Approach to Pediatric Laryngeal Stenosis

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Introduction

* Subglottis stenosis (SGS):
  * Most common acquired disorder of larynx in children
  * Most common disorder of larynx requiring tracheotomy in infants
  * 3rd most common congenital laryngeal disorder
  * Pediatric larynx is funnel-shaped, narrowest at subglottis
    * Prone to ETT injury
Pathophysiology

- Neonatal subglottis: 4 to 5 mm (coronal plane)
- Premature newborn subglottis: 3.5 mm
- Adult subglottis: 12 mm
- Narrowing of infant subglottic diameter by 1mm:
  * Decreases cross-sectional area by 75%  
  * Increases airway resistance 16-fold
- Narrowing of adult subglottic diameter by 1mm:
  * Decreases cross-sectional area by 30%  
  * Increases airway resistance 2-fold
Prior to 1960s:
- Most SGS due to trauma or infection (TB, syphilis, typhoid, diptheria)
- Surgical intervention limited to airway dilatation and tracheotomy

Early 1960s:
- Medical advances revolutionized care and survival of premature infants
  - Advent of long-term intubation for neonates
- Led to increased incidence of SGS
- Subsequently prompted evolution of airway reconstruction
Acquired SGS

- Cause of SGS related to ETT
- If pressure from ETT exceeds capillary perfusion pressure:
  - 1st hours to days after intubation:
    - Mucosal ischemia, edema, erosion, ulceration
    - Can lead to full-thickness cricoid cartilage destruction and granulation tissue
  - Subsequent 3 weeks:
    - Healing period and re-epithelialization, even with ETT still in place
Why do some intubated neonates develop SGS?

- May have congenitally small cricoid to begin with
- Excess ETT movement (poor tube fixation or sedation)
- Tube design and materials (PVC, uncuffed)
- Superimposed infection (biofilms)
- Uncontrolled GERD/LPR

Duration of intubation:

- Factor in SGS in older children and adults (>2 weeks)
- In neonates, after 1st week of intubation, degree of injury not proportional to duration of intubation (Quiney & Gould, 1985)

Surgical factors:

- High tracheotomy, extensive laser surgery, previous surgery
Neonatal SGS Trends

* 1971-1979: 0.9% to 8.3%
* 1980-1989: 0% to 4.2%
* 1990-1999: 0% to 0.63% (Walner et al. 2001, Chicago)

Why did the incidence of acquired SGS decrease?
* Advanced awareness of potential subglottic injury from ETT:
  * Ideal ETT is not the largest that fits, but rather the smallest that permits adequate ventilation (leak <25cm H2O)
* Improved ETT material, tube fixation, use of non-cuffed tubes, anti-reflux precautions, steroids
* Other pulmonary advances (e.g. use of surfactant) reduced duration of intubation
Definition: SGS with no prior known risk factors (e.g. intubation)

Accounts for 5% of all pediatric SGS cases

More mild, more likely to spontaneously improve, less likely to require surgery

Two histopathologic types:

* Cartilaginous (most common):
  - Larynx fills with mesenchyme (8th week); recannalizes (10th weeks); variable shape (elliptical most common)
  - Not responsive to dilatation or laser procedures

* Membranous:
  - Thickened subglottic mucosa (fibrous tissue, mucus gland dilatation)
  - Usually circumferential, soft to palpation
History

- **Grahne 1971:**
  - Division of anterior and posterior cricoid in children, with stent wired to metal tracheostomy tube
- **Evans and Todd 1974:**
  - “Laryngotracheoplasty”:
    - Castellated incision of anterior cricoid cartilage and upper trachea, stented for 6 weeks
- **Cotton and Seid 1980:**
  - Anterior cricoid split procedure
    - For neonates with anterior glottic or subglottic stenosis, to attempt to avoid tracheotomy
- **Holinger et al. 1987:**
  - 138 patients underwent anterior cricoid split; successful extubation in 77%
Laryngotracheal reconstruction:
- Cricoid cartilage divided and repaired with cartilage grafts to enlarge pediatric subglottic lumen

Cotton et al. 1989:
- 203 children who had LTR; 186 (92%) decannulated
- Graft survival and growth was confirmed over prolonged follow-up

LTR became procedure of choice for repair of SGS
LTR success rates lower for more severe cases, which prompted search for other procedures

Partial cricotracheal resection with primary anastomosis:
- Has become viable option for severe cases of pediatric SGS
- Damaged cricoid cartilage is partially excised, along with proximal tracheal rings, with end-to-end cricoid-to-trachea anastomosis

2005: Monnier et al.:
- 57 pediatric CTR procedures; 55 (96%) decannulated

2005: White et al.:
- 93 pediatric CTR procedures
  - 55 as salvage procedure after failed LTR
- 87 (94%) decannulated
Clinical Scenarios

- Infant in ICU who has failed multiple extubation attempts
  - Could be other reasons for failed extubation besides SGS
  - Airway assessment may have to wait until child medically stable; this might have to occur after tracheotomy already placed
- Child seen in office with tracheostomy tube already present, with diagnosis of “airway obstruction”:
  - Airway evaluation required to determine site of obstruction
- Child intubated for random elective procedure, with post-extubation stridor
- Mild to moderate SGS in unintubated child:
  - Often asymptomatic until URI causes stridor; recurrent croup
- Diagnosis of SGS requires airway endoscopy
Since ETT is most common etiology for acquired SGS, ask relevant questions:

- Was child a premature newborn (<36 weeks)
- Was child ever intubated?
  - If so, why? For how long? Multiple intubations? Difficult intubations? What size tube?
Three basic functions of larynx:

1) Respiration
2) Protection:
   - Prevention of aspiration
   - Clearance of secretions
3) Phonation

Ask relevant questions related to all 3 functions
1) Respiration

- If the child has a tracheostomy tube, the most immediate question that should be addressed is: WHY?
- Need to know child’s pulmonary status
- If the patient is on ventilator, when is it estimated that he/she will be weaned?
- If patient is on CPAP/BiPAP via tracheostomy:
  - Can CPAP/BiPAP be delivered by another means?
  - Would subglottic pressure improve with decannulation?
- If patient has O₂ requirement:
  - Does this preclude removal of the tracheostomy tube?
- Pulmonary clearance is important
2) Aspiration

- Successful LTR requires more than merely establishing adequate subglottic airway
  - Significant aspiration must be avoided
  - If child already aspirates, this may be exacerbated by LTR (especially posterior graft)
    - Aspiration is relative contraindication
  - Two types of aspiration: food and saliva
    - Both types must be addressed pre-operatively
    - Child with no pre-operative identified aspiration may still aspirate after reconstruction, but this is usually temporary and self-limited
2) Aspiration: Oral Feeds

- Need to know patient’s feeding status
- If child takes p.o. diet:
  - Any difficulties with feeding or weight gain?
- If child is feeding-tube dependent: why?
  - Oral aversion (former premature newborns)
  - Neurological discoordination of swallowing/lack of laryngeal sensation
  - Vocal fold immobility:
    - Cardiac surgery: unilateral TVF paralysis
    - Chiari malformation: bilateral TVF paralysis
    - Anatomical defect: laryngeal cleft, TE fistula
2) Aspiration: Oral Feeds

- To evaluate potential aspiration of oral feeds:
  - Clinical feeding evaluation
  - Blue dye test
  - Modified barium swallow study
  - Functional endoscopic evaluation of swallowing ± sensory testing (FEEST)
  - Bronchoalveolar lavage (BAL)

- Management:
  - Altering texture (thickened feeds)
  - Swallowing therapy
  - NPO
2) Aspiration: Saliva

- **Salivary aspiration:**
  - Suspected in cases of chronic lung disease
  - Recurrent aspiration pneumonia
  - More likely in children with underlying neurological disability (CP, syndromes)

- **Diagnosis mainly based upon clinical suspicion**
  - Chest CT
  - Salivagram

- **Management:**
  - Anti-cholinergics
  - BoTox salivary gland injections
  - Salivary duct/gland surgery
  - Injection laryngoplasty (in cases of unilateral VF paralysis)
3) Phonation

- In work-up of SGS, voice quality is assessed primarily to provide clues regarding status of the airway
  - Strong voice: implies TVFs are able to contact
    - Does not imply TVFs can abduct properly
  - Hoarse voice: TVF immobility, scarring, edema, lesion
  - Aphonia: 100% obstruction (SGS, trachea, large tube)

- Good phonation is important, but is not required for successful airway reconstruction
  - Voice quality has regularly been sacrificed (or ignored) at expense of improving respiration
Physical Examination

Overall appearance is important:
- Failure to thrive vs. appearing healthy
- Any other anatomical problems:
  - Cleft lip/palate, dysmorphic facial features

Congenital syndromes:
- Higher risk of SGS (e.g. Down syndrome)
- Midface hypoplasia (nasopharyngeal & soft palate obstruction)
- Micrognathia (base of tongue/epiglottis obstruction)
Since GER thought to seriously compromise airway healing, every patient should receive a work-up (European 2003)

- Work-up: pH and impedance probes; **EGD with biopsy**, gastric scintiscan

**If work-up negative:**
- Still give reflux medication 1 month prior, and 6-12 weeks after, open airway surgery

**If work-up positive:**
- Treat with 1-2 months medication, and repeat study

**If severe GER:**
- Consider pediatric surgery referral for Nissen fundoplication
Flexible Nasolaryngoscopy

- All patients with suspected laryngeal airway obstruction should have flexible scope while awake:
  - Nasal cavities, nasopharynx, soft palate, tongue base
  - Assessment of supraglottic dynamic function
  - Assessment of vocal fold movement essential
Rigid Endoscopy

* The most accurate way to diagnose SGS
  * Thorough inspection of entire airway to rule out other lesions
* ETTs used to grade severity of SG
Subglottic Sizing Chart

Myer et al. 1994
Posterior Glottic Stenosis

- Scarring between posterior aspect of TVFs
- Also results from prolonged intubation
- May occur in isolation, or with SGS
- May closely mimic bilateral TVF paralysis
- Rigid endoscopy with TVF and arytenoid palpation required
- If associated with SGS, both need to be addressed to achieve decannulation
Posterior Glottic Stenosis

Table 1. Classification of Posterior Glottic Stenosis (Bogdarian & Olson).

<table>
<thead>
<tr>
<th>Type</th>
<th>Description</th>
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<tbody>
<tr>
<td>TYPE I</td>
<td>Glottis – interarytenoid scar, normal posterior commissure</td>
</tr>
<tr>
<td>TYPE II</td>
<td>Interarytenoid scar and posterior commissure scar</td>
</tr>
<tr>
<td>TYPE III</td>
<td>Posterior commissure scar involving a cricoarytenoid joint</td>
</tr>
<tr>
<td>TYPE IV</td>
<td>Posterior commissure scar involving both cricoarytenoid joints</td>
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Pre-operative Plan

* **Airway algorithm:**
  * Define each airway problem
    * Determine each problem’s importance
    * Determine ease/difficulty in fixing problem
    * Co-existing problems must be identified and addressed

* **Multi-disciplinary input:**
  * Neonatologist, intensivist, pulmonary, GI, cardiology, 1º care
  * Parents actively involved in decisions
Management also depends upon maturity of subglottic injury:
- Acute/subacute injury may be amenable to endoscopic repair
- Mature stenosis more likely to require open surgery
Tracheotomy

* Often most appropriate first step in neonate with SGS
  * Allows time for child to grow and gain weight
  * Allows optimization of pulmonary status prior to reconstruction (BPD)
  * Allows immature stenosis to mature

* Drawbacks:
  * Morbidity:
    * Tracheitis, bleeding/granulation tissue, mucus plugging, accidental decannulation
  * Parental concerns:
    * Constant vigilance, fear of exposing to germs, need for equipment and nursing staff, sheer exhaustion and anxiety
    * Delayed language acquisition skills
Neonate who has failed extubation, and other causes besides SGS have been ruled out:

- If mild SGS, reintubate with smaller tube, steroids, Abx, remove granulation tissue, try to extubate again
- GERD assessment and empiric treatment
- If Mod-severe SGS, can try anterior cricoid split
- If Mod-severe SGS, can try thyroid ala graft SSLTR
- If severe SGS, can perform tracheotomy
  - After tracheotomy, without the ETT to act as a stent, stenosis often becomes more severe with scar contraction and you can get view of true extent of damage
  - Can also get better by removing the irritating agent
**Endoscopic Treatment**

- **Serial dilatations:**
  - Not effective for mature, acquired stenoses
  - May be effective for early, immature subglottic stenosis
    - May “buy time” in newborn with SGS
    - May be useful if airway in danger of restenosing after open reconstruction

- **Durden & Sobol 2007:**
  - 10 neonates/infants (2-12 months) with acquired Grade 2 and 3 SGS from ETT
  - 70% avoided tracheotomy
Endoscopic Treatment: Laser

- Adult SGS: CO₂ laser 4-quadrant incisions followed by dilatation (Shapshay et al. 1987)
  - Often worsens original problem in children

- Appropriate pediatric laser use
  - Granulation tissue from early ETT injury
  - Subglottic hemangioma
  - Suprastomal tracheal cartilage collapse/granulation
  - Very mild, isolated stenosis (<1 cm vertical scar)

- Things to avoid:
  - Lasering circumferentially
  - Lasering in posterior commissure/interarytenoid area
Mitomycin C

- Antibiotic derived from *Streptomyces caespitosus*
- Anti-neoplastic and anti-proliferative properties (inhibitis fibroblast proliferation)
- Used regularly by ophthalmologists for decades
- Did not reduce granulation tissue in randomized, controlled trial after LTR for SGS (Hartnick et al. 2001)
- Long-term effects not known:
  - Agarwal & Morrison 2006: First case in world literature of laryngeal CA in non-smoking adult who had mitomycin C application after glottic web lysis
Open Surgery

- Recommended for mature SGS when conservative approaches have either failed, or are inappropriate
- Recommended for all Grade 3 and 4, and most Grade 2 SGS
- May be required in patients who do not have tracheostomy, if SGS with exercise intolerance and lack of airway growth
- Many approaches to pediatric LTR
- Traditional basic tenets:
  - Widen cricoid
  - Minimal excision of scar tissue (minimizing new raw surfaces)
  - Restoration of skeletal support with autogenous costal cartilage
  - Allow epithelialization of raw surfaces from adjoining respiratory epithelium
Concept: To avoid tracheotomy in premature newborns who have SGS:

Requirements:

- No other significant airway disease
  - Upper airway, pulmonary (off ventilator, O2, absence of infection)
- Weight > 1,500 gm

After repair: nasotracheal intubation x 5-7 days

Becoming less popular in favor of endoscopic techniques

Trend: small thyroid ala graft
SGS: Open Surgical Choices

- **Grade 1**
  - Primarily no surgery required

- **Grade 2:**
  - Usually requires LTR with anterior cartilage graft to cricoid

- **Grades 3 and 4**
  - LTR with anterior and posterior graft
    - (occasionally just posterior cricoid split)
  - Partial cricotracheal resection with posterior cricoid graft

- **Posterior glottic stenosis (with SGS):**
  - Posterior cartilage graft
LTR Pre-Requisites

* Original indication for intubation must no longer be present
* There must be no other significant co-existing airway obstruction
* Larynx must not be “reactive” or edematous
* Stenosis should consist of mature scar tissue
* Any GERD/LPR must be well-controlled
  * Use perioperative anti-reflux medication empirically
* Any eosinophilic esophagitis must be treated and resolved
* Use post-operative antibiotics; peri-extubation steroids
* Good general respiratory and neurological condition
* Age not important, but child should be thriving and of reasonable size (10 kg, except for cricoid split)
* High likelihood of decannulation once SGS repaired
Single vs. Double-Stage

* **Single-stage:**
  * If there is pre-existing tracheostomy tube, it’s removed at time of reconstruction
  * If there is not pre-existing tracheostomy tube, one is not placed
  * Stenting is with ETT: avg. 7 days
    * I do 3 – 5 days

* **Double-stage:**
  * If there is pre-existing tracheostomy tube, it remains in place even after reconstruction, and removed at later date
  * If there is not pre-existing tracheostomy tube, one is placed at time of reconstruction, and removed at later date
  * Stenting is with stand-alone stent, either above or attached to tracheostomy tube: avg. 3-4 weeks
    * I do 2 weeks
Tracheal Approach - Video

* **LTR.APROACH**
Advantage:
- Only one procedure needed
- Shorter duration of stenting

Disadvantages:
- It has to work; otherwise, need to re-intubate, re-perform tracheotomy
- Requires post-op sedation, expert ICU care

Single-stage advisable when:
- SURGEON COMFORT
- Primarily only when anterior graft needed but can be done with posterior
  - Grade 2 and mild Grade 3 stenosis
  - Useful for suprastomal cartilage collapse
- Useful for children who did not require pre-op tracheostomy tube
- High likelihood that LTR will be successful
Double-Stage LTR

**Advantages:**
- Allows longer duration of stenting
- Patient can be awakened immediately post-op, and can go home
- Don’t have to re-intubate or replace tracheostomy tube: it’s already in

**Disadvantages:**
- Can take months between reconstruction and decannulation
- When stent is in place, patient is 100% dependent on tracheostomy tube

**Double-stage advisable when:**
- Severe stenoses (Grades 3 and 4)
- Complex reconstruction (multiple levels, salvage)
- Questionable pulmonary function
- Airway with history of being highly reactive
- Children for whom reintubation is difficult (craniofacial or vertebral anomalies)
VIDEO DS LTR

* DSLTR VIDEO
Anterior Graft

- Appropriate for most grade II, and some mild III SGS
- Requirements: Intact posterior cricoid with no posterior glottic stenosis
- Usually done in single-stage
- Graft choices:
  - Thyroid cartilage (easy to harvest; smaller graft) (Fayoux et al. 2006)
  - Costal cartilage (widely used, up to 4cm length, sturdy and easy to carve; requires separate incision, adds time)
Thyroid Ala SSLTR
Costal Cartilage Graft
Costal Cartilage Rib Graft
Posterior Graft

- Used in combination with anterior graft for Grade 3 and 4 SGS
- Used in combination with anterior graft for Grade 2 SGS with associated posterior glottic stenosis
- Can be used for isolated posterior glottic stenosis
- 0.05 to 1.00 mm distraction recommended for each year of age, up to 1 cm
- Success rate for PGS decannulation: 83% (Rutter & Cotton)
Endoscopic Posterior Graft

- Inglis et al.:
  - 10 patients
- Procedure:
  - Suspension microlaryngoscopy
  - CO$_2$ laser to divide posterior cricoid
  - Costal cartilage graft (6-10mm) placed
  - Tracheostomy decannulation within 2-3 days
- Indications:
  - No more than mild Grade III SGS
  - Impaired TVF mobility from scar (PGS)
  - TVF paralysis
- 80% success rate
- Works best for PGS, less effective than anterior graft for SGS
LTR: Before and After
Disease-Specific Outcomes

* Hartnick et al.:
  * 199 children with SGS; overall decannulation rate: 84%
* Single-stage LTR decannulation rate:
  * Grade 2 SGS: 82% (100% overall)
  * Grade 3 SGS: 79% (86% overall)
  * Grade 4 SGS: 67% (100% overall)
* Two-stage LTR decannulation rate:
  * Grade 2 SGS: 85% (95% overall)
  * Grade 3 SGS: 37% (74% overall)
  * Grade 4 SGS: 50% (86% overall)

* Relative low success rates for Grade III and IV, with need for multiple revision surgeries, is what prompted partial cricotracheal resection
Partial Cricotracheal Resection with 1° Anastomosis

- Stenotic subglottic segment resected, with end-to-end anastomosis from trachea to posterior cricoid
- Used regularly in adults; has been adapted to children

**Advantages:**
- No need for graft
- Degree of stenosis does not impact outcome
- Faster time to decannulation compared to multi-stage LTR

**Disadvantages:**
- Anastomotic dehiscence
- RLN injury
- Possible interference with laryngotracheal growth
- Limitation in options for future reconstructive surgery
Best candidates:
- Severe SGS (Grades III and IV)
- No associated glottic disease
- Margin of ≥ 4mm of normal airway beneath TVFs
- Persistent circumferential stenosis following previous attempts at airway reconstruction

If resection up to level of TVFs required:
- Expect marked glottic edema
- Expect tracheostomy or T-tube afterwards

If glottic stenosis also present:
- May need to add posterior cricoid split and longer-term stenting (“extended” PCTR)
Best way to approach acquired SGS is to prevent it

When SGS occurs:

- Individualize management depending on:
  - Maturity of stenosis
  - Grade of stenosis
  - Co-existence of other sites of airway obstruction
  - Co-existence of other respiratory/GI/cardiac/neurology abnormalities
- Multi-disciplinary approach required
- Surgical techniques constantly evolving

One of the most challenging and rewarding areas of pediatric otolaryngology