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VCFS News: Your Information Resource



Peer Group Announcement and Update

Hi. My name is Raymond Cheng, and I am the director of the peer group program. For the past few months, I have been honored with organizing and hosting the groups. They have become incredibly successful, with new participants coming in every week. As someone who also has VCFS, it has been a dream and goal of mine to help those with similar needs, and to be able to change so many lives is a dream come true. The peer groups have gotten nothing but positive feedback, with numerous participants unable to contain their excitement, and coming back as frequent members.

In fact, they have been so successful that we have changed the frequency of the groups from every other week to every week. There are two groups currently: one on Wednesday at 5PM EST, and Thursday at 8PM EST. We can imagine there will be another slot opening soon.

We hope that more participants will be interested in the groups. Their only purpose is for the participants to make friends and socialize. Every session so far has been interesting and engaging, with a variety of topics to chat about, upcoming events and/or holidays to look forward to, and much more. There have been laughs and deep connections. There will potentially be games implemented as well. It is an hour of fun and conversations. We hope to see you join us, and as always we look forward to hosting the next groups to come!

To speak with me and to find out more about the groups or joining one, contact me by email at raymond.chang@vcfscenter.org. I'd love to speak with you.

Matthew Rontal is a Facial Plastic Surgeon in Royal Oak, Michigan, a suburb of Detroit. Dr. Rontal completed medical school at the University of Michigan and his internship at the Albert Einstein College of Medicine Medical Center in the Bronx, and a Fellowship at the Hospital of the University of Pennsylvania in Philadelphia. His residency in Otolaryngology was completed at the University of Michigan Hospitals in Ann Arbor. He completed a Fellowship in craniofacial surgery at Upstate Medical University where he was exposed to velo-cardio-facial syndrome under the tutelage of Dr. Shprintzen. He began his practice working with the world-renowned craniofacial surgeon, Dr. Ian Jackson, working with him until his retirement in 2010. He joined the Rontal Clinic in 2010 and is a faculty member at Oakland University William Beaumont School of Medicine. Dr. Rontal has lectured locally, regionally, nationally, and internationally. He is highlighted on the website of the Virtual Center for Velo-Cardio-Facial Syndrome in the link to his lectures at the Burlo Garofolo Children's Hospital in Trieste, Italy where he has also demonstrated his surgical approach to velopharyngeal surgery in children with VCFS.



Q&A with Dr. Rontal

- 1. We were told that our 6-year-old child who has VCFS needed an operation to eliminate VPI and nasal speech. We have seen three surgeons, and we got three different recommendations. The first recommended an operation called a pharyngeal flap, a second doctor said that a pharyngeal flap was dangerous because of sleep apnea and a sphincter pharyngoplasty was recommended, and a third doctor said to try speech therapy for 6 months and to come back after. What should I do?*

In my opinion a 6-year-old with VCFS who has VPI will need a surgical correction in almost all cases. This is related to the anatomy that is specific to the syndrome, and also the lower muscle tone that affects all of the muscles in the body in people with VCFS. Speech therapy cannot resolve VPI according to the leading experts who have studied VPI in both VCFS and in people with cleft palate.

In terms of which procedure to choose, we believe strongly that superiorly based pharyngeal flap is the best choice when compared with than sphincter pharyngoplasty or other procedures. The pharyngeal flap can be tailored to the dimensions of the velopharynx (the region where the palate and walls of the throat work together during speech). Perhaps most importantly, the pharyngeal flap can be placed high enough in the velopharynx to function appropriately without causing obstruction of the airway in the portion of the pharynx just behind the soft palate. Sphincter pharyngoplasty is lacking in sufficient width or bulk, especially in VCFS cases and cannot be positioned high enough in the velopharynx to resolve the VPI. When done appropriately, pharyngeal flaps can be tailored to the defect.

Secondary arguments can be considered further as well. Many claim that sphincter pharyngoplasty is “dynamic” - that the muscles within the reconstruction move during speech to close the velopharynx. This has been clearly disproven by Dr. Ysunza. The muscles in the tissue used for reconstruction have no residual function after surgery and even if they did move, their movement would be the opposite of that which is desired during speech. The sphincter procedure places an insufficient and arbitrary bulk of tissue in the general vicinity of where it may be useful with little control and random success. In VCFS it is very unlikely to be useful because the velopharynx is very wide and the tissue used in the sphincter is too thin and narrow to provide adequate closure of the velopharynx.

As far as sleep apnea after pharyngeal flap, this has been addressed through prior removal of adenoid and tonsil tissue. Studies with large numbers of patients have shown that the risk of sleep apnea is reduced to 1% or less if tonsils and adenoids are removed first.

2. What is the success rate for operations that are meant to eliminate VPI?

Success rates in the medical literature are variable. Part of the reason for that is that the measurement of success from one study to the next is not standardized. Some centers require that success is defined as complete and total elimination of hypernasality. Others use more vague measures of improvement in speech. Still others use objective measurements of nasality that may or may not be correlated with actual observation of speech.

Overall, the success rate in most studies is usually around 85-95%. In my practice and in our center, the success rate is in the 95% range as defined by total correction of VPI as observed by experts in surgery and speech pathology based on postoperative nasopharyngoscopy and assessment of nasal air escape during non-nasal speech samples. Using ranking scales such as normal-mild nasality-moderate-severe or “improvement” scales that are useless. We use the approach of direct observation of the closure of the velopharyngeal mechanism and the presence or absence of resonance and airflow through the nose several months after surgery because a lesser result is still abnormal speech carrying some degree of isolation from the rest of the speaking world.

3. How long is a child hospitalized after pharyngeal flap surgery?

On average, the postoperative stay is 2 nights in my practice. When a child demonstrates that he or she will support themselves with oral intake of fluids, we feel comfortable with discharge. A few kids are ready on post op day 1. A few take a few to several days to be willing to swallow sufficiently after surgery. The vast majority are ready on postoperative day 2.

4. *Is there anything special that needs to be done before surgery to eliminate VPI in a child with VCFS?*

As mentioned earlier, adenotonsillectomy would be done prior to pharyngeal flap. There are two reasons for this. First and foremost, pharyngeal flap surgery has the effect of blocking some of the airway between the oral cavity and nasal cavity. The tonsils sit just below this location, and the adenoids just above. After surgery, this is likely to cause too much obstruction of the airway that could result in developing obstructive sleep apnea. Another reason is that the incisions in the throat for pharyngeal flap includes the place where the adenoids sit. The adenoids cannot be cut so when adenoids are present, the location of the pharyngeal flap will be too low to be effective for speech.

Another important need is to visualize the course of the internal carotid arteries in the back of the throat. It is well known that these major arteries that supply blood to the brain can often be in the operative field in people with VCFS and there is a risk of damaging the arteries that could result in a major complication. By knowing the location of the arteries before the surgery, it is possible to avoid damaging them thereby eliminating the complication. This is done by doing either MRA (Magnetic Resonance Angiography) or CTA (Computed Tomography Angiography).

Another issue is the other aspects of speech often found in VCFS. Articulation of sounds is often very severely impaired in children with VCFS. Even in the presence of VPI, the articulation disorder can be corrected by approaching the impairment correctly. The more progress there is in normalizing articulation prior to surgery, the better the child will sound after surgery. Speech therapy may still be needed after surgery but will often be easier to implement with corrected VPI.

5. *Are all operations to treat VPI the same? If not, how do you know what operation to do?*

Surgical procedures for VPI all attempt the same thing - to partially obstruct the velopharynx (the region where the palate and walls of the throat work together during speech) such that complete closure can be performed quickly and efficiently by the patient during normal speech. However, the pharyngeal flap achieves this result more reliably than the sphincter pharyngoplasty. Furthermore, the procedure should be tailored to the particular velopharyngeal gap and function in an individual patient. In other words, the gap needs to be measured using imaging as described in the previous newsletter by Dr. Ysunza.

6. *As a surgeon, how do you know what to do for a patient with VPI? Do you always do the same thing for every patient?*

We determine the extent and dimensions of how wide the pharyngeal flap needs to be based on measurements of the velopharynx and its function during speech in each individual patient. Measurements are created via video multi-view X-ray before surgery and nasopharyngoscopy before surgery. The information from both of these exams is mandatory for us to plan our approach for every patient.

7. *Is pharyngeal flap surgery more difficult in someone with VCFS than someone who does not have VCFS?*

VCFS presents unique challenges in surgery. The velopharynx (the region where the palate and walls of the throat work together during speech) is wider and the movement of the walls of the throat is less in VCFS. This demands a much broader flap in most cases. In addition, the tissue used in surgery is thinner and requires greater care in surgery. We also need to avoid damaging the internal carotid arteries which are displaced in people with VCFS which is not true for other people with VPI who do not have VCFS.

8. *What is the goal of surgery for VPI? How do you know if it is successful?*

The goal is complete elimination of hypernasality while avoiding risk to the patient. Because many people with VCFS have other anomalies that could be of concern during surgery such as congenital heart disease, the possibility of adverse reactions to medications that might be used during anesthesia, and the risk for airway obstruction, the importance of anticipating any possible problem related to the syndrome must be taken into account.

9. *At what age would you recommend doing surgery in a child with VCFS?*

Surgery should be performed when a child has grown such that the mouth and throat are large enough allow surgery to be done, and as soon as a child is able to participate with the preoperative exams as well as pre- and post-operative speech therapy. Usually this occurs around after 4 years of age. Most importantly, the child must be using speech to communicate. If a child's language development is severely delayed, it would be impossible to get the necessary information from preoperative evaluations that would guide the surgeon to know the specifics of how to approach the operation.



Extra, extra, read all about it!

In our next newsletter, we speak once again to our very own Ali Oppenheimer about the release of her second book, “Ali Always Said NO.”



The Virtual Center for VCFS