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Noisy Breathing: Stridor in Children with Low Trunk Tone

by Susan Agrawal

A surprising number of children with cerebral palsy or other conditions affecting muscle tone have very noisy breathing when they inhale, commonly called stridor. While this fact has been noted anecdotally by both doctors and parents, surprisingly little research has been done on the subject. No medical articles have ever been published that focus exclusively on stridor in children with hypotonia (low trunk tone) or cerebral palsy.

Stridor from Non-Congenital Laryngomalacia

Most children with inspiratory stridor are diagnosed with laryngomalacia, a condition in which the tissues in the larynx or voice box periodically block the airway, creating a noisy sound upon inhalation. Laryngomalacia is classically thought of as a benign disorder of infancy, one that typically worsens in the first six months of life but ultimately disappears by 18-24 months. In 99% of these babies, no treatment is needed and the infants thrive without complications. Infants with very severe laryngomalacia may develop respiratory problems such as retractions or blue spells, and may struggle with feeding problems. In these infants, surgery may be indicated, typically a supraglottoplasty or aryepiglottoplasty, and in rare cases a tracheotomy.

A clinically similar condition to congenital laryngomalacia affects children with neurological disorders, and particularly those who have low muscle tone (hypotonia) in the trunk or neck. One author postulates how this may occur:

The term *laryngomalacia* is used imprecisely in children with CP [cerebral palsy]. In the laryngomalacia of infancy, it is the cartilages of the airway themselves that are overly pliable and collapse centripetally during inspiration, whereas in the “laryngomalacia” of CP, the floppiness of the airway is caused by soft tissue laxity, not by too pliable cartilage. Neurologic abnormalities probably predispose to the development of this soft tissue laxity. The reflex causing upper airway dilation during inspiration may be absent in these patients, resulting in relative airway narrowing in inspiration. Respiratory muscle and diaphragmatic movement may be too forceful and not properly modulated. Together, these factors result in an increased velocity of airflow through the upper airway, causing so-called suction traction on soft tissues by the Bernoulli principle. Suction traction initially causes stretching of the soft tissues over supraglottic cartilaginous structures. As the soft tissues become progressively more

redundant, they prolapse into the airway, eventually bringing with them the underlying supraglottic cartilages (the arytenoid, corniculate, and cuneiform cartilages). Suction traction also distorts the shape of the epiglottis, causing it to become “omega-shaped” and to prolapse posteriorly.¹

While somewhat difficult to understand, the author proposes that a floppy airway is the problem for most children with non-congenital laryngomalacia. Over time, the combination of a floppy airway and improper breathing due to muscle tone cause the tissues of the larynx to fall into the airway, closing it off and causing stridor.

Another Possible Disorder: Pharyngomalacia

Another rarely diagnosed but probably common condition is pharyngomalacia or pharyngeal collapse. The pharynx, the part of the airway situated at the back of the mouth above the larynx, can also collapse, contributing to stridor. It is thought that many children with laryngomalacia, particularly those with floppy trunks, may also have a pharynx that collapses upon inspiration. In the past, not all bronchoscopies looked at the pharynx, leaving pharyngomalacia a rarely diagnosed condition. The number of children with pharyngomalacia is likely to increase dramatically once it becomes standard practice to look at both the larynx and pharynx in children with stridor and hypotonia.

An early article published in 1997 introduced the idea of pharyngomalacia and hypotonia, making a connection between children with laryngomalacia who lacked the typical anatomical deformities congenital laryngomalacia (such as extra tissue in the larynx) and who also exhibited pharyngeal collapse.² Of these children, 67% had neurological symptoms, with hypotonia the most common abnormality. More than half of these children had been treated with typical surgical means, mostly supraglottoplasty, but none was cured. Several children ended up needing BiPAP, and approximately half required tracheotomies. The authors ultimately posit a neurological explanation for the concurrent laryngeal and pharyngeal collapse. Another case report on a child with Down syndrome similarly links the “generalized floppiness” of children with Down syndrome to pharyngomalacia or hypopharyngeal collapse.³

Stridor from Laryngeal Dystonia

Yet another possible cause of stridor in children with muscle tone disorders is laryngeal dystonia. Simply put, dystonia is an involuntary contraction of agonist and antagonist muscles, usually causing twisting and arching in the trunk of children with dystonic cerebral palsy. In these children, the muscles of the larynx may also be affected by dystonia, causing stridor when inhaling.⁴ Treating the underlying dystonia with baclofen or gabapentin, or directly treating the vocalis muscle with botox may completely eliminate stridor.

Laryngeal dystonia is not easily distinguished from laryngomalacia. It should be suspected in children with underlying dystonia and in children whose stridor disappears completely while asleep or under anesthesia.

Surgical Complications for Children with Neurological Disorders

Several studies have noted that children with cerebral palsy, hypotonia, or other neurological disorders do not respond well to the typical surgeries used to correct stridor, particularly the supraglottoplasty. One study of Taiwanese children noted very poor outcomes for children with cerebral palsy, with minimal improvement as compared to children without cerebral palsy receiving the same types of surgeries.⁵ Another study, which boasts a 100% resolution of symptom rate in children without any neurological disorders or other abnormalities, shows that all children with shortterm unfavorable results were neurologically impaired, and the children who were ultimately considered surgical failures all had other abnormalities, most commonly neurological in origin.⁶ Ultimately, only 57% of children with associated abnormalities showed resolution of their symptoms within six months of surgery, compared to 100% with no other abnormalities. Some authors go so far as to suggest that a tracheotomy is the procedure of choice for children with severe laryngomalacia and hypotonia.⁷

Typical complications of supraglottoplasties include dysphagia (trouble swallowing) and aspiration, since the redundant tissues of the larynx removed during surgery may actually protect some children from aspiration. These complications also tend to be more prominent in children with hypotonia or neurological disorders.

In addition, the presence of pharyngomalacia often leads to poorer results from surgery. The pharynx should be checked for collapse on inspiration before surgery is undertaken.

Suggestions for Treating Children with Moderate to Severe Stridor and Neurological Disorders

For children with mild to moderate stridor that does not cause blue spells, feeding problems, fatigue, or a need for oxygen, attempts should be made to reduce the stridor using lifestyle modifications and medications. Children with severe stridor that causes additional symptoms may benefit from BiPAP, CPAP, or surgical interventions. While some children may have success with standard laryngomalacia surgeries such as a supraglottoplasty, some may unfortunately require a tracheotomy.

The following suggestions may help to reduce stridor.

- ❑ **Reduce or eliminate any medications that reduce muscle tone or cause weakness.** Tone-reducing medications are very commonly used in children with cerebral palsy and similar conditions to treat seizures, irritability, or hypertonia. These medications, however, may also reduce muscle tone in the throat, leading to a floppy larynx and/or pharynx. Reducing or eliminating these medications may dramatically reduce stridor. Some of these medications include Baclofen, Zanaflex, benzodiazapenes such as Valium, Ativan, or Klonopin, and narcotics. Alternatives may include a targeted baclofen pump, localized botox injections, and non-sedating medications for mood and pain.

- ❑ **Reduce or eliminate dystonia.** Children with generalized dystonia may also have dystonia of their airways. Reducing generalized dystonia by using baclofen or gabapentin may eliminate stridor. Children with localized dystonia of the larynx may benefit from botox injections.
- ❑ **Treat reflux aggressively.** Reflux, and particularly reflux that reaches the level of the pharynx or larynx, can cause swelling and irritation of the airway, thereby worsening stridor. Reducing reflux by using motility medications, acid suppressants, GJ tubes, or surgery will in turn reduce stridor.
- ❑ **Eliminate airway inflammation and irritation.** Airway inflammation and irritation frequently leads to worsened stridor. Some children may benefit from a daily nasal steroid such as Nasonex or Flonase. Others may need a stronger oral steroid such as decadron during periods of croup or colds. In some children, simply flushing the nasal passages with saline may help reduce inflammation and irritation. Children who aspirate also tend to have significant airway inflammation. In these children, reduction of secretions through medication or suctioning, a fundoplication surgery, or a GJ tube may reduce airway inflammation, thereby reducing stridor.
- ❑ **Use positioning.** Many children with cerebral palsy have increased stridor in certain positions, such as when the head is leaning back or the upper body is reclined. Positioning the head in an upright or forward-leaning posture may improve stridor.
- ❑ **Give it time.** In many children, stridor improves as they grow and their airway becomes bigger and stronger.

¹ Gordon Worley, *et al.* Laryngeal Dystonia Causing Inspiratory Stridor in Children with Cerebral Palsy. *Laryngoscope* 2003;113:2194.

² Patrick Froehlich, *et al.* Discoordinate Pharyngolaryngomalacia. *International Journal of Pediatric Otorhinolaryngology* 1997;39:9-18.

³ Lee Shing Yan Robert, *et al.* Obstructive sleep apnea syndrome secondary to pharyngolaryngomalacia in a neonate with Down syndrome. *International Journal of Pediatric Otorhinolaryngology* 2005;69:919-21.

⁴ Worley, 2192-5.

⁵ Kuo-Sheng Lee, *et al.* CO2 laser supraglottoplasty for severe laryngomalacia: A study of symptomatic improvement. *International Journal of Pediatric Otorhinolaryngology* 2007;71:889-95.

⁶ Craig W. Senders, *et al.* Laser supraglottoplasty for laryngomalacia: are specific anatomical defects more influential than associated anomalies on outcome? *International Journal of Pediatric Otorhinolaryngology* 2001;57:235-44.

⁷ David J. Kay, *et al.* Laryngomalacia: A Classification System and Surgical Treatment Strategy. *ENT-Ear, Nose & Throat Journal* 2006;328-31; 336.