Surgical Management of Pediatric Subglottic Stenosis

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Overview

- Pediatric subglottic stenosis
  - Patient presentation and work-up
  - Medical management
  - Surgical intervention
Stridor

- A harsh, high pitched musical sound that results from turbulent airflow through the upper airway

- Etiology may range from mild illness to severe, life-threatening situation
Stridor Etiology

- Congenital
- Inflammation
- Trauma
- Foreign bodies
Stridor Presentation

- Variable age of onset
- Patient typically presents with sudden onset of symptoms
- Acquired stridor (inflammation, trauma, foreign bodies) is more likely than congenital stridor to require airway intervention
Congenital Stridor

- Eighty-five percent of children under 2.5 years presenting with stridor have a congenital etiology
- Often not present at birth
- Typically presents prior to four months of age
Assessing Stridor

- Determination of respiratory phase in which sound is noted
  - Inspiratory
  - Biphasic
  - Expiratory
Inspiratory Stridor

- Result of supraglottic obstruction
- High-pitched
Biphasic Stridor

- Result of extrathoracic tracheal obstruction including
  - Glottis
  - Subglottis
- Intermediate pitch
Expiratory Stridor

- Result of intrathoracic tracheal obstruction
- Associated with prolonged expiratory wheezing
Congenital subglottic stenosis

- Third most common cause of stridor in the neonate.
- Involves narrowing of the subglottic lumen in the absence of trauma.
- Full term neonate with lumen of less than 4 mm, or 3 mm in premature infant.
Subglottic airway

- The subglottic airway is the narrowest area of the newborn’s airway since it is a complete, nonpliable, nonexpandable ring.
Subglottic stenosis

- Caused by failure of the trachea to recannalize during embryogenesis.
- Approximately 5% of children will require smaller size ET tube than expected for age.
- Children with Down syndrome have high incidence of congenital airway narrowing.
Subtypes

- **Membranous** – usually circumferential, soft and dilatable.
  - Submucosal fibrosis
  - Submucosal gland hyperplasia
  - Granulation tissue

- **Cartilaginous** – has a more variable appearance.
  - Normal shaped cricoid with decreased lumen
  - Abnormally shaped cricoid with lateral shelves
  - Elliptical shape
Patient Presentation

- If severe, will have respiratory distress at birth.
- In milder cases, may present in first few weeks of life.
- Most become symptomatic by three months of age due to increased activity and ventilatory requirements.
Symptoms of upper airway obstruction predominate.

Inspiratory stridor progresses to biphasic as obstruction worsens.

Exacerbated by URI, and these children tend to have recurrent croup.
Office evaluation

- Onset, duration, severity, feeding abnormalities, failure to thrive, recurrent URI/croup
- Positional effects on stridor
- Voice quality
- Symptoms of reflux
- Complete head and neck exam
- Flexible laryngoscopy
Diagnosis

- Differential
  - Congenital
    - Laryngeomalacia
    - Tracheomalacia
    - VC paralysis
    - Cysts
    - Clefts
    - Vascular compression
    - Mass
Differential

Infection/Inflammation
- Croup
- GER
- Tracheitis

Neoplastic
- Subglottic hemangioma
- Recurrent respiratory papillomas

Foreign body
Flexible Laryngoscopy

- Best performed with
  - Unanesthetized child
  - Upright position
  - 1.9mm laryngoscope

- Scope should be passed through both nasal passages

- Evaluate vocal cord mobility
Definitive diagnosis

- Rigid endoscopy
- Imaging studies usually not necessary.
  - Neck films
  - Fluoroscopy
  - CT/MRI
Rigid endoscopy

- Evaluate the subglottis and glottis for fixation, scarring, granulation, edema, paralysis or paresis, and other abnormalities. Evaluate the distance and caliber of the stenosis. Apply the Myers and Cotton staging system only to circumferential SGS. Glottic stenosis and SGS often coexist and must be considered when reconstruction is planned.
Grading Systems for SGS

- Cotton-Myer (1994)
- McCaffrey (1992)
Grading Systems for SGS

- Cotton-Myer
  - Based on relative reduction of subglottic cross-sectional area
  - Good for mature, firm, circumferential lesions
  - Does not take into account extension to other subsites or length of stenosis
Table 2. *The Myer-Cotton Subglottic Stenosis Grading Scale*

<table>
<thead>
<tr>
<th>Grade</th>
<th>From</th>
<th>To</th>
<th>Examples</th>
</tr>
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<tbody>
<tr>
<td>Grade I</td>
<td>No Obstruction</td>
<td>50% Obstruction</td>
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<tr>
<td>Grade II</td>
<td>51% Obstruction</td>
<td>70% Obstruction</td>
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<tr>
<td>Grade III</td>
<td>71% Obstruction</td>
<td>99% Obstruction</td>
<td></td>
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<tr>
<td>Grade IV</td>
<td>No Detectable Lumen</td>
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The Myer-Cotton subglottic stenosis Grading Scale was developed at Cincinnati Children’s Hospital Medical Center. This scale helps group patients of similar severity of disease and is used to standardize the measuring of subglottic stenosis for discussion among those caring for these patients.
ET tubes of various sizes are placed sequentially.

Leak test performed

When 10-25 cm H$_2$O leak pressure achieved, this is patient’s tube size.

Compare to normal values for age.
The McCaffrey system classifies laryngotracheal stenosis based on the subsites involved and the length of the stenosis. Four stages are described: **stage I** lesions are confined to the subglottis or trachea and are less than 1 cm long, **stage II** lesions are isolated to the subglottis and are greater than 1 cm long, **stage III** are subglottic/tracheal lesions not involving the glottis, and **stage IV** lesions involve the glottis.
Grading Systems for SGS

- McCaffrey
Management of SGS

- Medical
- Observation
- Tracheostomy
- Airway expansion procedure
Management of SGS

- **Medical**
  - Diagnosis and treatment of GER
  - Pediatric – consultation with primary physician and specialists (pulmonary, GI, cardiology etc.)

- **Adult**
  - Assess general medical status
  - Consultation with PCP and specialists
  - Optimize cardiac and pulmonary function
  - Control diabetes
  - Discontinue steroid use if possible before LTR
Management of SGS

Observation

- Reasonable in mild cases, esp. congenital SGS (Cotton-Myer grade I and mild grade II)
  - If no retractions, feeding difficulties, or episodes of croup requiring hospitalization
  - Follow growth curves
  - Repeat endoscopy q 3-6 mo
Management of SGS

- **Tracheostomy**
  - Often the initial step in treatment of pediatric acquired SGS
  - May be avoided in patients with congenital SGS
  - Allows time for the infant to mature
    - Lungs – BPD
    - Wt. – 10 kg (Cotton)
  - 2%-5% mortality in children
    - Accidental decannulation and plugging
Grade I stenosis

- Usually will grow out of stenosis.
- Treatment is medical
- May have recurrent croup
- If surgery needed, may try endoscopic, dilation, or laser (CO2 or KTP)
Grade II

- Surgery is needed secondary to respiratory distress.
- May try endoscopic, dilation, or laser.
- May require open procedure
High Grade

- Refers to grade III or IV lesions
  - Laryngotracheal reconstruction
    - Anterior
    - Anterior and posterior
    - Anterior, posterior, and lateral
  - Partial cricotracheal resection
Preoperative planning

- Treat GER/EER before attempting reconstruction.
- Assess full extent of stenosis.
- Order CT scan with 3D reformats if total length of stenosis remains undetermined after rigid bronchoscopy.
- Treat any respiratory infections with antibiotics, and steroids
Evaluation for reflux

- Signs of extraesophageal reflux are noted, and include post-cricoid edema, ventricular effacement, and follicular bronchitis.
- BAL for lipid-laden macrophages
Intervention

- Goal of intervention is
  - to have an adequate airway to allow for normal activity without the need for tracheostomy
  - Single stage procedure, or two stage procedure with minimal postoperative morbidity, and minimal hospital stay. (Cable et al)
Cotton’s Stages of Reconstruction

- Stage 1 – complete evaluation of the airway
- Stage 2 – expansion of the subglottic lumen with preservation of function
- Stage 3 – stabilization of the expanded lumen framework (grafts and/or stents)
- Stage 4 – healing
- Stage 5 - decannulation
Surgery for SGS

I. Endoscopic
   - Dilation
   - Laser

II. Open procedure
   - Expansion procedure (with trach and stent or SS-LTR)
     - Laryngotracheoplasty
     - Laryngotracheal reconstruction
Management of SGS

How do you decide which procedure to perform

- Status of the patient
  - Any contraindications
    - Absolute
      - Tracheotomy dependent (aspiration, severe BPD)
      - Severe GER refractive to surgical and medical therapy
    - Relative
      - Diabetes
      - Steroid use
      - Cardiac, renal or pulmonary disease
Management of SGS

- Endoscopic
  - Dilation
    - Practiced frequently before advent of LTR
    - Requires multiple repeat procedures
    - Low success rate but an option for patients who cannot undergo LTR
Management of SGS

- Endoscopic
  - Laser
    - 66-80% success rate for Cotton-Myer grade I and II stenoses
    - Factors associated with failure
      - Previous attempts
      - Circumferential scarring
      - Loss of cartilage support
      - Exposure of cartilage
      - Arytenoid fixation
      - Combined laryngotracheal stenosis with vertical length >1cm
Laser excision of subglottic web
Laser excision of subglottic web
Management of SGS

- Grade III and IV stenoses require an open procedure
Anterior cricoid split

- Patient weight > 1500 grams
- Failure to extubate in identified SGS
- Oxygen requirement < 30%
- No active respiratory infection
- Good pulmonary and cardiac function.
ACS
ACS

- Remain intubated 7-10 days
- Ab and antireflux meds while intubated.
- Complications include reintubation, pneumothorax, pneumomediastinum, subcutaneous emphysema, wound infection, and persistent SGS.
- Success rate of 58-100%
Single Stage LTR

- Surgical correction with short period of stenting.
- Two stage procedure still necessary for patients with poor pulmonary reserve, or multilevel stenosis.
- Grade II and selected grade III SGS.
SSLTR

- Same approach as for ACS
- Remove ET tube when air leak at 20 cm H₂O.
Gustafson et al. Retrospective chart review at tertiary care hospital.

- 200 pediatric patients, 96% decannulation rate.
- 29% required reintubation, 15% needed trach
- 4% remained trach dependent
- Anterior/posterior vs. anterior or posterior, higher rates of reintubation
- 70% Grade I/II
- EBM C
Gustafson et al

- Age greater than four, less complications after extubation and less need for sedation. (48 hours)
- Increased duration of stenting did not affect outcome. Follow leak pressure. 20 cm H₂O
- Moderate to severe tracheomalacia may be contraindication
Complications

- Younis et al. Retrospective chart review. 46 patients underwent A/P SSLTR. 35 Grade III/VI.
- 83% decannulation rate
- EBM C

<table>
<thead>
<tr>
<th>Table 4. Complications not related to age or type of stenosis</th>
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<tbody>
<tr>
<td>Complication</td>
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<tr>
<td>Reintubation</td>
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<tr>
<td>Failure</td>
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<tr>
<td>Anterior collapse</td>
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<tr>
<td>Pneumonia</td>
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<tr>
<td>Subcutaneous emphysema</td>
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<tr>
<td>Bleeding, hematoma</td>
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<tr>
<td>Graft failure (resorption)</td>
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<tr>
<td>Atelectasis</td>
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<tr>
<td>Withdrawal</td>
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<td>Poor voice</td>
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### Table 6. Tips for success and causes of failure of posterior graft single-stage laryngotracheal reconstruction

<table>
<thead>
<tr>
<th>Factors affecting success</th>
<th>Factors affecting failure</th>
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<tbody>
<tr>
<td>Preoperative flexible and rigid endoscopy to identify all airway problems</td>
<td>Graft-related problems</td>
</tr>
<tr>
<td>Surgical experience</td>
<td>Oversized graft</td>
</tr>
<tr>
<td>At least 3 years of SSLTR and/or 10 cases</td>
<td>Intraluminal sutures</td>
</tr>
<tr>
<td>Control of comorbidities</td>
<td>Demuded perichondrium</td>
</tr>
<tr>
<td>Gastroesophageal reflux</td>
<td>Instability</td>
</tr>
<tr>
<td>Pulmonary disorders</td>
<td>Resorption</td>
</tr>
<tr>
<td>Cardiac disease</td>
<td>Stent-related problems</td>
</tr>
<tr>
<td>Neurologic deficits</td>
<td>Wrong size: endotracheal tube mismatched to patient</td>
</tr>
<tr>
<td>Metabolic derangements</td>
<td>Inadequate duration: 14 to 21 days needed</td>
</tr>
<tr>
<td>Attention to procedural detail</td>
<td>Gastroesophageal reflux</td>
</tr>
<tr>
<td>Graft perichondrium toward the lumen</td>
<td>Poor muscle tone or degeneration</td>
</tr>
<tr>
<td>Graft bevel toward the lumen</td>
<td></td>
</tr>
<tr>
<td>Submucosal (buried) absorbable sutures</td>
<td></td>
</tr>
<tr>
<td>Tight-fitting graft in the posterior cricoid split</td>
<td>Prolonged paralysis</td>
</tr>
<tr>
<td>Graft level with the intraluminal mucosa</td>
<td>Keloid formation</td>
</tr>
<tr>
<td>Superior limit below or at the interarytenoid space</td>
<td>Infection or inflammation</td>
</tr>
<tr>
<td>Inferior limit not beyond the cricoid</td>
<td>Missed airway pathology</td>
</tr>
<tr>
<td>Boat-shaped graft: 4 to 5 mm wide, 2 to 3 mm thick, 1 to 2 cm long</td>
<td>Laryngotracheomalacia</td>
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<td></td>
<td>Anterior wall collapse</td>
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<td></td>
<td>Vocal cord fixation</td>
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<tr>
<td></td>
<td>Interarytenoid stenosis or fixation</td>
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<td></td>
<td>Glottic stenosis</td>
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SSLTR, single-stage laryngotracheal reconstruction.
LTR with stenting

- Anterior – adequate for isolated anterior subglottic stenosis
- Anterior/posterior – for circumferential or posterior SGS
- Anterior/posterior/lateral – for complete SGS
LTR

- Introduced in 1972 by Fearon and Cotton.
- Widely used
- Tracheostomy and stent in place for several months
Same approach as ACS.
May perform posterior split if needed. Must be aware of esophageal mucosa to avoid inadvertent injury.
Stenting/tracheostomy short term (4-6 weeks) or long term (>2 months)
Duration of stenting

- Duration of stenting dependent on:
  - Amount of rigidity in the area of stenosis
  - Distortion of anatomy
  - Propensity for keloid formation/hypertrophic scar
  - Stability of grafts
  - Scar contracture
Complications

- Dysphagia
- Aspiration
- Granulation tissue
- Dislodgement of stent
Granulation tissue
Factors leading to failure

- Choi et al, retrospective chart review at tertiary care children’s hospital.
  - 17 patients requiring 42 LTRs
  - 2 perioperative deaths, 15 successfully decannulated.
  - 27 failed procedures
  - 24 of 27 failed procedures, at least one cause could be found for failure.
  - EBM-C
Factors leading to failure

- **Preoperative**
  - Inadequate assessment of post. SGS

- **Intraoperative**
  - Stent
    - Duration, length, type
  - graft

- **Postoperative**
  - Keloid formation, GERD, suprastomal/infrastomal collapse, poor follow-up, slipped or broken stent
Stents

- Aboulker, Montgomery T-tube, silastic swiss roll (portex and finger cot - no longer used). All have their own limitations, complications.

- **Aboulker is rigid, providing stenting and less collapsibility.**

- Swiss roll causes granulation tissue, gentle pressure.
  - used less often

- Montgomery stent for older children with adequate distance between glottis and stenosis.
  - Associated plugging, with airway obstruction
  - Used less often
Aboulker and Montgomery stents
Aboulker – most frequently used stent
Montgomery T tube

- Lumen with small caliber, easily occluded
- Used less frequently than Aboulker stent.
Cartilage

- Cartilage is better material because they have a lower rate of resorption, are easy to carve, and are viable without a vascular pedicle. They also retain bulk even without functional use.
- Rib and auricular most commonly used.
- Can not use grafts for lateral splits
Graft material

- Auricular cartilage
- Thyroid cartilage
- Hyoid bone
- Rib cartilage
- Irradiated cartilage
LTR with stenting

- Procedures requiring long term stenting falling out of favor.
- SSLTR or two-stage LTR preferred
- CTR another option for high grade stenosis
Cricotrachael resection

- First reported in 1970 and popularized in the 90s.
- More technically challenging than split procedures.
- Can be used as salvage for failures.
Success rates

- White et al, retrospective chart review of 100 consecutive patients at tertiary care center.
  - 96 total patients, 89 with Grade III/IV stenosis
  - 94% decannulation rate
  - Vocal cord dysfunction was only significant risk factor for failure to decannulate after 1 surgery.
  - MRSA and pseudomonal infections may play a role in failure, but cohort too small.
  - EBM-C
CTR

- Best candidates are those with severe SGS, without associated glottic pathology and with at least 4mm in healthy airway below the vocal folds and above the stenosis.
Exposure for CTR
CTR – Line of resection in relation to recurrent laryngeal nerve
- Elevation of perichondrium from anterior cricoid arch to avoid recurrent laryngeal nerve injury
CTR – anterior cricoid arch excised
CTR – removal of soft tissue of posterior cricoid plate
CTR – optional partial laryngofissure for increased luminal diameter
CTR – dissection of party wall
Completed CTR
CTR – completed reconstruction with stay sutures
CTR – posterior anastomosis

**Figure 1** Posterior anastomosis suture placement. A, Cotton/Monnier technique; B, Grillo technique, C, described technique.

**Figure 2** Anterior views of posterior anastomosis suture placement.
Figure 4  Preoperative bronchoscopy shows superior extent of stenosis (A) and postoperative bronchoscopy shows widely patent subglottis (B) in the same patient.
CTR complications

- Anastamosis failure
- Granulation tissue
- RLN injury
- Arytenoid prolapse
- Restenosis
- Wound infection
- Need for further procedures
- Re-intubation
Postoperative Care

- Intensive care unit
- Intensivist familiar with these cases
- Patients with trach and stent
  - Abx
  - Antireflux
  - Trach care teaching
  - Often discharged in several days
  - Repeat endoscopy q 3-4 weeks for stent evaluation
  - Stent duration
    - Depends on purpose
      - Hold graft in place – as little as one weeks
      - Counteract scar formation – months to a year
Postoperative Care

- ACS or SS-LTP
  - More intense care
  - Intubated 7-14 days with ETT as stent
  - Broad spectrum abx
  - Antireflux
  - Chest physiotherapy and log rolling
  - May need paralysis
  - Extubate when audible air leak at 20 cm H20
  - Decadron 1mg/kg 12hrs prior to extubation and 5 days postextubation
Conclusions

- Fiberoptic laryngoscopy and direct laryngoscopy/bronchoscopy essential for diagnosis.
- Choice of procedure dependent on grade of stenosis, ability of surgeon, and diligent post-operative care.
- High decannulation rates, but may require multiple procedures.
Bibliography

- Cable et al. Pediatric airway reconstruction: Principles in decision-making and outcomes at The University of Iowa Hospitals and clinics. *Ann Oto Rhinol Laryngol* 113:2004; 289-293