Sunday, August 4, 2019

Breakout Session E29 • 10:45-11:45 am • Chantilly Ballroom West

Feeding Difficulties and Aspiration in CHARGE Syndrome

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Presenter Information

Dr. Hart is a faculty member in the Division of Otolaryngology at Cincinnati Children's Hospital Medical Center. She has fellowship training in the management of pediatric airway disorders and specializes in the management of children with complex aerodigestive disorders. She is a member of the CHARGE Clinic at Cincinnati Children's and cares for multiple children with CHARGE syndrome every month.

Presentation Abstract

Feeding difficulty affects up to 90% of children with CHARGE syndrome and aspiration in seen in 60-70% of children. Both dysphagia and aspiration can have significant impact on a child's health and quality of life. We will discuss anatomic, developmental and behavioral factors that contribute to feeding and swallowing difficulties. We will also review how aspiration is diagnosed and discuss management options.

Learning Objectives

- Understand anatomic, developmental and behavioral considerations that contribute to feeding difficulty in children with CHARGE syndrome.
- Recognize risk factors for aspiration and discuss diagnostic considerations and management options.

Dysphagia and Aspiration in CHARGE Syndrome

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August 2019
Dallas, TX

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Disclosures

- None

Topics to be Covered

- Feeding Difficulty in CHARGE
  - Anatomic considerations
  - Developmental considerations
  - Behavioral Considerations
- Aspiration in CHARGE
- Management Options
  - Therapy
  - Medical Options
  - Surgical Options

Feeding difficulties in CHARGE

- Up to 90% of children have some form of feeding difficulty
- Failure to thrive is common
- Aspiration present in 60-75%
- Can improve over time
- Multi-factorial

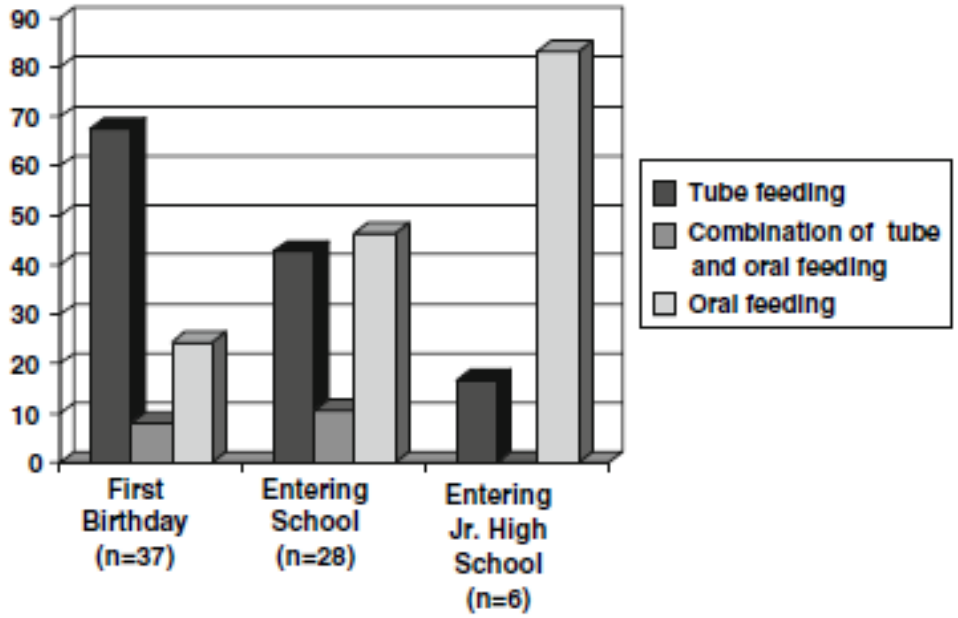
Feeding Difficulties

Dobbelsteyn, et al (2008):

- 39 children with CHARGE
- 92% required NG feeds at some point
- 77% s/p g-tube
- 62% remained feeding tube dependent

Feeding Difficulties

- By school age, < half ate exclusively by mouth
- Aspiration & GER associated with ongoing need for tube feeds
- Cranial nerve dysfunction significant contributor



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Feeding Difficulty: Major Considerations

- Cranial nerve abnormalities
- Structural abnormalities
- Behavioral issues

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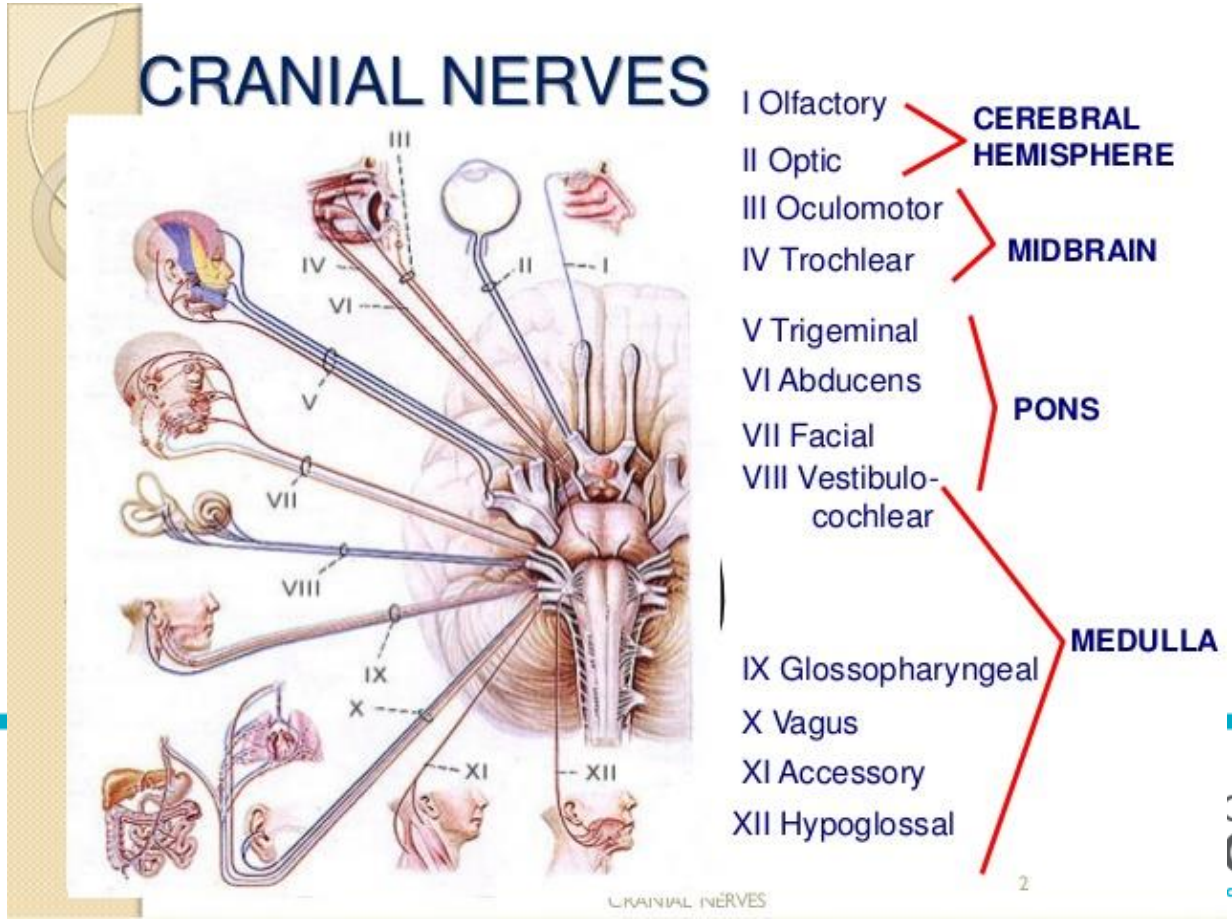
Cranial nerves

- Abnormalities present in 80-90% of patients



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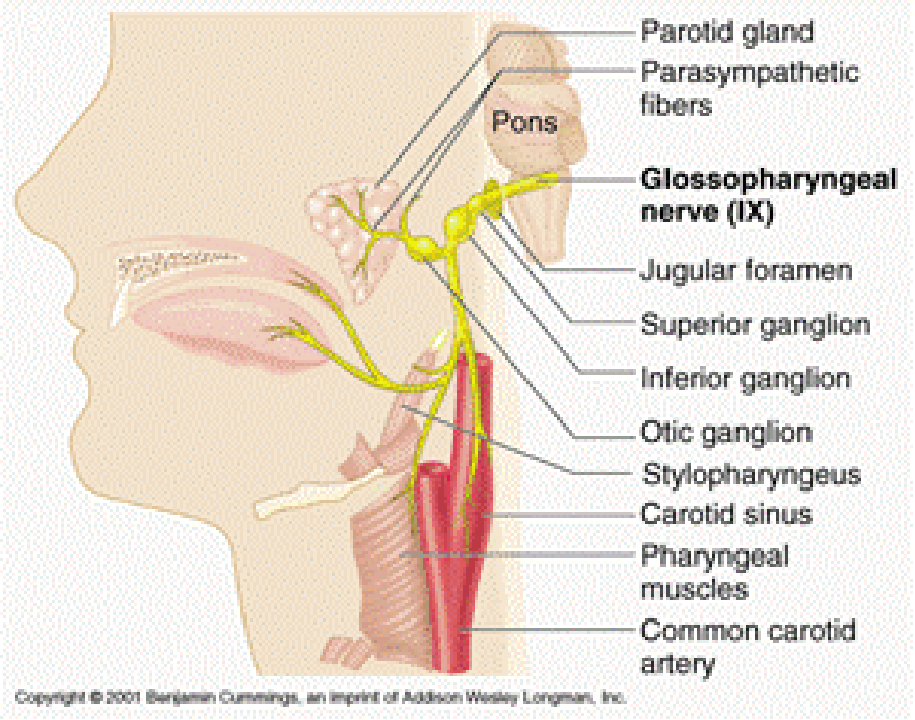
Cranial Nerve Abnormalities

- Negatively impact feeding development
  - Weak sucking/chewing
  - Swallowing difficulty
  - Gastroesophageal reflux
  - Aspiration

Cranial Nerves: IX

Glossopharyngeal:

- Sensory: pharynx, palate, posterior 1/3 of tongue
- Special sensory: taste for posterior 1/3 of tongue



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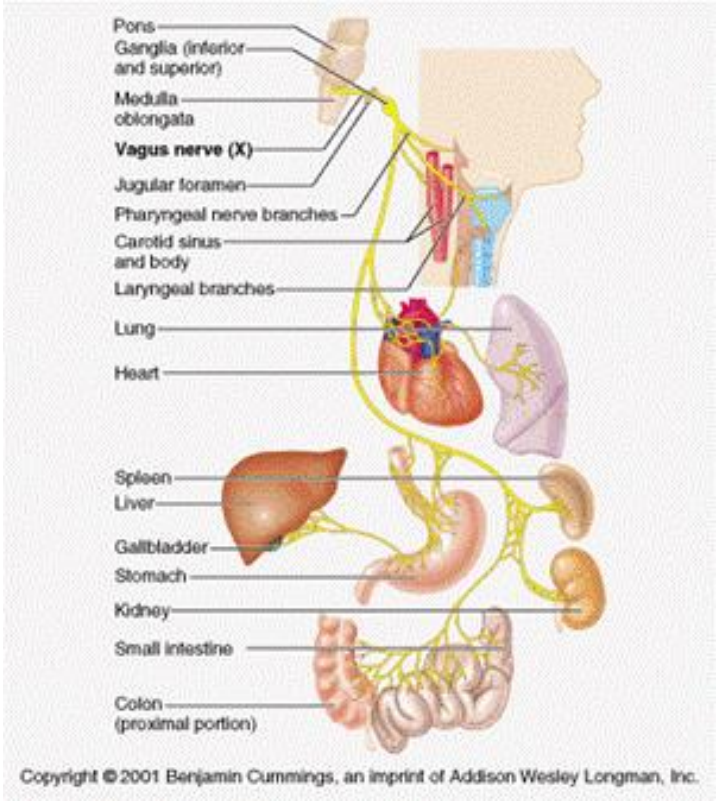
Cranial Nerves: X

Vagus:

- Motor: pharynx and larynx
- Sensory: larynx

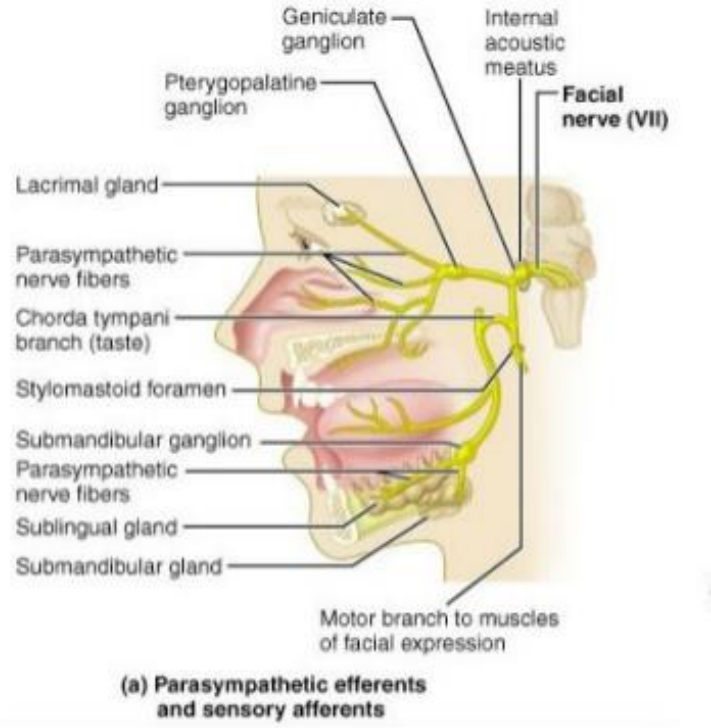
- Abnormalities of IX/X result in:
  - Inability to coordinate suck/swallow
  - Gagging
  - Aspiration



Cranial nerves: VII

Facial nerve:

- Can have symmetric bilateral facial palsy
  - Motor: Muscles of facial expression; contributes to hyoid/laryngeal movement
  - Special sensory: taste for anterior 2/3 of tongue
  - Innervation of submandibular and sublingual glands



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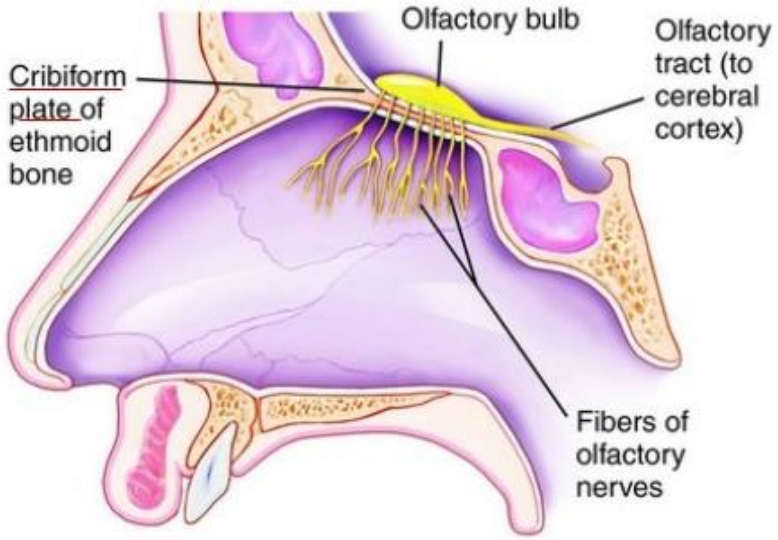
Cranial nerves: I

Olfactory nerve:

- Special sensory: smell
- Absent or reduced sense of smell
- Present in >80%
- Can be hard to assess
- Complicates learning to eat
- Safety issue (smoke, rotten food)



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Structural Abnormalities

- Can impact feeding directly
- Can create feeding difficulty indirectly
  - Increased work of breathing
  - Increased caloric needs

Structural abnormalities

Choanal atresia :

- 50-60% of patients
- Unilateral or bilateral
- Interferes with breathing
- Interferes with ability to eat by mouth

Structural abnormalities

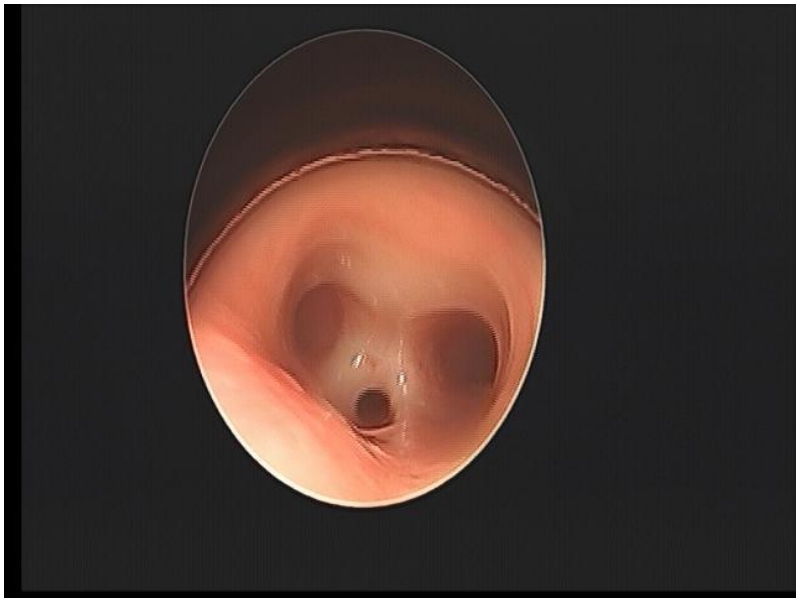
Cleft lip/palate

- Present in 25%
- Inadequate suck
- Interferes with swallowing

Structural abnormalities

Tracheoesophageal Fistula/Esophageal atresia

- Present in 15-20%
- TEF: Connection between trachea and esophagus
- EA: Incompletely formed esophagus
- Both prevent normal oral feeding until repaired



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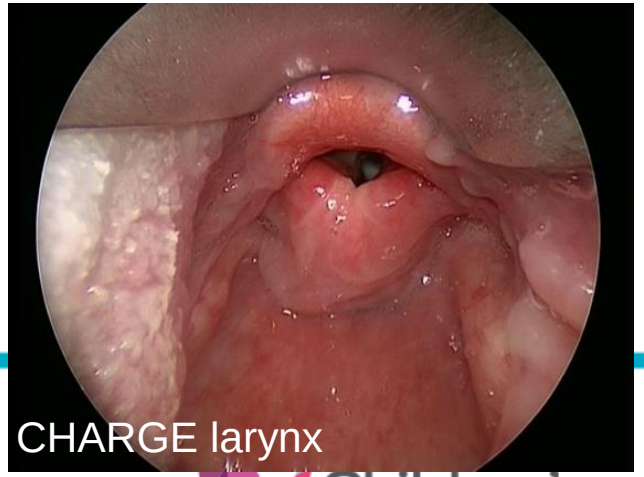
Structural Abnormalities

Larynx (Voicebox)

- Prominent, anteriorly placed arytenoids
- Shortened aryepiglottic folds
- Foreshortened appearing vocal folds
- Many with decreased sensation


Normal larynx


CHARGE larynx

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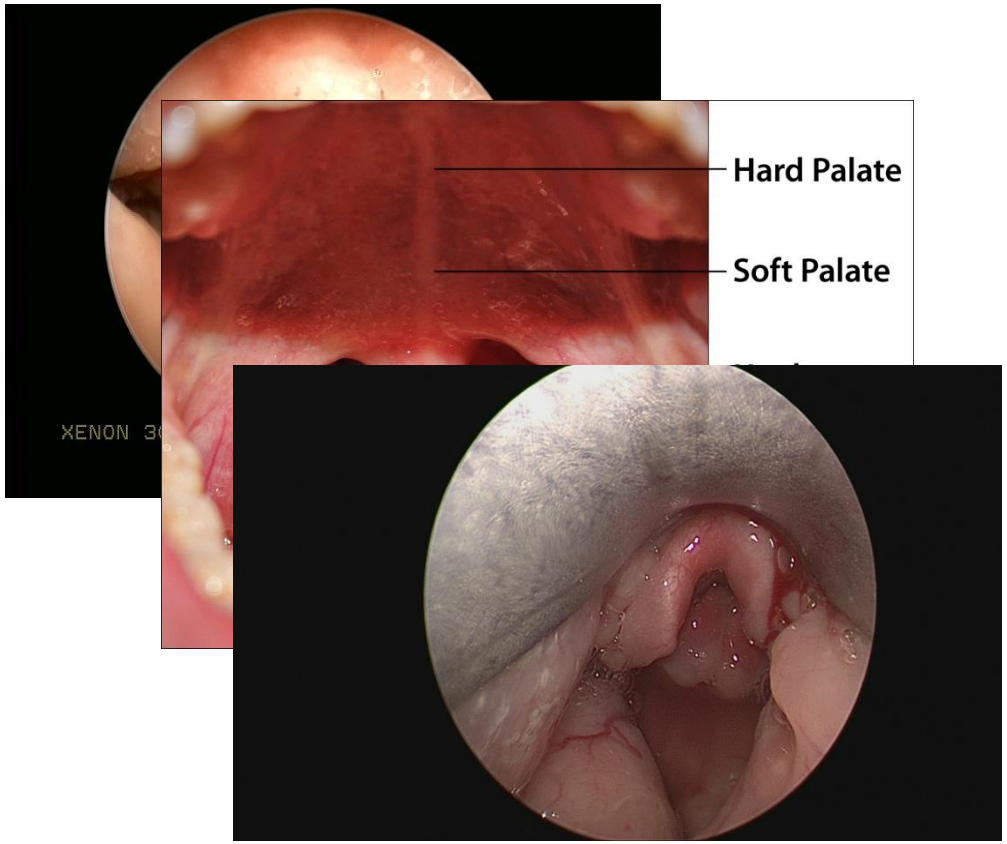
Upper Airway Obstruction

- Increased work of breathing
- May worsen risk for aspiration
- May require tracheotomy



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Other Structural Abnormalities

Congenital Heart Disease

- Present in 75-80%
- Severe impairment can impact oral feeding
- Vocal fold paralysis following cardiac surgery
- Additional risk for aspiration

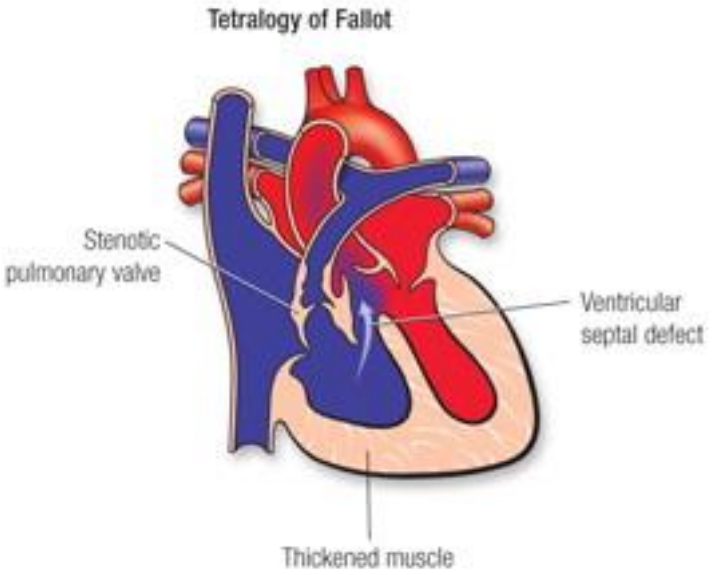
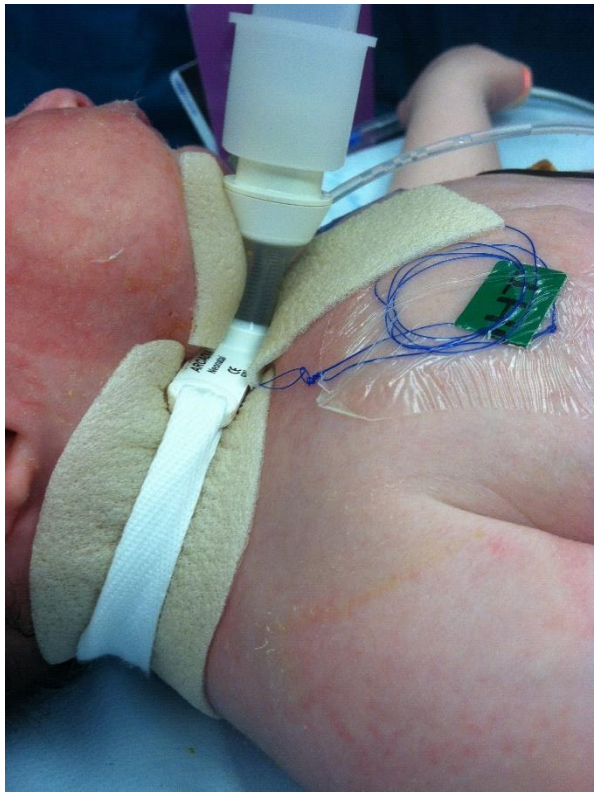


Image: heart.org

Tracheotomy-dependent Patients

- Swallowing dysfunction is commonly reported in adults and children with tracheotomy tubes
- 50-87% of adult patients demonstrate aspiration
- Not well studied in children
- Children with long-term tracheotomies who are NPO are at high risk for developing feeding issues



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Swallowing Dysfunction in Pediatric Trach Patients

- Multiple factors may contribute
- Tracheostomy tube itself
- Underlying condition
- Comorbidites:
  - Former premature infants
  - Neurologic deficits
  - Global developmental delay

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Role of Passy Muir Valve

- One way valve that allows an increase in subglottic pressure, assisting with speech and cough production
  - May improve swallowing
  - May decrease aspiration
  - May assist in management of secretions

Role of PMV in Dysphagia

- In adult patients, complete occlusion of tube has been shown to improve swallowing function
- Theory
  - PMV closes at the end of inspiration and prevents air leakage
  - Allows for generation of normal subglottic pressures during swallow
- Mixed evidence about the use of PMV
  - Some studies show decreased aspiration
  - One study showed 9 of 20 patients aspirated with and without PMV

Role of PMV in Dysphagia in Kids

- Only one study, 12 patients
- Mixed group
  - Age at trach ranged from 1 day to 9 years
  - Varied indications
- VSS performed with PMV and sham PMV
- Results–No significant difference
  - Thin liquids: 5 improved, 4 got worse, 3 had no change
  - Puree: 1 improve, 1 got worse

Ongkasuwan J et al. Laryngoscope 2014;124:1469-1474

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Behavioral Feeding Issues

Hudson, et al (2016)

- 20 parents of CHARGE patients interviewed about feeding behaviors (age 2-32 years, average 10 years)
- All reported difficulties chewing and most needed close supervision during meals
- 95% reported overstuffing
- 75% reported food packing in cheeks

These things can all increase aspiration risk

Swallowing Assessment

- Require multi-disciplinary evaluation
  - Speech language pathology
  - Otolaryngology
  - Pulmonary
  - PT/OT

Goals of Feeding Evaluation

- Determine safety of feeding
- Aspiration risk
- Compensatory strategies
- Diet Modification
- Therapy recommendations

Oral Motor Evaluation

- Oral sensory—Keep in mind cranial nerve issues
  - Hypersensitivity
    - Sensitive to touch, gagging, texture refusal, resistance to utensils
  - Hyposensitivity
    - Pocketing, inefficient bolus movement, limited OM movements
  - Impaired oral tactile discrimination
    - Difficulty differentiating between various textures simultaneously

Oral-Pharyngeal Skills

- Oral anatomy
- Oral motor patterns
  - Symmetry
  - Lip retraction, lip pursing hyposensitivity
  - Tongue protrusion, thrust, retraction or hypotonicity
  - Wide jaw excursions, phasic biting or limited range of motion
- Developmentally appropriate skills
  - Bottle feeding
  - Cup Drinking
  - Utensil Use
  - Transitioning to solids
  - Advancing textures

Instrumental Evaluations

- Most children with CHARGE will undergo at least one, likely multiple
- Obtain to answer specific questions
- Specific test chosen is dependent upon the question being asked
- Some patients need multiple tests
- Tests are often complimentary

Videofluoroscopic Swallowing Study: VSS

- aka “Modified Barium Swallow”
- Collaborative study - Radiologist/Speech Pathologist
- “Gold standard” for evaluation of dysphagia
- Determines if airway protection is deficient and **WHY** the problem exists

Pediatric FEES

- Otolaryngologist
  - Endoscopy
- Speech pathologist
  - Determines food textures to be tested
  - Feeds or assist with feeding
- Joint development of recommendations
- Added benefit of assessing vocal fold movement

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Swallowing Parameters

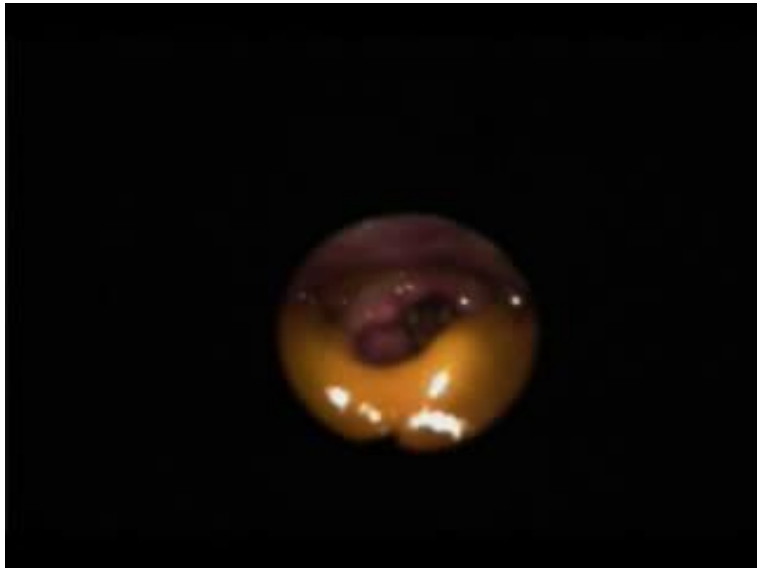
- **Secretion management**
- Swallowing Response Time
- Pharyngeal clearance
- Laryngeal Penetration
- **Aspiration**
- **Protective Reactions**
- Responses to compensatory strategies

In Trach Patients: “Dye” Testing

- Artificial food coloring added to liquids, foods (not enteral feeding bags), saliva
- In trach patients, can monitor presence of food color via suctioning
- If coloring is present when suctioning, infer aspiration
- Issues with reliability, validity
- Evidence questionable as to from above or below in some cases

Aspiration

- Material passing into the airway
- Can aspirate liquids, food, saliva or reflux
- Coughing/choking with feeds
- Can be silent (no cough response to material entering airway)



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Aspiration in CHARGE

- Aspiration observed in 60% of children on either/or:
  - Video Swallow Study (VSS)
  - Flexible Endoscopic Evaluation of Swallowing (FEES)
- Factors that contribute:
  - Cranial nerve dysfunction
  - Swallowing dysfunction

Saliva Production

- 1-1.5 Liters per day in adults
- Major Salivary Glands
  - Parotid
  - Submandibular
  - Sublingual
- Minor Salivary Glands
  - Small glands lining oral cavity

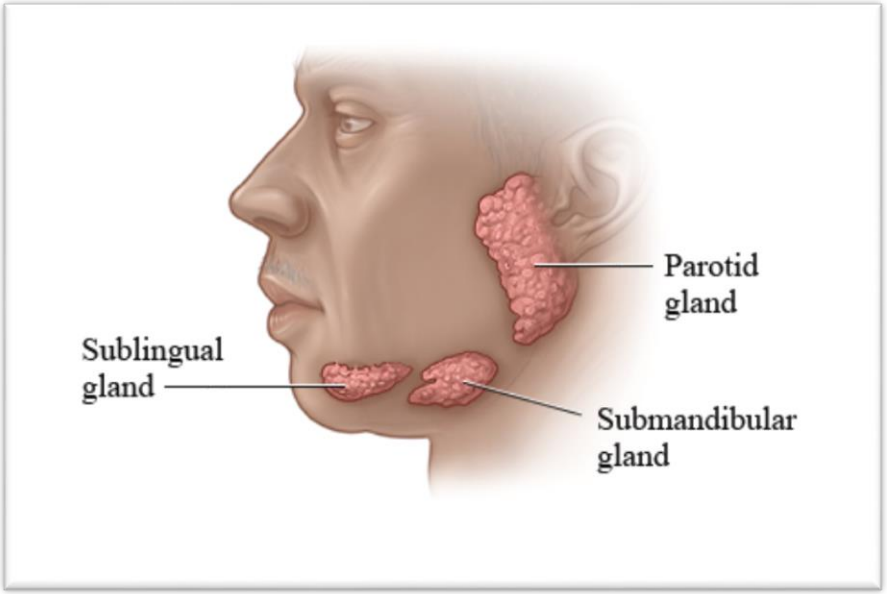


Photo credit: webmd.com

Function of Saliva

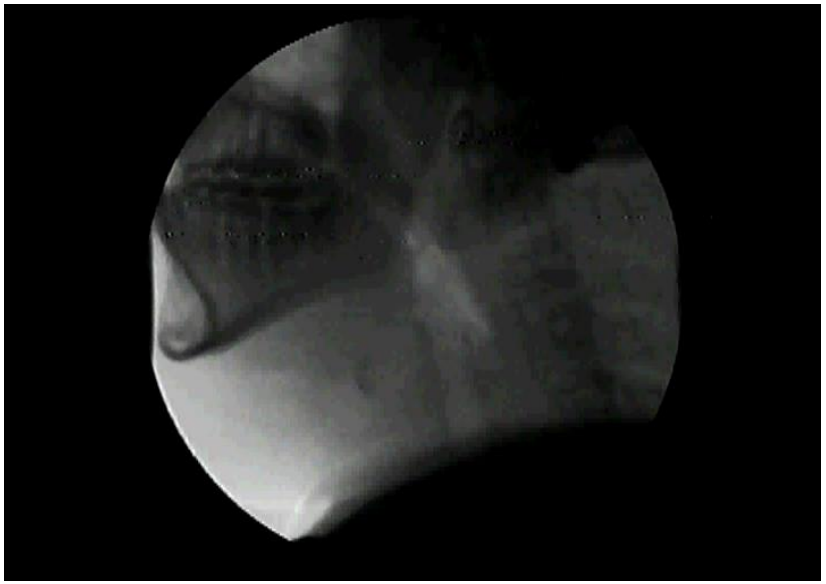
- Moisten oral mucosa
- Moisten and cool food
- Assist in dissolving food
- Buffers acid
- Enzymes in saliva initiate digestion
- Antibacterial properties

Causes of Drooling/Aspiration

- Poor oral motor control
- Inefficient swallow or decreased frequency of swallow
- Developmental delay
- Malocclusion or structural problem with mouth
- Mouth-breathing / upper airway obstruction
- Postural problem

NOT a problem of OVERPRODUCTION

Aspiration: VSS & FEES



VSS

FEES

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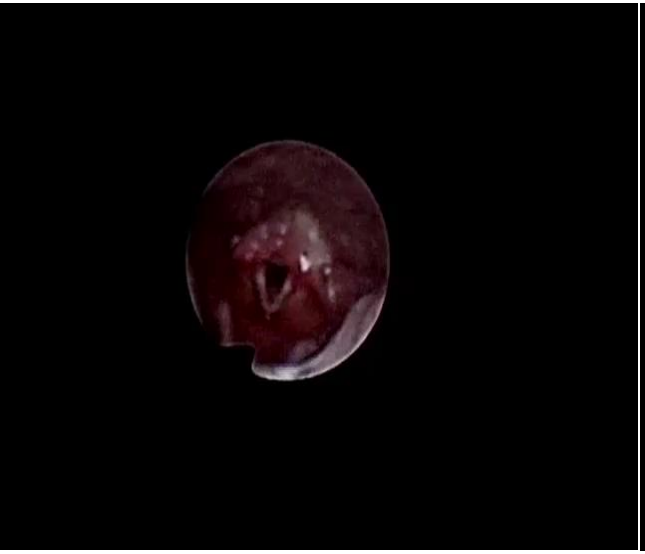
FEES in CHARGE

- Wide variety of impairment



Essentially normal

Moderate impairment

Severe impairment

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Aspiration: Why do we care?

- Frequent suctioning
- Noisy breathing/choking
- Recurrent pneumonia
- Oxygen requirement
- Long term damage to the lungs

Aspiration: Feeding Options

- Dependent on severity of aspiration
- Thickened feeds, modified swallowing techniques
- Nasogastric tube feeding
- Gastrostomy tube placement

Drooling/Aspiration:Treatment Options

- No Treatment
- Therapy
  - Oral motor therapy
  - Rehabilitative
- Medical Treatment
- Surgical Treatment

**We usually progress in a stepwise fashion

No Treatment

- Insignificant problem
- NO aspiration present
- Anticipated improvement
- Drooling is a low priority

Therapy Options

- Oral motor therapy
  - Verbal cueing
  - Tongue mobility
- Correct situational factors
  - Positional devices
  - Oral devices to improve / mimic lip competency
- Behavioral therapy
  - Instruction, prompting, positive reinforcement
  - Negative social reinforcement
  - Cueing techniques
  - Self-management procedures

Medications

- ~80% effective when first started

- Glycopyrrolate (Robinul)
- Transdermal hyoscine (Scopolamine)
- Botulinum toxin A (Botox ®)

Glycopyrrolate (Robinul)

- Effective in 95% of patients – initially
- Side effects lead to discontinuation in 20%
- Common side effects:
  - Dry mouth
  - Thick secretions
  - Urinary retention
  - Flushing

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Scopalamine Patch

- 24 – 72 hour delay in response
- Drowsiness is common
- Unable to focus / accommodate making reading difficult

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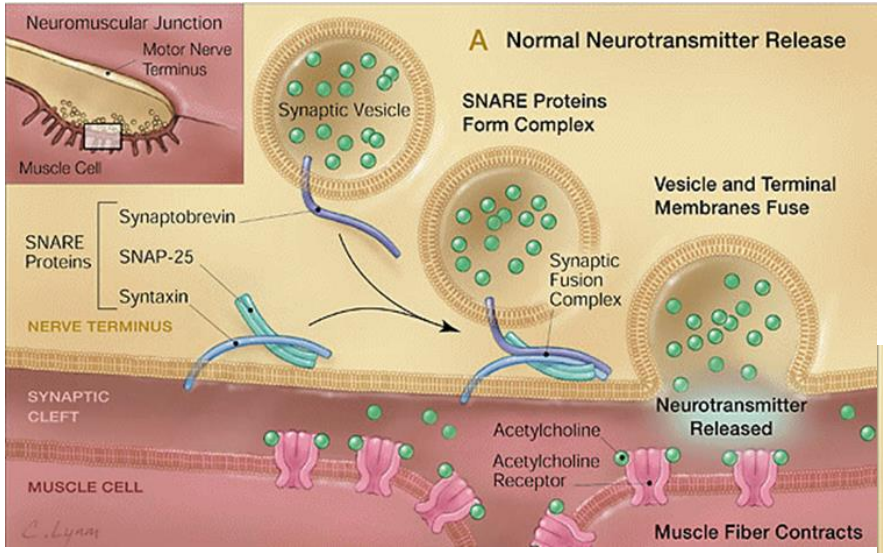
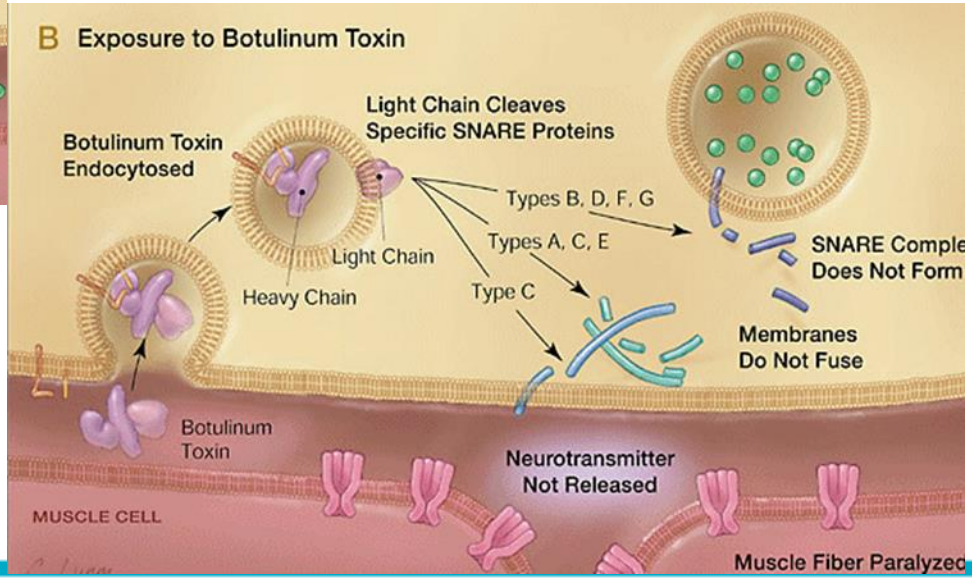
Botulinum A

- Injected directly into glands
  - Parotid
  - Submandibular
- Decreases saliva production by blocking release of neurotransmitters
- Usually lasts a few months

Botulinum A



Prevents release of neurotransmitter



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Botulinum A

- Use ultrasound to inject directly into glands

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Botulinum A Results

- Study of 97 children
- Effective in 62%
- Partially effective in 8%
- Not effective in 29%
- Duration of effect: average of 4 months (range <1 month to 18 months)

Botulinum A Complications

- Dysphagia most common (10%)
  - Aspiration Pneumonia
- Thickened secretions (1.5%)

Medical Treatment Complications

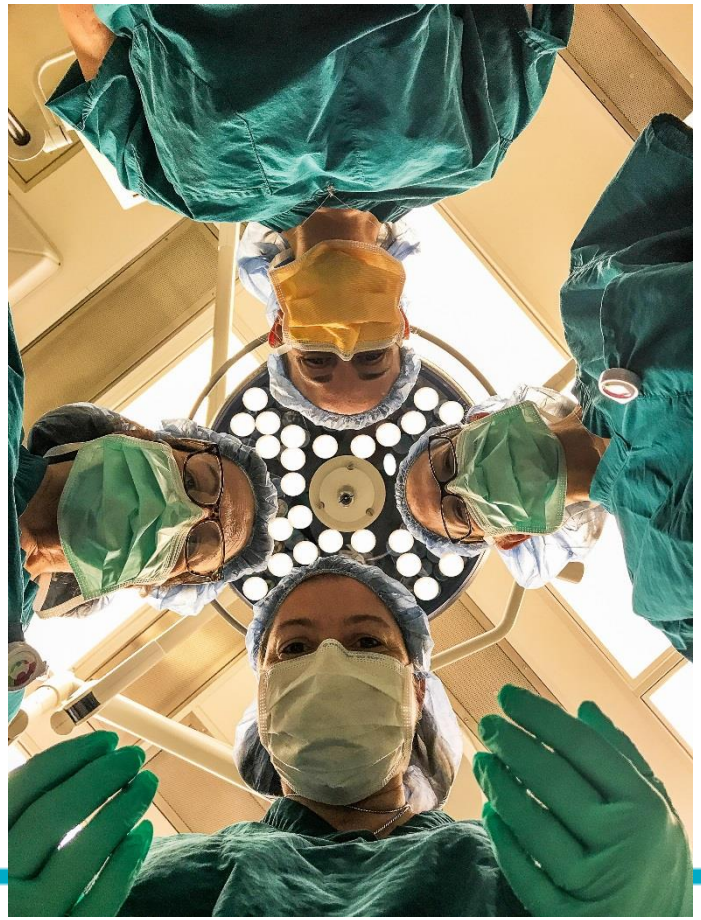
- Medical treatment options provide short term results
- Drugs become less effective with time and often need to increase dose of medication
- Increasing dose increases side effects

Medical Treatment Complications

- Glycopyrolate
  - Flushing, constipation, urinary retention, irritability, excessive dry mouth, behavioral changes
- Transdermal scopolamine
  - Reaction to adhesive
  - Pupil dilation secondary to touching the patch and then the eye
- Botulinum Toxin
  - General weakness, respiratory failure, death have been reported

Surgical Treatment

- Indications
  - Failure of medical treatments
  - Family or patient doesn't want to undergo/maintain medical treatment
  - Profuse drooling
  - Aspiration with pulmonary complications



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Surgical Options

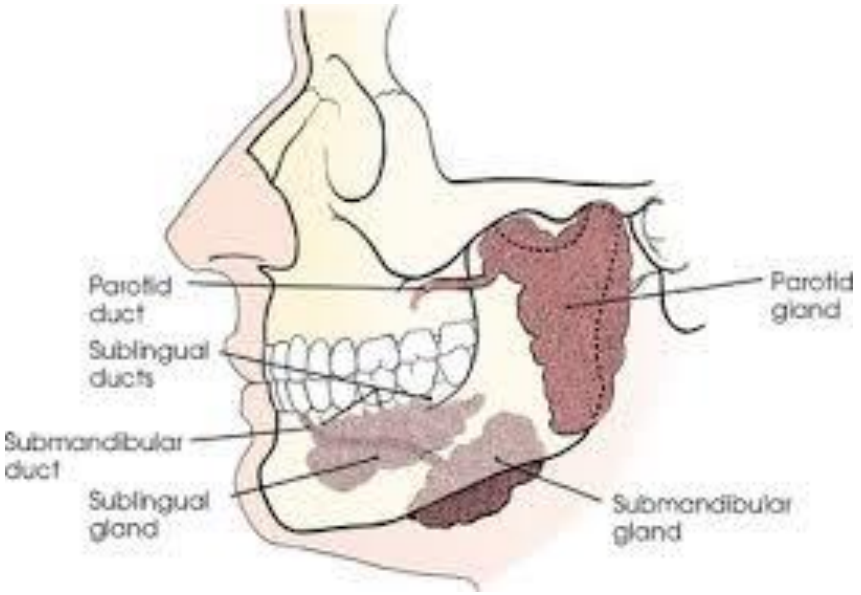
- Salivary duct relocation
  - Not appropriate if aspirating secretions
- Ligation of salivary ducts
  - Parotid and/or Submandibular
- Excision of glands
  - Submandibular
- Tympanic neurectomy
  - Decrease salivary flow
- Combination

Surgical Options—Success Rates

- Salivary duct relocation
  - 80% long term control
- Ligation of salivary ducts (4 ducts)
  - 30% long term satisfaction
- Tympanic neurectomy
  - 75% with good control

“Drool” Procedure

- Bilateral Parotid duct ligation and bilateral submandibular gland excision
- Common combination
- 87% successful
- Complications:
  - Injury to nerve that controls lower lip
  - Dry mouth (10%)



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Surgical Options—Complications

- Salivary duct relocation
  - Mucocele formation
  - Aspiration
- Ligation of salivary ducts (4 ducts)
  - Mucocele formation
- Tympanic neurectomy
  - Decreased taste sensation
  - Nerve regrowth/reconnection

Surgical Options Continued

- Tracheotomy
  - May be necessary in up to 30% of children with CHARGE (Roger 1999)
  - Does not prevent aspiration but makes it easier to suction and keep lungs clean
  - More likely if a child has upper airway obstruction or chronic lung disease

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Surgical Options Continued

- Laryngotracheal separation
  - Larynx (voicebox) separated from trachea (windpipe)
  - Only treatment that is 100% effective in preventing aspiration
  - Unable to talk
  - Usually only done when lungs extremely damaged due to aspiration

Summary

- Swallowing/feeding issues are very common in children with CHARGE
- Issues are multi-factorial and require multi-disciplinary management
- Aspiration is one of the biggest concerns and should be managed aggressively
- Management is dependent upon severity of dysfunction

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Questions?

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