Neurofibromatosis Type 1 and Velopharyngeal Insufficiency Kimberly Marrs, B.S., & Heather Thompson, Ph.D., CCC-SLP

What is velopharyngeal insufficiency (VPI)?

Velopharyngeal insufficiency (VPI) is a resonance disorder that occurs during speech that results from the inability of the velum (or soft palate) to meet with the nasopharynx (or the back of the throat). When breathing through the nose, the velum is lowered and moves away from the nasopharynx, allowing air to exit the nose. When speaking, the velum is raised and lowered, to allow the individual to produce different speech sounds. For example, to produce a "b" sound, the velum must be raised, forcing all of the air out of the mouth, and to produce an "m" sound, the velum must be lowered, allowing air to flow out of the nose. When the velum closes while talking, air cannot come out of the nose. When a person has VPI, there is an incomplete closure between the velum and nasopharynx, forcing sound into the nasal cavity. VPI results in nasal sounding speech qualities called *hypernasality* and *nasal air emission* (Kummer, 2020).

What causes VPI?

One of the most common causes of VPI is cleft palate. Individuals with cleft palate may have been born with a space in the roof of the mouth, they may have a short velum, or a velum that does not move as well as it should. Other causes of VPI include differences in the tonsils and adenoids, oral cavity tumors, or deficits that take place after surgery or head injury (Kummer, 2011).

How is VPI relevant to Neurofibromatosis Type 1 (NF1)?

Research has shown that individuals with NF1 may have hypernasality and/or VPI (Alivuotila et al., 2010; Cosyns et al., 2011; Pollack & Shprintzen, 1981; Saal et al., 2004; Thompson et al., 2010, 2013). One study found VPI to occur in 7.4% to 52% of patients with NF1 (Zhang et al., 2012). The exact cause of hypernasality and/or VPI in individuals with NF1 is unknown. Further research is needed.

How is VPI diagnosed?

An evaluation of velopharyngeal function begins with a speech-language pathologist or otolaryngologist (ENT). A diagnosis of VPI will be determined by examining the nose and throat with procedures called nasopharyngoscopy or videofluoroscopy (Kummer, 2019). Nasopharyngoscopy is when a small camera on a wire or scope is inserted into the nose to look at the velum and the back of the throat. Videofluoroscopy is an x-ray that takes pictures of the velum and back of the throat during speech. During videofluoroscopy procedures, a barium solution can be used to coat the nose and throat, allowing them to be seen and to determine if the velum is short or if it does not move as it should. Speech-language pathologists and otolaryngologists can work together to care for patients with VPI.

Why is it important to get an evaluation?

VPI can have significant effect on a person's ability to be understood by others (Kummer, 2019). An early evaluation is important as people with VPI may not only have poor communication effectiveness, but the presence of VPI may also increase the risk of social or emotional concerns (Johnson et al., 1999; Kummer, 2019; Lallh & Rochet, 2000; Stelck et al., 2011). Communication concerns such as VPI may also lead to challenges in communication in school or work settings.

How is VPI treated?

If determined that the speech disorder is due to VPI, an otolaryngologist, speech-language pathologist, and/or plastic surgeon may decide on treatment options in conjunction with the patient. Research is helpful to guide clinical decision-making. One option for improving VPI due to a structural difference may be a speech appliance called an obturator. Speech therapy cannot correct a structural difference, but it may be recommended after surgery to improve articulation for individuals who have difficulty producing sounds in a way that is consistent with the language(s) spoken.

Where can I receive more information about VPI?

Individuals who are concerned about resonance can reach out to their doctor or speech-language pathologist. Other resources for information about resonance and/or VPI include:

- 1. <u>American Speech-Language-Hearing Association (ASHA)</u>
- 2. <u>American Cleft Palate-Craniofacial Association (ACPA)</u>

<u>References</u>

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