Surgical Management of Advanced and Recurrent Subglottic Stenosis

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Grand Rounds Presentation
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Subglottic stenosis (SGS) is the narrowing of the airway below the true vocal folds.

- Congenital
- Acquired

Subglottis:

- Anterior commissure: 10mm
- Posterior commissure: 5mm

Normal lumen is 4.5 to 5.5 mm

- <4.0mm in full term infant is considered stenotic
- <3.5mm in premature infants is considered stenotic
Predisposing Factors

- Subglottis most susceptible to injury
  - Complete circular cartilage
  - Narrowest part in neonates
  - Lined by respiratory epithelium
    - Fragile and easily disrupted
Laryngeal Function

- Breathing
- Protect from aspiration
- Phonating
- Coughing
- Valsalva
- Maintaining PEEP
Anatomical Considerations

- The larynx is proportionally larger in children.
- The narrowest portion of the pediatric airway is the subglottis.
  - Adult is the glottis
- Positioning of Larynx
  - Pediatric $\rightarrow$ 2\textsuperscript{nd} cervical vertebrae
  - Adult $\rightarrow$ 6\textsuperscript{th} cervical vertebrae
- Normal laryngeal function is important to prevent aspiration as the pediatric epiglottis is more floppy and less functional.
Embryology

- Larynx develops from 4\textsuperscript{th} and 5\textsuperscript{th} arches
- Outgrowth of the primitive pharynx
- Laryngotracheal opening between arches
- Three masses form:
  - Hypobranchial eminence $\rightarrow$ Epiglottis
  - Paired arytenoid masses
- Recanalization occurs and failure leads to:
  - Atresia, stenosis, or web formation
- Arytenoid masses are separated by interarytenoid notch which creates a cleft if it is not obliterated
Poiseuille’s Equation

- Resistance = \( \frac{n \times L}{r^4} \)
- Therefore it is important to remember that as the airway narrows, breathing becomes more difficult.
- This effect is worse in children because they already have a smaller airway than adults.
Congenital SGS

- Secondary to inadequate recanalization of laryngeal lumen (3rd month gestation)
- Involved in 5% of SGS cases
- Subclassification:
  - Membranous
  - Cartilaginous
- Varying Degrees:
  - Complete atresia
  - Localized Stenosis
  - Webs
Glottic web
Membranous form

- Fibrous tissue hypertrophy
- Hyperplastic mucous glands
- Not an inflammatory cause
- Usually circumferential
- Often 2-3 mm below true vocal cords
- Can extend superiorly to involve the glottis
Cartilaginous Form

- More variable
- Most often involves cricoid cartilage
- Grows posteriorly from anterior cartilage as a sheet
- Leaves a small posterior opening
Localized Congenital SGS
Acquired SGS (95% of cases)

- Prolonged intubation
- Direct blunt trauma
- Fume/smoke inhalation
- Caustic Lye ingestion
- Chronic Infections
  - TB, Syphilis, leprosy, typhoid fever, etc.
- Chronic Inflammatory diseases
  - Sarcoid, Lupus, Wegner’s, GERD, RA
- Laryngeal Neoplasms
Intubation trauma

- Causes 90% of acquired SGS in children and neonates
- Incidence following intubation is 0.9-8.3%
  - 44% for low birthweight neonates and children with RDS
- Vast improvement from 60’s and 70’s where the incidence was 12-20%
- Produces pressure necrosis at the site of contact with the airway
  - The duration of intubation
  - The size of tube
  - The number of intubations
Intubation Trauma

- Pressure necrosis of cuff
  - Worse with high pressure low volume
  - Microcirculation ceases at 30 mm Hg
- Duration of Intubation (Whited 1985)
  - 2-5 days → 0-2% stenosis
  - 5-10 days → 4-5% stenosis
  - >10 days → 12-14% stenosis
Pathophysiology

Tissue Injury → Necrosis, Edema, and Ulceration → Mucociliary stasis

Secondary infection and perichondritis → Granulation tissue proliferation and scarring

Laryngeal dysfunction and increased susceptibility to injury → Airway, voice, and feeding abnormalities
Signs and Symptoms
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<tr>
<th>Question 1</th>
</tr>
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<tbody>
<tr>
<td><strong>Subglottic</strong></td>
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<tr>
<td><strong>Glottic</strong></td>
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<td><strong>Supraglottic</strong></td>
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Common Signs and Symptoms

1) Airway
   - Biphasic stridor, dyspnea, air hunger, retractions

2) Voice
   - Abnormal cry, hoarseness, aphonia

3) Feeding
   - Dysphagia, recurrent aspiration pneumonia
Question 1

- **Subglottic**
  - hoarse/aphonic voice, inspiratory/biphasic stridor, ± cough

- **Glottic**
  - muffled/throaty voice, inspiratory stridor, feeding problems, no cough

- **Supraglottic**
  - hoarse/husky voice, biphasic stridor, barking cough
Initial Presentation

- **Congenital SGS**
  - At birth if moderate or severe

- **Acquired SGS**
  - Usually within 2-4 weeks of trauma/insult

*Either type in its mild form can be asymptomatic and present after a subsequent insult to the airway*
Workup
History and Physical Exam

- Birth history (Prematurity)
- Intubation history
- Feeding/Voice/Breathing difficulties?
- Reflux?
- Infections?
- Autoimmune diseases?
- Other systemic symptoms?
Flexible Nasopharyngolaryngoscopy

- **Nose/Nasopharynx**
  - Pyriform aperture stenosis
  - Choanal atresia

- **Supraglottis**
  - Structure abnormalities
  - Laryngomalacia

- **Glottis**
  - VC mobility
  - Clefts/webs/masses

- **Immediate subglottis**
Radiologic Evaluation

- **Plain film**
  - Quick and cost effective in children
- **CT**
  - Very specific for site and length of involvement
- **MRI**
  - Good for surgical planning
- **Ultrasound**
  - Excellent for quick assessment of the levels and diameter of airway
Soft Tissue Film
Workup/Initial Evaluation

- Carretta et al. (2006)
  - Compared preoperative CT findings with intraoperative rigid endoscopy findings
  - Rigid Endoscopy most reliable diagnostic entity
- Rigid Endoscope is the gold standard
  - Allows better visualization of the vocal cords
  - Better assessment of stenotic levels

*Must be performed delicately to prevent mucosal irritation and stenosis exacerbation*
Rigid Endoscopy

- **Important intraoperative findings**
  - Outer diameter of largest ET tube or bronchoscope that can be passed
  - Location sites and length of stenosis
  - Other airway anomalies
  - Reflux changes
**Additional workup**

- **Labs**
  - PPD, RPR, C-ANCA, ANA, etc
- **24-hour PH monitoring (dual probe)**
- **EGD with biopsy**
  - If esophagitis is suspected
- **Modified Barium Swallow**
- **Functional Endoscopic Evaluation of Swallowing**
  - Test airway sensation especially if child has aversion to food.
- **PFT’s**
  - Used to compare Pre-op and Post-op pulmonary function
GERD and SGS

- **Koufman et al. (1991)**
  - 73% of 32 patients with LTS had abnormal lower pH probe results
  - 67% had abnormal upper pH probe results

- **Walner et al. (1998)**
  - 74 pediatric patients with SGS had 3 times greater incidence of GER than the general pediatric population

*Therefore all patients should be treated with PPI therapy even if they are not symptomatic to prevent recurrence (Burton, 1997)*
Staging
Question 2

- Why are these numbers important?
  - 0
  - 51
  - 71
  - 100
Cotton-Meyer Grading of SGS

- I – 0-50% narrowing
- II – 51-70% narrowing
- III – 71-99% narrowing
- IV – Complete obstruction with no lumen
- Cotton-Myer grading system for subglottic stenosis

<table>
<thead>
<tr>
<th>Classification</th>
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<tr>
<td>Grade I</td>
<td>No Obstruction</td>
<td>50% Obstruction</td>
</tr>
<tr>
<td>Grade II</td>
<td>51% Obstruction</td>
<td>70% Obstruction</td>
</tr>
<tr>
<td>Grade III</td>
<td>71% Obstruction</td>
<td>99% Obstruction</td>
</tr>
<tr>
<td>Grade IV</td>
<td>No Detectable Lumen</td>
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Laryngeal Stenosis Grading

- **Grade I**
  - Less than 70%
- **Grade II**
  - 70-90%
- **Grade III**
  - More than 90% with identifiable lumen
- **Grade IV**
  - Complete obstruction...No lumen
Grading Systems for SGS

- Lano (1998)
  - Based on subsites involved
  - Does not take into account length of stenosis or lumen diameter
    - Stage I – one subsite involved
    - Stage II – two subsites involved
    - Stage III – three subsites involved
Grading Systems for SGS

- McCaffrey (1992)
  - Based on subsites (trachea, subglottis, glottis) involved and length of stenosis
  - Does not include lumen diameter
- Grade I: Confined to the subglottis or trachea and are less than 1cm long
- Grade II: Isolated to the subglottis and greater than 1cm long
- Grade III: Sublottic and tracheal lesions not involving the glottis
- Grade IV: Glottic involvement
Management
Goals of Management

- To produce:
  1. Adequate airway
  2. Competent Larynx
  3. Acceptable voice

*Ultimately the goal is to treat the stenotic segment while preserving native normal segments*
History of SGS Treatment

- Early 1900’s most SGS was secondary to chronic infection and was treated primarily with tracheotomy
- 1950’s and 60’s – increase incidence intubation practices with high tracheotomy mortality
- 1980’s Cotton described Anterior Cricoid Split
- The last 15-20 years has brought about SS-LTR which allows removal of the tracheotomy tube shortly after surgical repair
- Endoscopic techniques have also emerged as viable options for initial treatment
Management Options

- Tracheotomy
- Endoscopic Management
  - Dilation
  - Laser excision of stenotic areas
- Anterior Cricoid Split
- LTR (single or two stage)
- CTR
Tracheotomy

- Maintains adequate airway
- Use smallest tube that permits ventilation
- Allows air leakage to prevent pressure injury and preserve phonatory function.
- Usually just temporary
  - If cannot decannulate by 2y/o consider endoscopic or open repair

*Must keep in mind suprastomal granulation tissue when considering decannulation*
Endoscopic Repair

- **Dilation**
  - Useful early in disease process
  - Local steroid injections can help reduce scarring
- **Scar excision with laser**
  - Become increasingly popular
  - Minimal damage to normal surrounding tissue
  - Avoids bleeding, edema, and the need for a tracheotomy in some cases
- **Useful in Grade I or II stenosis but often requires multiple procedures**
Outcomes with Endoscopic Repair

- Herrington et al. (2006) showed that 70% of patients undergoing dilation needed repeated procedures.
- Recommend usefulness in grade I or II stenosis before considering open interventions.
Anterior Cricoid Split

- 1980 – described by Cotton
- Alternative for Tracheotomy
- Procedure
  - Splits cricoid and first 2 tracheal rings
  - Neck is closed with ET tube in place to act as a stent for healing.
  - Intubated, sedated, paralyzed in ICU for 7-14 days
- Patients must have adequate pulmonary function to permit decannulation
- Best indicated for mild anterior narrowing
Anterior Cricoid Split
Strict Criteria

- Extubation failure ≥2 occasions
- Weight >1500g
- No assisted ventilation 10 days prior
- Oxygen requirement <30%
- No CHF for >1 month
- No acute URI
- No antihypertensive meds >10 days
External Expansion Surgery

- Reserved for grade III and IV stenosis, or refractory grade II
- Combines laryngeal and cricoid split with cartilage grafts and stenting
- Success rates are greater than 90%
  - Success defined by decannulation
- Repair at youngest age possible:
  - Improved speech and language development
  - Decreased tracheotomy morbidity/mortality
Costal Cartilage Grafts

- Abundant
- Can obtain any size necessary
- Generally use the 5\textsuperscript{th} rib
- Stenting required for several days as suturing does not ensure that the graft stays in its place.
Approach to obtaining graft

Figure 11-15. Costal cartilage graft donor site: right lower ribs. The deep layer of perichondrium remains in the patient.
Other grafts

- Auricular cartilage
- Thyroid alar cartilage
- Hyoid bone
Anterior laryngofissure with graft

- Good for:
  - Anterior stenosis
  - Anterior wall collapse
- Perichondrium of the anterior graft is placed on the lumen side
  - Re-epithelialization
  - Barrier to infection
- Large external flange to prevent prolapse of graft into the airway
Anterior Grafts: Modified boat shape

FIGURE 87-4. Schematic drawing of an anterior costal cartilage graft. The technique allows flanges at each end of the graft to lie over the cut margin of the thyroid cartilage superiority and the tracheal wall inferiorly (cross-section A). Smaller notches along the middle edges of the graft remain outside the border of the laryngotracheal fissure (cross-section B).
Placement of anterior graft
Laryngofissure with posterior cricoid division +/- grafting

- **Indications:**
  - Posterior subglottic or glottic stenosis
  - Circumferential stenosis
  - Cricoid deformity

- **Key points**
  - Avoid complete laryngofissure to avoid damage to anterior commissure
  - Knots buried to keep them extraluminal
  - Patients often receive stenting 3-6 months
Posterior Grafts: Regular boat shape
Single-staged Laryngotratheal Reconstruction (SS-LTR)

- Allows for shorter stenting period
- Anterior graft, posterior graft, or both
- ET tube initially to support the graft
  - 2-4 days if Anterior graft only
  - 7 days if Posterior graft is used as well
- Best results if patient >4Kg and >30wks
Two-Staged LTR

- The main difference is that a more permanent stent is used to maintain the airway while the graft heals
  - Montgomery T-tubes (silastic)
  - Aboulker Stents (teflon)
- Stents can be left for months

*Considered to be inert and prevent tissue injury
Stents

- Counteract scar contractures and provide a scaffold for the airway.
- Can also hold grafts in place and increase success rates.
- Types of stents
  - T-tubes most common in adults but these can become blocked in children
  - Aboulker stents more commonly used in children for two stage procedures
  - ET tubes act as stents in the immediate perioperative period for SS-LTR
- Patients must be evaluated for granulation tissue prior to stent removal with fiberoptic exam
Montgomery T-tube Stent
Aboulker Stent
Aboulker Stent with wired-in tracheostomy tube
Granulation Tissue Formation

- Nouraei et al. (2006) studied stent colonization and its correlation with granulation tissue formation.
- Colonization with S. aureus and P. aeruginosa statistically showed increased rates of granulation tissue.
- What didn’t play a role?
  - Duration of stent placement
  - Polymicrobial colonization (oral flora)
- Currently recommend antibiotics for 1 week post-op with coverage for Staph and Pseudomonas.
SS-LTR vs Two-Stage LTR

- Saunders et al. (1999)
  - Patients undergoing Two-Stage LTR had
    - More severe stenosis (Grade 2.56)
    - Previous laryngeal surgery
  - SS-LTR patients
    - Less severe stenosis (Grade 2.14)
    - Fewer post reconstruction procedures
    - Higher decannulation rate
- Single stage procedure significantly better for
  - Number of postoperative procedures (p=0.006)
  - Decannulation rate (p=0.03)
SS-LTR vs Two-Stage LTR

- Smith et al. (2010)
  - 71 patients
    - 22 SS-LTR (average grade 2.1)
    - 62 Two-Stage LTR (average grade 2.9)
  - Operation specific decannulation rate was better for SS-LTR (91% vs 68%) however overall decannulation rate was not significantly different (100% vs 93%)
Decannulation Rates for LTR

- Younis et al. (2004)
  - Overall success rate of 86%
  - Anterior graft (100%)
  - Anterior and posterior graft (83%)
  - Revision cases (70%)

- Koltai et al. (2006)
  - 89% successful decannulation
Cricotracheal Resection (CTR)
Cricotracheal Resection (CTR)

- 1953 – Conley describes 1st CTR in adults
- 1978 – Savary described 1st CTR in children
- Best results in patients with isolated tracheal stenosis
- More difficult approach with subglottic stenosis
- Well tolerated in patients with grade III or IV stenosis
- CTR > LTR in grade IV stenosis
Fig 2. Anterior-posterior and lateral views of cricotracheal resection procedure. A) Midline vertical incision through cricoid cartilage and upper stenotic tracheal rings. Ultimate lines of transection are indicated by dashed lines. RLN — recurrent laryngeal nerve. B) Resection of cricoid cartilage anterior to cricothyroid joints. Posterior cricoid plate is exposed. C) Dissection of upper stenotic trachea away from esophagus. D) Distal trachea with membranous pedicled flap. Stenotic segment has been resected. E) Anterior thyrotracheal and lateral cricotracheal anastomosis.
CTR Failure Risk Factors: White and colleagues (2005)

- 96% of patients had grade III or IV stenosis
- 94% overall decannulation rate
- Statistically Significant Odds Ratio:
  - Vocal cord paralysis post-op decreased decannulation (OR 5.2)
- Statistically Insignificant Odds Ratio:
  - Down syndrome
  - Trach tube at time of CTR
  - Use of chin to chest sutures
- Eosinophilic Esophagitis was significant but not enough patients to analyze
  - They suggest EGD with biopsy preoperatively for all patients including PPI therapy 6 months post-op.
CTR Complications: Rutter and colleagues (2001)

- Described the following complications:
  - Anastomotic webbing
    - Almost all cases and usually asymptomatic
  - Arytenoid prolapse (45%)
    - 60% asymptomatic
    - 40% required partial laser arytenoidectomy
  - Restenosis (20%)
    - 11% were still trach dependent
  - Postoperative infection (5%)
  - Recurrent laryngeal nerve palsy (5%)
  - Anastomotic dehiscence (0%)
    - although previously has been reported
Outcomes of breathing, voice, and swallowing

- Jacquet and colleagues (2005) studied post-CTR outcomes

- Airway
  - 95% of patients had no exertional dyspnea

- Voice
  - 21% had no vocal abnormalities
  - 49% had mild dysphonia
  - Overall, 70% showed post-operative improvement

- Swallowing
  - 89% of patients with pre-operative swallowing problems showed post-operative improvement.
Ikonomidis et al (2010)

- Why push earlier surgery?
  - Improved quality of life
  - Decreased financial burden on families
  - Improved speech and language development
  - No chance of accidental death from plugging which was reported as 2% in this series
Ikonomidis et al (2010)

- Retrospective review of CTR in children <10kg
- Less than 10kg (36pts)
  - Average weight 8.8kg smallest was 4.4kg
  - <1y/o (11pts), 1-2y/o (18pts), 2-3 y/o (7pts)
  - Single stage in 27 patients with 100% decannulation
  - Two stage in 9 patients with 92% decannulation
- No significant difference between this group and the comparison group weighing >10kg (65pts) after cox regression analysis
Postoperative Care (Gupta et al. 2010)

- Requires ICU with specialized staff
- Nasotracheal Intubation 7-14 days
- Sedation and paralysis
- Steroids
  - 12 hours before and for 5 days after decannulation
- Leak test prior to extubation
- Precedex during tracheal extubation
- Antibiotics
  - 2 weeks if no stenting
  - Months if stenting is performed
- All get anti-reflux medications
- Enteral feeding
- Chest physiotherapy and log rolling
- High index of suspicion for nosocomial infections
Recommendations

- Tracheotomy is #1 option to acutely manage the airway.
  Oropharyngeal phase is voluntary; Esophageal Phase is not voluntary.
- Grade I and II Stenosis:
  - Endoscopic Repair
- Grade III or refractory cases:
  - LTR with anterior and/or posterior rib grafts
- Grade IV:
  - CTR is the treatment of choice
  - Earlier treatment is better to preserve or encourage normal functional development.
Summary

- Subglottic stenosis is a common problem in patients with prolonged intubation.
- Each patient needs an appropriate workup including history, physical, and laryngoscopic examination.
- Management is driven by grading.
- Always remember to maximize airway, voice, and swallowing.
# References