

Subglottic Stenosis

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Basic Science Review

Wound Healing

- involves three temporally overlapping stages
- inflammatory phase, proliferative phase and contraction or remodeling phase

Inflammatory Phase

- involves vascular constriction and then dilation
- coagulation and complement cascade is activated
- PMNs enter wound at 6 hrs post injury
- PMNs function to phagocytize debris and bacteria from wound

Inflammatory Phase

- Helper T cells are necessary for wound healing
- Macrophages enter wound within 48 hrs
- only cells that can fxn at low O₂ levels
- Macrophages are essential to wound healing

Inflammatory Phase

- growth factors are major regulators of healing
- they interact with cellular receptors to modify cell activities
- IL-1 directly stimulates fibroblast activity including proliferation and collagen synthesis
- IL-2 is produced by helper T cells

Inflammatory Phase

- structural components essential to wound healing include fibronectin, collagens, glycoproteins and glycosaminoglycans
- several types of collagen are found in the healing wound

Proliferative Phase

- lasts 10-14 days
- begins with re-epithelization
- the epithelial cells 1-2mm from the wound edge undergo phenotypic changes
- cell replication rate increases 17 fold
- epithelial cell migration is dependent on local humidity and oxygenation
- epithelial cells migrate much quicker when the wound is moist and occluded

Proliferative Phase

- Neovascularization is the next part of the proliferative phase
- macrophages secrete angiogenic factors
- endothelial migration results in capillary bud formation
- collagen deposition begins when fibroblasts enter the wound at 48-72hrs
- the collection of fibroblasts, inflammatory cells and capillary buds is referred to as granulation tissue

Wound Contraction and Remodeling

- Begins 6-7 days after injury and is maximal for 10 days
- eventually decreases the defect by 40-60%
- skin grafts and flaps can reduce contraction by 50-70%
- remodeling results in a scar with as much as 80% of the skin's original tensile strength

Subglottic Stenosis

- Congenital or acquired narrowing of the subglottic airway
- third most common congenital airway problem
- Otolaryngologist must be an expert at diagnosis and management
- can occur in adults and children

Anatomy

- infant larynx differs in size and position when compared to adult larynx
- the narrowest portion of the adult airway is the glottic aperture while in the infant it is the subglottis
- the infant larynx is higher in the neck
- the structures of the infant airway are more pliable and less fibrous making it susceptible to narrowing from edema

Embryology

- Respiratory system is outgrowth of primitive pharynx
- begins at 26 days after conception
- laryngotracheal diverticulum becomes separated from foregut by tracheoesophageal folds
- tracheoesophageal folds fuse to form tracheoesophageal septum
- septum divides foregut into ventral laryngotracheal tube and a dorsal esophagus
- failure of TE folds to fuse can cause TE fistula

Embryology

- Larynx develops from 4th and 5th branchial arches
- laryngotracheal opening lies between these two arches
- laryngeal aditus becomes T shaped by growth of three masses
- 1st mass=hypopharyngeal eminence which eventually becomes the epiglottis
- 2nd and 3rd masses are arytenoid masses. As these masses grow between 5-7th weeks, laryngeal lumen is obliterated

Embryology

- recanalization occurs in 10th week
- failure to recanalize = atresia or stenosis of larynx
- arytenoid masses separated by notch which eventually becomes obliterated.
- failure to obliterate can result in posterior laryngeal cleft which can cause severe aspiration in the newborn

Congenital Subglottic Stenosis

- Thought to be secondary to failure of laryngeal lumen to recanalize
- defined as subglottic diameter less than 4.0mm in full term infant
- normal full term newborn subglottic diameter = 4.5-5.5
- premature infant subglottic diameter = 3.5mm. If less than 3.5mm in premie = subglottic stenosis

Congenital Subglottic Stenosis

- Considered congenital if no previous history of intubation or trauma
- divided into membranous or cartilaginous types
- membranous is soft-tissue thickening from fibrous connective tissue or hyperplastic submucous glands
- membranous may also involve the vocal folds

Congenital Subglottic Stenosis

- Cartilaginous usually results from thickened or malformed cricoid
- usually forms large anterior subglottic shelf leaving only small airway posteriorly
- can be due to an elliptical shaped cricoid
- membranous type usually less severe than cartilaginous

Congenital Subglottic Stenosis

- severity depends on degree of subglottic narrowing
- symptoms can range from mild with picture of recurrent croup to severe with respiratory distress at delivery
- often associated with other congenital anomalies

Acquired Subglottic Stenosis

- Numerous causes including intubation, trauma, infection/inflammation, thermal or caustic injuries
- most common cause is endotracheal intubation
- since more very premature infants are surviving, incidence of acquired subglottic stenosis has increased

Acquired Subglottic Stenosis

- Reported incidence in intubated patients = 1-8%
- pathogenesis not completely understood
- one theory includes mucosal pressure leading to ulceration leading to chondritis and finally deposition of fibrous material
- less results in weakened cartilage framework and firm scar

Acquired Subglottic Stenosis

- risk factors in neonates include prolonged intubation, size of endotracheal tube, increased motion of tube, repeated intubations, birth weight less than 1500g, infection, presence of NG tubes and GERD
- many feel most important factor is length of intubation

Acquired Subglottic Stenosis

- There is no “safe” period for intubation
- premies tolerate intubation better than adults due to more yielding and pliable tissues
- it has been suggested that tracheotomy be considered after 50 days of intubation in neonates

Acquired Subglottic Stenosis

- Ideal endotracheal tube size allows air leak at pressure of 20cm H₂O
- absence of audible air leak is indicative of excessively large tube
- tube motion can cause abrasion and trauma to mucosa
- tube should be carefully secured and patient adequately sedated

Acquired Subglottic Stenosis

- Repeated intubations should be minimized. No routine tube changes.
- Better education and care of intubated infants has lead to a decrease in incidence of acquired subglottic stenosis
- routine use of surfactant also appears to have lowered the incidence

Diagnosis

- Typically present with stridor and respiratory distress
- stridor is biphasic
- diagnosis begins with complete history
- question parents about: duration, progression, hx of prematurity, birth trauma, hx of intubation, feeding problems, change in voice or cry, recent trauma or foreign body exposure

Diagnosis

- Examine child at rest and when agitated
- auscultate over nose, mouth, neck and chest
- quality of child's voice should be noted
- flexible fiberoptic examination should be performed
- special attention paid to vocal cord motion
- subglottis can sometimes be seen below the cords

Diagnosis

- Radiographic evaluation includes AP and lateral views of neck
- narrowed subglottic airway suggests stenosis or croup
- airway fluoro can be helpful
- Ba swallow can help r/o vascular compression
- CT has not been helpful in assessing pediatric airway

Diagnosis

- Gold standard remains rigid endoscopy under GA
- Magnification with Hopkins telescopes very helpful in defining pathology
- palpation of cricoarytenoid joints imp
- wait few minutes after removing ET tube to look for edema that tube was stenting

Diagnosis

- No universal classification system
- in past, measurements were done subjectively or using various instruments
- most commonly used system today is Cotton's
- percentage of obstruction and anatomic location were assigned grade I-IV based on perceived percentage of obstruction

Diagnosis

- This system is dependent on skilled judgement
- Myer, Conner and Cotton have proposed system based on standardized endotracheal tube sizes
- the ET tube that will pass thru the lumen and has normal leak pressures is compared to the expected age-appropriate tube size

Diagnosis

- The maximum percentage of airway obstruction is determined and assigned a grade:
- Grade I <50% obstruction
- Grade II 51-70% obstruction
- Grade III 71-99% obstruction
- Grade IV no detectable lumen

Management

- begins with prevention
- control of risk factors is essential
- although controversial, many feel that significant GERD should be treated prior to any surgical intervention
- Halstead recently demonstrated that significant GERD is an important cofactor in many pediatric airway disorders, particularly subglottic stenosis

Management

- Acquired subglottic stenosis is typically more severe than congenital and more likely to require surgical intervention
- many of these patients will require a tracheotomy while awaiting definitive therapy

Management

- Mild stenosis (Grades I and II) can usually be treated with endoscopic techniques such as dilation and CO2 laser resection
- factors associated with failure include: previous attempts at endoscopic repair, loss of cartilaginous support, exposure of cartilage during laser resection, bacterial infection, posterior inlet scarring, glottic involvement, vertical scar length of $> 1\text{cm}$

Management

- Endoscopic dilation has had disappointing results
- Endoscopic laser resection for Grades I and II stenosis have success rates ranging from 66-80%

Management

- More severe stenosis (Grades III and IV) usually require open surgical approach
- contraindications include: inability to tolerate GA, persistent need for tracheotomy, significant GERD, an ICU not equipped to handle the post-operative care

Management

- Some of more popular procedures include: anterior cricoid split, laryngotracheoplasty (either stented or one-stage), and end-to-end anastomosis
- anterior cricoid split usually used in neonate who has failed extubation instead of doing tracheotomy

Anterior Cricoid Split

- Criteria include: extubation failure on two occasions due to laryngeal pathology, weight >1500g, no assisted ventilation 10 days prior, O₂ requirements <30%, no CHF one month prior, no infection
- performed after DL and B ahs confirmed diagnosis
- all other airway pathology must be r/o

Anterior Cricoid Split

- Vertical incision thru cricoid, first two tracheal rings and lower thyroid cartilage
- stay sutures on either side
- drain placement
- remains in ICU intubated and sedated for 7-14 days based on infant's weight

Laryngotracheal Expansion

- Involves scar division with distraction of edges with interposition of graft to widen the airway
- several techniques depending on severity and location of stenosis
- laryngotracheoplasty (LTP) can be done in two stages with a stent or a single stage using the ET tube as a stent
- One-stage LTP is gaining popularity

Laryngotracheal Expansion

- Anterior laryngofissure with anterior lumen augmentation - good for stenosis that does not involve the glottis and has good cartilage support
- Laryngofissure with division of posterior cricoid - used in pts with glottic involvement or significant cricoid deformity
- Laryngofissure with anterior and posterior grafting - as above but significant posterior stenosis

End-to-End Anastomosis

- Indicated if severe cricoid deformity causing grafting likely to fail
- most say there must be 10mm of normal airway below glottis but Cotton says can resect up to vocal folds but expect prolonged edema
- technically difficult due to close proximity of vocal folds and risk to recurrent nerves

End-to-End Anastomosis

- Stenosis <4cm can be resected with laryngeal release and cervical tracheal mobilization
- stenting is not required
- can be performed with a tracheostomy tube in place or as a single stage
- Monnier reports good success with high grade lesions - decannulation rate of 93%

Post-operative Care

- Require specialized care in ICU
- if two-staged, hospitalization stay is shorter
- if single-staged LTP or ACS, stay intubated in ICU for 7-14 days
- requires heavy sedation with or without paralysis
- extubation done when adequate airleak or after certain period

Complications

- Infrequent but can include (in decreasing order of frequency): atelectasis, pneumonia, malpositioned ET tube, accidental extubation, occluded ET tube, wound infection, granulation tissue, TC fistula
- Complications specific to prolonged sedation required for single-stage procedures include narcotic withdrawal and transient muscle paralysis

Outcomes

- The goal is decannulation
- success is dependent on cause, number of previous failed attempts, status of the remainder of airway and severity of stenosis
- Cotton reports overall success = 92%, Grade II = 97% Grade III = 91% and Grade IV = 72%

Outcomes

- Many authors report that a functional voice is restored in most patients
- MacArthur reports on 12 pediatric patients who underwent LTR
 - 78% had altered anatomy
 - 44% had altered function
 - 100% had decreased voice quality
- conclusion = children with high grade stenosis are at risk for poor voice outcome after LTR