lymphadenopathy, including tuberculosis or lymphoma.

Case

We have now addressed the different areas of the airway and how they can lead to narrowing in an approach that will hopefully allow us to think about the anatomic structures in a manner that provides us with an organized differential diagnosis, brief management, and natural history for many of the common causes of chronic stridor. Now let's apply this to our case.

Charlie is a 4-month-old with chronic stridor. You expand your history and physical to consider each item on the differential.

Laryngomalacia is high on your differential. Charlie has isolated inspiratory stridor present since the first few weeks of life. It is aggravated by sleeping in the supine position and also with feeding or exertion.

Considering vocal cord paralysis, Charlie has no symptoms of choking, or respiratory distress. On exam, they do not have a weak cry and has no associated focal neurological signs.

Thinking about subglottic stenosis, you ask about a history of intubation. Charlie has never had an ET-tube or any interventions of the respiratory tract. This rules out acquired subglottic stenosis, but we are unable to rule out congenital subglottic stenosis without visualization.

You examine the neck for hemangiomas in the beard distribution, and do not see any. This makes subglottic hemangiomas less likely.

With regards to the intrathoracic airways, our patient has isolated inspiratory stridor. This does not sound as much like tracheomalacia and/or bronchomalacia due to the lack of expiratory noises.

You discuss the differential with the family, telling them that the most likely diagnosis is laryngomalacia, but you refer them for a flexible laryngoscopy. This will allow a specialist to visualize the upper airway to below the glottis and determine the etiology of stridor! If this did not yield the diagnosis, they could proceed to advanced imaging studies to look for things such as extrinsic compression.

Charlie's family is quickly seen by ENT and the laryngoscopy confirms the diagnosis of laryngomalacia. They reassure the family that the Charlie has no respiratory distress, no evidence of failure to thrive and no other concerning symptoms and no surgical intervention will be required. Therefore, you can follow up with the patient to monitor the progression of stridor, growth parameters, and vitals. 6 months later Charlie's stridor has disappeared.

Take home points

In general, when thinking about the potential etiologies of chronic stridor, we should consider the onset, natural history, and associated symptoms. The general steps to take when evaluating stridor can be broken down as follows:

1. Assess whether this is truly stridor. We must ensure that we are hearing a high-pitched harsh noise without our stethoscopes. We also want to assess whether the stridor is inspiratory, expiratory or biphasic. From here, we can start to think about what could be causing this turbulent flow.
2. Assess whether there are concerning symptoms of respiratory compromise, failure to thrive, infection, or foreign body aspiration as in all pediatric patients and if observed, provide urgent management.
3. Assess of onset of stridor. Has this noise been present since birth or within the child's early life? Has this noise increased?
4. Assess associated symptoms. Is this patient in respiratory distress? Is the issue worse with activity? Does the stridor change with positional changes? Does the patient have other syndromic abnormalities or hemangiomas?
5. Work through an organized approach to identify the potential etiologies of airway narrowing

This concludes our PedsCases podcast on an approach to chronic stridor. Thanks for listening!

Resources

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