musc Children's Health
Medical University of South Carolina

Pediatric Vocal Fold Immobility

Anthony Ghanem, MD

Changing What's Possible | MUSCkids.org

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- I have no disclosures

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Objectives

- Understand the different etiologies of VF immobility
- Formulate a proper treatment strategy

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Outline

- Overview on anatomy and physiology
- Etiology of VFI
- Diagnostic evaluation
- Management
- Take home messages

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Embryology of the pediatric larynx

- The larynx is essential for a newborn's ability to breath, feed and cry
- Laryngeal development in the embryonic period (first 8 weeks)
- Maturation during the fetal period

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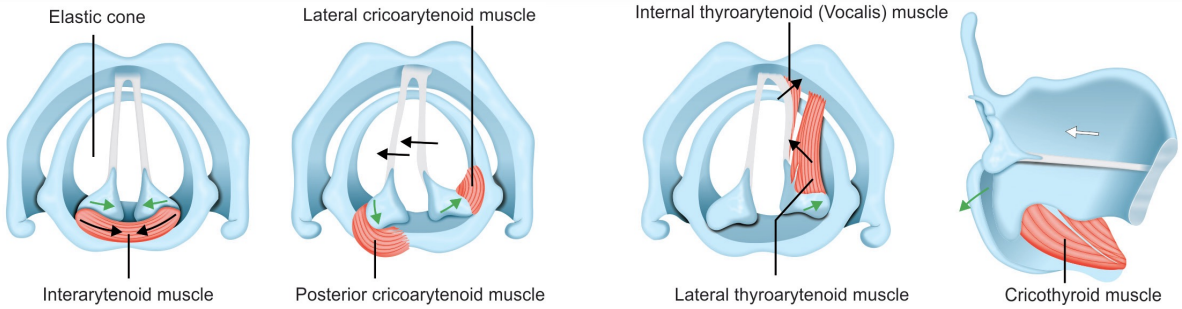
Anatomy



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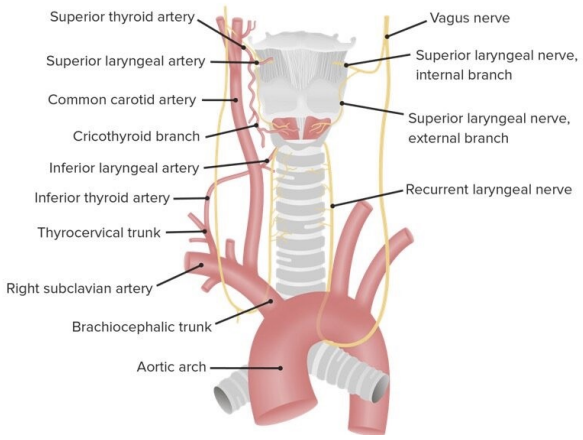
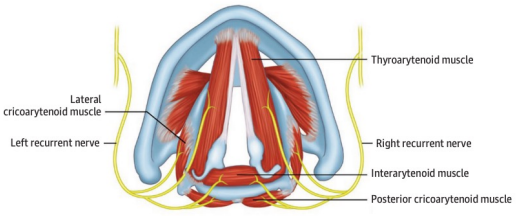
Anatomy





Hidgway et al., *JAMA Otolaryngology Head and Neck Surgery*, 2021

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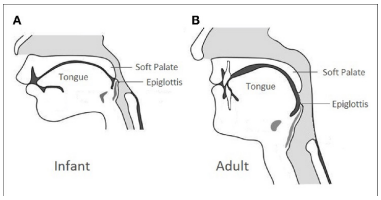
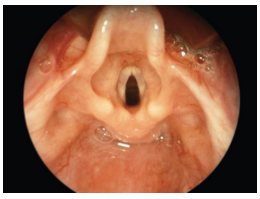
Infant vs. Adult larynx

- Infant larynx is located cephalad compared to its eventual descended position
  - Epiglottis near soft palate
- Size is 1/3rd of an adult larynx
- Vocal folds
  - ½ membranous
  - ½ cartilaginous





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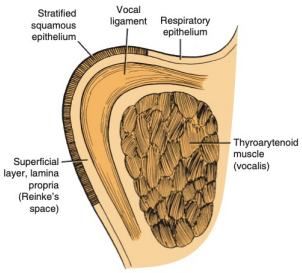
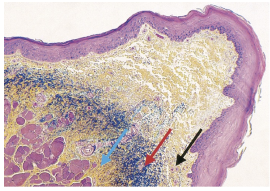
Vocal folds

Age	Overall length (mm)	Membranous	Cartilaginous	M/C
Newborn	2.5 – 3.0	1.3 – 2.0	1.0 – 1.4	1.1 – 1.8
Adult female	11 – 15	8.5 - 12	2.0 – 3.0	3.3 – 4.5
Adult male	17 – 21	14.5 – 18	2.5– 3.5	4.7 – 6.2





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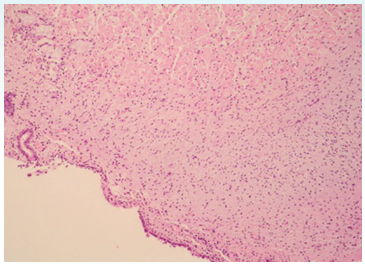
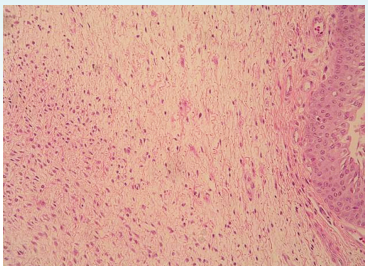
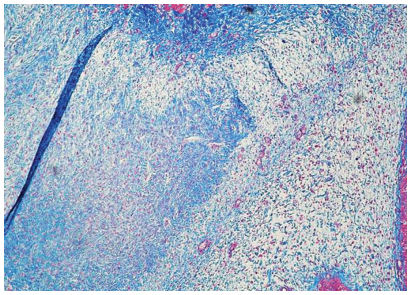
2 days



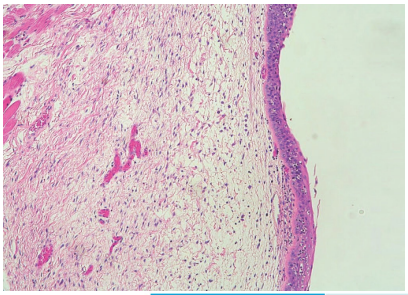
2 months



7 years



13 years



Hartnick et al. *Laryngoscope* 2005

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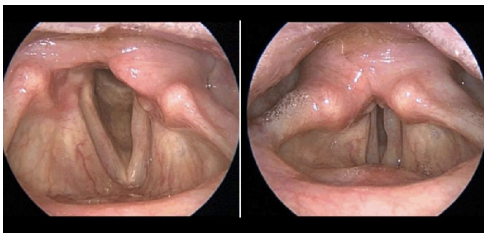
Vocal fold immobility

- Paresis
- **Paralysis**
- Fixation/tethering

- Unilateral or Bilateral ~ 10% of congenital laryngeal disease
  - 2nd most common cause of neonatal stridor
  - Bilateral in neonates
  - Early in life without gender predilection



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Etiology of VFI

- Congenital vs. acquired
- Neurologic vs. mechanical

- Unilateral VFP ~ 27-78%
  - Iatrogenic (13-81%)
  - Left > right

- Bilateral VFP ~ 22-73%
  - Idiopathic (17-75%)
  - Neurologic (20-55%)

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Etiology

Characteristic	No. (%) (N = 404)
Immobility	
Left	270 (66.8)
Right	32 (7.9)
Bilateral	102 (25.2)
Etiology	
Cardiac surgery	278 (68.8)
Idiopathic	85 (21.0)
Neurologic	30 (7.4)
Mixed[a]	5 (1.2)
Miscellaneous[b]	6 (1.5)

Jabbour et al. 2014

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Etiology

- Iatrogenic/trauma
  - Any procedure that comes in proximity to the vagus or RLN
    - ~ 8% left VFP after PDA ligation *(Zbar, 1996)*
    - Up to 25% in infants < 1150 g *(Smith, 2009)*
  - Posterior fossa trauma
  - Closed head injuries
  - Instrumentation of the larynx/hypopharynx (ETT)
  - Birth trauma
  - Vincristine neuropathy

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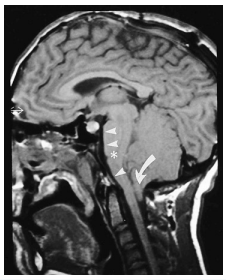
Etiology

- Central Nervous System
  - Mostly due to brainstem pathology
  - Most common is ACM (type 2 > type 1) → bilateral VFP
  - Complex respiratory problems in ACM
  - Leukodystrophies
  - Hydrocephalus
  - Perinatal hypoxia
  - ALS



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Etiology

- Peripheral Nervous System
  - Charcot-Marie-Tooth is most common
  - Myasthenia gravis
  - Myotonic dystrophy
  - Spinal muscular atrophy
  - Benign congenital hypotonia
- Idiopathic
  - First or second most common cause of VFP
  - Viral neuropathy?
- Neoplasia
  - Tumors of the skull base, neck or mediastinum
  - Very rare cause of VFP in children
- Cardiovascular
  - Vascular rings
  - Ortner syndrome

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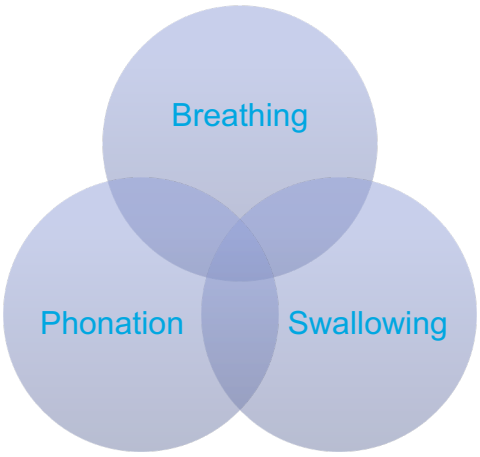
Signs and symptoms



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Signs and symptoms

Unilateral	Bilateral
Stridor	Stridor
Dysphonia	Increased work of breathing
Dysphagia	Cyanosis
Aspiration (often silent)	Glottic incompetence (in adductor paralysis)

Irace et al, *Laryngoscope*, 2019

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Differential diagnosis

- Any obstruction from the nasal tip down to the trachea!

- Pyriform aperture stenosis
- Choanal atresia
- Laryngomalacia
- Laryngeal webs
- Subglottic stenosis
- Subglottic hemangioma
- Tracheal stenosis

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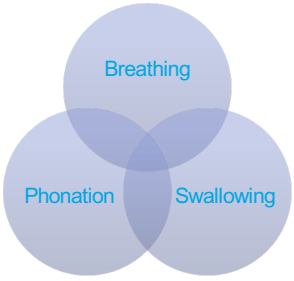
Diagnostic evaluation

- Thorough history
  - Symptoms
  - History of surgeries or trauma (including birth)
  - Detailed medical history (neuro/cardio/congenital anomalies)
  - Prematurity, ICU stays, intubation
- Examination
  - ABC
  - Listen and assess voice/breathing
  - Signs of increased work of breathing
  - Head and neck exam
  - Chest auscultation
  - **Awake flexible laryngoscopy**



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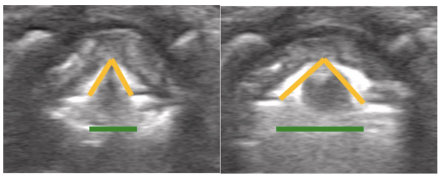
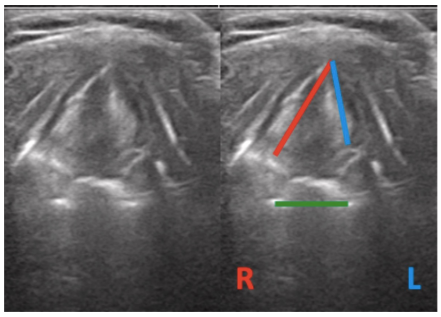
Laryngeal Ultrasound

- Low risk imaging option
- Better tolerated by young patients
- Sensitivity 91%
- Specificity 97% *Hamilton et al. 2021*





Deshpande et al. 2021

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Diagnostic evaluation

- Once VFP has been established → look for a cause
- MRI: brain, brainstem and skull base down to the mediastinum
- CT scan of the neck and chest in older children
- Swallowing studies
  - Barium swallow
  - FEES
- Rigid laryngoscopy and bronchoscopy

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Laryngeal EMG

- Useful in distinguishing vocal fold fixation from neurogenic paresis
- Relies on percutaneous placement of EMG needles for monitoring → sedation necessary in pediatric population
- Optimal examination between 4 weeks and 6 months of the expected injury
- Identifies normal innervation, absence of innervation, reinnervation and synkinesis
- Helpful in determining poor prognosis and treatment plan

Rickert SM et al. Laryngoscope. 2012;122(1):158-61.

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AANEM PRACTICE TOPIC

CONSENSUS STATEMENT: USING LARYNGEAL ELECTROMYOGRAPHY FOR THE DIAGNOSIS AND TREATMENT OF VOCAL CORD PARALYSIS

MICHAEL C. MUNIN, MD,[1] YOLANDA D. HEMAN-ACKAH, MD, MS,[2,3] CLARK A. ROSEN, MD,[4] LUCIAN SULICA, MD,[5] NICOLE MARONIAN, MD,[6] STEVEN MANDEL, MD,[7] BRIDGET T. CAREY, MD,[8] EARL CRAIG, MD,[9] and GARY GRONSETH, MD[10]

- If prognostic information is required on ultimate vocal fold mobility in a patient with vocal fold paralysis that is >4 weeks and < 6 months
- LEMG may be performed to clarify treatment decisions in a patient with vocal fold immobility that is presumed to be caused by RLN
- The individual parameters of the LEMG study that determine return of vocal fold motion include
  - Active voluntary MUP recruitment
  - Presence of polyphasic MUPs within the first 6 months after injury

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Diagnostic evaluation

Diagnostic tool	Sensitivity, %	Specificity, %	Additional comments
Flexible laryngoscopy	NR	NR	NA
Laryngeal ultrasonography	84[16]	95[16]	Infants (mean age, approximately 1-3 mo) with vocal fold motion impairment
	95[18]	88[18]	Neonates (median age, approximately 15 d) with vocal fold paresis
Laryngeal electromyography	100[19]	92[19]	Children (median age, approximately 12.5 y) with unilateral vocal fold paralysis
Magnetic resonance imaging	67[20]	86[20]	Children and adult patients (age range, 1-78 y) with peripheral nerve pathologic findings
Computed tomography	NR	NR	NA

Ridgway et al. 2021

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Prognosis

- Largely dependent on the cause of the paralysis and the presence and nature of muscle reinnervation
- Spontaneous recovery in children usually occurs around 6 to 12 months after injury

Outcome	Value (N = 404)
Resolved, No. (%)[a]	113 (28.0)
Ongoing, No. (%)	249 (61.6)
1 Evaluation/unclear records, No. (%)	42 (10.4)
Time to resolution, mo[a]	
Mean	7.9
Median (range)	4.3 (0.4-38.7)
Clinical resolution, proportion (%)[b]	72/249 (28.9)
Follow-up, mo	
Mean	33.2
Median (range)	26.0 (0.6-120.5)
Total group with no symptoms, No. (%)	185 (45.8)

[a] Resolution evidenced by laryngoscopy.

[b] Patients in the ongoing group who were asymptomatic at last follow-up.

Jabbour et al. 2014

Prognosis

Variable	Resolved, Proportion (%)	Time to Resolution, mo	
		Mean	Median (Range)
Immobility			
Left	72/270 (27)	7.2	3.8 (0.5-38.7)
Right	8/32 (25)	6.3	3.8 (0.4-22.9)
Bilateral	33/102 (32)	11.6	7.5 (0.7-38.1)
Etiology			
Cardiac	68/278 (24)	6.3	3.9 (0.5-38.7)
Idiopathic	34/85 (40)	11.1	5.9 (0.7-34.2)
Neurologic	8/30 (27)	9.9	2.3 (0.4-38.1)
Mixed	0/5 (0)	...	...
Miscellaneous	3/6 (50)	5.8	3.3 (1.6-12.6)

Jabbour et al. 2014

Outline

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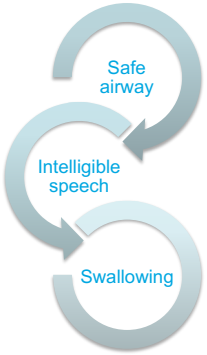
Management

- Factors to be considered
  - Etiology of the paralysis
  - Prognosis
  - Unilateral or Bilateral
  - Severity of symptoms
  - Associated conditions



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Unilateral VFP

- **Speech therapy** is advocated as the first-line treatment
  - Limited reports on efficacy of speech therapy in the pediatric population
- **Injection laryngoplasty** provides an immediate and temporary solution to improve glottic closure
  - May need multiple periodic injections

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Injection laryngoplasty

- Prevent the risk of chronic aspiration and feeding tubes
- Different types injectables
  - Gelfoam
  - Collagen
  - Calcium Hydroxyapatite
  - Carboxymethylcellulose
  - Autologous fat

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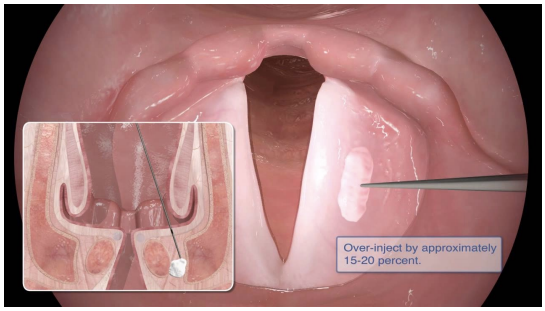
Injection laryngoplasty



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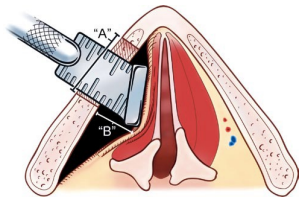
Laryngeal framework surgery

- More permanent solution in patients with poor VF mobility prognosis
- Medialization thyroplasty vs. arytenoid adduction



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Laryngeal reinnervation

- Offers a permanent solution
  - No foreign body implant
  - Consistent outcomes

→ Helps medialize the the vocal fold
→ Prevents atrophy of laryngeal muscles
→ Improves the tone of the affected VF

- Ansa cervicalis to RLN *(Crumley 1991)*
- NMP implantation into adductor laryngeal muscles *(Goding 1991)*

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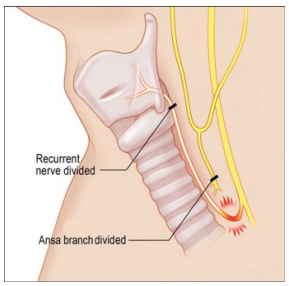
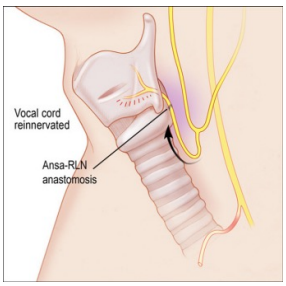
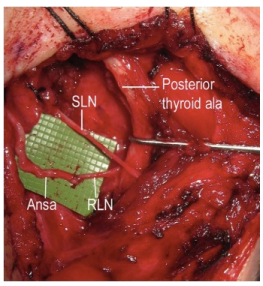
Laryngeal reinnervation







Chhetri et al. 2012

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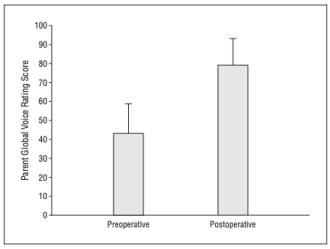
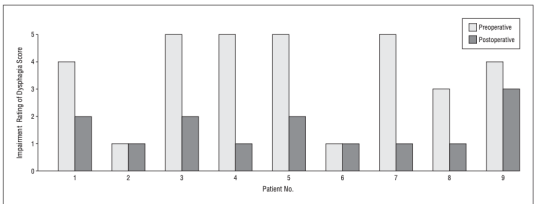
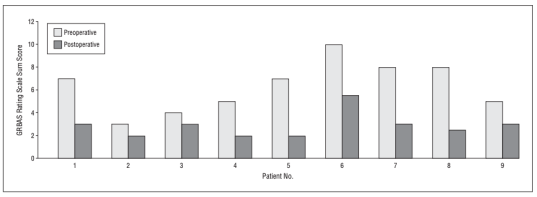
Laryngeal Reinnervation for Paralytic Dysphonia in Children Younger Than 10 Years

Marshall E. Smith, MD; Nelson Roy, PhD, CCC-SLP; Dan Houtz, MA, CCC-SLP







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Bilateral vocal fold paralysis

- The primary goal is to relieve airway obstruction
- In earlier studies → tracheostomy was the standard of care
- Prior to decision
  - Evaluate the cause of the VFP
  - Assess the patient for sleep apnea, lung disease of prematurity, GER
  - Assess neurologic status and swallowing
  - Severity of airway obstruction and work of breathing

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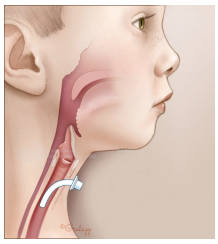
Tracheostomy

- A safe surgical step for BVFP
- Potentially reversible procedure
  - Maintains a stable airway
  - Possible spontaneous recovery
- Repeat examinations to detect return of function and plan future interventions
- ~ 50% of patients needing a tracheostomy require it to be in place for > 3 years *(Murty 1994)*



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Tracheostomy

- Speech acquired by means of a speaking valve or by covering the tracheotomy tube with the chin
- Removal of the tracheostomy is often desirable before children start attending school
- Parents should understand the trade-offs involved when opting for surgical intervention
  - Sacrifice voice
  - Sacrifice swallowing

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Static procedures

- Improve airway patency by enlarging the glottic aperture
  - Excising laryngeal tissue
  - Fixation techniques
  - Combination
- Woodman procedure in 1946
  - 20-40% decannulation failure with external approaches *(Lim 1985, Ossoff et al. 1990)*
- Thornell introduced endoscopic arytenoidectomy in 1952

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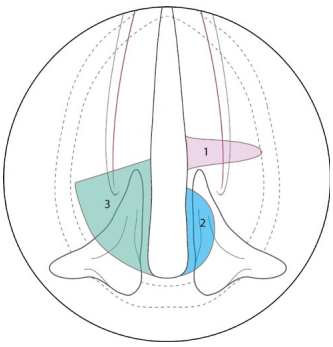
Static procedures

- Development of the CO2 laser provided new possibilities
  - Posterior cordotomy
  - Arytenoidectomy
- Improved outcomes and decannulation rate



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Laser cordotomy for the treatment of bilateral vocal cord paralysis in infants

Aude Lagier, Richard Nicollas*, Mélanie Sanjuan, Lafont Benoit, Jean-Michel Triglia

International Journal of Pediatric Otorhinolaryngology

- 11 patients < 2 years old with bilateral VFP
  - 4 needed a tracheostomy
- Laser posterior partial cordotomy
  - Decannulation after 1 session (n=2)
  - Avoided tracheostomy (n =5)
  - Repeat cordotomy followed by decannulation (n=2)
- Need for second procedure due to scarring and granulation

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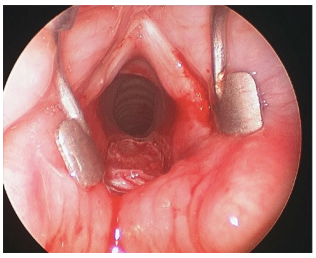
Posterior cricoid split with rib graft

- Posterior enlargement of the interarytenoid space
  - Laryngofissure vs endoscopic
- 28.6% decannulation with endoscopic technique *(Inglis et al. 2017)*
- Risk of dysphagia and tracheostomy
- Variation with AP cricoid split



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Laryngeal chemodenervation

Airway Augmentation and Maintenance Through Laryngeal Chemodenervation in Children With Impaired Vocal Fold Mobility

Marshall E. Smith, MD; Albert H. Park, MD; Harlan R. Muntz, MD; Steven D. Gray, MD†

- Botulinum toxin A injections for airway augmentation
  - TA injection more effective than CT muscle
- Treated 10 bilateral VFP pediatric patients
  - Helpful in providing a better airway in 4/10 patients
  - 50% received injections every 6-12 months
- Favorable candidates
  - Marginal airway obstruction attempting decannulation
  - Recurrent exertional stridor following decannulation

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Laryngeal reinnervation

- Considered the ideal form of rehabilitation for bilateral VFP to reanimate the vocal folds
- Goal is to restore both abductor and adductor movements
- No mucosal incision and no disruption of the laryngeal framework
  - Reduced risk of scarring
  - Potential improvement in swallowing and voice outcomes
- Different methods described
  - Phrenic – RLN
  - Phrenic – PCA muscle
  - Omohyoid NMP

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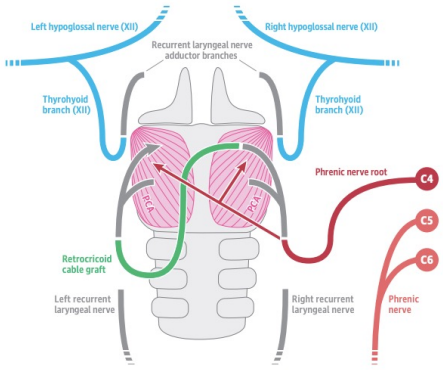
JAMA Otolaryngology–Head & Neck Surgery | Original Investigation

Bilateral Selective Laryngeal Reinnervation for Bilateral Vocal Fold Paralysis in Children

Janet W. Lee, MD; Nicolas Bon-Mardion, MD, PhD; Marshall E. Smith, MD; Jean-Paul Marie, MD, PhD

- All 8 patients decannulated (11.2 months)
- Follow-up period > 5 years

- Voice quality (GRBAS) improved in 6/8
- No swallowing impairment
- Bilateral inspiratory abduction in 5/8
- Unilateral abduction in 2/8



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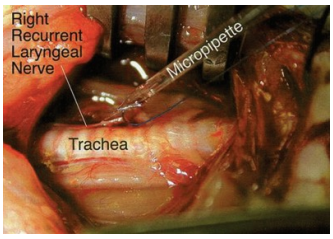
Future directions

- Induced pluripotent stem cells in rats *(Dirja 2016)*
  - Adipose derived stem cells *(Nishio 2016)*
  - Olfactory ectomesenchymal stem cells *(Saïd 2019)*

- Gene therapy *(Bijangi 2016) (Rubin, 2003) (Heavner, 2007)*



Rubin 2001

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Take Home Messages

- Vocal fold immobility is a sign and NOT a diagnosis
- Diagnosis and treatment of pediatric VFP should be carried out with special consideration of postnatal development of the laryngeal framework and special attention to the patient's predominating symptoms and age

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Thank you!

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