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FEATURE

Sniffs, Gasps, and Coughs

Irritable Larynx Syndrome Across the Lifespan

see also: Symptom Differences Between PVFM/VCD and Asthma | Focus on Divisions | References

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by Mary Sandage

Irritable Larynx Syndrome (ILS) (Morrison, Rammage & Emami, 1999) has as its central thesis the idea that we can develop neuroplastic changes in the brain when a threshold of tolerance to an irritation in the upper airway or larynx is passed, creating conditions which may include chronic cough and paradoxical vocal fold motion (PVFM). This model is appealing for patients who have been told by health care providers, likely out of frustration, that they should "just quit coughing" or "just breathe." It is helpful to consider that perhaps it isn't in the patient's head. Widdicombe and Singh (2006) recently published a review of the literature that addresses how cough can be "down-regulated," including voluntary suppression of cough as a likely but little-studied mechanism.

Little has been published about the speech-language pathologist's role in the assessment and treatment of PVFM and even less has been written about behavioral intervention for chronic cough. While these areas of assessment and treatment do not directly impact communication like dysphagia, no behavioralist knows the anatomy and physiology of the vocal folds and upper airway better than SLPs. While chronic cough and PVFM share many features, I will discuss them separately to honor the nuances of each.

Chronic Cough

Florence Blager (1988) was the first to describe a behavioral intervention for chronic or habitual cough. Cough becomes chronic if it lasts for more than three weeks, is not related to an active infection or disease process (Irwin, et al., 1998), and presents as a dry cough without productive mucous. Fatigue, change in social

function (e.g., avoiding restaurants), and trouble sleeping are common physiologic complications, with few reporting associated hoarseness. Patients may range in age from school-aged children to older adults, with the average length of coughing lasting from months to years. Most have tried many medical interventions without resolution of the cough.

Medical Management

In the absence of pulmonary disease or infection, postnasal drip, asthma, and/or gastroesophageal reflux disease (GERD) are considered the primary causes for chronic cough most of the time in a non-smoker (Irwin, et. al., 1998; Morice, et al., 2004). Children can develop chronic cough for many of the same reasons as adults; however, pediatric cough is less well understood. Medical management should include assessment and treatment for pulmonary disease, asthma, postnasal drip syndrome/rhinitis, gastroesophageal or laryngopharyngeal reflux disease (LPR), and extrathoracic obstruction prior to referral to an SLP. Endoscopic assessment of the larynx should be completed on all patients to rule out laryngeal pathology that may trigger cough without degrading voice quality, e.g., contact ulcer, and assess for clinical signs of reflux. Once the larynx has been visualized and the SLP is certain that all medical management has been fully explored, it is appropriate to move ahead with behavioral intervention to reduce or eliminate chronic cough.

The Role of the SLP

A thorough review of the patient's medical history is vital. Unfortunately, in my clinical experience, medical conditions triggering cough have sometimes been overlooked or under-treated, such as dysphagia or LPR, requiring referral back to the physician for further medical management. Discerning the pattern and frequency of cough as well as identifying trigger(s) for cough provide the necessary information to establish a behavioral plan. Ask detailed questions about the nature and pattern of the cough.

Intervention can be best described in three steps: improve the environment of the larynx to reduce irritation; train awareness of the sensations and behaviors that precipitate the cough; and implement a behavior that will delay/eliminate the cough before it happens to recalibrate the sensitivity threshold.

In addition to medical management to reduce laryngeal irritation that may trigger cough, it is important to counsel patients to eliminate other agents that may be exacerbating coughing such as menthol cough drops, gargling, dry environment, mouth breathing, and dietary components that exacerbate GERD or LPR. Encourage patients to hydrate and eat "wet" snacks (e.g., apple, melon, pear,

cucumber), swallow frequently, increase the humidity in the work or home environment, and breathe in through the nose.

Secondly, focused attention to the physiological precursors to cough is critical for success (e.g., dry tickle). Ask the patient to learn more about any environmental or behavioral influences (e.g., desk is under heating vent). Finally, several short periods per day of active cough suppression will build focused attention and enhance success for delaying or inhibiting cough. Patients are asked to choose from the following behaviors and use one or more instead of coughing: taking ice chips or cold water, hard swallow, hard swallow with laryngeal repositioning, or nasal inhalation, which keeps vocal folds open, humidifies air, and redirects attention away from cough.

Children, school-aged and older, also can benefit from this approach; however, they will likely require a longer generalization period. Follow-up at one-week and one-month intervals is important to refine the behavioral plan, facilitate carryover, and promote reduction/resolution of the cough.

Paradoxical Vocal Fold Motion

PVFM occurs when the vocal folds approximate or approach each other, particularly during inspiration, creating a narrow airway. Some patients produce stridor, the sound made when the vocal folds draw together during inspiration. PVFM is also called vocal cord dysfunction (VCD) and these terms are used interchangeably. PVFM continues to be frequently misdiagnosed as asthma or, if it co-occurs with asthma, it may not be recognized as a separate disorder.

This disorder is more common in females and has been documented in patients from infancy to geriatrics (Heatley & Swift, 1996; Maschka, et al.,1997), with most patients falling between 10 and 40 years of age (Kuppersmith, Rosen, & Wiatrak, 1993). PVFM in athletes is more prevalent (about 5%) than first thought (Rundell & Spiering, 2003).

The three primary causes for PVFM are upper airway sensitivity to laryngeal irritants (e.g., GERD/LPR, allergens/inhaled irritants, and post-nasal drip), psychological conditions, and laryngeal dystonia/dyskinesia (Mathers-Schmidt, 2001). Because the primary role of the upper airway or vocal folds is airway protection to ensure survival, the larynx is equipped with mucosal chemoreceptors, which, when irritated, signal the vocal folds to narrow or close. Irritating gas, such as from a chemical lab or factory; smoke; fumes (e.g., printed materials; vapor, e.g., sulfur dioxide, cleaning agents; mist; and dust) are documented triggers in patients with PVFM (Perkner et al., 1998). LPR may be a common, unrecognized trigger in children (Powell, et al., 2000).

Initially, PVFM was believed to be triggered primarily by psychological conditions. Caution must be taken to avoid indicating that the breathing disorder is "all in the head." This disorder has been described in infants and in children too young to demonstrate a conversion disorder or other psychiatric condition, which suggests that physiological triggers may play a primary role (Bless & Swift, 1993).

The neurological form of PVFM, diagnosed as episodic laryngeal dyskinesia or adductor laryngeal breathing dystonia, is rare and likely will not respond to behavioral methods, requiring medical intervention such as surgery, medication, botulinum toxin, or tracheostomy (Worley, Witsell, & Hulka, 2003; Maschka, et al., 1997). This form of PVFM will typically present as persistent inspiratory stridor during waking hours, with no reports of discrete breathing "attacks," and is often accompanied by dystonia or tremor in other parts of the body.

Medical Assessment

Like chronic cough, PVFM requires thorough medical assessment prior to referral to an SLP. The differential diagnoses that are considered during the initial medical work-up for reports of inspiratory stridor include asthma attack; gastroesophageal reflux; laryngeal pathology (e.g., laryngeal edema secondary to allergen, unresolved laryngomalacia, bilateral vocal fold paralysis, laryngeal mass); PVFM/VCD; and panic attack. An endoscopic assessment of the upper airway, completed by the SLP or an otolaryngologist, is necessary to rule out an obstruction, which would cause both inspiratory and expiratory stridor, particularly with exertion or laying flat on the back, during waking, resting, and sleeping. A thorough pulmonary assessment should be completed to definitively determine if the patient has asthma. A formal allergy assessment also will be warranted for patients who describe allergens as triggers for the breathing events. Patients with PVFM likely will report that they have only partially responded or not responded at all to empirical treatment for asthma and/or allergies. To make matters more confusing, about half of patients with PVFM also have asthma. There are distinct differences between asthma and PVFM/VCD (see Table on page 17).

The Role of the SLP

Extensive knowledge of likely triggers, medical management of those triggers, and behavioral methods for intervention are necessary to adequately serve this population. SLPs should allow patients to tell about their breathing difficulties, letting the stories unfold with some guidance and taking care not to lead the patient in a specific direction. Although these patients share common features, their unique presentations will inform a personalized behavioral treatment plan for recovery. The severity of the breathing challenge can be assessed using information about the frequency and intensity of the attacks. Frequency may

range from two attacks per month to 20 per day and a single "attack" may last from a few seconds to several minutes.

PVFM Treatment

Most patients will require medical management of the trigger(s) in order to support the breathing recovery exercises. The environment of the larynx must be improved to allow the breathing recovery exercises to be effective, eventually recalibrating the sensitivity of the larynx back to a typical threshold. Medical intervention may include reflux management, nasal drainage management, and asthma or allergy symptom control as warranted.

There are three basic steps for behavioral treatment: training awareness, training relaxed belly breathing, and finally, training the actual breathing recovery method. The behavioral program is described in detail in the literature (Mathers-Schmidt, 2001; Sandage & Zelazny, 2004). While the first step may appear unnecessary, I learned the hard way that patients are more successful with the program if their awareness to subtle changes in the body has been conditioned. This is accomplished through an exercise that is much like progressive relaxation; however, the objective is not necessarily to relax, but to develop the ability to attend to or notice very small changes in the body. This heightened attention trains patients to notice any physiologic sign that a breathing attack is imminent so that they can start the breathing exercise before the paradoxical behavior begins.

Secondly, the perception of air hunger promotes changes in breathing patterns that can exacerbate the feeling of breathlessness. Many patients with PVFM have extraneous neck and shoulder tension and rely on high-chest breathing. The goal is to re-establish belly expansion when inhaling, with belly collapse when exhaling while maintaining relaxed shoulders, neck, and jaw.

During the final step, the patient learns specific breathing patterns to use when feeling the onset of a PVFM attack to keep the vocal folds open during inhaling and exhaling. If the recovery exercise is started before the paradoxical behavior starts, the patient should be able to avoid an event completely, even if the onset occurs during extreme physical exertion as with athletes. The therapeutic process generally takes between three to five visits following the initial assessment.

SLPs are successfully applying behavioral methods to reduce or eliminate chronic coughing and PVFM. Once the patient experiences a few weeks without any cough or breathing difficulties, the patient may be referred back to the physician to determine if unnecessary medications can be discontinued, e.g., rescue inhalers, reflux medication. If behavioral intervention fails, the patient should be referred

back to the physician for consideration of other medical interventions as previously described.

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