November 20, 2015

• Diagnosis/Types
• Common craniofacial syndromes associated with cleft lip and/or palate
• Feeding
• Surgical Timelines
• Orthodontic treatment
• ENT treatment

February 19, 2016

• Velopharyngeal Function
  – Speech
  – Swallow
• Velopharyngeal Dysfunction
  – Anatomy
• Resonance Disorders
  – Posterior
  – Anterior
• Compensatory Articulation
• 22q11.2 deletion syndrome
Review: “Holes” in the Palate

• Fistula
  – “Hole” or open communication between hollow cavities
  • TEF (Tracheal Esophageal Fistula)
  • In the population with a Cleft Lip and/or Palate, a fistula denotes a post-operative breakdown of tissue
    – Due to;
      - Healing
      - Fingers/nipples/hard objects in mouth

Property of Dianne Altuna

Review: “Holes” in the Palate

• Fistula versus Alveolar Cleft
  – Fistula
    • Denotes surgical complication
    • “hole” posterior to incisive foramen

Property of Dianne Altuna

Review: “Holes” in the Palate

• Fistula versus Alveolar Cleft
  – Alveolar Cleft
    • Present until alveolar bone graft
    • “hole” anterior to incisive foramen
    • 7-9 years of age – Alveolar Bone Graft
    • This is when alveolar cleft is closed
    • Orthodontic treatment prior to ABG will increase size of alveolar cleft

Property of Dianne Altuna
“Holes” in the palate

• Is it impacting resonance?
  – Plug it
    • Compare anterior phrases to posterior phrases
      – Puppy, puppy, puppy, vs. Cookie, cookie, cookie
        » If air loss on “Puppy” and not on “Cookie” it is the Alveolar cleft
          • What other sounds may be impacted?
            • Tongue tip/alveolar sounds
        » If air loss on “Puppy” and “Cookie” may be velopharyngeal dysfunction

Velopharyngeal Function

• The coupling and uncoupling of the oral and nasal cavities during:
  – Speech
  – Swallow
• Involves multiple structures:
  – Velum
  – Pharynx
VELOPHARYNGEAL ANATOMY

• Intersection
  – Velum
  – Posterior Pharyngeal Wall
  – Lateral Pharyngeal Walls

Velar Muscles
  – Levator Veli Palatini Muscle
  – Musculus Uvulae
  – Tensor Veli Palatini Muscle
  – Palatoglossus Muscle
  – Palatopharyngeus Muscle

Pharyngeal Muscles
  • Superior Pharyngeal Constrictor
  • Middle Pharyngeal Constrictor
  • Inferior Pharyngeal Constrictor
  • Thyropharyngeus Muscle
  • Salpingopharyngeus Muscle
  • Stylopharyngeus Muscle
Neurological Input

- Cranial Nerves
  - V
  - IX
  - X
  - XI

Cranial Nerve X
- Important for Pharyngeal Phase of Swallow

VELOPHARYNGEAL FUNCTION

- Swallow
  - Reflexive, Brisk
  - If not achieved, results in nasal regurgitation
  - Nasal regurgitation is a neurological event
  - Pharyngeal flap will not assist with nasal regurgitation

- Speech
  - Speech is a dynamic process of opening and closing of the velopharynx
  - Speech is NOT STATIC
  - "Blowing" exercises WILL NOT strengthen the velopharynx
  - "Blowing exercises WILL NOT assist velopharyngeal function

VELOPHARYNGEAL FUNCTION

Velopharyngeal closure during swallow is reflexive

THEREFORE

Velopharyngeal closure during swallow does not mean velopharyngeal closure during speech
VELOPHARYNGEAL FUNCTION

Speech

• Velopharyngeal closure is required for all sounds with the exception of /m,n,ng/
• Feel the difference
  • /m/
  • /b/

VELOPHARYNGEAL FUNCTION

Plosives
Air is stopped and then released within the oral cavity
• Bilabials /p,b/
• Tongue Tip Alveolar /t,d/
• Velar /k,g/

Sibilants
Airstream is maintained over time with fine motor control of tongue for direction of air
• /s,z/

Affricates
Air is stopped, then maintenance of airstream
• /ch,j/

Fricatives
Airstream is maintained over time with fine motor control of oral articulators
• Upper teeth on lower lip /f,v/
• Rounded tongue placement /sh/
• Tongue between teeth /th/

VELOPHARYNGEAL FUNCTION

Velopharynx
Closed
• Plosives
• Affricates
• Sibilants
• Fricatives
• All Vowels

Velopharynx
Open
• Nasals
**Velopharyngeal Function, Resonance and Articulation**

/p/ - lips together, release pressure; requires VP closure
/m/ - lips together allow airflow through nose
/p/ - tongue tip behind upper teeth, allow airflow through nose
/p/ - lips together, release pressure, jaw and tongue motion for “diphthong”

**P U M P K I N P I E**

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**VELOPHARYNGEAL DYSFUNCTION**

An inability of the **Velum, Lateral Pharyngeal Walls and Posterior Pharyngeal Wall** to make closure for any reason during speech and/or swallow

**VELOPHARYNGEAL DYSFUNCTION**

Velopharyngeal dysfunction can be due to:

A. Velopharyngeal Mislearning  
   i.e. “Phoneme Specific Nasal Air Emission”

B. Velopharyngeal Incomptency  
   i.e. “apraxia”, neurological deficit

C. Velopharyngeal Insufficiency  
   i.e. Anatomical deficit
VELOHARYNGEAL DYSFUNCTION

Velopharyngeal Dysfunction can be found in Population with:
- Submucous Cleft Palate
  - Incidence of SMCP is 1/90 in general population
- Cleft Lip and Palate
  - 10-15% of repaired
- Cleft Palate
  - 10-15% of repaired
- Cleft Lip or Cleft Lip and Alveolus
  - 25% risk

VELOHARYNGEAL DYSFUNCTION

Velopharyngeal Dysfunction can also be found in Population with:
- No history of cleft palate
- No history of submucous cleft palate
- Adults S/P CVA, MVA, Progressive Neurological Disorders
- Children with severe to profound HI

VELOPHARYNGEAL DYSFUNCTION

Signs and Symptoms
- History of nasal regurgitation post cleft palate repair
- History of need for multiple placement of PE tubes
- Nasal Grimace
- Hoarse Vocal Quality
- Decreased Intelligibility
Resonance
The coupling and uncoupling of the oral and nasal cavities, controlled by the raising and lowering of the velum

Abnormal Resonance
- Hypernasality
  - Too much nasal airflow; carried on vowels
- Hyponasality
  - Too little nasal airflow
- Cul-de-sac Resonance
  - A combination of hyper and hyponasality
- Nasal Air Emission
  - Bursts of air noted on consonants
ARTICULATION

Articulation Errors

• Developmental
  – w/r, w/l
  – Blend reduction
  – Phonological errors

• Otitis Media
  – Fronting (t/k, d/g)
  – Final Consonant Omission
Articulation Errors

• Orthognathic Influence
  – Mid dorsum placement
  – Inverted dental/labial placement

Articulation Errors

• Compensatory Articulation
  – Glottal Stops/Pharyngeal Stops
  – Nasal Fricatives/Velar Fricatives
ARTICULATION AND RESONANCE DISORDERS

• Articulation Disorders (i.e. glottal stops) can cause velopharyngeal dysfunction and a resonance disorder
• Velopharyngeal Dysfunction can cause an articulation disorder
• Articulation Disorder can co-exist with a velopharyngeal dysfunction
• Articulation Therapy can improve Velopharyngeal Function

ARTICULATION AND RESONANCE DISORDERS

• Blowing therapy will do NOTHING
• Please do not use blowing therapy to “strengthen the velopharynx”
• Blowing cotton balls, ping pong balls, and bowling balls will not strengthen the velopharynx, or help with velopharyngeal closure for speech production
• Blowing therapy should never be a goal
• Blowing therapy will do NOTHING

WHAT TO DO?
Evaluation and Treatment

• Isolation
  – Imitatively
  – Spontaneously

• Nonsense syllables/final position
  – Vowel........+ phoneme
    • Imitatively
    • Spontaneously

Evaluation and Treatment

• Think outside of the box
  – Through away “developmental patterns”
    • Focus on /sh/ if the child is producing with nasal frication even if they are only 3 years of age
    • Many times, it is easier for the child to produce a later developing sound (i.e. /sh/) compared to an earlier developing sound that has maintained abnormal articulation patterns.

Evaluation and Treatment

• Think outside of the box
  – Phonological processing will not help
    • Child knows they need to create a “stop”/plosive. For some reason they feel they can only do this at the level of the glottis (i.e. glottal stop).
      – Paring an oral stop to a glottal stop will result in the use of the glottal stop
      – The child needs to learn PLACEMENT in the oral cavity
Evaluation and Treatment

- Think outside of the box
  - Be creative
    - Use carbonated water for gargle
    - Use pixie sticks (or crystal light crystals) to help with increasing sensory
    - Use refrigerator tubing to help with oral airflow ($10.50 for 100 feet)

Evaluation and Treatment

- Single words/final position
  - Imitatively
  - Spontaneously
- Nonsense syllables initial position
  - /phoneme/ ......+ vowel
    - Imitatively
    - Spontaneously

Evaluation and Treatment

- Single words/initial position
  - Imitatively
  - Spontaneously
- Single words/medial position
- Phrases
Treatment

• Placement, Placement, Placement
  – Same motor planning needed to walk, ride a bike
  – May need to teach how to use lips for /f/; where to make contact for /k,b/

Treatment

• Repetition, Repetition, Repetition
  – Phonological Processing will not help with this population
  – This population has been using CAD for many years, which means they DO NOT use their oral cavity
  – 75-100 repetitions of targeted sound in 30 minute session

ARTICULATION AND RESONANCE DISORDERS

• Blowing Therapy
  – Is a waste of time, effort and money
  – Has no correlation to VP function
  – VP function is NOT static
  – AT NO TIME SHOULD BLOWING THERAPY BE A GOAL
Treatment

• Therapy Suggestions for Placement
  — Plosives

Treatment

• Therapy Suggestions for Placement
  — Sibilants

Treatment

• Therapy Suggestions for Placement
  — Affricates
Treatment

• Therapy Suggestions for Airflow
  – Produce /h/ in isolation
  – Produce /h/ in isolation with lips rounded
  – Occlude nares by pinching
  – Use blowing activities to facilitate oral direction of airflow **ONLY**
    • as blowing will **NOT** strengthen vp mechanism

In the Trenches

• Provide 3-6 months of “good” therapy
• Refer to Craniofacial Team in area
• Speak to Speech Pathologist on Team
• Always be looking at whole child

Craniofacial SLP

• Assessment
  – Perceptual
  – Imaging
    • Nasopharyngoscopy
    • Videofluorography
Good Outcome

Good Articulation → Surgery → Good Articulation/ Appropriate Resonance

Need More Speech Outcome

Bad Articulation → Surgery → Residual Artic Deficits Can Continue to Cause VPD

Outcomes

• Improved Oral Placement
• Improved Velopharyngeal Function
• Improved Speech Intelligibility
CLEFT LIP +/- PALATE

- Known anatomical difference
  - Original Cleft Palate
- Hypoplasia of all tissue (muscle tissue)
  - Muscles of velum also affected
- Fluctuating Hearing Loss
- Possible Articulation Complications
- Possible Language Delays
- Age of Palate Repair
  - Increased risk of VPD if palatoplasty after 18-24 months of age

CASE STUDIES

22q11.2 Deletion Syndrome
- DiGeorge Syndrome
- Shprintzen Syndrome
- Velocardiofacial Syndrome
- Takao Syndrome

22q11.2 Deletion Syndrome
- Catch 22 Syndrome
- Conotruncal Anomalies Face Syndrome
- Sedlackova Syndrome
22q11.2 Deletion Syndrome

- 1:2,000 - General Population
- 1:1,800 - Live Births
- Autosomal Dominant
- "New Mutations"
- FISH Study vs. Chromosome
- 90% with 3-Mb deletion spanning 3 million base pairs of DNA
- Late Diagnosis

22q11.2 Deletion Syndrome

- Neonate - Infancy
  - Presence of Hyptonia
    - Global
    - Laryngeal/Pharyngeal
  - Presence of midline cardiac defects
    - VSD, AOS, Pulmonic Arteriosis or Stenosis, TOF, Truncus Arteriosis, Vascular Ring

22q11.2 Deletion Syndrome

- Feeding
  - If cleft palate, this is NOT driving force for feeding disorder
  - Hyptonia
    - Pharyngeal hypotonia
    - Decreased oral and pharyngeal sensation
  - Decreased oral and pharyngeal motor
  - GI motility problems
  - VPD
    - Nasal Regurgitation during feeding
    - O2/Heart rate decrease during feeding
  - Cardiac complications and feeding
  - Alternate method of feeding

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22q11.2 Deletion Syndrome

- Neonate - Infancy
  - Oral Facial/Craniofacial
    - Cleft Palate
    - Pierre Robin Sequence
    - VPD
    - Asymmetrical Crying Facies
  - Presence of Endocrine Disorders
    - Hypocalcemia
    - Hypothyroidism
    - Absent/Hypoplastic Thymus

- Toddler
  - Language Developmental Delay
  - Motor Developmental Delay
  - Laryngeal Findings
    - Upper airway obstruction
    - Vocal cord paresis/paralysis
  - Ear/Hearing
    - Recurrent OME
    - Ear findings
  - Ear/Hearing
    - Recurrent OME due to involvement of tensor veli palatini muscle and poor innervation (CN X)
    - Conductive hearing loss
    - Asymmetrical sensorineural hearing loss
  - Hypotonicity, poor core strength, decreased gross and fine motor development
  - Upper airway obstruction secondary to obtuse angle of mandible/hypertrophic adenoid/tonsils
  - Vocal cord paresis/paralysis
22q11.2 Deletion Syndrome

- Toddler
  - Eye Findings
    - Suborbital congestion
    - Narrow palpebral fissures
  - Oral/Facial Findings
    - Enamel hypoplasia
    - Flaccid/hypotonic facies
    - Long face
    - Prominent nasal bridge

22q11.2 Deletion Syndrome

- Toddler
  - Short Stature/Small for Age
  - Oral/Facial Findings
    - Enamel hypoplasia
    - Flaccid/hypotonic facies
    - Long face
    - Prominent nasal bridge

22q11.2 Deletion Syndrome

- Childhood
  - Psychiatric Findings
    - Autism Spectrum Disorder
    - Bipolar affective disorder
    - Impulsiveness
    - Obsessive compulsive disorder
    - Severe anxiety

- Later Childhood
  - Cognition/Learning Disorders
    - Math concepts
    - Reading for comprehension
    - Concrete thinking
    - Drop in IQ scores with age
    - ADD/ADHD
    - IQ 60-80
22q11.2 Deletion Syndrome

- Later Childhood
  - Cognition/Learning Disorders
    - Math concepts
    - Reading for comprehension
    - Concrete thinking
    - Drop in IQ scores with age
    - ADD/ADHD
    - IQ 60-80

- Teen/Early Adulthood
  - Psychiatric Findings
    - Early onset Schizophrenia
    - Rapid mood disorder
    - Social Immaturity
    - Phobias
  - Social Skills
    - Slower development in adolescents due to emergence of psychiatric disorders
    - Deficits in facial emotion recognition
    - Increased amygdala volumes?
Books/Resources

- Therapy Techniques for Cleft Palate Speech and Related Disorders by Karen Golding Kushner
- Educating Children with Velo-Cardio-Facial Syndrome (Genetics and Communication Disorders Series) by Donna Cutler-Landsman
- Cleft Palate & Craniofacial Anomalies: Effects on Speech and Resonance by Ann W. Kummer
- Cleft Palate Speech Management: A Multidisciplinary Approach by Robert J. Shprintzen and Janusz Bardach
- Velo-Cardio-Facial Syndrome Volume I by Robert J. Shprintzen and Karen J. Golding-Kushner
- Cleft Palate & Craniofacial Anomalies: Effects on Speech and Resonance by Ann W. Kummer
- Cleft Palate Speech Management: A Multidisciplinary Approach by Robert J. Shprintzen and Janusz Bardach
- Velo-Cardio-Facial Syndrome Volume I by Robert J. Shprintzen and Karen J. Golding-Kushner
- Child Neuropsychology 2005 11(1)

Books/Resources

- Velo-Cardio-Facial Syndrome Volume II by Robert J. Shprintzen and Karen J. Golding-Kushner
- Missing Genetic Pieces by Sherry Baker-Gomez
- A Different Life: Growing Up Learning Disabled and Other Adventures by Quinn Bradlee
- VCFSEF.org or 1.855.800.VCFs
- www.acpa-cpf.org
- Ruscello, Dennis. An Examination of NSOME for Children with Velopharyngeal Inadequacy. Seminars in Speech and Language 29(4)
- Lof, Greg. Controversies surrounding use of NSOME for Childhood Speech Disorders. Seminars in Speech and Language 20(4)

QUESTIONS????????