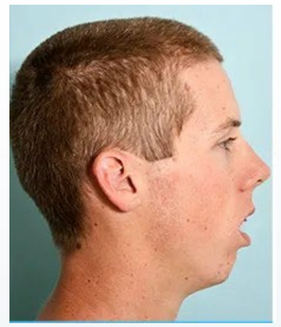
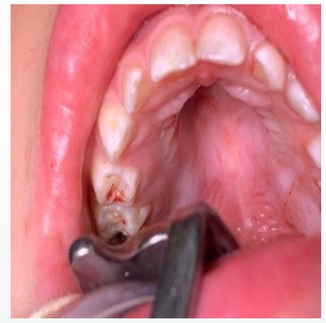
The CGTriad: A Sleep Apnea Phenotype

Becca Maginot, DDS, MS, Garrett Sherman, APRN, FNP-C, Max Beranek, Student, Alena Taing, BA, Nina Yoshpe, MD, FACS, FAAP

Department of Otolaryngology - Head and Neck Surgery, Miller Children's and Women's Hospital Long Beach





"We must see the children and we must treat them."

Prepared for The Society for Ear Nose and Throat Advancement in Children (SENTAC)

Introduction

Dr. Christian Guilleminault (C.G), the father of Sleep Medicine, termed the phrase ***Obstructive Sleep Apnea (OSA)*** and provided the field of medicine with an invaluable tool used to quantify the severity of sleep apnea. This was developed with his colleague, Dr. William Dement, and known as the ***Apnea-Hypopnea Index (AHI)***.

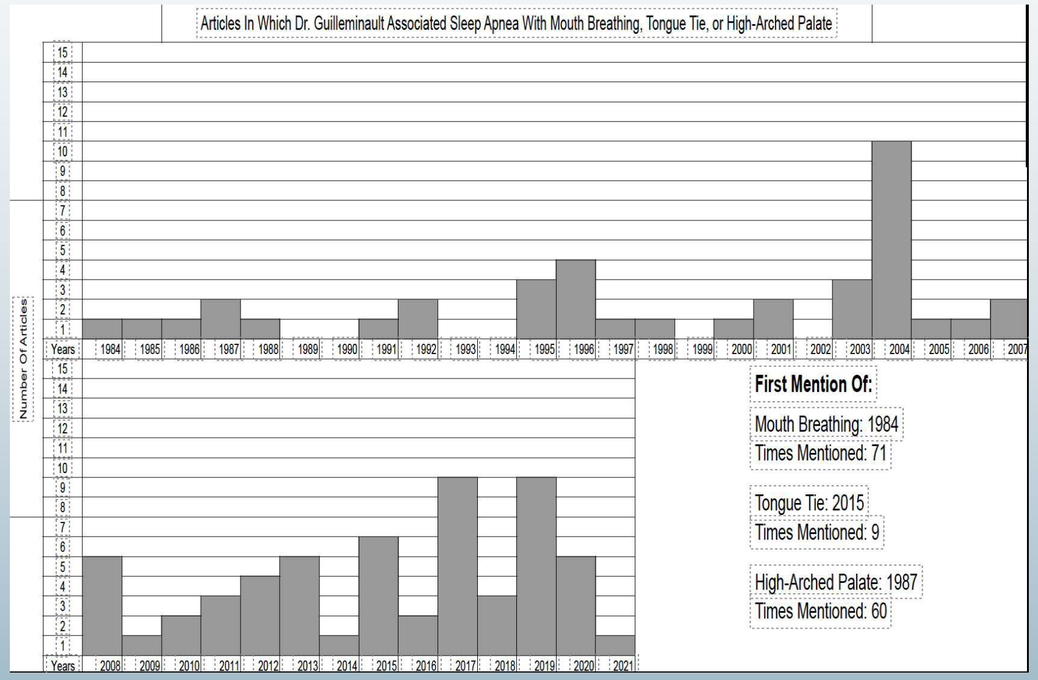
In addition to his tenure at Stanford University, Dr. Guilleminault co-authored hundreds of articles during his illustrious career in the field of Sleep Medicine and co-founded the journal *Sleep* - the Sleep Research Society's official publication. His work is known throughout medicine for collaboration amongst multidisciplinary teams; most notably, the fields of Otolaryngology, Orthodontia and Dentistry.

Objective / Study Design

The purpose of this retrospective study is to honor the work of C.G and identify an easy-to-recognize "triad" of symptoms that contribute to the development of OSA across the lifespan.

Methods

- PubMed research library was referenced for key terms (i.e., Christian Guilleminault, Obstructive sleep apnea, Mouth breathing, Ankyloglossia (Tongue tie), High arched palate)
- 800 articles were identified and reviewed for relevance
- 80 articles met search criteria and were included in this study



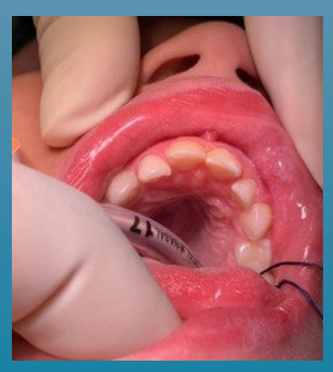
C.G: A Visionary, Collaborator, and Unifier of disciplines

Results

Of the articles that met critera:

- **Mouth breathing** was cited an astounding 71 times and mentioned as far back as 1984
- **High-arched palate** (or dome shaped palate) was referenced second most (n=60) between the years of 1987-2019, and
- **Tongue tie** (ankyloglossia), a newer concept, was referred to in nine separate articles starting in 2015

Through these findings, the ***CG Triad*** was developed with the intent of educating clinicians on physical exam findings that increase the risk of developing OSA.



Conclusion

Dr. Guilleminault's work spanned over 40 years, and his 800+ articles (authored or co-authored), included the terms *mouth breathing, ankyloglossia (tongue tie), and high arched palate* throughout his literature (~10% of the time!)

It is our hope that these three, readily identifiable exam findings, will help clinicians promptly identify those at high risk of OSA allowing for early intervention from the multi-disciplinary team (e.g., ENT, DDS, OMFS, Family and Sleep medicine).

Going forward we hope to investigate the role of genetics (i.e., parental exam findings) in the development of OSA amongst their offspring. This will allow clinicians to be forward thinking and intervene at the youngest age possible.

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Acknowledgments

Thank you Dr. Guilleminault and colleagues for years of diligent work in the field of sleep medicine and paving the way for a multidisciplinary approach to managing obstructive sleep apnea. We'd also like to thank Dr. Audrey Yoon for her rigorous work in Pediatric Dentistry and assisting with study design.

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SCENT SOUTHERN CALIFORNIA EAR, NOSE, AND THROAT

Delayed Sublingual Edema Following Neonatal Mandibular Distraction Osteogenesis

E. Marin Miller, BS[1]; W. Nicholas Jungbauer Jr, BS[1]; William W. Carroll, MD[1]; Phayvanh P. Pecha, MD[1]

[1]Medical University of South Carolina, Department of Otolaryngology - Head and Neck Surgery, Charleston, SC, USA.

Background

- Micrognathia affects 1 in 1500 births and is the precipitating factor in Pierre Robin Sequence (PRS).[1]
- PRS is characterized by a hypoplastic mandible, glossoptosis, airway obstruction, and often a U-shaped cleft palate.[2]
- Mandibular Distraction Osteogenesis (MDO) is a well-accepted surgical treatment for patients with PRS.[3]
- Documented long term complications of MDO include increased sagittal overbite, tooth injury, premature coronoid ossification, inappropriate vector of distraction, dentigerous cyst, and temporomandibular joint injury.[4-8]
- Recently, early in 2022, Morovic et al. described 2 cases of PRS patients post-MDO with hypertrophied sublingual glands requiring surgical excision.[9]

METHODS

Retrospective case series of three patients diagnosed with PRS presented with delayed onset sublingual swelling following uncomplicated neonatal MDO by three different surgeons. The patients presented between August 2019 and September 2021. Retrospective chart review was used to identify patient demographics, chronicity and description of symptoms, and imaging results.

CASES

The three patients presented at 2, 4, and 12 months following MDO for micrognathia secondary to PRS with intermittent sublingual swelling associated with sialorrhea and feeding difficulties. Patient demographics are detailed in **Table 1**. There was no associated recent illness, fevers, or purulent drainage. All three children underwent MRI imaging which demonstrated asymmetric sublingual gland edema **(Figure 1; Table 2**). The edema was located on the left sublingual gland in two children and was bilateral in the third**. Figure 2** demonstrates bilateral sublingual swelling in Patient 2. The symptoms continue to recur 25.5 ± 3.3 months (range, 22.3 – 28.9) postoperatively and all are being managed conservatively.

CASES

Table 1: Patient demographics.

Patient	Sex	Age at distraction (months)	Syndrome	MDO complications	Months post-MDO symptoms developed	Follow up length (months)
1	Female	1.8	None	-	4	22.3
2	Male	0.6	Stickler	Left hardware failure	2	25.4
3	Male	1.8	22q11.2	-	12	28.9

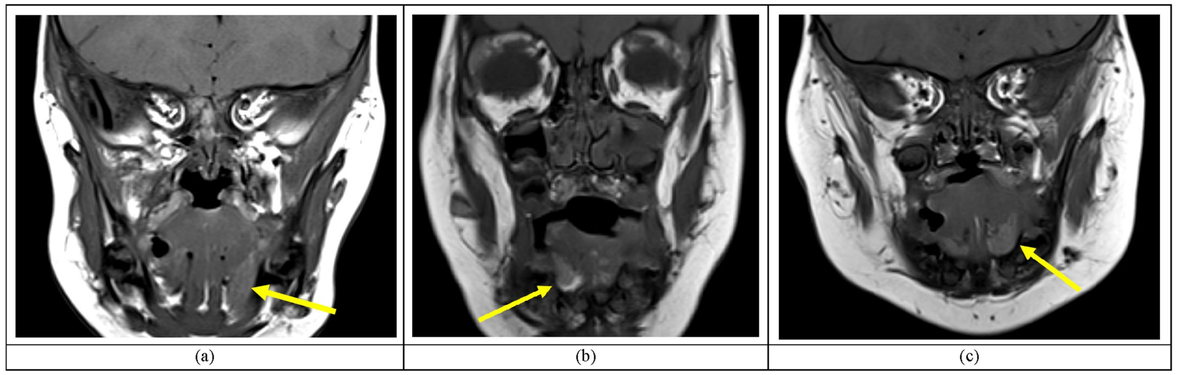

Figure 1: Coronal noncontrast MRIs. (a) Patient 1, demonstrating prominence and mild to normal T1 hyperintensity of the left posterior sublingual gland. (b) Patient 2, demonstrating right anterior sublingual prominence. (c) Patient 3, demonstrating left sublingual gland prominence in Patient 3.

Table 2: Patient Symptoms and Laterality

Patient	Symptoms	Laterality
1	Intermittent sublingual swelling a few times per week with sialorrhea and complete resolution between episodes	Left >Right
2	Bilateral intermittent sublingual swelling, alternating sides, occurring once per week with sialorrhea that interferes with oral intake	Bilateral
3	Intermittent sublingual swelling with sialorrhea	Left

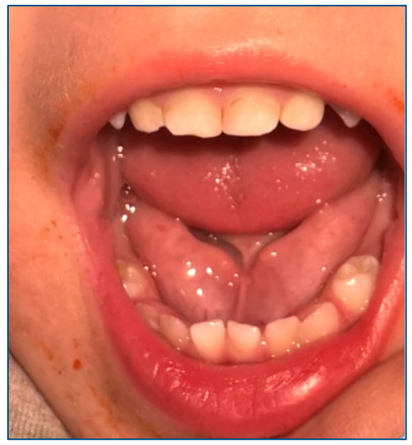

Figure 2: Patient 2 at 24 months demonstrating an episode of bilateral sublingual edema

DISCUSSION

- Conservatively managed fluctuating floor of the mouth swelling has yet to be described as a complication of MDO.
- Differential diagnosis includes: lymphovascular lesion, ranula, sialocele, salivary duct injury, and most likely, sublingual gland hypertrophy.[10-15]
- Unable to confirm diagnosis without biopsy/excision.
- Morovic et. al. described 2 cases of infants with PRS who presented status post MDO with sublingual swelling.[9] Both infants underwent surgical excision which confirmed the diagnosis. In contrast to ours, these cases were described as static lesions. It is possible our patients are also experiencing fluctuating sublingual gland hypertrophy. If our cases are the same in etiology, we present conservative management such as glycopyrrolate and watchful waiting, as appropriate treatment in this rare MDO sequelae.

CONCLUSION

The present case series describes an unusual and possibly underreported sequela following MDO in PRS infants. Chronic delayed onset intermittent sublingual edema is a possible long-term complication following neonatal MDO and further studies should explore the incidence and etiology of this finding.

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Infected Thyroglossal Duct Cyst in a Neonate: A Report of a Rare Case

Brandon Tapasak[1]; Dang-Khoa Nguyen[2]; Santino Cervantes, MD[1,2]
[1]University of Central Florida College of Medicine, Orlando, FL, [2]Nemours Children's Health, Orlando, FL

Introduction

- Most common congenital cervical anomaly and the most common pediatric mass, with a prevalence of 7% in the worldwide population
- Thyroglossal duct usually involutes by the tenth week of gestation, but if it persists, secretion from the epithelial lining causes inflammation and cyst formation [1]
- Differentiating features of a TGDC include a close association with the posterior aspect of the hyoid located midline in the suprahyoid neck or paramidline in the infrahyoid neck [2]
- Most common presenting symptom is an asymptomatic mid-line neck mass, but other symptoms include a sore throat, pain, dysphagia, and hoarseness [3]
- Complications of a TGDC include infection from oropharyngeal organisms and airway obstruction from a rapidly enlarging cyst, but these are most commonly lingual TGDC [4]
- Few reports have characterized the presentation of a TGDC in a patient less than 1 year old and even fewer have diagnosed an infected cyst in a patient this age [5,6]
- We report a case of an infected TGDC in a 3-week-old neonate, while exploring the rarity of this presentation and reviewing management strategies of these patients

Case Report

- A 3-week-old neonate presented to the Emergency Department in respiratory distress with a 5-day history of worsening nasal congestion and upper airway obstruction after upper respiratory infection
- Physical exam was significant for a large, firm, well-circumscribed midline neck mass with pink overlying skin (Figure 1,4)
- An ultrasound revealed a large midline neck mass measuring 3.1×4.2×3.2 cm abutting the hyoid, with internal echogenicity consistent with a thyroglossal duct cyst, causing posterior tongue compression of the airway (Figure 2,3)
- The patient was emergently taken to the operating room for incision and drainage, where she underwent a difficult intubation due to superior-posterior tongue displacement and global supraglottic edema
- Discharged on postoperative day 5 on a course of Augmentin after cultures grew methicillin-sensitive Staphylococcus aureus with anticipation of surgical cyst excision in 6 months
- The patient was followed closely and started on cephalexin prophylaxis to prevent recurrence of infection
- Following definitive resection, the neck mass was confirmed to be a TGDC

Mean Age at Presentation	Standard Deviation of Age at Presentation	Age Range at Presentation	Mean Infection Frequency
6.6 years	±3.7 years	6.5 weeks – 20 years	25.3%

Table 1. Summary of pediatric TGDC literature review.

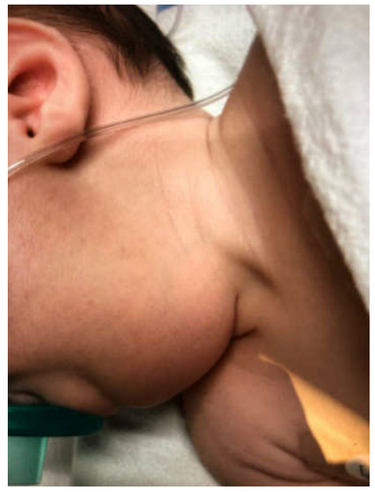

Figure 1. Clinical image of neck mass.

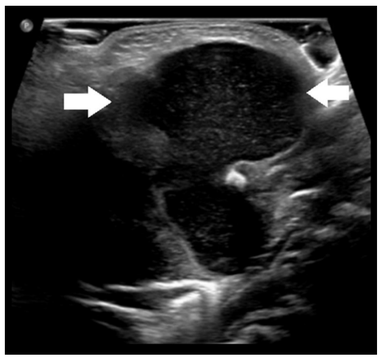

Figure 2. Midline ultrasound neck longitudinal view of neck mass (between arrows).

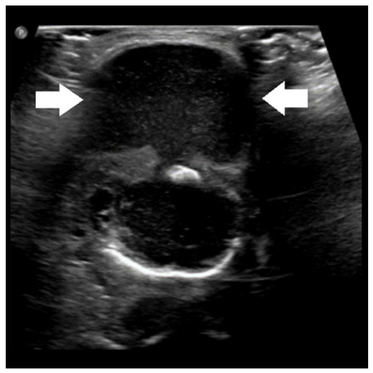

Figure 3. Midline ultrasound neck transverse view of neck mass (between arrows).

Discussion

- Literature review of pediatric TGDC using the electronic database Medline via PubMed (2000-2021) was performed to determine the age of presentation and infection frequency (Table 1)
- TGDC most often presents as an asymptomatic midline cyst in children, although some studies have found a thyroglossal fistula is most common [7,8]
- Older children (5 years old or older) are more likely to present with an infected TGDC [3]
- Incision and drainage are thought to decrease the duration of inflammation, only affect the localized tissues, and lessen the scarring and destruction from the extensive inflammation due to the cyst itself [9,10]
- We support the use of incision and drainage when the airway is at risk of obstruction with Sistrunk procedure later
- Risk factors increasing recurrence are the presence of infection, age less than 2 years old, cyst lobulation, intraoperative rupture, incomplete excision, dermal involvement, and cases with fistulas [1]

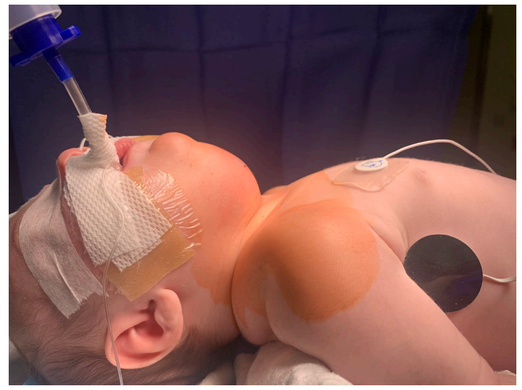

Figure 4. Image of neck mass prior to incision and drainage.

Conclusions

- Although TGDC rarely presents in the neonatal period, clinicians should keep the diagnosis in their differential when evaluating an asymptomatic midline neck mass
- Serious complications such as infection and airway obstruction can occur if not treated promptly
- Although use of only antibiotics has been the traditional treatment regimen for infected TGDC, incision and drainage should also be considered when there is risk of airway obstruction
- While the Sistrunk procedure has become the standard surgical treatment for TGDC, there is an increased risk for recurrence in patients with clear risk factors such as infection and young age, and more management guidance is needed to reduce the risk for these patients

Contact

Brandon Tapasak
University of Central Florida College of Medicine
6850 Lake Nona Blvd., Orlando, FL 32827
btapasak@knights.ucf.edu
321-505-5802

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7: Pediatric Cholesteatoma Associated with Congenital Aural Atresia and Stenosis

Olivia Kalmanson, MD, MS; Christian Francom, MD; Owen Darr, MD; Steven Hamilton, MD

Background

Pediatric cholesteatomas exhibit more aggressive behavior than those of adults. Pediatric-specific data is lacking regarding cholesteatomas associated with congenital aural atresia and stenosis.[1-3]

Objective

Identify presenting features, associated surgical treatment, and report outcomes in pediatric patients with congenital aural atresia (CAA) or stenosis (CAS) and acquired canal cholesteatoma.

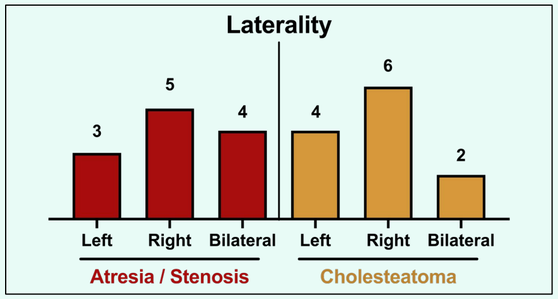
Methods

Retrospective chart review at a single tertiary care children's hospital of all pediatric patients with congenital aural atresia or stenosis with associated cholesteatoma between Jan 1, 2003 – Oct 15, 2018.

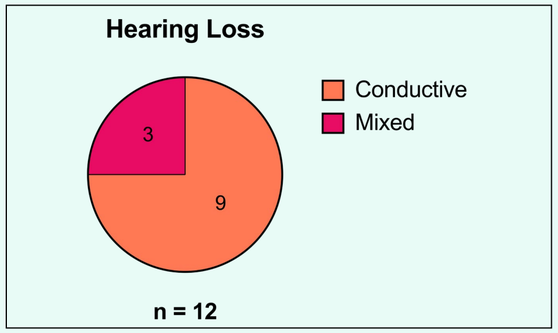
Results

- 12/278 patients with CAA/CAS developed acquired cholesteatoma
- 66% Male / 33% Female
- Average age at first surgery:
  11.3+/-3.7 years

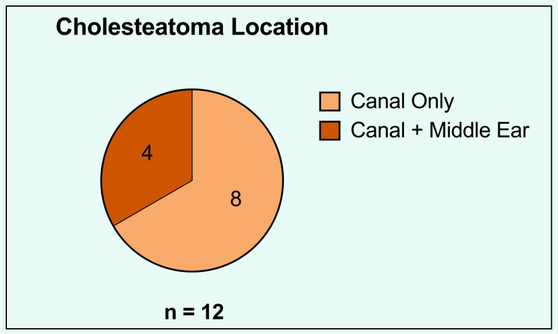
Results



Cholesteatoma development in these patients matched the side of CAA/CAS. Patients with bilateral CAA/CAS developed right, left, or bilateral cholesteatomas.



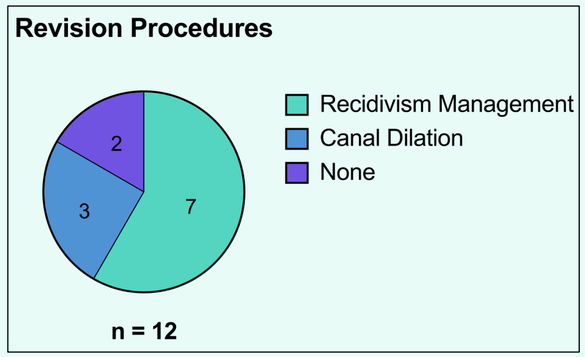
All patients with CAA/CAS and associated canal cholesteatoma exhibited hearing loss, of which 75% was conductive and 25% was mixed.



All patients exhibited canal cholesteatoma, with 33% extending into the middle ear or mastoid cavity. There were no cases of isolated middle ear cholesteatoma in this population.

[1]Kuo CL, Shiao AS, Yung M, Sakagami M, Sudhoff H, Wang CH, Hsu CH, Lien CF. Updates and knowledge gaps in cholesteatoma research. Biomed Res Int. 2015;2015:854024. [2]Preciado DA. Biology of cholesteatoma: special considerations in pediatric patients. Int J Pediatr Otorhinolaryngol. 2012 Mar;76(3):319-21. [3]Dornhoffer JL, Friedman AB, Gluth MB. Management of acquired cholesteatoma in the pediatric population. Curr Opin Otolaryngol Head Neck Surg. 2013 Oct;21(5):440-5.

Results



All patients underwent primary surgery. 10/12 required at least one revision: 7 for recidivism management and 3 for revision canalplasty. The need for revision surgery was not associated with extension of cholesteatoma into the middle ear or mastoid.

Conclusion

Acquired canal cholesteatomas were uncommon in patients with congenital aura atresia or stenosis. However, the need for revision surgery was common among those patients with cholesteatoma, suggesting that early screening is still indicated to prevent morbidity from delayed diagnosis.

Rare Case of Nasal Vault Oncocytic Cystadenoma

Aarti Agarwal MD[1], Kathleen McClain DO[2], Karen Banker PA-C[2], Kudakwashe Chikwava MD[3], Udayan K. Shah MD MBA[1,2,4]

[1] Department of Otolaryngology - Head and Neck Surgery, Sidney Kimmel Medical College, Thomas Jefferson University, Philadelphia, PA, USA
[2] Division of Otolaryngology, Department of Surgery, Nemours Children's Hospital - Delaware, Wilmington, DE, USA
[3] Department of Pathology, Nemours Children's Hospital - Delaware , Wilmington, DE, USA
[4] Department of Pediatrics, Sidney Kimmel Medical College, Thomas Jefferson University, Philadelphia, PA, USA

Introduction

- Oncocytic papillary cystadenoma is a rare benign tumor generally seen in the minor salivary glands. Oncocytes are large, irregularly shaped cells with granular eosinophic cytoplasm that are rich in mitochondria.
- Cysts are generally asymptomatic unless they progress causing obstruction with symptoms related to their anatomic location and proximity to other structures.
- We report a pediatric patient with a painless cyst of the nasal vestibule for which pathology showed an oncocytic cystadenoma.

Case Presentation

- 10 yr old female with a bulge on the left side of her nasal vestibule that increased in size and began to cause discomfort with a visible external abnormality. Was prescribed oral clindamycin in the ED which decreased the tenderness and size of this lesion, but it remained persistent.
- On exam, fullness above the left nasal tip was seen, with a yellow, compressible cyst above the lower lateral cartilage filling the vestibule (Fig 1).
- MRI showed a 1.2 cm x 0.9 cm x 1.2 cm rim-enhancing cyst in the left anterior nasal cavity with bright T2 signal (Fig 2).
- Excision in the OR was accomplished with endoscopic assistance. Lesion grossly was an ovoid tan fluctuant thin-walled cyst with minimal colorless serous fluid. Microscopy revealed a multilocular complex cyst lined by stratified epithelial cells with extensive tubule formation focal squamous metaplasia and rare mucinous cells (Fig 3). Features consistent with seromucinous oncocytic cystadenoma.
- Was evaluated 3 weeks postop with resolution of the external nasal deformity.

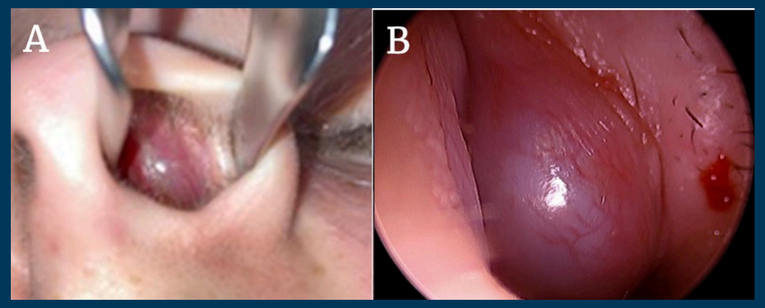


Figure 1: External (A) and internal (B) nasal exam of lesion.

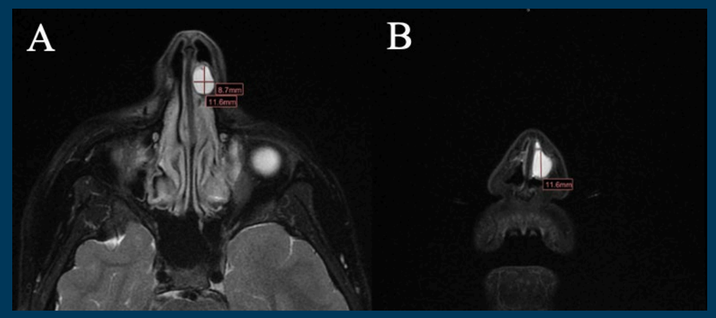


Figure 2: T2-weighted Magnetic Resonance Imaging (MRI) with lesion seen as hyperintense in the left nasal vault. A) Axial Cut B) Coronal Cut

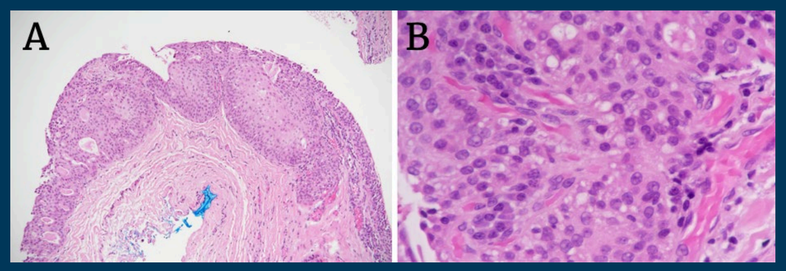


Figure 3. Low and high-power photomicrographs showing the cyst lininig stratified epithelial cells with extensive tubule formation and focal squamous metaplasia (3A) as well as rare mucinous cells (3B) in the left nasal vault. A) Axial cut B) Coronal cut

Conclusion

- The WHO defines oncocytic cystadenomas as cystic lesions lined by oncocytic epithelium with occasional luminal papillary projections. These tumors are lined by an epithelial bilayer which is comprised of inner columnar eosinophilic or oncocytic cells surrounded by smaller basal cells.
- Oncocytic Cystadenomas are benign tumors of the salivary glands that only cause symptoms related to their anatomic location and proximity to other structures.
- Generally, they are treated with surgical excision with rare recurrence. Our patient went to the operating for excision and on short-term follow-up visit did not have any recurrence.
- While rarely reported in the pediatric patient, given the presence of minor mucinous gland tissue in this location, it is feasible that oncocytic cystadenomas could occur anywhere along the upper aerodigestive tract.
- This case demonstrates the need to consider a broad range of surgical pathologies in anterior nasal vault tumors, and supports the value of nasal endoscopic approaches for excision of these lesions.

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DIAGNOSIS & MANAGEMENT OF RIGA-FEDE DISEASE: A CASE SERIES

Catherine G. Nguyen, MS; Emily E. Wikner, MD; Thomas Q. Gallagher, DO; Laurie A. Birsch, DDS; David H. Darrow, MD, DDS

EVMS Department of Otolaryngology

Norfolk, VA

Children's Hospital of The King's Daughters

EVMS Eastern Virginia Medical School

Background

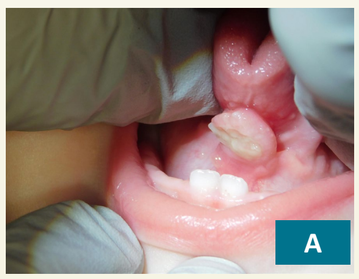
Mucosal lesions of the oral cavity are common in children. However, the sudden appearance of ulcerated or hyperplastic mucosa on the tongue or floor of the mouth of a nursing or developmentally-delayed child should alert the otolaryngologist to the diagnosis of **Riga-Fede disease (RFD)**.[1,2] This disorder is characterized by mucosal trauma resulting from the repetitive protrusion of the tongue against the incisors. It is often described histologically as **"traumatic ulcerative granuloma with stromal eosinophilia" (TUGSE)**.[3] Although the lesion begins as a benign ulceration, it may progress into a mass of hyperplastic, painful, and/or bleeding tissue that mimics malignancy.

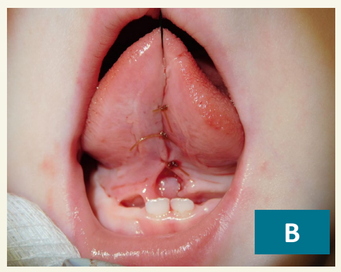
Successful management of RFD depends on correct identification of the disorder and coordinated intervention between the pediatric otolaryngologist and the pediatric dentist. We present a series of cases that demonstrates the diverse presentations and management options for RFD.

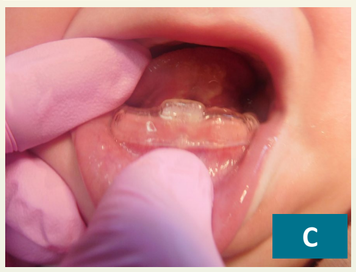
Case Summaries

Case 1 (Figures A-D)

An 8 m/o M presented with a lesion of the ventral tongue and floor of the mouth (A) that first appeared 2 months prior, coinciding with eruption of his lower incisors. Mother reported that the area had been mildly painful while nursing, with occasional bleeding, but overall had not interfered with nursing. Surgical excision was performed (B) in coordination with his dentist who fabricated an appliance (C) to protect the soft tissue from further irritation. At follow-up, despite some compliance issues, there was no recurrence of the ulceration or the mass (D).







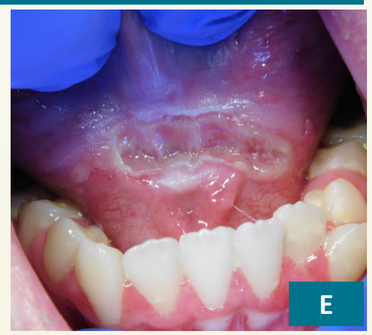
Summary of Cases

	Mechanism of trauma	Physical exam findings	Treatment	Follow up (months)
Patient 1	Tongue movement over newly erupted teeth while nursing	1.5 X 1 cm lesion in midline at junction of floor of mouth with ventral tongue	Surgical excision, protective appliance	3 No recurrence
Patient 2	Habitual tongue thrust due to developmental delay (cerebral palsy)	Chronic 1.5-2 cm ulcer with raised borders in anterior floor of mouth	Surgical excision, occlusal adjustment, protective appliance	98 Multiple recurrences
Patient 3	Recent changes in feeding mechanics (sippy cup)	1 cm mass of ventral tongue with central ulceration and surrounding mucosal hyperplasia	Surgical excision; referral to pediatric dentist; parents chose observation	6 No recurrence
Patient 4	Habitual biting/sucking due to developmental delay (Noonan syndrome)	Mild whitish mucosal ulceration of ventral tongue without erythema	Referral to pediatric dentist; parents chose observation	3 No change
Patient 5	Repetitive rubbing against upper teeth	2 X 2 X 3 cm firm, nontender, irregular mass of anterior left tongue without	Surgical excision	3 No

Case Summaries (cont.)

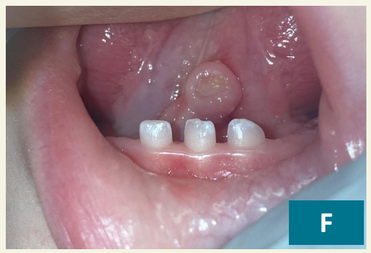
Case 2 (Figure E)

A 10 y/o M with history of cerebral palsy and developmental delay presented with recurrent ulceration thought to be aphthous but limited to the floor of mouth (E). He was referred for a recurrence that did not respond to his typical course of medical therapy. Surgical excision was performed and pathology revealed only chronic inflammation. Although the lesion healed completely, there were multiple recurrences over the next several years. At an office visit, he was noted to thrust his tongue repeatedly and a diagnosis of RFD was made. Despite adjustment of the incisal edges of the lower front teeth and fabrication of a protective splint, his ulcers continue to recur.



Case 3 (Figure F)

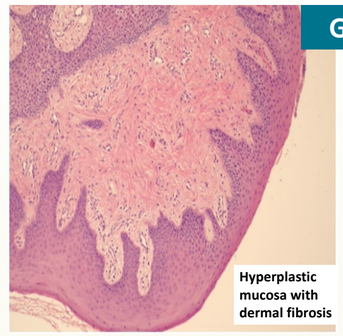
A previously healthy 15 m/o M presented with a 3-month history of a "canker sore" under his tongue. Mother reported no preceding trauma or feeding issues, but patient had started using a new, firmer sippy cup. The lesion gradually grew and developed central discoloration and heaped up granulation tissue (F). Excision was performed and final pathology revealed pyogenic granuloma. The ulcer recurred post-op and referral was made to a pediatric dentist for fabrication of a protective appliance.

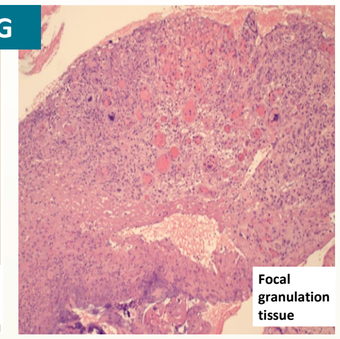


Case Summaries (cont.)

Case 5 (Figure G)

A previously healthy 9 y/o F presented with a slowly growing dorsal tongue mass that was first noticed 4 months prior. No history of systemic symptoms or trauma. Ultrasound performed prior to referral was nondiagnostic but suggested a fluid-filled mass. MRI at our facility suggested possible lymphovascular malformation. Excisional biopsy revealed a final pathology report of TUGSE (G). Mother reported, retrospectively, noting a sore at the tip of patient's tongue that she had been rubbing against her upper incisors.





Discussion

Five patients presenting with ulcerations or masses of the tongue or floor of the mouth were diagnosed with RFD over a nine-year period. In two of the children, a change in feeding mechanics over 2 and 5 months, respectively, preceded appearance of the mass. Three of the children manifested oral habits resulting in repeated trauma to the mucosa.

Once identified, **RFD** is best managed by removing the pathology and eliminating the etiology. **Surgical excision** restores normal mucosal appearance and function. Recurrence is avoided by reducing dental trauma through **alteration of feeding mechanics**, **occlusal adjustment**, **use of protective appliances**, **or dental extraction**.[1-3]

Interdisciplinary care between the **otolaryngologist** and **pediatric dentist** is paramount for optimal functional and anatomic outcomes in RFD/TUGSE.

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Coblation as an Effective Tool for Treating Subglottic Cysts and Hemangiomas in Pediatric Patients

Ana Khatiashvili[1], Lara Reichert[2]
1 Albany Medical College, 47 New Scotland Ave, Albany, NY, USA
2 Division of Otolaryngology, Albany Medical College, Albany, NY, USA

Background

Subglottic cysts and hemangiomas are rare but life-threatening conditions in pediatric patients. Subglottic cysts are generally associated with premature infants with a history of prolonged endotracheal intubation[1], while subglottic hemangiomas are congenital vascular lesions that grow rapidly and are uncommon head and neck tumors in pediatric patients. Both conditions can present with generalized respiratory symptoms such as stridor[2]. Early diagnoses and treatment are crucial in avoiding airway compromise[3].

Methods

Medical records of 3 pediatric patients from June 2021 to June 2022 were reviewed. All cases involved the use of coblation as a treatment for subglottic cysts or hemangiomas. Patients' presentation, surgical intervention(s), and postoperative course were analyzed.

Discussion

Subglottic cysts (SGC) are hypothesized to be formed by the damage inflicted by an endotracheal tube in preterm infants.[4] Clinical presentations are gradual and most often include stenosis, stridor, failure to thrive, and possibly respiratory failure. Diagnosis is made by performing laryngoscopy[5] and standard treatment is the use of CO2 laser. [6] Recurrence of SGC is frequent and observed in 25-70% of patients.[7]

Infantile subglottic hemangiomas (SGH) are rare benign congenital tumors. They often present with stridor, along with symptoms of cough and cyanosis. [8] When treating these subglottic conditions coblation should be considered as an effective tool.

Coblation, uses low-heat polar radiofrequency energy conducted through a plasma field and can accurately reduce tissue volume.[9] Notably, coblator operates at

Case Presentations

Case #1

Patient is a 4-month-old male with a history of 24-week premature gestation. He presented with an inspiratory stridor, subcostal retractions and failure to gain weight properly. Direct laryngoscopy and bronchoscopy (L&B) demonstrated near complete obstruction by subglottic cysts (Fig. 1) which were removed by coblation (Fig. 2). Subsequent L&B demonstrated excellent healing and only a small amount of early stenosis that was balloon dilated.

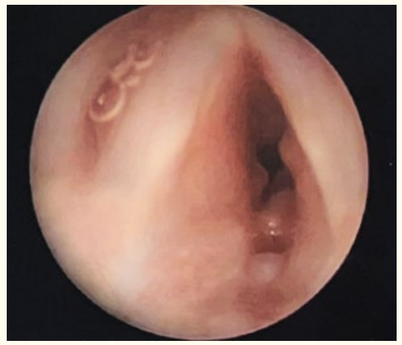

Fig. 1. Multiple subglottic cysts with near complete obstruction.

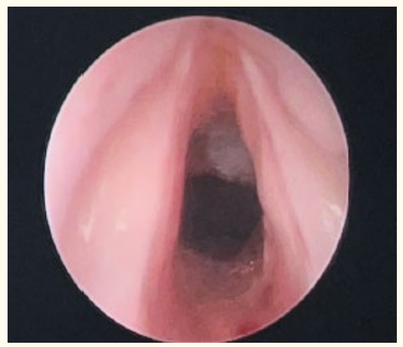

Fig. 2. Subglottis immediately after coblation.

Case #2

Patient is a 5-month-old male with a history of 28-week premature gestation who presented with difficulty breathing and stridor. Patient was reported to suffer severe subcostal retractions and cyanosis in his lips, requiring supplemental oxygen. L&B demonstrated multiple suglottic cysts obstructing 95% of the airway (Fig. 3). Coblation was used to reduce the cysts after which a normal sized airway was achieved

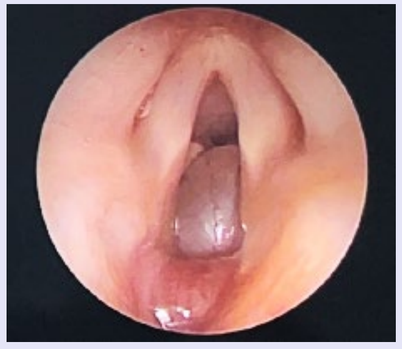

Fig. 3. Large subglottic cyst obstructing 95% of the airway.

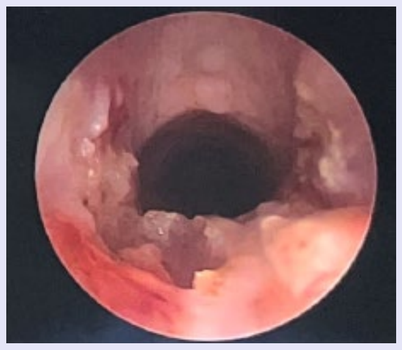

Fig. 4. Normal sized airway established after coblation.

Case #3

Patient is an 8-month-old female with a history of multiple hemangiomas and respiratory symptoms. Direct L&B demonstrated a large pharyngeal hemangioma (Fig. 5), with several more exophytic lesions along the tracheal wall. Hemangioma was removed by coblation (Fig. 6), with good reduction in bulk and no bleeding. Balloon dilation was used in order to improve patency.

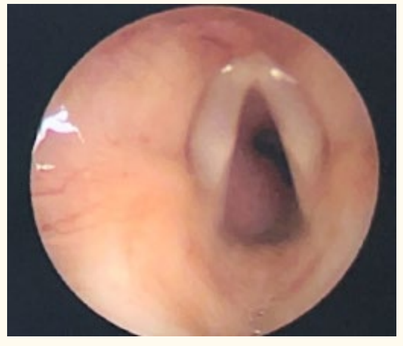

Fig. 5. Large Hemangioma obstructing 75% of the airway.

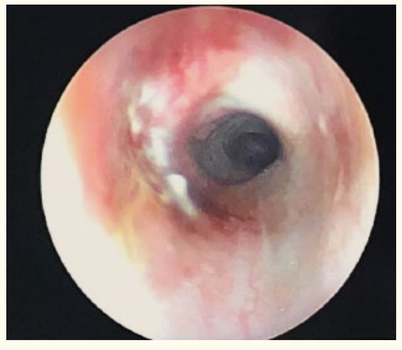

Fig. 6. Patent airway achieved with the use of coblation.

Discussion Cont.

which is considerably lower than 400-600 °C achieved by cautery and CO2 lasers. This does not only minimize damage to the surrounding tissue, reduce bleeding and provide potential for reduced scar formation, but almost completely eliminates the risk of airway fire.[10]

All three patients in our case study were treated with coblation. After the procedure there was a significant improvement in their breathing and feeding without any signs of stridor. Subsequent endoscopies have demonstrated slight stenosis at the treatment sight in only one of the patients, which was successfully balloon dilated. Overall, all three patients showed excellent responses to coblation with no recurrence of cysts and hemangiomas.

Summary

All 3 cases responded well to coblation. Subsequent endoscopies have shown excellent healing with no additional cyst or hemangioma formation with minimal stenosis.

The results of the study suggest that coblation may be a safer and more effective alternative to cautery and CO2 lasers for the treatment of pediatric subglottic cysts and hemangiomas.

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Pediatric Single Sided Deafness (SSD) at Nemours Children's Hospital, Delaware

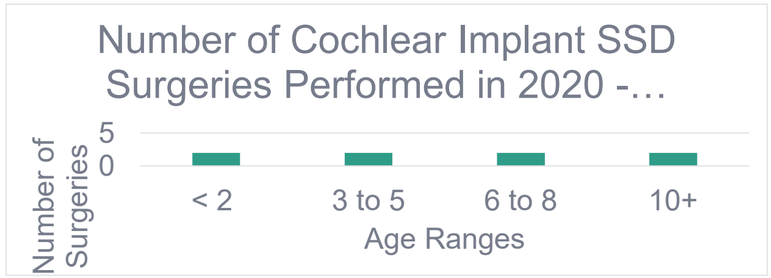
Introduction

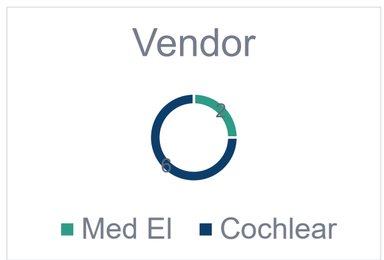
Cochlear implant surgery provides a way for pediatric patients with severe-to-profound single sided deafness (SSD) to access binaural hearing. The purpose of this investigation was to review outcomes for children with SSD implanted at the Nemours Children's Hospital in 2020 and 2021. Our program consists of Audiology, Otolaryngology, Psychology, Therapy Services, and Social Work. We collaborate with patients' school districts and other various service providers to help achieve improved auditory benefit and outcomes.

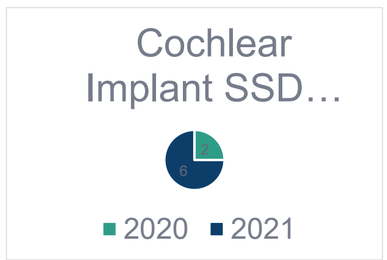
Methods

A retrospective case review from a tertiary pediatric (<18 years) hospital for children with SSD was completed to assess outcomes with pre and post cochlear implant surgery. Eight pediatric patients were implanted with either a Cochlear Americas or MED EL device. Assessment questionnaires (i.e, LittleEars, PEACH, or APAHB), audiometric thresholds, word recognition, and sentence in noise testing were attempted pre and post cochlear implant surgery.

Demographics







Results

Patient	Etiology	Age @ Implantation	Side	Vendor	Pre-PTA	Post-PTA	Pre-SRT	Post-SRT	Pre-Q	Post-Q	Pre Word Recognition	Post Word Recognition	Pre Sent Recognition	Post Sent Rec	Plan
1	CMV	1y9m	L	Med El	82 dBeHL	47dBHL	N/A	40dBHL	LittleEars Met (33)	N/A	CNT	CNT	CNT	CNT	CPA LittleEars PSI WIPI
2	Unknown Sudden Onset	14y7m	R	Cochlear	72dBHL	28dBHL	NR @ 100 dBHL	30 dBHL	DNT	Loss to f/u	68%	Loss to f/u	11%	Loss to f/u	APHAB CNC BKB-SIN
3	CMV	1y1m	R	Cochlear	83dBeHL	CNT	CNT	CNT *possible 70 dBHL SAT	LittleEars Met (11)	N/A	CNT	CNT	CNT	CNT	CPA Cortical Thresholds LittleEars PSI WIPI
4	EVA	5y8m	L	Cochlear	75dBHL	42dBHL	85dBHL	45dBHL	DNT	DNT	0%	48%	0%	DNT	PEACH CNC BKB-SIN
5	Unknown Progressive	4y4m	R	Cochlear	NR @ 110 dBHL	32dBHL	CNT	25dBHL	DNT	DNT	0%	92%	CNT	DNT	PEACH PBK BKB-SIN
6	EVA	8y0m	R	Med El	52 dBHL	40dBHL	55 dBHL	35dBHL	PEACH Typical (95.45)	DNT	52%	100%	12.7 SNR Moderate SNR Loss	DNT	PEACH CNC BKB-SIN
7	TBI	12y9m	L	Cochlear	100dBHL	28dBHL	95dBHL	20dBHL	DNT	DNT	16%	60%	6%	91%	APHAB CNC BKB-SIN
8	Labyrinthitis	7y2m	R	Cochlear	83dBHL	20dBHL	85dBHL	20dBHL	DNT	DNT	0%	84%	DNT	DNT	PEACH CNC BKB-SIN

Outcome Measure Questionnaires

- **LittleEars** is designed to screen the auditory development in children with normal hearing and in children with hearing loss who have received a cochlear implant(s) (CI) or hearing aid(s) (HA). It covers auditory development in the first 2 years after a CI or HA fitting (up to 2 years of hearing age) or in hearing children up to 2 years of age.
- **Parents' Evaluation of Aural/Oral Performance of Children (PEACH)** is a questionnaire designed to record how your child is hearing and communicating with others when using his/her hearing aids and/or cochlear implant.
- **Abbreviated Profile of Hearing Aid Benefit (APHAB)** is a 24-item self-assessment inventory in which patients report the amount of trouble they are having with communication or noises in various everyday situations. Benefit is calculated by comparing the patient's reported difficulty in the unaided condition with their amount of difficulty when using amplification. The APHAB produces scores for 4 subscales: Ease of Communication (EC), Reverberation (RV), Background Noise (BN), and Aversiveness (AV)

Conclusions

Cochlear implant surgery is a viable treatment option for pediatric SSD. Open set speech and improvement in background noise can be achieved. Thorough counseling prior to surgery and throughout follow up post-surgery appointments on expectations is vital to achieving successful outcomes to avoid loss to follow-up or patient dissatisfaction.

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Improving Audiology and Otolaryngology Post-Tube Follow-Up Appointments in the Delaware Valley

K Banker, P Barth, B Burke, S Francese, M Morrow, S Oliet, M Stone, J Verhoff

NEMOURS CHILDREN'S HEALTH

Background / Description

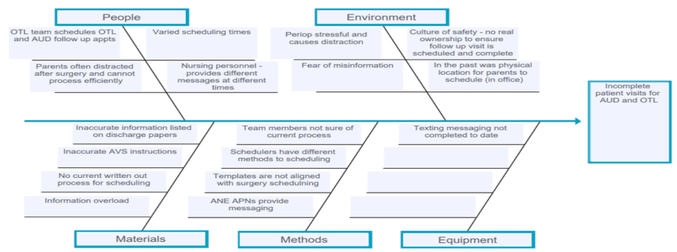
- Most common otolaryngology (ORL) procedure is bilateral myringotomy tube (BMT) or ear tube placement
- Standard of care is a follow up audiogram with Audiology (AUD) within 6 weeks and follow up with ENT at 6 months following tube placement
- Patients lost to follow up may experience adverse events including hearing loss and unneeded oral antibiotics
- We have an opportunity to improve follow-up care for patients post ear tube placement in the Delaware Valley

Aim

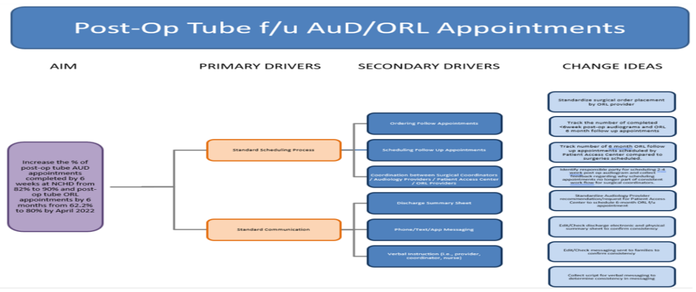
- Increase completed Audiology (AUD) six-week post-tube appointments from 72% to 92% within 6 months
- Increase scheduled Otolaryngology (ORL) six-month post-tube appointments from 63% to 80% within 6 months

Design / Strategy

- A fishbone diagram was used to identify barriers for success in the current postoperative scheduling process



- A driver diagram was created to identify steps within the pre- and post-surgical processes that would help increase the percentage of postoperative appointments scheduled and completed



- Outcome, process, and balance measures were formulated to determine the success of interventions

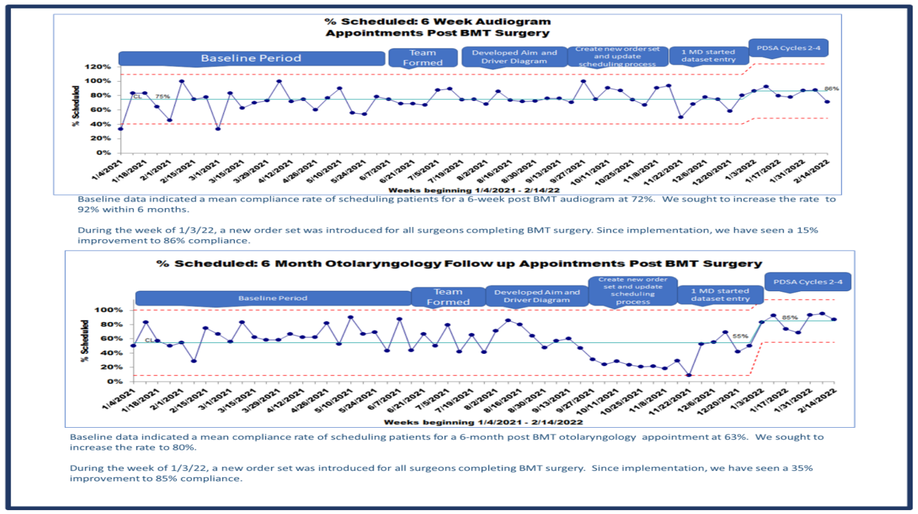
OUTCOME MEASURES:
- % of patients who have completed an audiogram with audiology 6 weeks after (BMT) surgery
- % of patients who have scheduled a six-month follow-up appointment with otolaryngology following BMT surgery

PROCESS MEASURES:
- % BMT patients with scheduling work queue orders for AUD and ORL appointments
- % work queue orders contacted by Access scheduling team

BALANCE MEASURES:
- Template utilization for physicians, physician assistants, and audiologists
- Impact to Access Center

Changes Made

- 12/8/2021- Post operative instructions modified to reflect appropriate follow-up audiogram scheduled within 6 weeks and ORL office follow-up within 6 months.
- 12/16/2021- PDSA cycle #1 initiated- new manually entered post-op orders directing follow-up appointments to be scheduled by the Access Center (Limited to 1 surgeon)
- 1/7/2022- PDSA cycle #2 changed manual order entry to a postoperative order set expanded live for all ORL surgeons following BMT procedures



Lessons Learned

- Different departments have different workflows and resource allocation – important to bring everyone affected into the conversation for best outcomes
- Creation of quality control measures to ensure that follow-up appointments are being completed appropriately and in a timely manner
- Sometimes a complex problem can be addressed with a simple solution!

Challenges / Barriers

- Difficulty obtaining accurate baseline data due to halt in elective procedures and office appointments due to COVID 19
- Shifting scheduling responsibility to Access Center caused significantly increased workload for outbound scheduling team
- Data collection challenging due to difficulty in pulling data and keeping up-to-date because of delay between surgery and scheduling of appointment

Next Steps

- Expand postoperative orders and scheduling process to other surgeries requiring follow-up
- Continue conversations with scheduling groups to address any unintended consequences
- Periodic data pulls to ensure that best practices are being achieved

Concluding Materials

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Thank you to our mentor, Judith Guidash!

Mission:ZERO

Repair of septal perforation and saddle nose deformity in an adolescent with psychiatric comorbidities

Annica C. Eells, MD[1]; Brittany E. Howard, MD[1]; Stephen F. Bansberg, MD[1]

[1]Mayo Clinic Department of Otolaryngology, Division of Facial Plastic Surgery; Phoenix, AZ, USA

BACKGROUND

Septal perforation due to intranasal button battery placement is a well-documented occurrence in the pediatric population. Crusting, epistaxis, congestion/obstruction, and whistling on inspiration are symptoms commonly associated with perforations. Perforations in the pediatric population may affect nasal development, resulting in loss of dorsal support, columellar retraction, and poor tip support

We present a case of an adolescent with a large septal perforation and aesthetic deformity as a consequence of button battery damage sustained during childhood. Perforation repair and functional aesthetic rhinoplasty were planned as a two-stage procedure. The patient's reconstructive course was also complicated by acute suicidality and ongoing depression.

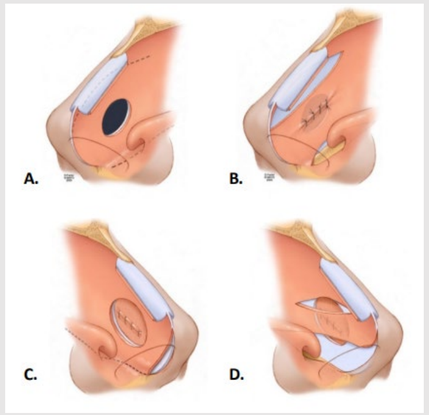

Figure 1

Image showing septal perforation repair. A. Dashed lines represent incisions to develop left-sided superior and inferior bipedicled flaps. B. Superior and inferior flaps advanced and sutured. C. Dashed line represents extension of right hemitransfixion incision to create an inferior bipedicled flap. D. Right inferior flap advanced to oppose left suture line.

CASE

A 16-year-old male presented initially to author (B.E.H.) for saddle nose deformity. He had a history of button battery placement at age 4 which led to septal perforation and subsequent progressive nasal deformity (Figure 3).

The patient was subsequently referred to the senior author (S.F.B.) for evaluation regarding perforation repair. The perforation measured 1.8 cm in length and 1.5 cm in height. The anterior-superior perforation margin was 0.8 cm from the internal valve angle. The perforation was closed through an endonasal approach utilizing bilateral, longitudinally-oriented mucosal flaps supported with an autogenous temporalis fascia interposition graft (Figure 1). Complete closure was achieved.

Reconstructive septorhinoplasty was planned nine months after the perforation repair; however, the patient experienced suicidal ideation that required hospitalization and deferment of surgery. The scheduling of reconstructive surgery was delayed until the patient committed to establishing mental health care. When this was achieved, reconstructive surgery was successfully performed 16 months following the perforation repair (Figure 2). The patient's postoperative course was uneventful.

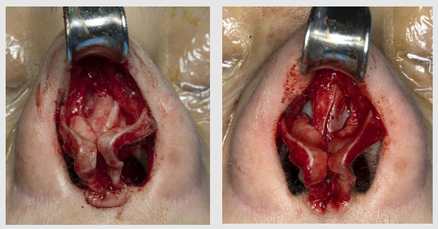

Figure 2

Intraoperative pre- and post-reconstruction of nasal septal deformity. Saddle nose and internal nasal valve deformity were corrected with spreader grafts and upper lateral cartilage re-suspension. External nasal valve and nasal tip corrected with caudal extension graft, caudal septum re-suspension, and rim grafts. Costal cartilage graft was taken and utilized for the aforementioned purposes.

RESULTS

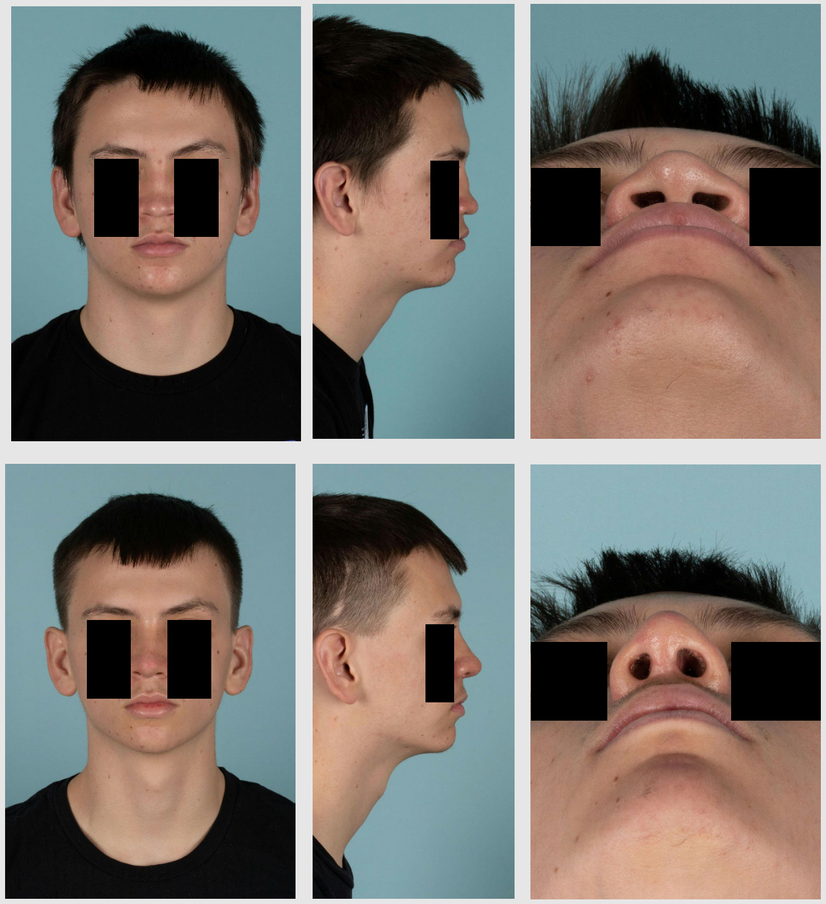

Figure 3

Preoperative photographs, followed by postoperative result at 2 weeks. Preoperative projectometer measurement showed a projection of 22 mm and rotation 19 mm with postoperative projection of 26 mm and rotation 21 mm.

CONCLUSIONS

The literature regarding pediatric septoplasty and perforation repair has not demonstrated a significant contraindication to early repair if surgery proceeds conservatively[1]. High perforation closure rates can be achieved in the adolescent population with repairs utilizing nasal mucosal flaps supported with an interposition graft[2]. The presence of saddle deformity, usually associated with larger perforations and/or those due to an inflammatory etiology, increases the reconstructive challenge. We have noted large perforation repairs can further destabilize the dorsum[3]. Costal cartilage grafting can optimize the functional and aesthetic deformities common to these noses. This case highlights our two-staged approach to reconstruction of a large septal perforation and several aesthetic deformities. An endonasal approach for perforation closure with delayed reconstructive rhinoplasty allows for perforation healing and postoperative dorsal stabilization. This avoids the possibility of an exposed costal cartilage graft and subsequent risk of infection that would be present with concurrent operations.

Our patient's postoperative course following perforation repair was complicated by depression and suicidal ideation. A detrimental effect of rhinoplasty performed at the time of psychiatric distress has been documented[4]. This risk is greatest during the early postoperative period. Caution is advised before proceeding with elective facial plastic surgery in a patient with an acute psychiatric condition. Extensive discussion occurred with this patient and his family on surgical and mental health risks before secondary surgery was scheduled. Second stage surgery was performed only after assurance and agreement on mental health care was reached.

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The Role of 3D Modeling in Cochlear Implantation

Alyssa Leong,[1] Monika Edejer,[1] Ellen Smith Au.D,[2] and Daniela Carvalho MD MMM[2,3]

[1] University of California, San Diego, USA [2] Rady Children's Hospital of San Diego, USA
[3] Department of Otolaryngology, University of California, San Diego, USA

Abstract

Cochlear implantation (CI) can provide hearing for pediatric patients with congenital severe to profound sensorineural hearing loss (SNHL), but it can have variable to poor outcomes in patients with significant cochleovestibular abnormalities. We report the case of an infant with cochleovestibular abnormalities whose surgery planning was done with 3D modeling of the temporal bones. Permission has been granted by the family. An ex 29-week preemie was diagnosed with congenital bilateral profound sensorineural hearing loss. The patient was also diagnosed with Nicotinamide Adenine Dinucleotide. Imaging of the patient's temporal bone revealed bilateral hypoplastic cochleas. A 3D model of both temporal bones was used to assess the best surgical approach and best electrode array for his anatomy. He was implanted at 10 months of age with a Cochlear Americas CI612 implant with Contour Advance Electrode. He had a right sided gusher controlled with temporalis muscle. We were able to insert 16 and 15 electrodes into the patient's left and right cochleas, respectively. There were 13 positive NRT responses on the left and 15 on the right. Soundfield testing at 3 months post-activation revealed responses in the normal hearing range (left) and mild hearing loss range (right). He is responding to all Ling sounds and is starting to imitate animal sounds. 3D modeling can be used as an additional tool for CI in patients with cochlear abnormalities. These challenging cases require all possible resources to achieve the best outcomes with cochlear implantation.

Introduction

Cochlear implantation is an excellent treatment option for patients with severe to profound sensorineural hearing loss. However, the outcomes with a CI can be influenced by several factors, including cochlear anatomy, cochlear nerve diameter and developmental issues. We present a case of a patient with Nicotinamide Adenine Dinucleotide (NAD) deficiency, with bilateral SNHL due to abnormal cochleas e cochlear nerves, who underwent bilateral cochlear implantation.

Case presentation

We present a case of an ex 29-week preemie who spent 2 months in the NICU and failed newborn hearing screening bilaterally. He had cor triatriatum, ASD, has prolonged QT and kidney issues. He was then diagnosed with Nicotinamide Adenine Dinucleotide (NAD) deficiency due to the patient's heart and kidney abnormalities. The causes for NAD deficiency disorder include genetic or environmental reasons that lead to various congenital malformations in these organs as well as the vertebrae and limbs. There have been cases reported of SNHL in NAD deficiency disorder patients, hinting at a possibility of a syndromic relationship. It should also be noted that the patient was premature, another added risk factor for SNHL.

Further testing revealed bilateral profound SNHL. He was fit with hearing aids at 6 months of age, with no benefit. Imaging of the temporal bones revealed significant cochleovestibular abnormalities (FIgure 1). The patient had bilateral hypoplastic cochleas, with the right cochlea being much smaller than the left side. His lateral semicircular canals (SCC) were absent, his posterior SCCs were hypoplastic, and he had smaller than normal vestibules. A MRI of the patient's IACs and brain showed the patient likely had Birman IV bilaterally. This indicated that although both ears were potential candidates for bilateral cochlear implantation (CI), it could result in possible poorer outcomes with cochlear implantation.

He was evaluated by the Rady Children's Hospital of San Diego multidisciplinary cochlear implant team and considered a candidate for bilateral simultaneous CI. Due to his diagnosis of NAD and abnormal cochleovestibular anatomy, the temporal bone CT scan images were used to build a comprehensive 3D model of each cochlea in order to analyze each cochlea and determine the most appropriate implant array for his unique anatomy (Figure 3). The length of the cochleas were 9mm for the right side and 9.8mm for the left (when measuring it straight). As the left side had a more bulbous tip, there was also a measure with a possible "curved implant", which measured 14.9mm. Different electrode arrays were then used to match the 3D model and it was determined that the best option was to use a Cochlear Americas CI612 cochlear implant with Contour Advance electrode, keeping the stylet in place to optimize the length of insertion within his malformed cochleas.

Bilateral simultaneous cochlear implantation was performed when the patient was 10 months of age. There was, as expected from the 3D modeling, partial placement of the electrode arrays. 16 electrodes were placed in the left cochlea and 15 electrodes were placed in the right cochlea. NRTs were present for 13 electrodes on the left and 5 electrodes on the right. All exhibited normal impedances. An AP skull x-ray showed the CI electrode arrays were in an excellent position inside the cochleas (Figure 2). The cochlear implants were initially activated with six and seven extracochlear electrodes disabled on the left and right ear, respectively. He is responding to all Ling sounds in both ears at 4 feet of distance and soundfield testing is in the normal to mild range. He is recognizing and enjoying songs, understands simple commands and is saying several simple words.

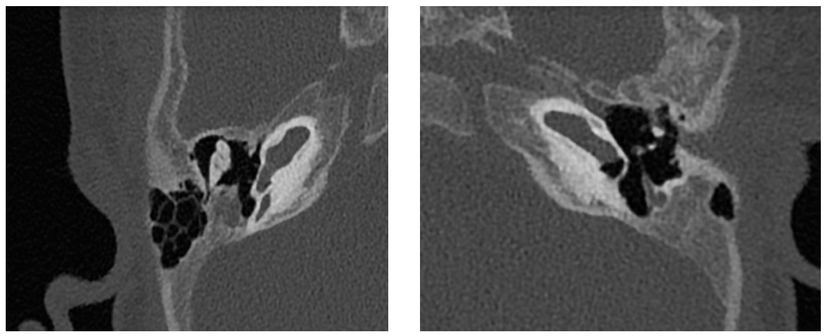

Figure 1: CT of the temporal bone, axial cuts, showing bilateral hypoplastic cochleas

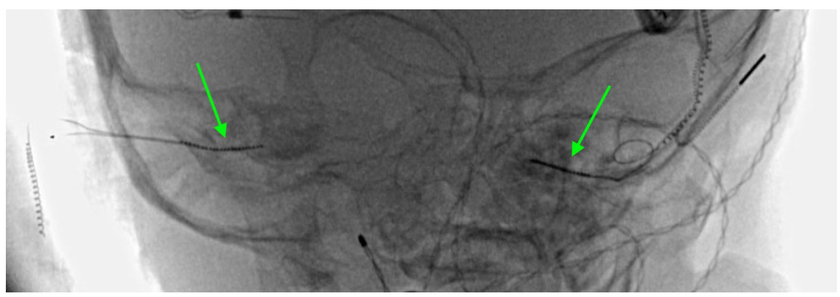

Figure 2: Intraoperative X-Ray showing both electrode arrays in the cochleas (green arrows)

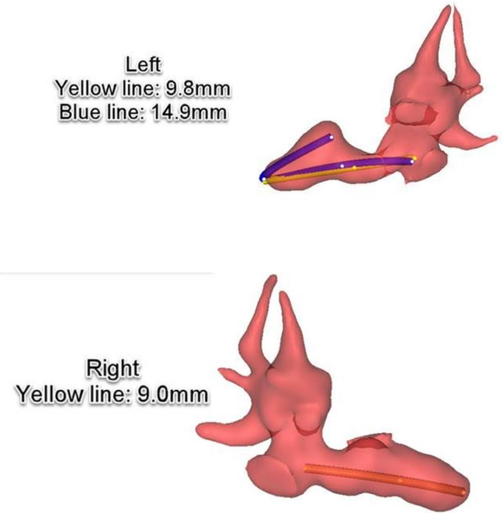

Figure 3: Bilateral cochleovestibular 3D reconstruction model with masurements of the cochlear lengths

Conclusion

3D modeling can be used as an additional tool for patients with cochlear abnormalities. It can aid in selecting the appropriate electrode array and developing a surgical strategy to optimize the implant placement in cases of complex anatomy. Availability of these 3D reconstruction techniques is becoming more widespread and could become routine for surgical planning in patients with significant cochlear malformations.

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Does REM AHI predict persistent OSA after pediatric adenotonsillectomy?

Caroline M Fields, BS; Nicolas S Poupore, BS; Husein Smaily, MD; Jenna H Barengo, MD; Shaun A Nguyen, MD, MA; Jacqueline Angles, DO; Clarice S Clemmens, MD; Phayvanh P Pecha, MD; William W Carroll, MD

Medical University of South Carolina, Department of Otolaryngology – Head and Neck Surgery, Charleston, SC, USA

ABSTRACT

Objective: The utility of REM AHI in the management of pediatric obstructive sleep apnea (OSA) is not fully understood. The purpose of this study was to evaluate the relationship of REM AHI to postoperative persistence of OSA in children undergoing adenotonsillectomy.
Methods: A single-institution retrospective chart review was performed. Children under the age of 18 that received an adenotonsillectomy for OSA with both a preoperative and postoperative polysomnogram (PSG) were eligible for inclusion. Children with craniofacial or neuromuscular disorders or a tracheostomy were excluded. The primary outcome was the persistence of OSA after adenotonsillectomy defined as a postoperative obstructive apnea-hypopnea index (oAHI) ≥ 1.5. REM-predominant OSA was defined as a ratio of REM/NREM AHI ≥2. Patients with persistent and resolved OSA were compared.
Results: A total of 353 patients were included, with 143 (40.5%) being female and 14.7% having obesity. 232 (65.7%) had postoperative persistent OSA. The preoperative REM AHI, REM AHI minus NREM AHI, and REM AHI minus oAHI of children with persistent OSA did not differ significantly from children with resolution of OSA (24.2 [32.3] vs. 20.4 [29.6], p=0.142; 15.7 [24.3] vs. 15.5 [24.2], p=0.664; 12.4 [18.8] vs. 12.1 [20.4], p=0.810). Rates of persistence were not different between those with REM-predominant OSA and REM-independent OSA (63.8% vs. 70.7%, p=0.218). Analyses based on severity of preoperative OSA (mild, moderate, and severe) also did not yield significant difference in these two groups. No statistical differences were seen in the rates of postoperative OSA when stratifying patients with a threshold REM AHI set at 20, 30, 40, 50, and 60.
Conclusion: This study suggests that preoperative REM AHI may be a poor predictor of OSA persistence after adenotonsillectomy. Further study is needed to help characterize how pre-operative REM AHI should impact clinician's decision making and recommendations.

METHODS

- Retrospective chart review
  - Children under the age of 18 that received an adenotonsillectomy, adenoidectomy, or tonsillectomy to address OSA from 2015 to 2021
- Selection Criteria
  - Inclusion: primary sleep surgery performed at this institution, at least one preoperative PSG, at least one postoperative PSG
  - Exclusion: syndromic abnormalities involving the head and neck, craniofacial or neuromuscular disorders, presence of tracheostomy, ventilator dependence
- Outcomes
  - Primary: persistence of OSA after adenotonsillectomy defined as a postoperative oAHI≥1.5
  - Secondary: necessity of additional therapies to address continued symptomatology of OSA
- Variables
  - Mild OSA defined as oAHI of 1.5 to <5, moderate OSA as an oAHI of 5 to <10, severe OSA as an oAHI of ≥10
  - REM-predominant OSA defined as REM AHI/NREM AHI ≥2, REM-independent OSA as REM AHI/NREM AHI <2
- Statistical Analysis
  - Significance of p-values <0.05 determined for continuous variables by an unpaired t-test for normally distributed and Mann-Whitney for non-normally distributed values
  - Significance of p-values <0.05 determined for nominal variables by a chi-square test
  - SPSS 27.0

RESULTS

Table 1. Comparison of PSG Parameters by Postoperative Persistence

Characteristic	Postoperative Persistent OSA (AHI ≥ 1.5)	Postoperative Resolution of OSA (AHI < 1.5)	P-value*
	Mean ± SD or Median [IQR]	Mean ± SD or Median [IQR]	
Preoperative PSG variables for all patients	(N = 232)	(N = 121)	
REM AHI	24.15 [32.3]	20.40 [29.6]	0.142
(REM AHI – NREM AHI)	15.70 [24.30]	15.50 [24.20]	0.664
(REM AHI – overall AHI)	12.35 [18.77]	12.10 [20.35]	0.810
Preoperative PSG variables for mild OSA preoperatively	(N = 28)	(N = 29)	
REM AHI†	8.95 ± 7.1	8.61 ± 4.85	0.789
(REM AHI – NREM AHI)	7.20 [7.37]	7.00 [6.50]	0.780
(REM AHI – overall AHI)	5.75 [5.90]	5.80 [5.16]	0.798
Preoperative PSG variables for moderate OSA preoperatively	(N = 71)	(N = 41)	
REM AHI	17.20 [7.4]	18.60 [8.8]	0.261
(REM AHI – NREM AHI)	13.60 [9.90]	14.70 [8.50]	0.398
(REM AHI – overall AHI)	9.80 [7.80]	11.50 [8.50]	0.268
Preoperative PSG variables for severe OSA preoperatively	(N = 133)	(N = 51)	
REM AHI	41.20 [33.9]	46.50 [36.2]	0.451
(REM AHI – NREM AHI) †	25.33 ± 27.28	28.78 ± 24.70	0.433
(REM AHI – overall AHI) †	19.82 ± 22.15	23.13 ± 20.20	0.354

N = number of patients; IQR = interquartile range; SD = standard deviation; OSA = obstructive sleep apnea; AHI = apnea-hypopnea index; PSG = polysomnogram; REM = rapid eye movement; NREM = non-rapid eye movement; BMI = body mass index; † = failed normality test.
*Significance of P-values < 0.05 determined for continuous variables by an unpaired t-test for normally distributed and Mann-Whitney for non-normally distributed values.

Table 2. Crosstabulation of Postoperative Persistence by Preoperative Sleep Stage Predominance

Characteristic	Postoperative Persistent OSA (AHI ≥ 1.5)	Postoperative Resolution of OSA (AHI < 1.5)	Total	P-value
	N (%)	N (%)	N	
	(N = 232)	(N = 121)		
Sleep stage predominant OSA by preoperative REM AHI/NREM AHI ratio				0.218
REM-independent OSA (REM AHI/NREM AHI ≤ 2)	70 (70.7)	29 (29.3)	99	
REM-predominant OSA (REM AHI/NREM AHI ≥ 2)	162 (63.8)	92 (36.2)	254	

N = number of patients; OSA = obstructive sleep apnea; AHI = apnea-hypopnea index; REM = rapid eye movement.
*Significance of *P*-values < 0.05

Figure 1. REM related OSA

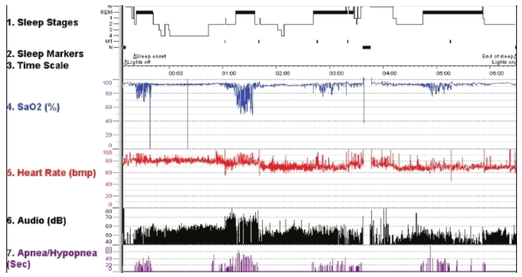

Table 3. Crosstabulation of Postoperative Persistence by Cutoffs of Preoperative REM AHI

Characteristic	Postoperative Persistent OSA (AHI ≥ 1.5) (N = 232)	Postoperative Resolution of OSA (AHI < 1.5) (N = 121)	Total	P-value
	N (%)	N (%)	N	
Preoperative REM AHI set at 20				0.468
REM AHI < 20	98 (63.6)	56 (36.4)	154	
REM AHI ≥ 20	134 (67.3)	65 (32.7)	199	
Preoperative REM AHI set at 30				0.401
REM AHI < 30	135 (64.0)	76 (36.0)	211	
REM AHI ≥ 30	97 (68.3)	45 (31.7)	142	
Preoperative REM AHI set at 40				0.415
REM AHI < 40	161 (64.4)	89 (35.6)	250	
REM AHI ≥ 40	71 (68.9)	32 (31.1)	103	
Preoperative REM AHI set at 50				0.516
REM AHI < 50	179 (64.9)	97 (35.1)	276	
REM AHI ≥ 50	53 (68.8)	24 (31.2)	77	
Preoperative REM AHI set at 60				0.794
REM AHI < 60	197 (65.4)	104 (34.6)	301	
REM AHI ≥ 60	35 (67.3)	17 (32.7)	52	

N = number of patients; OSA = obstructive sleep apnea; AHI = apnea-hypopnea index; REM = rapid eye movement.
*Significance of *P*-values < 0.05

CONCLUSIONS

- The preoperative REM AHI, REM AHI minus NREM AHI, and REM AHI minus oAHI of children with persistent OSA did not differ significantly from children with resolution of OSA
- Analyses based on preoperative severity of OSA also did not yield a significant difference between these two groups
- Rates of persistence were not different between those with REM-predominant OSA and REM-independent OSA
- These results suggest limited utility of REM AHI in prognosticating risk of postoperative OSA

SUMMARY

- There are no clinical guidelines currently in place that advise physicians on how to interpret a high REM AHI preoperatively.
- No difference in preoperative REM AHI was found between those with persistent and resolved OSA postoperatively.
- Further research is needed to determine the significance of REM related OSA in children and its impact on the efficacy of other approved treatments for pediatric OSA.

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Drug-induced sleep endoscopy impacts intraoperative decision making in young children with obstructive sleep apnea

Emma K. Landes BA[1], Rebecca Z. Lin BA[1], Judith E.C. Lieu MD MSPH[1], Katherine Dunsky MD[1]
[1]Washington University School of Medicine Department of Otolaryngology

Washington University in St. Louis School of Medicine
Biomedical Research

Background

- Untreated OSA has long-term consequences including cardiovascular disease, metabolic disorders, and developmental problems.[1]
- Drug-induced sleep endoscopy (DISE) is performed using a flexible endoscope to evaluate the upper airway while under anesthesia to simulate a sleep state that preserves airway dynamics, thereby enabling identification of the area of obstruction.
- Studies of DISE-directed surgery have not compared differences in surgical management between patients with and without DISE.
- Children <2 years of age have a higher risk of perioperative respiratory complications[2] and are more likely to have sites of obstruction other than adenotonsillar hypertrophy.

Goal and Hypothesis

Goal: To identify the impact of DISE on surgical decision making in young children.
Hypothesis: If DISE was performed, the surgical plan may deviate from adenotonsillectomy.

Methods

- We reviewed a consecutive cohort of patients with OSA under 3 years old between July 2018 and August 2021.
- Patients were identified with ICD-10 codes: sleep-disordered breathing (G47.9), newborn sleep apnea (P28.3), and sleep apnea (G47.30).
- Charts were reviewed for demographics, medical history, polysomnography (PSG) findings, DISE Chan-Parikh Score[3], surgeries, and outcomes.
- Chi-squared and independent-sample t tests were performed with IBM SPSS 28.0.

Table 1. Patients with diagnosed OSA with DISE and without DISE reviewed for possible surgical interventions

Surgery details	No DISE (n=56)	DISE (n=51)	*p* value
Surgery			**0.023**
No, n (%)	10 (17.9)	2 (3.9)	
Yes, n (%)	46 (82.1)	49 (96.1)	
Adenotonsillectomy	23 (45.1)	26 (51.0)	0.552
Adenoidectomy	13 (25.5)	13 (25.5)	1
Tonsillectomy	2 (3.9)	5 (9.8)	0.24
Supraglottoplasty	7 (13.7)	18 (35.3)	**0.011**

*2 children underwent DISE twice

Table 2. Differences in pre- and post-intervention PSG measurements of patients with DISE and without DISE

PSG Difference	No DISE (n=21)	DISE (n=26)	(CI), *p* value
AHI (mean ± SD)	-17.7 ± 54.3	-19.3 ± 33.6	(-24.4, 27.7), 0.900
oAHI (mean ± SD)	-32.1 ± 60.4	-18.2 ± 32.3	(-41.6, 13.8), 0.319
O2 Nadir (mean ± SD)	4.6 ± 8.9	9.8 ± 11.1	(-11.2, 0.7), 0.086

Table 3. Outcomes following intervention of patients with DISE compared to those without DISE

Outcomes	No DISE (n=56)	DISE (n=51)	*p* value
Persistent OSA	26 (61.9)	27 (54.0)	0.445
Complications	2 (4.5)	5 (9.8)	0.328
Bleeding	0 (0.0)	0 (0.0)	
Dehydration	0 (0.0)	3 (5.9)	0.102
Post-op admission	7 (15.9)	9 (17.6)	0.821
Readmission	0 (0.0)	4 (7.8)	0.058
Other complication	1 (2.3)	2 (3.9)	0.647

Results

- We identified 107 children with sleep apnea under age 3:
  - Half of these children underwent DISE.
- The mean age in years of patients with DISE was older than those without DISE (1.99 vs 1.62; p=0.013).
- There were no significant differences in weight percentile, sex, race, ethnicity, insurance type, prematurity, birth/perinatal events, chronic medical problems, smoke exposure, or prior ENT surgeries between the two groups.
- The mean pre-intervention O_2 nadir of patients with DISE was lower than those without DISE (74.0 vs 78.6; p=0.016).
- Children who had DISE were more likely to undergo any surgery to address sleep apnea; and of the surgical options, they were more likely to undergo supraglottoplasty.
- There were no significant differences in outcomes or changes in pre- to post-intervention PSGs between the two groups.

Conclusions

- Patients with DISE were more likely to undergo supraglottoplasty compared to patients without DISE.
- Laryngomalacia may be missed in children who do not have overt stridor when awake.
- Surgeons should consider utilizing DISE for young children with OSA as they may have sites of obstruction other than adenotonsillar hypertrophy.

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The changing landscape of pediatric salivary gland stones: a half-century systematic review

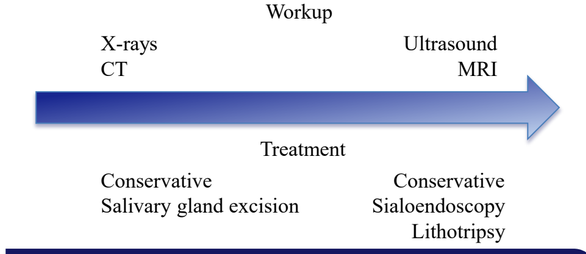
Tanya Chen MD[1], Rachel Szwimer MSc[2], Sam J Daniel MD FRCSC[3]

[1]Department of Otolaryngology – Head and Neck Surgery, University of Toronto [2]Faculty of Medicine, McGill University [3]Department of Otolaryngology - Head and Neck Surgery, Montreal Children's Hospital

Introduction

Up to 3% of sialolithiasis is attributed to the pediatric population. If left untreated, sialolithiasis can lead to pain and swelling when eating, infection, and ultimately surgical. Sialolithiasis remains the most common reason for salivary gland excision. This surgery carries a risk for facial nerve injury and results in an unsightly scar for children. As technology has evolved and treatment paradigm has shifted, there is lacking a consensus on this change amongst children with sialolithiasis.

Figure 1. Change over time in workup and treatment of sialolithiasis



Objective

The objective of this review was to systematically assess the evidence for pediatric sialolithiasis, including its demographic characteristics, diagnosis, and treatment.

Methods

A literature search was performed on Medline and Embase databases from inception to Dec 4, 2020. Demographic data, symptoms, stone characteristics, diagnostic modality and intervention were collected data points. Complications were classified as minor or major. Minor complications were self-limited symptoms. Major complications were significant symptoms that required further intervention, infections requiring antibiotics, or ductal complications including avulsion, perforation, or stenosis.
Quality assessment was performed with a modified version of a tool for quality appraisal of case reports with bias domains of selection, ascertainment, causality, and reporting.

Results

41 out of 604 articles were included. Only 8 (19.5%) studies included more than 10 patients.

Over the years, an increasing number of articles were published on pediatric sialolithiasis whereby 70.7% (n=29/41) of articles were published after the year 2000.

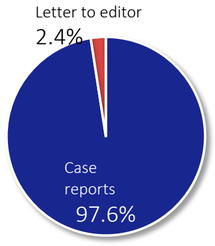
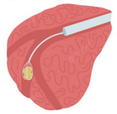


Table 1. Characteristics of salivary gland stones, diagnostic and therapeutic interventions

	Count (n)	% of total reported
Affected gland (n=246)		
Parotid	32	13.0
Submandibular	210	85.4
Sublingual	4	1.6
Number of stones (n=142)		
Single	101	71.1
Multiple	41	28.9
Location in gland (n=121)		
Distal	69	57.0
Proximal	29	24.0
Intraglandular	11	9.1
Hilum	12	9.9
Imaging modality		
Plain radiograph	36	23.4
Sialography	22	14.3
CT	13	8.4
Ultrasound	73	47.4
MRI	0	0.0
None	10	6.5
Intervention		
Intraoral sialolithotomy	88	31.5
Gland excision	21	7.5
Interventional sialoendoscopy	89	43.3
Extracorporeal shock wave lithotripsy	24	8.6
Conservative management	11	3.9
Spontaneous passage or resolution	8	2.5
Not reported	4	1.4

Results

Table 2. Recommendations for interventions in pediatric sialolithiasis

	Indications	Limitations	Complications
SIALOENDOSCOPY 	Proximal or distal stones	Large stones (>5mm) Distal stricture Cost Surgical expertise	Duct injury Failure of stone passage
INTRAORAL EXCISION 	Distal stones, easily visible in oral cavity	Proximal stones Mostly requires GA in children	Failure of stone passage Ductal stricture, ranula
ESWL 	Proximal stones Non-invasive/non-surgical	Distal stones Specialized equipment Needs ultrasound	Failure of stone passage
SIALECTOMY 	Large, multiple stones Proximal stones Recurrent Last line	Increased morbidity	Xerostomia Neuropraxia
CONSERVATIVE	Should always be first line	Failure requires intervention	Failure of stone passage

Average stone size: 7.7mm [range 1.3mm - 3.5cm]
98.7% of all reported stone sizes were <1.0cm.

The most common source of bias was due to reporting, due to missing details regarding the patient's investigations or treatments that would make reproducibility difficult.

Figure 2. Quality breakdown of articles

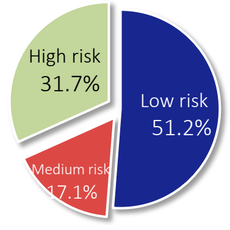
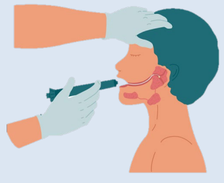


Table 3. Complications according to intervention

Intervention	Complication (n)			
	Minor	Major	None	N/A
Intraoral sialolithotomy	8 (10.3%)	2 (2.6%)	68 (87.1%)	10
Extracorporeal shock wave lithotripsy	7 (29.2%)	6 (25.0%)	11 (45.8%)	0
Interventional sialoendoscopy	4 (5.1%)	10 (12.8%)	64 (82.1%)	11
Gland excision	0 (0%)	0 (0.0%)	16 (100%)	5
Conservative management	1 (12.5%)	0 (0.0%)	7 (87.5%)	3
Spontaneous passage or resolution	0 (0%)	0 (0.0%)	5 (100%)	3

Conclusion

Pediatric sialolithiasis is uncommon and poorly studied despite its potential morbidity but is not so different from the adult population. Sialoendoscopy is slowly becoming the leading diagnostic and therapeutic modality, alongside ultrasound over radiographs. However, specialized equipment, surgical experience, and anatomical factors must be considered. There is an increasing research interest in this field and further larger-scale high-quality studies are warranted to follow the future innovations.



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Superabsorbent Polymer (SAP) Obstruction Presenting As Periorbital Cellulitis and Nasal Abscess

Meghana Chanamolu[1], Maxwell Bergman MD[2], Amy Manning MD[2], Prashant Malhotra MD[2], Kevin Liu MD[2], Tendy Chiang MD[2,3]

[1] Northeast Ohio Medical University, Rootstown, OH, USA
[2] Department of Pediatric Otolaryngology, Nationwide Children's Hospital, Columbus, OH, USA
[3] Center of Regenerative Medicine, Abigail Wexner Research Institute, Nationwide Children's Hospital, Columbus, OH, USA

Introduction

- SAPs are small, round, non-compressible balls with a smooth, slippery surface and are currently marketed as toys for young children[1]
- Comprised of groups of copolymers giving them a high-water absorption capacity making SAPs a potential cause of foreign body obstruction in children[2].
- Previous reports of nasal, bronchial, auditory canal, and bowel obstructions have been noted and they led to symptoms such as dehydration and pneumonia that required hospitalization and operative removal[6,7].
- The radiolucent properties of SAP can cause obstructions to be misdiagnosed as congenital abnormalities, infections, neoplasms, systemic diseases, or trauma[3].
- Nasal obstruction can be detrimental as secretions from mucosal epithelium cause further expansion, leading to edema and congestion[4].
- Patients with SAP-induced nasal cavity obstruction can present with rhinorrhea, epistaxis, facial edema, poor appetite, fever, and facial pain.
- Their gel-like properties make SAP challenging to retrieve, as they are prone to fragmenting into smaller pieces[5].
- **Misdiagnosis in this case can be exceedingly detrimental as nasal infections are treated with topical medications and irrigations, which introduce more moisture into the nasal airway and allowing for SAP expansion and increased injury to nasal mucosa[1]**.
- **Delays in SAP retrieval also place the patient at risk of aspiration or ingestion, which is known to cause local damage, obstruction, and hospitalization[6,7]**.
- In this article we present a unique case in which the physical and imaging presentation of SAP -related nasal obstruction manifested as periorbital cellulitis and nasal abscess.
- A delay in diagnosis led to a prolonged clinical course, extensive local mucosal damage, multiple operative debridement's and nasal vestibular stenosis.

Case

A 2-year-old presented to an ED with two days of right eye swelling, pain, and rhinorrhea. Ocular examination showed mild inferior periorbital edema and conjunctival redness. She was diagnosed with **conjunctivitis** and discharged on a course of Augmentin. The following day, she returned to the ED for worsening symptoms, again diagnosed with acute conjunctivitis. Over the next 24 hours the patient developed increased discharge, erythema, and edema of her right face. She was brought to the emergency department and was initially suspected to have **preseptal orbital cellulitis**. Upon inspection of her right naris, a large, translucent foreign body was noted and described as a "green jelly-filled ball." CT imaging demonstrated a right anterior nasal cavity rim-enhancing hypodense collection presumably phlegmon/**forming abscess**. There was mild mucosal thickening of the medial right maxillary sinus and overlying nasolabial fold. The radiology report also noted "no discrete radiodense foreign object identified". ENT saw her the next morning and underwent urgent operative removal of the foreign body polymer the following day. Intraoperatively, a clear polymer was occluding the right naris and the obstructing material was removed in fragments. There was a distinct lack of any purulent drainage or abscess during the procedure, but there was robust granulation. At her two-week follow-up appointment, the patient was suffering from frequent epistaxis, rhinorrhea, congestion, and periorbital edema. Endoscopy showed dry mucosa, granulation tissue, and crusting/bleeding in the area of the original obstruction. Because of the granulation tissue density, the patient was taken to the operating room six days later for debridement. She returned to clinic two weeks after her debridement surgery and was found to have significant crusting and firm narrowing of her right nares, concerning for vestibular stenosis. Subsequent intraoperative examination confirmed the diagnosis of vestibular stenosis, and she required two subsequent scar tissue division procedures along with Kenalog injections.

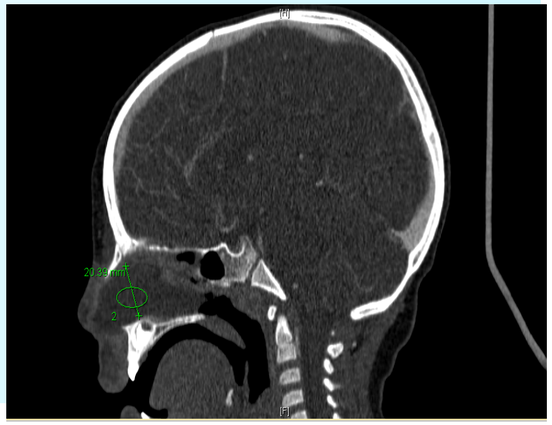

Image 1. Sagittal CT demonstrating extent of initial suspected occlusion that led to ambiguous presentation. An right anterior nasal cavity rim-enhancing hypodense collection measuring 4.9 cm x 2 cm x 1.7 cm.

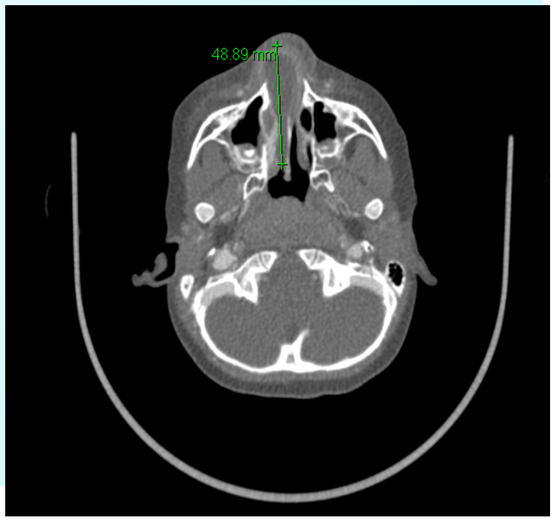

Image 2. Axial CT demonstrating extent of initial suspected occlusion that led to ambiguous presentation. An right anterior nasal cavity rim-enhancing hypodense collection measuring 4.9 cm x 2 cm x 1.7 cm.

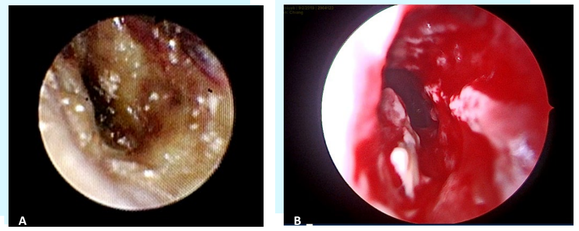

Image 3. A.) Granulation and crusting of nasal tissue that was present during follow-up visit. To extensive to remove in office. B.) Intraoperative scope showing nasal vestibule opening after removal of the granulation tissue.

Discussion

- The extent of soft tissue injury observed in our patient is due to the expansile properties of SAPs and the resultant mucosa pressure necrosis.
- Foreign body insertion into nasal cavities are particularly detrimental given the extensive moisture available and small confined space.
- CT findings often include rim-enhancing hypodense mass, lack of radiopaque foreign body, and surrounding soft tissue inflammation.
- Nasal mucosa atrophy due to pressure necrosis can result in recurrent epistaxis, crusting, and rhinorrhea, as observed in our patient.
- Due to the malleability and friability of SAP, bedside removal was unsuccessful thereby necessitating intraoperative removal
- If SAP fragments were left undisturbed, particles would likely re-adhere to the nasal epithelium leading to epithelial atrophy, recurrent epistaxis, and delayed healing
- The long duration of foreign body obstruction resulted in pressure necrosis, extensive granulation tissue, and vestibular stenosis, which has required multiple surgical interventions including repeated debridement, nasal aperture dilation, and scar division.
- Acquired causes of vestibular stenosis include infection, foreign body reaction, and iatrogenic and traumatic injury
- Nasal epithelial trauma leads to migration, proliferation, and differentiation of inflammatory cells and surrounding cells which causes tissue remodeling the tissue and contracture of the nasal airway.
- When removing obstructing foreign bodies such as SAP, suctioning, chemical cauterization, and soft tissue manipulation can further traumatize the vestibular epithelium thereby leading to scarring and further vestibular stenosis

Conclusion

SAP represent a significant hazard to young children. They are easily placed as foreign bodies and their expansile properties can cause significant local injury to the nasal, auditory, respiratory, or gastrointestinal tissues. This case report demonstrates a particularly severe nasal mucosal injury with resultant vestibular stenosis. Our case also highlights the difficulty in timely diagnosis SAP obstruction, as it is not readily visible on diagnostic imaging and physical symptoms can mimic other, more common, presentations. Parents and health professionals should be aware of SAP dangers and keep foreign body obstruction in the differential diagnosis in suitable clinical scenarios.

Contact

Meghana Chanamolu
401 South Main Street,
Apt. 211B,
Akron, Ohio 44303
5673719751
mchanamolu@neomed.edu

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AUGUSTA UNIVERSITY
MEDICAL COLLEGE OF GEORGIA

Pediatric Mandibular Malignancies: A Comprehensive SEER Analysis

Aditya Devarakonda BS, Harrison Cowart BS, Nicholas Drury MD, Heather Koehn MD MMHC

31

SENTAC

Abstract

Background: Mandibular malignancies are rare entities in pediatric otolaryngology. Prior studies have characterized histological tumor subsets, but we offer a contemporary analysis of the myriad of malignant pediatric mandible tumor subtypes using the Surveillance, Epidemiology, and End Results (SEER) registry.

Methods: SEER 18 registry data was collected using the 0-18 years age range and C41.1 (mandible) ICD-O-3 site code. Univariate Cox proportional hazard ratios (HR) were calculated for overall survival (OS) and disease-specific survival (DSS) among patient demographics, tumor characteristics, and treatment groups. Kaplan-Meier survival curves were generated for OS and DSS for significant groups.

Results: Of 541 patients identified, 64 patients met inclusion criteria. The median age of diagnosis was 13.0, and the median survival was 8.7 years. Osteosarcoma was the most common diagnosis (n = 22). Race, sex, odontogenic origin, or treatment modality did not affect OS or DSS. However, p-values for age <13 years and sarcomatous histology approached significance in OS. 'Distant' staging was associated with significantly elevated HR of 6.28 and 5.29 for OS and DSS, respectively ($p < 0.05$). Kaplan-Meier curve demonstrated lower OS and DSS for 'distant' stage versus 'localized' and 'regional'. Sarcomas exhibited lower DSS ($p < 0.05$) than non-sarcomas on the curve.

Conclusion: To the best of our knowledge, we report the largest analysis of pediatric mandibular malignancies in the literature. 'Distant' stage and sarcoma subtype tumors were associated with decreased survival, while age of diagnosis had a notable survival trend. Clinical suspicion and early diagnosis are paramount for improved survival.

Objectives

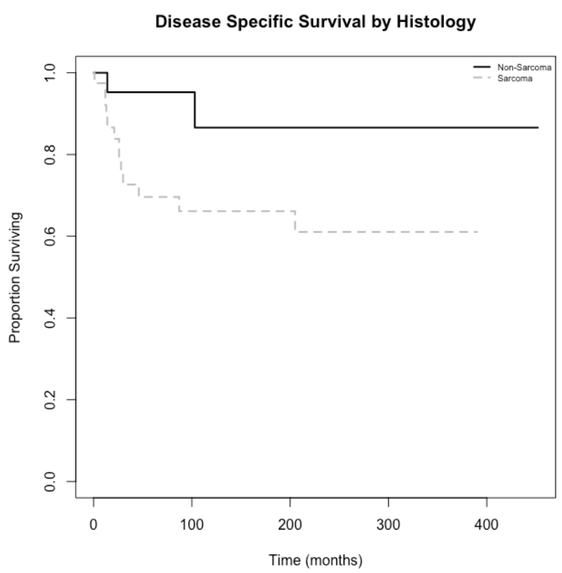
- Quantify pediatric mandibular tumor patient demographics to establish baselines for clinicians and researchers to compare
- Calculate hazard ratios for patient qualities, tumor characteristics, and treatment groups
- Identify patient qualities, tumor characteristics, and treatment modalities that correlate with disparities in survival

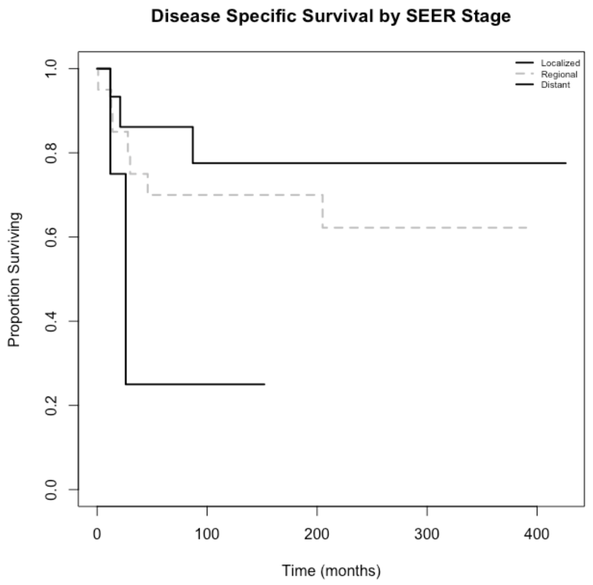
Methods

- Surveillance, Epidemiology, and End Results (SEER) 18 registry data was collected using the 0-18 years age range and C41.1 (mandible) ICD-O-3 site code.
- Univariate Cox proportional hazard ratios (HR) were calculated for overall survival (OS) and disease-specific survival (DSS) for 64 patients.
- Patient demographics (age at diagnosis, race, gender), tumor characteristics (staging, histologic subgroup), and treatment groups (surgery, chemotherapy, and/or radiotherapy) were used as variables.
- Kaplan-Meier survival curves were generated for OS and DSS for significant groups.

Results

- The median age of diagnosis was 13.0 years, and the median survival was 8.7 years.
- Osteosarcoma was the most common diagnosis (n = 22), followed by Ewing sarcoma (n = 13) and malignant ameloblastoma (n = 10).
- 'Distant' staging was associated with significantly elevated HR of 6.28 and 5.29 for OS and DSS, respectively ($p < 0.05$)
- Kaplan-Meier curve demonstrated lower OS and DSS for 'distant' stage versus 'localized' and 'regional'. Sarcomas exhibited lower DSS ($p < 0.05$) than non-sarcomas on the curve.





Conclusion

- To the best of our knowledge, we report the largest analysis of pediatric mandibular malignancies in the literature covering a wide variety of tumor subtypes.
- Race, gender, and treatment modalities were not associated with any significant effects on DSS or OS.
- 'Distant' stage and sarcoma subtype tumors were associated with decreased survival, while age of diagnosis had a notable survival trend.
- Clinical suspicion and early diagnosis are paramount for improved survival.
- Future multi-institutional studies are needed to fully elucidate these associations due to the rarity of these malignancies.

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	Median Survival	Disease Specific Survival (DSS)		Overall Survival (OS)	
	Months	HR (95% CI)	p	HR (95% CI)	p
Race					
White	140	Base	--	Base	--
Black	69	1.26 (0.39 – 4.12)	0.697	1.42 (0.52-3.84)	0.492
Other	73	1.21 (0.26 – 5.65)	0.805	1.20 (0.26-5.46)	0.816
Sex					
Male	104	Base	--	Base	--
Female	94	1.01 (0.36 – 2.85)	0.979	0.89 (0.35 – 2.27)	0.812
Histology					
Non-sarcoma	114	Base	--	Base	--
Sarcoma	87	1.38 (0.90 – 17.76)	0.068	2.55 (0.84 – 7.69)	0.097
Subtype					
Non-odontogenic	87	Base	--	Base	--
Odontogenic	276	3.92e-09 (0 – Inf)	0.998	0.329 (0.074-1.46)	0.143
SEER Stage					
Localized	140	Base	--	Base	--
Regional	144	1.74 (0.45 – 6.75)	0.421	1.27 (0.44 – 3.67)	0.664
Distant	26	6.28 (1.23 – 32.07)	0.0272*	5.29 (1.23 – 22.66)	0.025*
Age					
< 13 years	103	Base	--	Base	--
≥ 13 years	114	0.47 (0.17 – 1.33)	0.157	0.42 (0.16 - 1.33)	0.07
Radiation					
No radiation	109.5	Base	--	Base	--
Radiation	103	1.50 (0.48 – 4.73)	0.489	1.60 (0.57 – 4.47)	0.374
Surgery					
No surgery	124.5	Base	--	Base	--
Surgery	104	2.50 (0.33 – 19.02)	0.376	2.91 (0.39 – 21.78)	0.299
Chemotherapy					
No chemotherapy	234	Base	--	Base	--
Chemotherapy	103	2.05 (0.58 – 7.28)	0.27	1.80 (0.64 – 5.07)	0.269
Statistical significance (p < 0.05) denoted by *.					

Excision of a Thyroglossal Duct Cyst with Unique Findings Concerning for Malignancy

Luciano Venturino, MS3[1]; Dr. Bethzaida Suarez Gonzalez, MD[2]; Dr. Lara K Reichert, MD MPH[2]
[1] Albany Medical College, Albany, NY
[2]Albany Medical Center, Department of Otolaryngology Head and Neck Surgery, Albany, NY

Abstract

Background: Thyroglossal duct carcinoma is an uncommon diagnosis encompassing about 1% of all thyroglossal duct cysts (TGDC). The purpose of the study is to discuss a unique case of TGDC. It is essential to differentiate between TGDC and carcinoma as management of the latter often requires more aggressive treatment.
Methods: Case Presentation
Results: We present a case of a 4-year-old male with several weeks of a midline neck mass with overlying erythema and mild tenderness, elevated thyroglobulin, and an ultrasound suspicious for TGDC but given concerning features of deep soft tissue extension a CT was recommended. The CT of the neck showed bilateral level 2 and 5 lymphadenopathy with a solid midline neck mass just superior to the thyroid gland that was concerning for malignancy to radiology team. The mass was excised using the Sistrunk procedure, and the final pathology was consistent with an inflamed benign thyroglossal duct cyst with no evidence of thyroid tissue or malignancy.

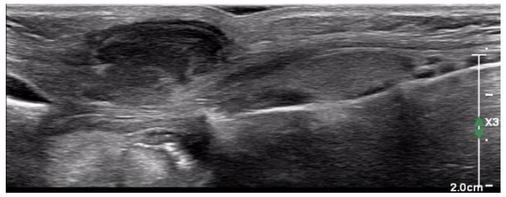

Figure 1: Neck ultrasound - Hypoechoic lesion in the midline of the neck suspicious for TGDC. However, there also appeared to be further extension of this lesion into the deep soft tissues.

Patient History

A 4-year-old male patient presented with 2 weeks of midline neck swelling and an abnormal thyroglobulin of 70.9. The TSH, free T4 and WBC were normal. Physical examination showed a non-toxic appearing patient with a non-tender midline neck ovoid lesion measuring 2-3 cm below the level of the hyoid that was not particularly mobile. He had not experienced any fevers or weight loss.

Neck ultrasound revealed a hypoechoic lesion in the midline of the neck suspicious for TGDC; however, there appeared to be further extension into the deeper soft tissues with internal vascularity for which a CT of the neck was recommended. The CT neck revealed an enhancing solid midline mass just below the level of the hyoid bone measuring 1.3 x 1.9 x 1.0 cm. Findings were concerning for a for a TGDC with ectopic thyroid versus TGDC carcinoma. Nonspecific enlarged bilateral level 2B/5A lymph nodes were also found. Due to patient's new onset symptoms, abnormal labs, imaging findings, and potential for malignancy, recommendation was made for removal with the Sistrunk procedure, which the patient underwent 2 weeks after initial consultation.

Pre-Operative Imaging

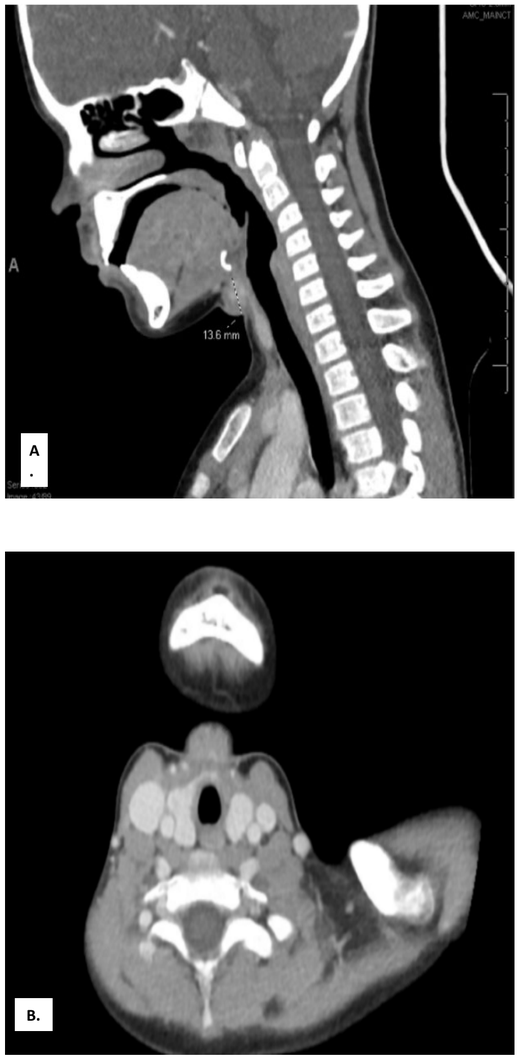

Figure 2: Pre-operative CT of the neck revealing an enhancing solid midline mass just below the level of the hyoid bone. – A. Sagittal view. B. Axial view.

Surgical intervention

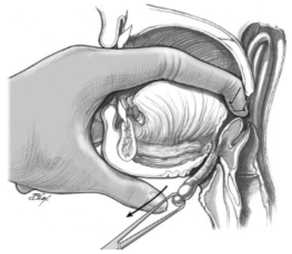

Figure 3: Sistrunk procedure.

Intraoperative findings were consistent with a 2 x 2 cm mass in the midline neck with obvious inflammation, abnormal purple discoloration and altered anatomy. During the procedure, the cyst partially ruptured, and orange gelatinous material extruded. The tract of the neck mass was followed to the hyoid bone with a midline portion of the hyoid removed in the usual fashion for a Sistrunk procedure.

Pathology results

Final pathology was consistent with an inflamed benign thyroglossal duct cyst with no evidence of thyroid tissue or malignancy. The GRAM, GMS, AFB stains were all negative for organisms.

Post-operative course

On post-operative day 7, the patient was taken to the emergency department due to swelling around the incision site in the absence of fevers, chills or breathing issues. Ultrasound revealed a seroma measuring 3.0 x 1.4 x 3.5cm, which was needle aspirated at bedside with 1 mL of serous fluid and the patient was discharged from the emergency department with a course of antibiotics. He was seen for follow up approximately 2 weeks later and appeared to be doing well with well-healed surgical incisions.

Discussion

This report presents a unique case of a pediatric TGDC with high concern for malignancy from radiology and primary care teams; thus, an expedited surgical intervention was coordinated. It is important to differentiate between these as there are differences in management, especially when assessing higher risk TGDC carcinoma cases. While TGDC can be treated with the Sistrunk procedure, management of malignancy may include additional thyroidectomy, followed by radioactive iodine therapy and/or thyroid-stimulating hormone suppression.

Contact

Contact: Lara Reichert, MD MPH
Albany Medical College
Department of Surgery
Division of Otolaryngology
Albany, NY
larakreichert@gmail.com

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Poster #36

Evaluation and Management of Juvenile Nasopharyngeal Angiofibromas

F. Zaman[1], A. Vescan[2], E. J. Propst[3], P. Muthusami[3], M. Gete[2], N. E. Wolter[3]

1. Temerty Faculty of Medicine, University of Toronto, Toronto, Canada
2. Department of Otolaryngology Head and Neck Surgery, Sinai Health System
3. Department of Otolaryngology – Head and Neck Surgery, The Hospital for Sick Children

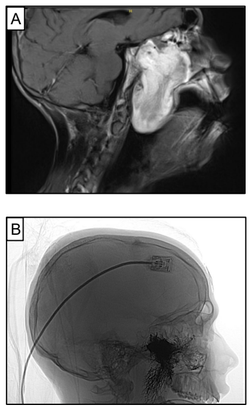
SickKids THE HOSPITAL FOR SICK CHILDREN

Otolaryngology – Head & Neck Surgery UNIVERSITY OF TORONTO

Abstract

Background: Juvenile nasopharyngeal angiofibromas (JNA) are rare, vascular tumors occurring almost exclusively in adolescent males. JNAs can rapidly grow and invade adjacent structures. This study's purpose is to explore the management and outcomes of JNAs over time. Methods: Single institution, retrospective chart review included pediatric patients with JNA diagnoses who presented from 2000/01/01 to 2022/02/01. Results: Thirty-five JNAs were seen at our hospital. Symptoms were present for a median (IQR) of 5.8 (3-12) weeks before referral. The median (IQR) age at surgery was 14 (12.6-15.2) years. Seventy-one percent (24/35) were Radkowski stage 2A or 2B. Nineteen (54%) were treated with open surgery (OS) and 16 (46%) were managed endoscopically (ES). Preoperative embolization was used in 91% (31/35) of cases. The median (IQR) intraoperative blood loss during OS was 675 (398-1050) ml compared to 500 (113-1700) ml during ES (p=0.38). The median (IQR) length of stay was shorter for ES (2.0 (1.0-2.0) days) compared to OS (4.0 (3.8-5.5) days, p=0.0003). The median (IQR) follow up was 3.2 (1.7-4.3) years. One patient with a Radkowski stage 3B tumor treated with OS had residual disease requiring post-operative radiation. One patient with a Radkowski stage 2C treated endoscopically had a recurrence. Conclusions: Management of JNAs has shifted towards endoscopic management. While this has resulted in a statistically significant decrease in length of stay, we did not find a difference in intraoperative blood loss. Despite the excellent visualization afford by endoscopic surgery, recurrence is still possible and vigilant postoperative monitoring is required.

Introduction



Background

- Juvenile nasopharyngeal angiofibroma (JNA) is a rare, benign, vascular tumor that occurs almost exclusively in adolescent males between the ages of 9-19 years old[1]
- Commonly presents with painless unilateral nasal obstruction, recurrent unilateral epistaxis, and a nasopharyngeal mass[2]
- Surgical resection is the primary treatment and can be done through endoscopic, endoscopic assisted (also known as a hybrid) or open surgical approaches[3, 4]
- Different surgical factors, preoperative considerations, tumour factors, and post-operative considerations are all important to explore in an institution's experience treating JNA[1, 5-7]

Objective

To describe the management and outcomes of open and endoscopic resection of juvenile nasopharyngeal angiofibroma (JNA)

Fig. 1. Sagittal view of Radkowski 2C Juvenile Nasopharyngeal Angiofibroma. A) Sagittal MRI T1 sequence. B) Sagittal fluoroscopic image obtained during preoperative embolization fluoroscopy using ethylene-vinyl alcohol copolymer -based liquid embolic agent.

Methods

- Retrospective cohort study
- Data Sources:
  - Electronic patient records (EPR)
  - EPIC
- Inclusion criteria: pediatric patients aged 0 to <18 years old who were diagnosed with JNA. Charts from January 1, 2000 to February 1, 2022

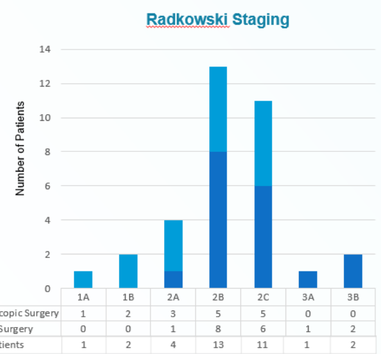
Primary Outcome: Rate of recurrence

Secondary Outcomes:
- Tumour stage
- Types of treatment
- Perioperative blood loss
- Length of hospital stay
- Years followed up

Results

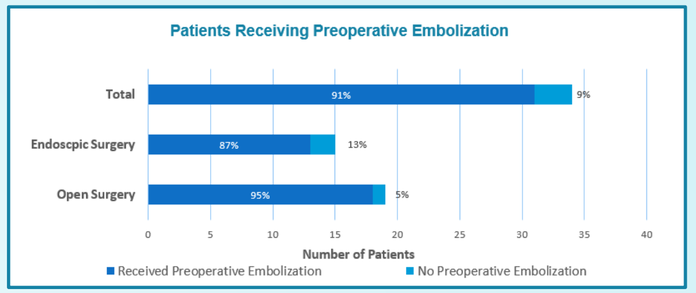
Table 1. Patient Demographics

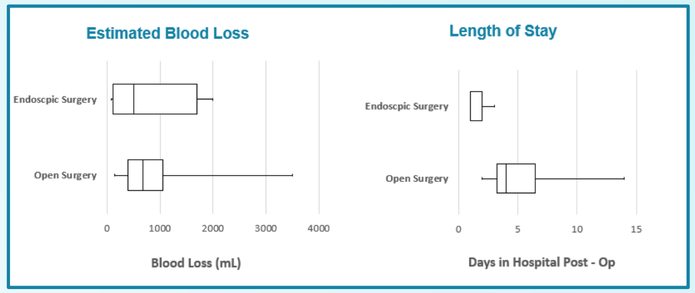
Number of Patients	35
Age at Surgery, years [median (IQR)]	14 (12.6-15.2)
Preoperative embolization	31 (91%)
Surgery Type	
Endoscopic Surgery	19 (54%)
Open Surgery	16 (46%)
Follow up, months [median (IQR)]	10 (6-20)

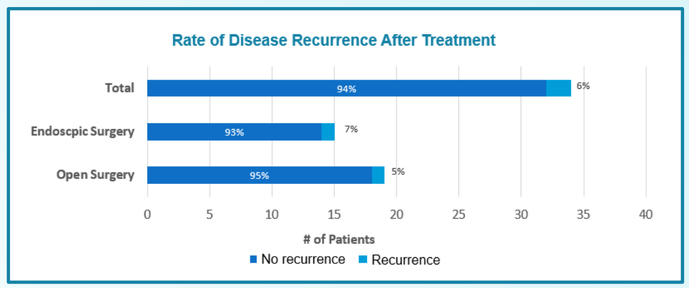


- Symptoms were present for a median (IQR) of 5.8 (3-12) weeks before referral

Results







Conclusions

- Surgical management of JNAs is shifting towards endoscopic resection
- Endoscopic approach has significantly reduced length of stay in hospitals post operation
- No significant difference in estimated blood loss between the open or endoscopic approaches were seen
- Recurrence of disease is still possible with both open and endoscopic surgical approaches with similar rates

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37. Pediatric Vallecular Cysts

Jessie G Jiang BS, BA[1], Sarah Gitomer MD[2], Brian Herrmann MD[2]

[1]University of Colorado School of Medicine, Aurora, Colorado, USA.

[2]Children's Hospital Colorado, CU Department of Otolaryngology - Head and Neck Surgery, Aurora, Colorado, USA

Children's Hospital Colorado

Affiliated with University of Colorado Anschutz Medical Campus

BACKGROUND

Congenital vallecular cysts (VC) are rare but result in health consequences such as feeding and respiratory issues. They are most common in neonates and infants and symptoms often present within the first week of life. Thus, early diagnosis and proper management is crucial, however, the number of studies on vallecular cysts and their associated symptoms is limited.[1-2]

OBJECTIVE

To characterize the cases of pediatric VC treated at our institution and contribute to existing literature on its management. We hypothesize that like previous studies, age of diagnosis will be related to symptoms of VC, with more severe symptoms prompting earlier diagnosis.

METHODS

A retrospective chart review of pediatric patients with VC from 2005-2022 was performed at a tertiary care children's hospital. Data on diagnosis characteristics, associated symptoms, and surgical treatment type was collected.

RESULTS

CHILDREN WITH VALLECULAR CYSTS (N = 19)	Mean (Range) / %
Age at diagnosis	3.3 (0-17 yrs)
Duration of symptoms	4.5 mo (0-3 yrs)
Race	
White	16%
Asian or Pacific Islander	16%
African American	16%
Hispanic or Latino	32%
Other	21%

RESULTS

Figure 1

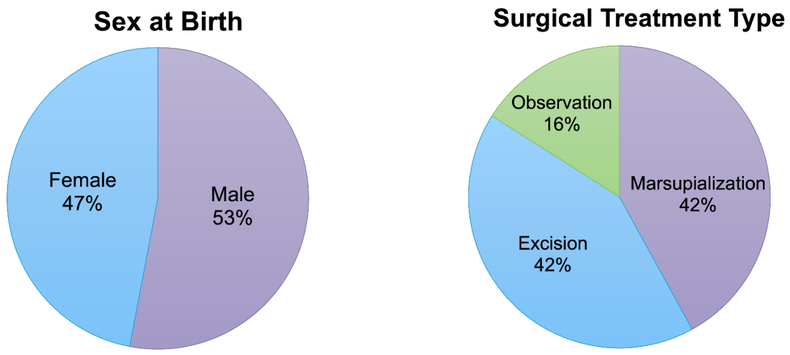
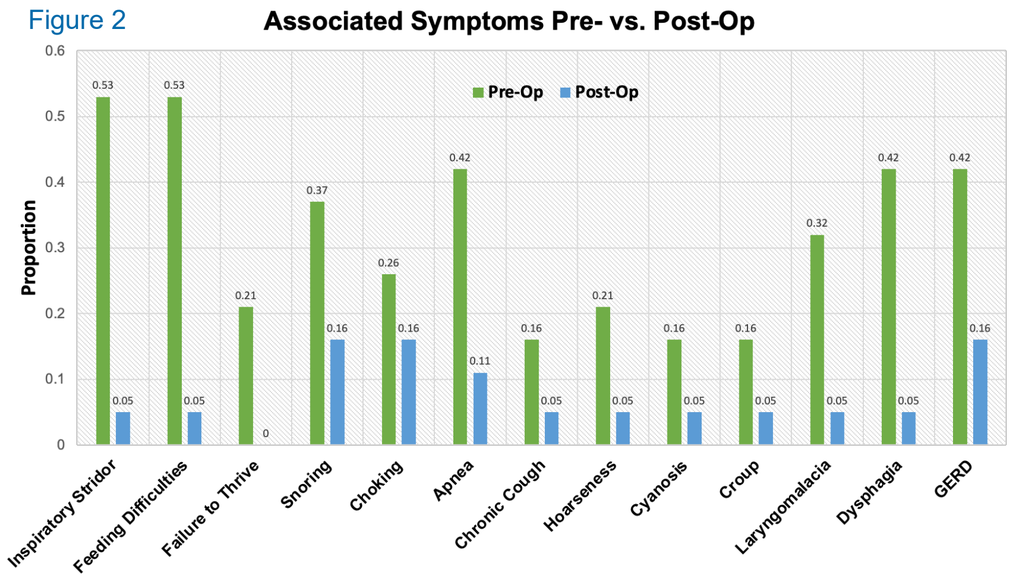


Figure 2



- The most common preoperative symptoms were inspiratory stridor, feeding difficulties, apnea, GERD, and dysphagia (Fig. 2)
- The most common post-operative symptoms included snoring, choking, and GERD (Fig. 2)
- Two (11%) patients had recurrence of the VC, both of which had their first VC excised
- In general, diagnosis before 1 year of age corresponded with more severe associated symptoms than diagnosis at an older age
- Symptom presentation varied between children less than 1 year and older than 1 year:

Age at diagnosis	Most common symptoms
≤1 year N = 13	Feeding difficulty (77%) Inspiratory stridor (69%) Cyanosis (50%) Dysphagia (46%) Laryngomalacia (46%)
> 1 year N = 6	Apnea (50%) GERD (50%) Allergic Rhinitis (50%)

CONCLUSIONS

This is the largest up-to-date review on pediatric VC. Our findings are consistent with previous studies suggesting that feeding difficulties and inspiratory stridor are the most common presentations of VC. Our preliminary data also provide new insight into the differences in symptom presentation between age groups and suggest that marsupialization is more effective in treating symptoms and preventing recurrence.

DISCLOSURES

No relevant disclosures

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Gold Laser Tonsillectomy: Effect of Energy on Postoperative Complications

Wooyoung Jang, BS[1], Cynthia Schwartz, MD[2], Jad Zeitouni, BBS[1], Akshay Raghuram, BS[1], Yusuf Dundar, MD[2]
[1]Texas Tech Health Science Center School of Medicine, Lubbock, TX, [2]University Medical Center, Department of Otolaryngology, Lubbock, TX

Abstract

The Gold Laser combines a suction tip and a flexible quartz laser fiber and is the tool of choice for many surgeons for tonsillectomies, adenotonsillectomies, and adenoidectomies.

In this retrospective study, we compared the amount of energy (kJ) utilized by the Gold Laser to postoperative complication rates. We found that there were significant correlations between higher kJ delivered and incidence of major bleeding, emergency center visits, readmission, and maximum postoperative pain scores. Furthermore, residents displayed higher energy use compared to attendings, which also differed by PGY.

Otolaryngologists utilizing the Gold Laser should be mindful of the amount of energy used and aim to use less when possible.

Introduction

The use of the Gold Laser (LF-40 GOLD Laser System, medical Energy, Inc., Pensacola, Florida, USA) in tonsillectomy and adenotonsillectomy has been shown in previous studies to be just as effective and safe compared to traditional cold steel dissection and coblation[1]. The Lightforce Gold Laser hand piece, with its suction tip and flexible quartz laser fiber, allows the surgeon to cut soft tissue, coagulate bleeds, and clear smoke, blood, secretions, and irrigation fluid with a single instrument[3].

While safe, the amount of energy delivered (kJs) varies from case to case. Currently, there are no studies regarding how the amount of energy delivered (kJs) influences the incidence of postoperative complications. In this retrospective study, we compared the effect of energy delivery by the Gold Laser on pain, clinic communications, EC visits, readmission, dehydration, bleeding, and other complications.

Methods and Materials

A retrospective chart review was performed on patients who underwent tonsillectomy, adenoidectomy, and adenotonsillectomy from 6/1/2017 to 6/1/2022.

Patients whose surgeries were performed without the Gold Laser or those with incomplete records were excluded. A total of 420 patients were included in the study. Demographic data is found in Table 1.

Statistics were performed using GraphPad Prism 9.4.1 (Dotmatics, Boston, Massachusetts, USA). D'Agostino and Pearson tests indicated that the data was not normally arranged. Therefore, appropriate nonparametric tests were selected. For univariate analyses, the Mann-Whitney test was performed. For multivariate analysis, the Kruskal-Wallis test was performed.

Results

The amount of energy used in adenoidectomies, tonsillectomies, and adenotonsillectomies differed (p=<0.0001) due to the need to remove additional tissue. Therefore, subgroup analysis was performed. There was a significant correlation between higher kJ delivered and the incidence of several postoperative results in both the tonsillectomy group and the combined group. These included major bleeding requiring cauterization in the operating room (p=0.0311, 0.0123), emergency center visits (p=0.0131, 0.0050), and readmission (p=0.0210, 0.0135) (Table 2). However, no significance was seen in relation to the number of clinic phone calls, dehydration, postoperative oral fluid intake, and minor bleeding.

The adenotonsillectomy group did not display significance in major bleeding, emergency center visits, or readmission.

Discussion

While the combined group and the tonsillectomy group showed that increased kJ led to a significant (p=0.0311, 0.0123) increase in major bleeding rates, the adenotonsillectomy group failed to show significance (p=0.1075), and the adenoidectomy group did not meet the criteria for statistical evaluation. While it did not reach significance, the combined group had lower kJ values being associated with minor bleeding, suggesting that there may be a minimum amount of kJs needed to ensure appropriate cautery and prevent minor bleeding.

However, in general, higher kJ values were associated with less desirable outcomes, such as emergency center visits in the combined and tonsillectomy groups (p=0.0131, 0.050), more clinic phone calls in the tonsillectomy group (p=0.0025), more readmissions in the combined and tonsillectomy groups (p=0.0210, 0.0135), and decreased PO intake in the adenotonsillectomy group (p=0.0040).

From a training standpoint, having a resident present led to significantly (p=<0.0001, <0.0001, 0.0091) higher kJ delivery in the combined, tonsillectomy, and adenotonsillectomy groups, indicating that residents may be somewhat more heavy-handed about their tonsillectomies than attendings.

Conclusions

The amount of energy delivered by the Gold Laser during adenoidectomy, tonsillectomy, or adenotonsillectomy showed significant correlation to several postoperative complications, most notably bleeding requiring cauterization in the OR.

As a result, clinicians utilizing the Gold Laser during these procedures should be mindful about the amount of kJ they use and aim to use less energy when possible. However, surgeons should still aim to always achieve adequate hemostasis at the time of surgery.

Table 1. Patient demographic information

	All	Adenoidectomy	Tonsillectomy	Adenotonsillectomy
Gender				
Female	223 (53.1%)	5 (23.8%)	111 (65.3%)	107 (46.7%)
Male	197 (46.9%)	16 (76.2%)	59 (34.7%)	122 (53.3%)
Age (years)	12.5 (±11.0%)	4.4 (±3.8)	21.3 (±11.8)	6.8 (±4.7)
Race				
White	375 (89.3%)	19 (90.5%)	162 (95.3%)	194 (84.7%)
Black	28 (6.7%)	1 (4.8%)	6 (3.5%)	21 (9.2%)
No Reply	13 (3.1%)	1 (4.8%)	1 (0.6%)	11 (4.8%)
American Indian or Alaska Native	2 (0.5%)	-	-	2 (0.9%)
Asian	2 (0.5%)	-	1 (0.6%)	1 (0.4%)
Weight (kg)	47.7 (±33.3)	22.0 (±11.2)	71.9 (±31.9)	32.1 (±23.0)
Indications†				
Recurrent tonsillitis	228 (33.0%)	1 (3.2%)	132 (62.3%)	95 (21.1%)
Obstructive sleep apnea	122 (17.6%)	2 (6.5%)	9 (4.2%)	111 (24.7)
Sleep disordered breathing	275 (39.7%)	6 (19.4%)	62 (29.2%)	207 (46.0%)
Adenoiditis	54 (7.8%)	21 (67.7%)	-	33 (7.3%)
Peritonsillar abscess	5 (0.7%)	-	4 (1.9%)	1 (0.2%)
Dysphagia	4 (0.5%)	-	1 (0.5%)	3 (0.7%)
Halitosis	4 (0.5%)	-	4 (1.9%)	-
Speech delay	1 (0.1%)	1 (3.2%)	-	-

† Indications may have an n greater than the total number of patients, because some patients had multiple indications, for example, patients who met the criteria for both obstructive sleep apnea and recurrent tonsillitis.

Table 2. Univariate Analysis

	Combined data for adenotonsillectomies, tonsillectomies, and adenoidectomies.				Adenoidectomies alone.				Tonsillectomies alone.				Adenotonsillectomies alone.			
	Average kJ	Median kJ	N	p	Average kJ	Median kJ	N	p	Average kJ	Median kJ	N	p	Average kJ	Median kJ	N	p
Energy delivery (kJ) with no bleeding or major bleeding only				***0.0311***				-†				***0.0123***				0.1075
Major bleeding	9757	8684	11		-	-	-		10905	11751	6		6057	7053	8	
No bleeding	7176	6549	386		4049	3611	20		6122	5934	150		8214	7861	216	
Emergency Center Visit				***0.0131***				-‡				***0.0050***				0.5222
Yes	8063	7674	54		-	-	-		7829	6769	28		8314	8110	26	
No	7063	6492	366		4102	3613	21		5988	5855	142		8120	7826	203	
Readmission				***0.0210***				-‡				***0.0135***				0.6870
Yes	8376	7989	35		-	-	-		8472	7065	17		8284	8110	18	
No	7084	6516	385		4102	3613	21		6049	5914	153		8130	7826	211	
Resident Present				***<0.0001***				0.8621				***<0.0001***				***0.0091***
Yes	7775	7364	215		4167	3727	12		7052	6561	86		8675	8494	117	
No	6579	6049	205		4015	3611	9		5512	5140	84		7584	7142	112	
Total PO < 150 mL				0.2984				-†				-				***0.0040***
Yes	9312	10438	7		3723	3723	1		-	-	-		12080	12995	5	
No	7954	7623	106		8365	8365	2		8212	8159	6		7881	7632	94	

* Nausea, vomiting, et cetera.
† There was only one episode of minor bleeding associated with adenoidectomy alone, and there were zero episodes of major bleeding associated with adenoidectomy alone. Only one patient took in less than 150 mL. Therefore, p values could not be calculated for these categories.
‡ Zero patients who underwent adenoidectomy alone presented with dehydration, to the Emergency Center, required unplanned readmission, or presented with other complications.
‡ All tonsillectomy patients who were admitted overnight drank greater than 150 mL.

Contact

Dr. Yusuf Dundar, MD
TTUHSC Department of Otolaryngology
3601 4th Street, STOP 8315
Lubbock, TX 79430-8315
Yusuf.Dundar@ttuhsc.edu
806.743.4115

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PRESENTER:
Virali Shah, BS, MBA

ABSTRACT:

Subglottic stenosis (SGS), narrowing of the upper trachea, can be an acquired condition in pediatric patients. Presenting with varying degrees of dyspnea and stridor, acquired SGS is most commonly due to intubation. Airway stenosis is not often considered a surgical complication. No literature on acquired SGS after endoscopic sinus surgery exists. We present a unique case of SGS in a 13-year-old patient with juvenile nasopharyngeal angiofibroma.

OBJECTIVES:

- The goal of this case report is to shed light on the detection and management of SGS post-operatively
- This report also reviews other risk factors for SGS development, the rising role of endoscopic surgery in pediatric population, and effective treatment options

BACKGROUND:

- Pediatric subglottic stenosis can arise from idiopathic, congenital, or acquired causes
- Most common cause of SGS is acquired from prolonged intubation; estimated incidence of 11%
- Significant risk factors for acquired SGS development post-intubation
  - Use of less sedation
  - Longer duration of intubation
  - Endotracheal tube size
  - Co-existing acid reflux

41: ACQUIRED SUBGLOTTIC STENOSIS AFTER SURGERY IN THE PEDIATRIC POPULATION: A UNIQUE CASE OF JUVENILE NASOPHARYNGEAL ANGIOFIBROMA

Virali Shah, MBA[1]; Dr. Jordon, Grube, DO[2]; Dr. Lara Reichert, MD, MPH[2]
[1]Albany Medical College
[2]Albany Medical Center; Department of Otolaryngology-Head and Neck Surgery

CASE:

13-year-old male presenting with progressive dyspnea 6 weeks after endoscopic skull base surgery for juvenile nasopharyngeal angiofibroma (JNA)

HISTORY AND PHYSICAL:

- Original presentation for JNA: 1-week history of nasal congestion, mild epistaxis, and decreased visual acuity
- CT and MRI: Large skull base mass within his pterygopalatine fossa with extension into infratemporal fossa, cavernous sinus, middle fossa, nasopharynx, and bilateral nasal cavities
- Urgent intubation required prior to preoperative embolization due to respiratory instability
- Proceeded with endoscopic skull base surgery for tumor resection

OPERATIVE PROCEDURES:

- Combined endoscopic and endoscopic-assisted open sublabial approach to the right infratemporal fossa, pterygopalatine fossa, and middle cranial fossa with right Caldwell-Luc procedure
- Right modified medial left maxillectomy, bilateral total ethmoidectomy, bilateral sphenoidotomy, posterior septectomy, bilateral middle turbinate reduction
- Resection from ventral skull base; Histopathology confirmed JNA

Postoperative Course:

Hospital Stay:
No complications
Visual symptoms resolved

1-Week Post-Op:
Difficulty with nasal breathing and mild brown-bloody nasal discharge; underwent bilateral rigid nasal endoscopy with debridement

2-Week Post-Op:
Doing well

6-Week Post-Op:
Difficulty breathing in school for 2 weeks, observed by brother too, momentary cessation in breathing 3x/day during class, reduced exercise tolerance, stridorous breathing at baseline; flexible laryngoscopy showed significant narrowing below vocal cords

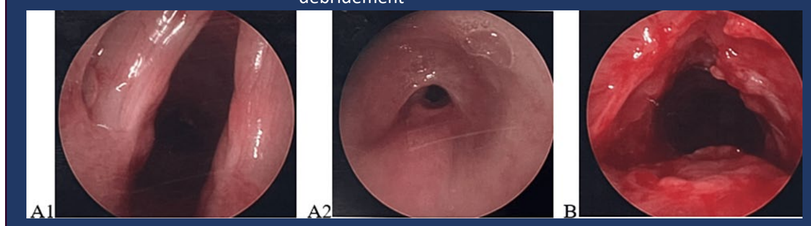

Figure 1. Rigid Bronchoscopy photos demonstrating stenosis before (A1, A2) and after dilation (B)

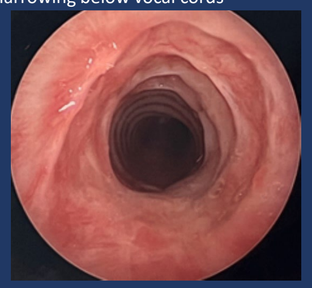

Figure 2. Rigid bronchoscopy results 2 weeks after initial dilation procedure

SGS Management:

- Anterior rhinoscopy: negative; bilateral rigid nasal endoscopy: well healing sinuses and no residual tumor
- Flexible laryngoscopy: significant narrowing below vocal cords = SGS
- Urgent intraoperative bronchoscopy:
  - Grade 3 subglottic stenosis extending into cervical trachea; 3 cm segment of stenosis; serial dilation to grade 1 subglottic stenosis; triamcinolone circumferential injection
- Repeat balloon dilation and injection at 2-weeks
  - Resolution of biphasic stridor, dyspnea, and exercise intolerance
- 1-month, 3-month, and 6-month follow-up: patient doing well

DISCUSSION:

- 90% of trauma leading to SGS is from prolonged endotracheal intubation; other forms of trauma: penetrating, blunt, and strangulation injuries are less common
- This is the first ever reported case of acquired SGS after endonasal surgery; possibility of SGS in absence of traditional risk factors
- Surgeons should maintain high degree of clinical suspicion in pts presenting with dyspnea/stridor weeks or months post-op
- Endoscopic skull base surgery has increased in pediatric populations
- Important to recognize possible SGS complication after endoscopic skull base surgery
- Treatment options have improved for SGS, ranging from endoscopic balloon dilation to cricoid split to laryngeal reconstruction

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Surgical Technique for Columellar Reconstruction in Midline Cleft Repair

Kelly M. Atherton, MSCR[1]; Corin M. Kinkhabwala, MD[2]; Krishna G. Patel MD-PhD[2]; Phayvanh P. Pecha, MD[2]
[1]College of Medicine, Medical University of South Carolina, Charleston, SC [2]Department of Otolaryngology – Head & Neck Surgery, Medical University of South Carolina, Charleston, SC

INTRODUCTION

- Estimated incidence of craniofacial clefts: 1/600-1000 live births.[1-4]
- Midline facial clefts (MFC) are a rare subset of craniofacial clefts, defined as a congenital vertical cleft through the upper lip.
- Surgical repair complicated by deformed/absent nasal cartilages, septal cartilage and columella.
- Lack of consensus around surgical technique.

CASE PRESENTATION

A total of four children with MFC presented to our craniofacial clinic between December 2010 and November 2021 (Table 1), with one patient requiring creation of a neo-columella, presented below.

A 22-month-old female presented with a large midline cleft lip and palate. Medical history included G-tube dependence. The patient was acutely transferred to our institution due to an episode of respiratory distress. A tracheostomy was performed before cleft lip repair.

The patient had an absent cartilaginous septum and columella. Palatal shelves were present with a midline complete cleft palate.

Intraoperatively, the patient was discovered to have a severely retracted prolabium and premaxilla (Figure 1). The premaxilla and prolabial remnants were attached inferiorly to the nasal cavity (Figure 2) as they were too retracted to mobilize into a more anterior position. Recovery was uneventful, and the patient was discharged on post-operative day three.

At the one-week post-operative visit, the neo-columella and lip incision remained intact without hypertrophy or signs of infection. At the eight-month post-operative visit, the midline incision appeared well approximated with a well healed scar (Figure 3), with tissue created for neo-columella to act as a scaffolding for future augmentation in this area.

OPERATIVE TECHNIQUE

- Trial of taping to ensure no airway compromise with lip closed.
- Modified straight-line design: tissue denoted as "L" flap within a Millard cleft lip repair was designed along the lateral lip edges with modifications that included dry vermilion and upper lateral lip epithelium.
- Bilateral mirrored flaps designed with intention to rotate 90 degrees medially to create nasal floor and attachment site for the neo-columella.[5]
- After incisions, orbicularis oris freed from alar and piriform attachments in a supraperiosteal plane within the midface.
- Modified "L" flaps rotated, advanced, and distal ends sutured together at the midline.
- Lateral lip edges approximated at midline and closed in multi-layered fashion: mucosa, orbicularis oris, and external skin.
- Neo-columella created by back cutting along midline internal nasal skin to create a turn-out "V"-shaped hinge flap. (This detaches the columellar skin from any membranous septum within the nose.)
- Hinge flaps were used to create a tubed neo-columella connected to the nasal floor previously created by the "L" flaps.
- In future iterations, the most distal end of the elevated "L" advancement-rotation flaps may be sutured vertically along the neo-columella to create larger nasal side walls. In this case, there was a vestigial prolabial remnant that was filleted for insertion onto the right maxillary alveolar ridge. The above-described multi-layered lip closure was performed after the neo-columella was inset.

Table 1. Patient characteristics. *case presentation patient.

Patient	Age at Procedure	Gender	Diagnosis	Associated Abnormalities
1	8 months	Female	Midline cleft lip and palate; deficient nasal dorsum and columella	Small for gestational age, failure to thrive, patent ductus arteriosus, lobar holoprosencephaly, absent corpus callosum, bilateral optic nerve coloboma, central DI, adrenal insufficiency, panhypopituitarism, hypothyroidism, microcephaly
2	9 months	Female	Midline cleft lip s/p repair; nasal hypoplasia; deficient nasal dorsum	Cerebral palsy, microcephaly, holoprosencephaly, epilepsy, developmental delay, scoliosis, Binder Syndrome, non-verbal status
3	10 months	Male	Midline cleft lip and primary palate; absent columella; absent caudal nasal septum	Left thumb polydactyly Wassel type II
4*	2 years	Female	Midline cleft lip and palate; absent cartilaginous septum; absent columella	Holoprosencephaly, 22q duplication, central diabetes insipidus (DI), metopic synostosis,

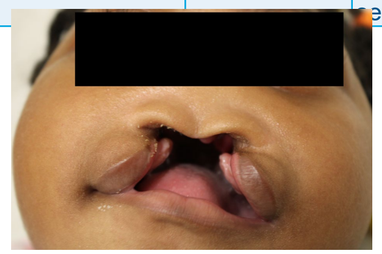

Figure 1. Pre-operative image.

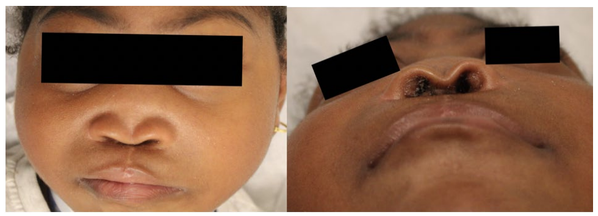

Figure 3. Eight-month post-operative images.

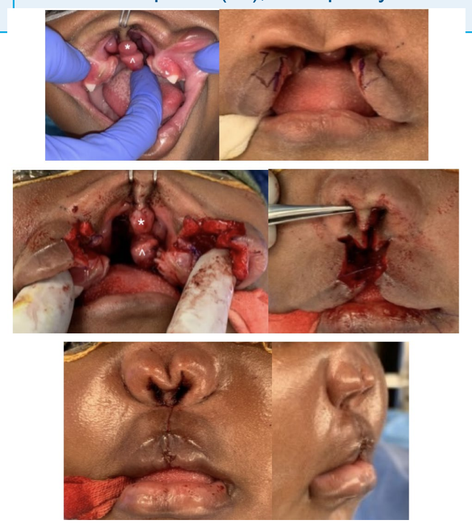

Figure 2. Intra-operative images. Prolabium denoted by *, premaxilla denoted by ^.

DISCUSSION

Midline Facial Clefts

Embryology: failure of mesodermal migration or failure of fusion of the medial nasal process, which arise from the frontonasal process.[6]

Causal factors: de novo chromosomal abnormalities and teratogens.[6]

Associated Abnormalities: neurologic malformations,[7] growth hormone deficiency, central diabetes insipidus, optic nerve hypoplasia, agenesis of septum pellucidum,[7] neonatal hypoglycemia,[8] holoprosencephaly, microcephaly, anencephaly.[7,9,10]

Associated syndromes and chromosomal abnormalities: Thurston syndrome, Pai syndrome,[9,11,12] Chromosome 18 p11.1 deletion,[10] Chromosome 20p11 deletion,[13] Chromosome 1p31 deletion,[14] and Chromosome 8p mosaic tetrasomy.[15]

Airway compromise is a serious consideration for performing and timing of repair. Midline cleft lip repair cannot be easily grouped with unilateral or bilateral lip repairs because the repair significantly narrows the airway comparatively, due to a lack of medial structures such as a premaxilla or prolabium. For this reason, MFCs are often repaired at a much later date, ideally past the time that the child is a predominately nasal breather.

Special considerations include reconstructing a columella if sufficient tissue is available to provide

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A Case Report of Bilateral Sensorineural Hearing Loss in Pediatric Tubulointerstitial Nephritis and Uveitis (TINU)-Atypical Cogan Syndrome.

Jess Rhee MSc[1], Jonathan Park MD FRCPC[2],
Lorne Parnes MD FRCSC[3], Peng You MD FRCSC[3]

[1] Medical Student at the Schulich School of Medicine and Dentistry, Western University; [2] Department of Pediatric Rheumatology, Western University; [3] Department of Pediatric Otolaryngology, Western University

Abstract

Background:
Tubulointerstitial nephritis and uveitis (TINU) syndrome is a rare, multisystem autoimmune disorder that causes inflammation of the uvea and renal tubules. Cogan's syndrome is an autoimmune condition that classically presents with interstitial keratitis as well as auditory and vestibular dysfunction. Overlap of the two syndromes have been previously described. Herein, we describe a pediatric case of TINU and atypical Cogan's with hearing loss.

Methods:
Case report and literature review

Results:
The patient was a 14-year-old Palestinian male who initially presented with abdominal pain, elevated CRP, and acute renal failure. A kidney biopsy revealed interstitial nephritis and he was started on prednisone. Prednisone was tapered off when nephritis improved, but several months later the patient reported decreased visual acuity and was diagnosed with uveitis. Subsequently, the patient noted hearing loss and was diagnosed with profound, bilateral sensorineural hearing loss refractory to medical therapy. While the patient denied vestibular symptoms, videonystagmography confirmed vestibular weakness in the right ear. The patient was diagnosed at a tertiary care center and treated with pulse dose methylprednisolone, infliximab, and methotrexate. The patient subsequently underwent cochlear implantation on the right to address his hearing loss.

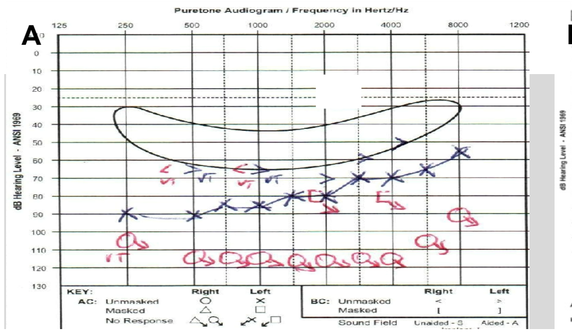
Conclusions:
This is the second documented case of hearing loss seen in TINU and atypical Cogan's. This case highlights the challenges of multisystem disease process and importance of providing multidisciplinary care. This case also showcases the importance of considering autoimmune causes of hearing loss in pediatric patients which require additional investigations and immunosuppressive therapy.

Introduction

Tubulointerstitial nephritis and uveitis (TINU) syndrome is a multisystem autoimmune disorder characterized by inflammation in the uvea and renal tubules[1].Cogan syndrome (CS) is classified primarily as a variable vessel vasculitis disease with potential systemic involvement[2]. Cogan's is typically characterized by ocular and vestibulouauditory symptoms[2]. Although both syndromes can occur at any age, both seem to frequently affect young adults and adolescents[1,2]. The diagnosis of TINU is based on the presence and combination of uveitis and renal involvement with renal biopsy consistent with acute interstitial nephritis[1]. The diagnosis of CS is based upon the presence of inflammatory eye disease and vestibuloauditory dysfunction. Table 1 demonstrates possible differential diagnoses with non-infectious inflammatory eye disease and acquired sensorineural hearing loss.

Materials and Methods

A relevant case of atypical TINU-Cogan Syndrome is presented. A literature search was conducted for peer-reviewed publications using the online search database PubMed on June 16, 2022. Search terms included: Cogan Syndrome, TINU, and atypical Cogan Syndrome in various combinations. Search results were limited to the English language. There were no restrictions for the year of publication, and additional articles were included following review of the reference list.



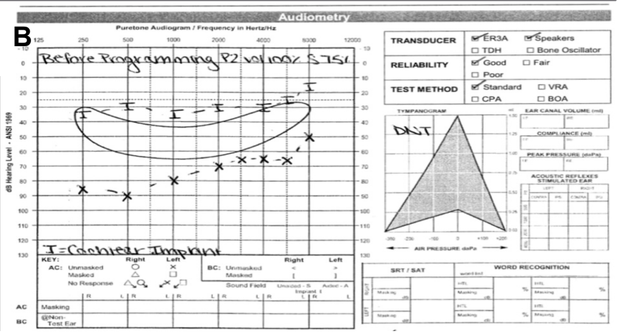


Figure 1A: Patient audiogram demonstrating bilateral sensorineural hearing loss, profound in the right ear.

Figure 1B: Patient audiogram demonstrating right cochlear implant initial post activation thresholds

Results

Case Presentation
In this case report, the patient is a 14 year old male of Palestinian ancestry. He initially presented on March 9, 2021 with right lower quadrant abdominal pain. Investigations showed elevated C-reactive protein (CRP) of 107mg/L, elevated creatinine of 150μM/L, normal urea of 3.4 mmol/L, and no hematuria or proteinuria. Subsequent kidney biopsy revealed interstitial inflammation. He was started on oral prednisone treatment and was responding well until he reported new onset of visual and auditory symptoms. He had evidence of uveitis and significant hearing loss in the right ear. Videonystagmography showed marked vestibular weakness on the right side but normal responses on the left side. He was started on infliximab, methotrexate, and folic acid as part of his medical treatment. Workup for toxoplasmosis, hepatitis, cat scratch fever, Lyme disease, syphilis, legionella, pneumocystis carinii, mycoplasma, viral and fungal infections were all negative. Genetic panel testing was also negative. Due to left severe and right profound hearing loss refractory to medical therapy, he underwent a cochlear implant procedure for his right ear. The right cochlear implant was successful and the average implant thresholds were 30 decibels. There is the potential for left ear implant if his hearing deteriorates further despite biological medical treatment.

Literature review
A total of one article was published about atypical Cogan Syndrome with TINU. The case case report presented similar symptoms of tubulointerstitial nephritis, uveitis, and vestibular failure requiring cochlear implantation[3]. High-dose corticosteroids (1mg/kg/day IV) has been shown to improve the odds of recovering hearing loss when given within 2 weeks of initial auditory symptoms[4]. When steroids are not effective, intolerance, of contraindications occur, DMARD therapy has been used as adjunct therapy[4]. Infliximab has been shown to have a 80% response rate in patients with CS and failed response of steroid and DMARD combination therapy[4,5].

Table 1. Differential for non-infectious inflammatory eye disease with acquired sensorineural hearing loss, modified from Brogan et al. 2012

Diagnosis	
Systemic Vasculitides and Autoimmune diseases	
• ANCA-associated vasculitides (GPA)	• PAN
• Kawasaki Disease	• Cogan's Syndrome
• Ectopic thymus	• SLE ± APS
• SS ± APS	• Primary APS
• IBD	• Relapsing polychondritis
• Autoimmune inflammatory arthritides	
Auto-inflammatory disorders	
• Sarcoidosis	• Behcet's Disease
• Cryopyrin-associated periodic fever syndromes	
Miscellaneous Causes	
• TINU syndromes	• Vogt-Koyanagi-Harada Syndrome
• Mitochondrial cytopathies	
• Susac's syndrome (retinocochleocerebral vasculopathy)	

Legend: granulomatosis with polyangiitis (GPA), polyarteritis nodosa (PAN), Sjogren's syndrome (SS), antiphospholipid syndrome (APS), systemic lupus erythematosus (SLE), inflammatory bowel disease (IBD)

Discussion

- This unusual presentation of atypical Cogan's syndrome with element of TINU is the second reported case
- Renal biopsy demonstrated acute interstitial nephritis and videonystagmography showed marked vestibular weakness on the right side and normal responses on the left side.
- At the time of write up, the patient received right cochlear implant and ongoing monitoring for his left sided hearing loss as well and on Infliximab every 8 weeks
- This case is the second of its kind reported in literature and highlights the complexity of symptomology of these multisystem diseases and the importance of timely treatment to prevent irreversible hearing loss

Conclusion

This case highlights the challenges of multisystem disease process and importance of providing multidisciplinary care. This case also showcases the importance of considering autoimmune causes of hearing loss in pediatric patients which require additional investigations and immunosuppressive therapy.

Corresponding author

Dr. Peng You, MD, FRCSC
Department of Otolaryngology-Head and Neck Surgery
London Health Science Centre
Schulich School of Medicine & Dentistry
Western University
peng.you@lhsc.on.ca

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Paediatric Sjögren's syndrome with bilateral parotid cysts: A case report.

Jamila Skinner, BSc, James Fowler, MD[1],
Jonathan Park MD FRCPC[2], Peng You, MD FRCSC[1]
[1]Department of Otolaryngology – Head and Neck Surgery, Western University, London, Ontario, Canada; [2] Department of Pediatric Rheumatology, Western University

Abstract

Introduction:
Sjögren's syndrome is an autoimmune disease characterized by the destruction of exocrine glands. Clinically, this results in the loss of tear and saliva production. Although xerophthalmia and xerostomia, also known as sicca, is a common presentation among adults, pediatric patients more often present with recurrent parotitis and glandular enlargement. Overall symptoms can vary, making initial diagnosis challenging. Approximately 80% of patients with Sjögren's syndrome experience parotid gland enlargement, however, salivary cysts are rare. Herein, we present a case of pediatric Sjögren's syndrome that presented as bilateral parotid cysts.

Methods:
A unique case of Sjögren's syndrome which presented with parotid gland enlargement is reported. A comprehensive search was conducted for cases of Sjögren's syndrome presenting with parotid gland enlargement in the English-language.

Results:
A 12-year-old female presented with a 2-month history of bilateral parotid masses. The patient denied any history of xerostomia, xerophthalmia, or constitutional symptoms. Imaging revealed bilateral complex cystic intraparotid masses. A right parotid gland biopsy was performed showing parotid gland parenchyma with dense lymphoplasmacytic infiltrate. Ultimately the presumptive diagnosis of Sjogren's syndrome was made

Conclusion:
We present a unique case of Sjogren's syndrome with bilateral intraparotid cysts. This case illustrates the importance of a thorough workup to aid in diagnostic certainty. Parotid cysts associated with Sjogren's are rare but should be considered within the differential diagnosis for pediatric patients with parotid swelling/mass.

Introduction

Sjögren's syndrome is an autoimmune disease whereby the destruction of exocrine glands results in the loss of tear and saliva production.

The diagnostic criteria for Sjögren's syndrome is both complex and controversial. The 2002 AECG criteria is the most commonly used diagnostic tool.

Diagnosis: 4 of 6 criteria (must include either IV or VI) or 3 of 4 objective criteria (III, IV, V, VI).

AECG Criteria:

I. Ocular symptoms (at least 1)
  I. Dry eyes for x 3 months.
  II. Ocular foreign body sensation.
  III. Use of artificial tears 3x per day.
II. Oral symptoms (at least 1)
  I. Dry mouth x 3 months.
  II. Recurrent or persistently swollen salivary glands.
  III. Need for liquids for swallowing dry foods.
III. Ocular signs (at least 1)
  I. Abnormal Schimer's Test (<5 mm/5 min).
  II. Positive vital dye staining of eye surface.
IV. Histopathology
  I. Biopsy showing focal lymphocytic sialadenitis.
V. Oral signs (at least 1)
  I. Unstimulated whole salivary flow (≤ 15 ml/ 15 min).
  II. Abnormal parotid sialography.
  III. Abnormal salivary scintigraphy.
VI. Autoantibodies (at least 1)
  I. Anti-SSA (Ro).
  II. Anti-SSB (La).

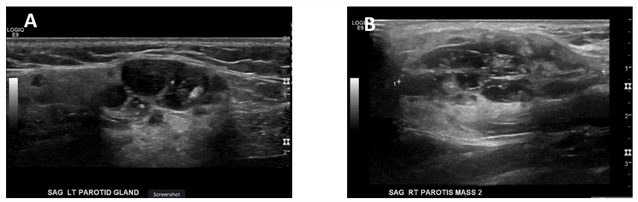

Figure 1: : Case presentation of Sjögren's syndrome. Sagittal ultrasound of left (A) and right (B) parotid glands depicting bilateral complex solid/cystic masses intraparotid glad masses.

Case

A 12-year-old female presented with a 2-month history of bilateral parotid masses. She reported no history of xerostomia, dry eyes, or B symptoms.

Ultrasound imaging revealed bilateral complex cystic intraparotid masses measuring 4.7 x 3.1 x 1.7 cm on the right with a second, more posterior and deep mass measuring 4.3 cm at its greatest diameter. The left mass measured 3.2 cm. **[Figure 1]**

Magnetic resonance imaging confirmed numerous cystic lesions on the parotid bilaterally. These were hyperintense on T2, hypointense on T1. **[Figure 2]**

Parotid gland parenchyma with dense lymphoplasmacytic infiltrate was found on parotid gland biopsy.

Given the lack of diagnostic certainty, serology was ordered. Work-up was negative for infectious etiology such as HIV, mumps, CMV, and EBV. Serology was positive for rheumatoid factors, as well as anti-Ro/SSA and anti-La/SSB antibodies consistent with a diagnosis of Sjogren's syndrome. The patient was subsequently treated with hydroxychloroquine.

Discussion

- Typical differential of parotid cysts include: mumps, HIV, lymphatic malformation
- Occurrence of parotid cyst with Sjögren's Syndrome has been reported but is very rare especially in the paediatric population
- Parotid swelling, if unpainful, should warrant imaging whereby in contrast, painful parotitis may first be treated with conservative measures such as massage, analgesics, and sialagogues

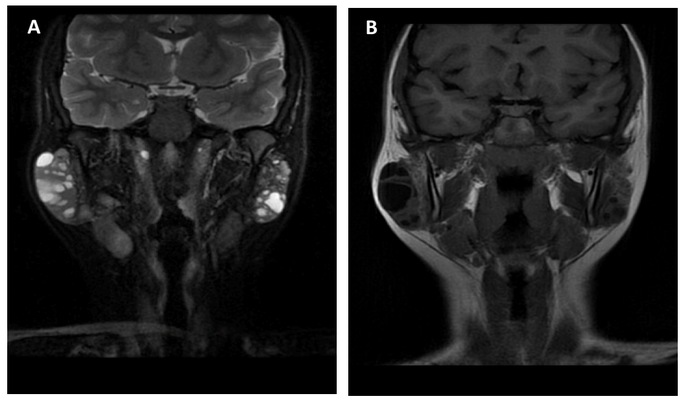

Figure 2: Case presentation of Sjögren's syndrome. T2 weighted **(A)** and T1 weighted **(B)** MR head/neck demonstrate numerous cystic lesions in the partoid glands bilaterally. Imaging criteria found no pathological lymph nodes.

Conclusion

This is a unique case of pediatric Sjogren's syndrome with bilateral intraparotid cysts. Our case illustrates the importance of thorough workup to aid in diagnostic certainty. Parotid cysts associated with Sjogren's are rare, but should be considered within the differential diagnosis for any patient with parotid swelling/mass.

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Topical Antibiotic Ear Drop Use at the Time of Tympanostomy Tube Placement: A Retrospective Analysis of the Pediatric Health Information System (PHIS) Database

Hailey Brigger[1], Matt Hall Ph.D.[2], Daniela Carvalho M.D., M.M.M.[3], Shelby Leuin M.D.[3], Wen Jiang M.D.[3]

[1]Vassar College, Poughkeepsie, NY, USA. [2]Children's Hospital Association, Lenexa, KS, USA.

[3]Department of Otolaryngology, UCSD; Rady Children's Hospital San Diego, CA, USA.

Introduction

Tympanostomy tube placement (TTP) is the most common ambulatory surgery in the United States, with 670,000 cases performed in children under the age of 15 each year.[1] This procedure is most commonly performed for recurrent acute otitis media, persistent middle ear fluid, or persistent ear infections despite antibiotic therapy.[2] Controversy exists regarding the intraoperative and perioperative use of antibiotic ear drops. Recent clinical practice guidelines published a recommendation against routinely prescribing antibiotic drops for tympanostomy tube otorrhea prevention, with perceived overuse of antibiotic drops and recommended saline drops.[3]. The purpose of this study is to assess tympanostomy tube complications and costs at a population level and compare current practice to recently published guidelines.

Table 1. Baseline patient characteristics

	Total	No antibiotic drops	Antibiotic drops	*p*
N Discharges (%)	**445808**	**164803 (37.0)**	**281005 (63.0)**	
Principal Diagnosis				
Acute otitis media	4836 (1.1)	1160 (0.7)	3676 (1.3)	<.001
Chronic otitis media	141787 (31.8)	57890 (35.1)	83897 (29.9)	
Unspecified otitis media	299185 (67.1)	105753 (64.2)	193432 (68.8)	
Age (y)				
<1	85508 (19.2)	31803 (19.3)	53705 (19.1)	<.001
1-4	294435 (66)	107869 (65.5)	186566 (66.4)	
5-9	50276 (11.3)	19065 (11.6)	31211 (11.1)	
10-14	12282 (2.8)	4797 (2.9)	7485 (2.7)	
15-18	3307 (0.7)	1269 (0.8)	2038 (0.7)	
Sex				
Male	262060 (58.8)	96830 (58.8)	165230 (58.8)	0.754
Female	183714 (41.2)	67966 (41.2)	115748 (41.2)	
Race/Ethnicity				
Non-Hispanic White	301904 (67.7)	114210 (69.3)	187694 (66.8)	<.001
Non-Hispanic Black	44722 (10)	15663 (9.5)	29059 (10.3)	
Hispanic	57355 (12.9)	19559 (11.9)	37796 (13.5)	
Asian	8005 (1.8)	3001 (1.8)	5004 (1.8)	
Other	33822 (7.6)	12370 (7.5)	21452 (7.6)	
Payor				
Private	253833 (56.9)	101083 (61.3)	152750 (54.4)	<.001
Government	168867 (37.9)	58794 (35.7)	110073 (39.2)	
Other	23108 (5.2)	4926 (3)	18182 (6.5)	
Disposition				
Home	417767 (93.7)	160914 (97.6)	256853 (91.4)	<.001
Home Health	85 (0)	26 (0)	59 (0)	
Skilled Facility	98 (0)	34 (0)	64 (0)	
Other	27858 (6.2)	3829 (2.3)	24029 (8.6)	
Complex Chronic Conditions, any	33159 (7.4)	12487 (7.6)	20672 (7.4)	0.007

Contact: Hailey Brigger, hbrigger@vassar.edu

Methods

- A retrospective cohort analysis using the Pediatric Health Information System (PHIS) database, for patients 0 - 18 yrs of age, with a principal procedure of myringotomy and tube placement from 1/1/2010 – 12/31/2020.
- PHIS is a comparative administrative database from 49 freestanding tertiary care children's hospitals.
- The main outcome measure was the use of antibiotic drops during the index encounter and the rate of tube re-insertion within 90 days. Pharmacy billing data was used to report on the administration of both topical antibiotic and topical antibiotic/steroid combination drops.
- Secondary outcomes included total hospital costs (estimated from charges using annual hospital-specific cost-to-charge ratios), 7-day all-cause return to the emergency department (ED) and 7-day all-cause readmissions.
- The Cochran-Armitage Trend Test was used to assess trends. A generalized estimating equation model was fitted for the multivariable mode for re-insertion of ear tubes within 90 days, accounting for patient clustering within the hospital (need input from statistician). Results are presented as adjusted odds ratios (AOR) with 95% confidence intervals. All statistical analyses were performed using SAS v. 9.4 (SAS Institute, Cary, NC) with statistical significance set at $p < .05$.

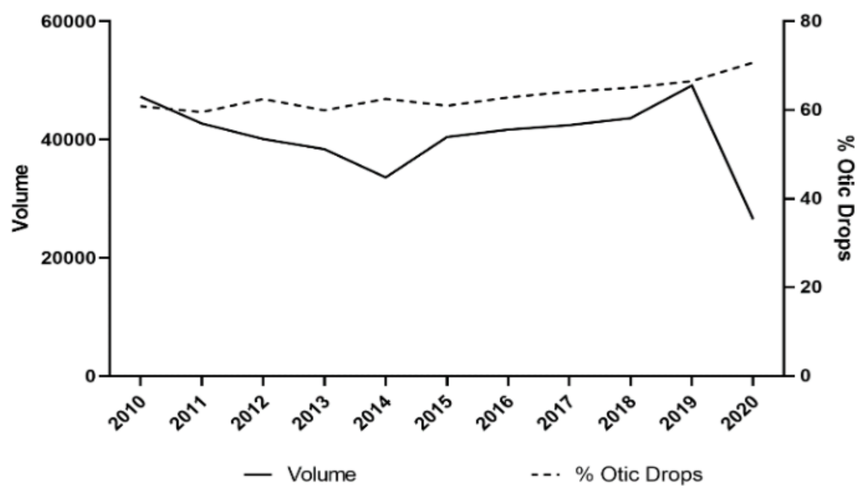
Table 2. Univariate Outcome

	Overall	No Antibiotic drops	Antibiotic Drops	p
Cost ($): Geometric Mean (Std Dev)	1872.3 (1.8)	2014.3 (1.8)	1793.7 (1.8)	<.001
Re-insertion of ear tubes within 90 days: N (%)	1702 (0.4)	836 (0.5)	866 (0.3)	<.001
Readmission within 7 days: N (%)	1106 (0.2)	435 (0.3)	671 (0.2)	0.103
Readmission within 30 days: N (%)	3546 (0.8)	1291 (0.8)	2255 (0.8)	0.488
ED visit within 7 days: N (%)	5445 (1.2)	1867 (1.1)	3578 (1.3)	<.001
ED visit within 30 days: N (%)	17734 (4)	6019 (3.7)	11715 (4.2)	<.001

Table 3. Multivariable analysis for re-insertion of ear tubes within 90 days

	Adjusted Odds Ratio (95% CI)	*p*
Treatment		
No antibiotic drops	Reference	
Antibiotic drops	0.6 [0.4, 0.9]	0.018
Principle Diagnosis		
Acute otitis media	Reference	
Chronic otitis media	0.8 [0.5, 1.4]	0.454
Unspecified otitis media	0.7 [0.5, 0.7]	0.018
Age	1.0 [0.9, 1.0]	0.600
Race/Ethnicity		
Non-Hispanic White	Reference	
Non-Hispanic Black	1.0 [0.8, 1.2]	0.685
Hispanic	0.8 [0.6, 1.1]	0.112
Asian	0.8 [0.5, 1.3]	0.302
Other	0.7 [0.5, 1.0]	0.040
Payor		
Private	Reference	
Government	1.0 [0.9, 1.3]	0.650
Other	0.8 [0.7, 1.1]	0.149
Complex Chronic Conditions	1.5 [1.2, 1.9]	<0.001

Figure 1. Temporal changes in case volume of tube placement during the study period, and the percent utilization of otic drops. The Cochran-Armitage test was used to assess trend, with p (trend) < 0.001.



Result

- **445,808** ambulatory surgery discharges met criteria; **37.0% no antibiotic drops, 63.0% antibiotic drops**
- For principal diagnosis, 1.1% had acute otitis media, 31.8% had chronic otitis media, the majority (67.1%) listed as "unspecified otitis media"
- Overall mean cost of was $1773.5. For encounters where antibiotics drops were used the mean cost was $1691.6, encounters where no antibiotics were used was $1923.2 ($p<0.001$)
- Antibiotic drops were associated with less rate of ear tube re-insertion with 90 days, with aOR 0f 0.6, ($p<0.018$)

Discussion

- Antibiotic drops were associated with less cost for the surgical encounter when compared to encounters without antibiotic use
- There appears to be a large gap between current practices and the new guideline recommendations
- There was a statistical difference of tube re-insertion in favor of antibiotic drops, however the absolute difference was small, 0.2%.
- Overall ED visit and re-admission rates were quite low

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Effectiveness of Prenatal Diagnosis in Micrognathia: A Systematic Review and Meta-analysis

Caroline M Fields, BS; Nicolas S Poupore, BS; April N Taniguchi, BS; Hussein Smaily, MD; Shaun A Nguyen, MD, MA; Phayvanh P Pecha, MD; Ryan D Cuff, MD; William W Carroll, MD

Medical University of South Carolina, Department of Otolaryngology – Head and Neck Surgery, Charleston, SC, USA

ABSTRACT

Objective: Studies evaluating the ability to diagnose and accurately predict the severity of micrognathia prenatally have yielded inconsistent results. This review aimed to evaluate reliability of diagnosing prenatal micrognathia and postnatal diagnostic congruence.

Methods: Per PRISMA guidelines, a systematic review using PubMed, Scopus, and CINAHL databases was performed. Studies using a subjective (radiologist's discretion) or objective (mandibular measurements) prenatal diagnosis of micrognathia via ultrasound with a confirmatory postnatal examination were included. Prenatal severity was defined using objective and subjective measures. Postnatal severity was defined as respiratory obstruction at birth requiring intubation or surgical intervention. Meta-analyses of proportions and relative risk were performed.

Results: A total of 16 studies with 2,753 neonates were included. The false-negative rate of prenatal micrognathia diagnosis predicting postnatal micrognathia diagnosis was 11.62% (95%CI 2.58-25.94); the false-positive rate was 2.19% (95%CI 0.24-6.01). Utilizing objective parameters, false-negative rates were statistically lower (0.20% [95%CI 0.00-0.70]), but not false-positives (1.37% [95%CI 0.71-2.22]). Patients determined to have severe micrognathia by prenatal imaging did not have a statistically significant increase in risk of being born with severe micrognathia (RR 3.13 [95%CI 0.59-16.55], p=0.180) (Figure 11).

Conclusion: The false-negative rate of micrognathia diagnosis via prenatal subjective determination was over 1 in 10, with objective measures improving accuracy. Jaw index and inferior facial angle were mainly used; however, each study used different cutoffs that were retroactively applied. This study highlights the need for a uniform objective criterion, potentially combining two measurements, to improve prenatal diagnosis and planning for postnatal care.

METHODS

- **Search Criteria:**
  - PubMed, Scopus, and CINAHL (3/3/22)
  - Search strategy: "micrognathia", "prenatal diagnosis", "ultrasound or MRI"
- **Selection Criteria:**
  - Inclusion: objective or subjective prenatal diagnosis of micrognathia, confirmatory postnatal examinations, measurement of prenatal severity, postnatal respiratory interventions
  - Exclusion: non-English, non-human studies, review articles, case reports <4 patients, and duplicates
- **Outcomes:**
  - Primary: efficacy of characterizing postnatal severity of micrognathia via prenatal diagnosis
  - Secondary: accuracy of prenatal diagnosis using subjective and objective methods
- **Quality Assessment:**
  - Risk of Bias in Non-Randomized Studies – of Interventions (ROBINS-I)
- **Statistical Analysis:**
  - Meta-analysis of single means: mean, N for each study, and standard deviation (Comprehensive Meta-analysis 3)
  - Meta-analysis of proportions: false positive and false negative (MedCalc 18.10.2)
  - Meta-analysis of relative risk: mild or severe micrognathia pre- and post-natally (MedCalc 18.10.2)

RESULTS

Figure 1. PRISMA Flowchart

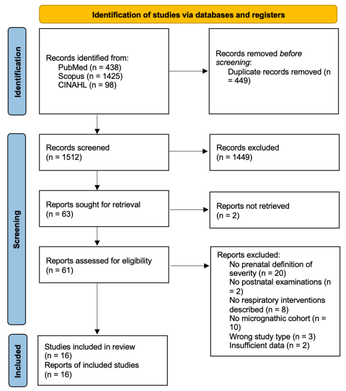


Figure 2. Relative Risk of Severe Micrognathia at Birth for Patients with Severe Micrognathia on Prenatal Imaging

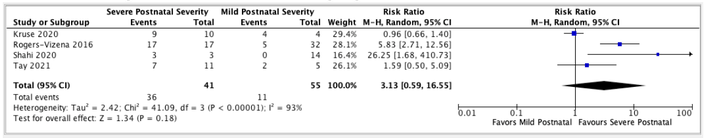


Figure 3. Relative Risk of Severe Micrognathia at Birth for Patients with Mild Micrognathia on Prenatal Imaging

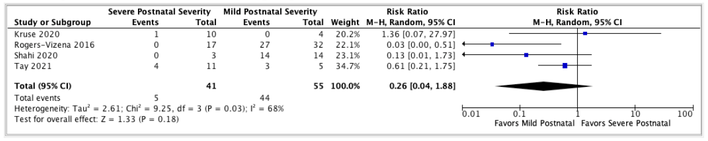


Figure 4. Proportion of False Negative Diagnoses by Subjective Determination on Prenatal US

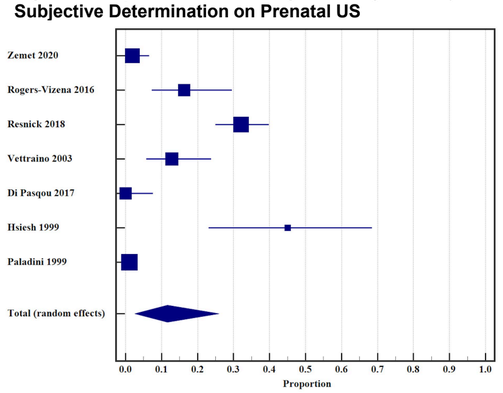
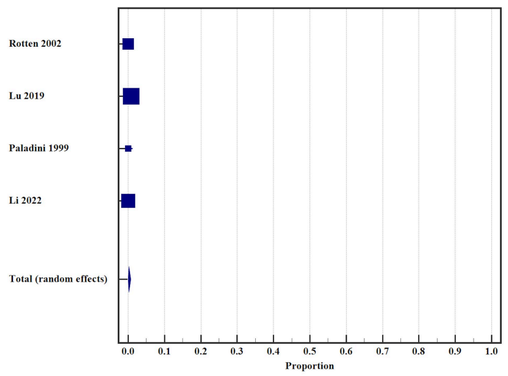


Figure 5. Proportion of False Negative Diagnoses by Objective Determination on Prenatal US



CONCLUSIONS

- This review demonstrated that when micrognathia was diagnosed subjectively, more than 1 in 10 cases of micrognathia were missed.
- Objective methods demonstrated an 11.41% decrease in the rate of false negative diagnoses.
- At present, the limited data from this study were inconclusive as to whether severity can be accurately predicted from prenatal imaging.
- A prospective observational trial using standardized normative values of mandibular measurements would better elucidate the accuracy of predicting micrognathia severity and diagnosis on prenatal imaging.

SUMMARY

- Currently, there are no guidelines that incorporate objective measures for use in examining the fetal facial profile on routine prenatal imaging.
- This review demonstrates that objective parameters may potentially increase the sensitivity of prenatal diagnosis.
- Furthermore, due to the paucity of literature regarding prenatal predictions of severity of micrognathia, it is difficult to ascertain the accuracy in prediction of severity as it relates to risk of postnatal respiratory obstruction.
- The heterogeneity of the current literature calls for the creation of a uniform, objective criterion for evaluating micrognathia on routine prenatal sonogram in order to optimize preparation for potential perinatal airway complications.

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Factors Associated with Eosinophilic Esophagitis in an, Urban Tertiary Care Pediatric Aerodigestive Population Undergoing Triple Endoscopy

Sheila Moran BS/BA[1], Cassidy Anderson BS[1], Risha Sheni BS[1], Daniel Li MD[2], Monica Azmy MD[3], Anthony M. Loizides MD[1], Christina J. Yang MD[1,3,4].

1. Albert Einstein College of Medicine, Bronx, NY
2. Department of Otolaryngology, Yale School of Medicine, New Haven, CT
3. Department of Otorhinolaryngology-Head and Neck Surgery, Montefiore Medical Center, Bronx, NY
4. Department of Pediatrics, Children's Hospital at Montefiore, Bronx, NY

Background

Triple endoscopy (flexible bronchoscopy, rigid direct laryngoscopy and bronchoscopy, and esophagogastroduodenoscopy with biopsy) is a key component in the diagnosis and management of pediatric aerodigestive complaints. Eosinophilic esophagitis (EoE) may present with cough, dysphagia for solids, or asymptomatic in patients screened prior to airway reconstruction. The factors associated with histopathologic diagnosis of EoE have not been elucidated.

Methods

Study Design

A retrospective chart review was performed of all pediatric patients, aged 0-21 years, who underwent triple endoscopy at an urban, tertiary care children's hospital from January 1, 2015 to December 31, 2019. Bivariate statistical analysis and multivariable regression were used to compare the demographics and clinical characteristics of patients with and without EoE.

Statistical Analysis

Descriptive statistics were used to summarize the overall sample. Bivariate statistical analysis was conducted to compare the baseline characteristics between patients with and without EoE and assess for potential predictors of EoE. Variables with alpha ≤ 0.1 on bivariate analysis were selected for inclusion in the multivariate logistic regression model.

Results

119 cases were included in the analysis. 15.97% (19) received a diagnosis of EoE following triple endoscopy. The most common indication for triple endoscopy was upper airway obstruction (47% found to have EoE and 24% without EoE). Patients with EoE were more likely to have a personal history of eczema (OR= 4.02 (1.3-12.49); p = 0.016) and dairy-free diet (OR = 0.26 (0.08-0.81); p = 0.02). Age, gender, recency of PO, and tracheostomy tube prior to the endoscopy were not found to be associated with increased odds of EoE.

Table 1. Demographics and clinical characteristics

		EOE (N=19)	No EOE (N=100)	p-value
Age, median (Interquartile range)		4.3(2.6-7.9)	3.6(2.0-6.4)	0.50
Male, n (%)		14(73.7)	62(62.0)	0.33
Race, n (%)				
	Non-Hispanic White	0(0)	7(7.0)	0.47
	Non-Hispanic Black	6(31.6)	26(26.0)	
	Hispanic	11(57.9)	45(45.0)	
	Other	2(10.5)	22(22.0)	
History of allergies, n (%)				
	No	7(36.8)	59(60.2)	0.06
	Yes	12(63.2)	39(39.8)	
History of asthma, n (%)				
	No	6(31.6)	32(32.3)	0.95
	Yes	13(68.4)	67(67.7)	
Rating of asthma, n (%)				
	Mild	9(69.2)	31(55.4)	0.71
	Moderate	4(30.8)	20(35.7)	
	Severe	0(0)	5(8.9)	
Family history of eczema, n (%)				
	No	8(42.1)	74(74.0)	0.01
	Yes	11(57.9)	26(26.0)	
Family history of atopy, n (%)				
	No	9(47.4)	52(52.0)	0.71
	Yes	10(52.6)	48(48.0)	
Started PO within last 6 months, n (%)		14(73.7)	86(86.9)	0.17
	No	5(26.3)	13(13.1)	
	Yes			
Diet includes gluten, n (%)				
	No	5(26.3)	24(24.5)	1.0
	Yes	14(73.7)	74(75.5)	
Diet includes diary, n (%)				
	No	8(42.1)	22(22.5)	0.09
	Yes	11(57.9)	76(77.5)	
Trach at time of exam, n (%)				
	No	11(61.1)	77(78.6)	0.14
	Yes	7(38.9)	21(21.4)	

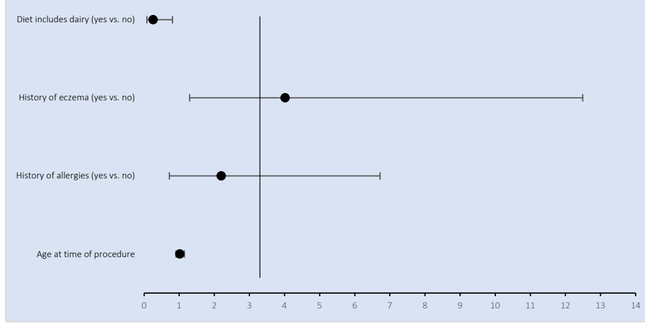
Table 2. Indications for Triple Endoscopy

	Whole sample (N=119)	EoE (N=19)
Indication for triple scope, n (%)		
Uncontrolled/persistent asthma	9 (7.6)	1 (5.3)
Chronic cough	12 (10.1)	3 (15.8)
Recurrent pneumonia	13 (10.9)	0
Recurrent croup	18 (15.1)	2 (10.5)
Dysphagia	28 (23.5)	4 (21.1)
TEF	4 (14.8)	1 (25.0)
Vascular Ring	1 (3.7)	0
Hematemesis	3 (11.1)	0
Unspecified	19 (70.4)	3 (75.0)
GERD	5 (4.2)	0
Airway Stenosis	29 (24.4)	9 (47.4)
Glottic stenosis	12 (41.4)	2 (22.2)
Subglottic stenosis	1 (3.5)	3 (33.3)
Tracheal stenosis	11 (37.9)	4 (44.4)
Multiple levels	5 (17.2)	0
Hemoptysis	3 (2.5)	0
Failure to thrive	1 (0.8)	0
ALTE/ BRUE	1 (0.8)	0

Table 3. Multivariable Analysis

Variable	Odds Ratio (95% CI)	p-value
Age at time of procedure	1.02 (0.91-1.15)	0.73
History of allergies	2.20 (0.72-6.72)	0.17
History of eczema	4.02 (1.3-12.49)	0.02
Diet includes dairy	0.26 (0.08-0.81)	0.02

Figure 1. Multivariable Analysis



Conclusions

In this retrospective, cohort study, a history of eczema and dairy-free diet were associated with increased odds of EoE in patients who underwent triple endoscopy in our population. Larger, multi-institutional studies are needed to identify early predictors of EoE in pediatric aerodigestive populations.

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Psychosocial Measures and Outcomes Among Caregivers of Children with Tracheostomies: A Systematic Review

Darlene E. Acorda, PhD, RN, CNE, CPNP-PC; Jennifer N. Brown, MPAS, PA-C;
Elton M. Lambert, MD; Karen DiValerio Gibbs, PhD, MSN/MPH, RN

Background

- Children with tracheostomies have complex medical issues with many requiring continuous medical care at home.
- Parents often assume the majority of care which can lead to a shift in family dynamics and a decrease in caregiver quality of life.
- To date, no systematic review has examined quantitative measures in this population.
- We aimed to identify instruments used to measure caregiver psychosocial outcomes after their child's tracheostomy

Methods

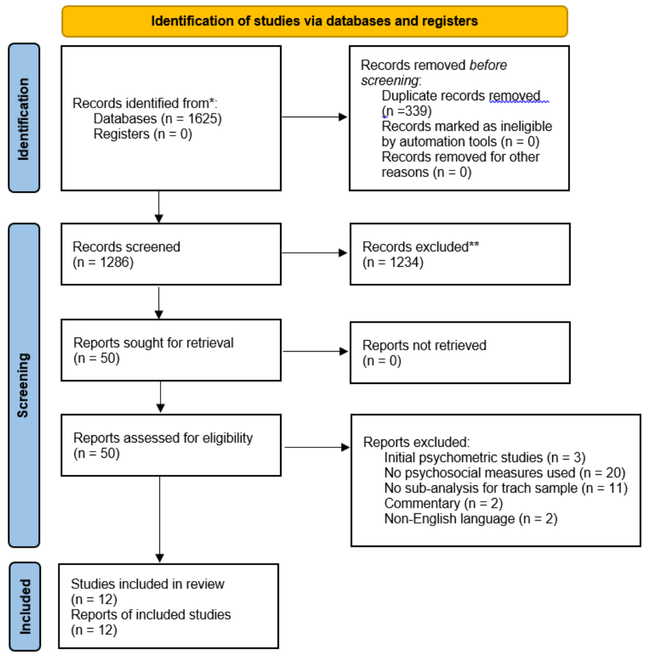
- Databases searched: Medline, CINAHL, EMBASE
- Studies that evaluated psychosocial outcomes in caregivers of pediatric patients with tracheostomy were included.
- Data was extracted using a form piloted by the authors.
- Author resolved conflicts through review and discussion.

Results

- Psychosocial constructs evaluated included: quality of life, stress, coping, family functioning, health status, decision conflict, and regret.
- The Pediatric Tracheostomy Health Status Index was the only instrument validated in this population.

Results

PRISMA Flow Diagram



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Supplemental table of Instruments and Outcomes

INSTRUMENTS IDENTIFIED:

- Pediatric Tracheostomy Health Status Index (PTHSI)
- Medical Outcomes Study Short Form (SF-12)
- Decisional Conflict Scale (DCS)
- Decision Regret Scale (DRS)
- Adult Caregiver Quality of Life Survey (ACQOL)
- Family Inventory of Life Events (FILE)
- Family Crisis Oriented Personal Evaluation Scale (F-COPES)
- Psychological General Well-Being Index (PGWBI)
- Impact on Family Scale (IOFS)
- Family APGAR
- Feetham Family Functioning Survey (FFFS)
- Chronicity Impact and Coping Instrument (CICI:PQ)
- PedsQL Family Impact Module
- Parenting Stress Index 4th edition Short Form (PSI-4-SF)

Discussion

- Caring for a tracheostomy-dependent child significantly impacts caregiver quality of life, adds profound stress, and affects family structures and functioning.
- The heterogeneity of measures identified in this review makes it difficult to compare findings within this population
- Scores on commonly used measures such as the PedsQL and IFS were comparable to those reported in caregivers of children with medical complexity and other chronic conditions
- Unclear if the psychosocial outcomes assessed by the measures in this review are specific to tracheostomy placement or the particular disease process of the child
- Lack of pre and post QoL assessments make it difficult to glean true impac tof tracheostomy on pre-existing psychosocial status

Limitations

- May have missed instruments used routinely in practice but have not been published in academic work
- Only English studies included and may have overlooked publications in other languages
- Heterogeneity of the instruments and lack of comparison to normative data make it difficult to compare findings

Conclusions

- Few quantitative studies have explored the psychosocial outcomes of caregivers of children with tracheostomies despite findings suggesting significant impacts on caregiver quality of life and family functioning
- More studies are needed in diverse caregiver populations using validated measures to understand the full impact of a tracheostomy on caregiver well-being
- Longitudinal studies are needed to examine the long-term impact and changes in the constructs identified in this review.

References

Access references by scanning QR code

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Deep Neck Space Infections Mimicking Kawasaki Disease: A Systematic Review

52 SENTAC

Henya Sandhaus, DO[1], Albert Botchway, PhD[2], Dana L Crosby, MD MPH[1], Arun Sharma, MD[1], Stacie R Gregory, MD[1]

Introduction

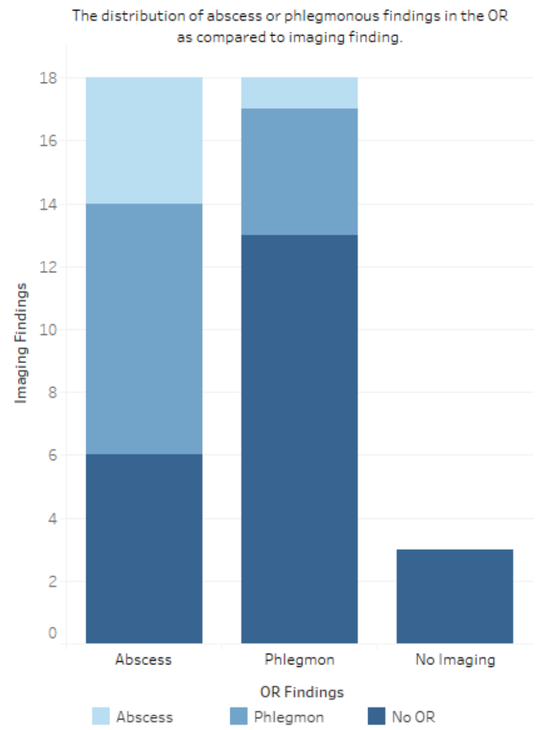
- Kawasaki Disease (KD), also known as Mucocutaneous Lymph Node Syndrome (MLNS), is an acute, self-limiting pediatric vasculitis primarily affecting the coronary arteries[1, 2]
- Early diagnosis is important for prevention of coronary artery aneurysm[3]. Despite diagnostic criteria from the American Heart Association, KD is a diagnosis of exclusion
- Otolaryngologic presentations of KD may mimic deep neck space infections (DNI) delaying diagnosis
- Few retrospective chart reviews have been performed in attempt to classify clinical, laboratory and imaging differences to help distinguish the two conditions[4-6]
- The goal of this systematic review is to quantitatively and qualitatively describe the atypical otolaryngologic manifestations of KD that may mimic DNI

Methods and Materials

- Systematic review of literature between 4/28/2020 and 5/4/2020
- Search criteria included KD or MLNS related to DNI or Otorhinolaryngology
- Prisma analysis for 45 qualitative and 34 quantitative analysis
- **Independent Variable**: Kawasaki disease
- **Covariates**: Age, gender, race,
- **Dependent variables**:
  - Symptoms present at hospital presentation
  - Symptoms developed during hospital stay
  - Abscess/phlegmon on imaging
  - Abscess/phlegmon intraoperatively
  - Location of lymphadenopathy/abscess/phlegmon
- Data analyzed using Fischer's exact stratified by location of DNI

		Mean ± SD or Frequency (%)
Age	Age	5.54 (3.64)
Gender	Male	31 (79.5)
	Female	8 (20.5)
Race	Asian	3 (7.7)
	Caucasian	8 (20.5)
	African American	2 (5.1)
	Unknown	26 (66.7)

		Imaging Report	
		Abscess	**Phlegmon**
Type Of Imaging	Ultrasound	0 (0.0%)	3 (16.6%)
	CT	15 (83.3%)	9 (50.0%)
	MRI	3 (16.7%)	4 (22.2%)
	Soft tissue x-ray	0 (0.0%)	2 (11.1%)
Imaging Results	Adenopathy	0 (0.0%)	10 (55.6%)
	Hypodensity	6 (33.3%)	4 (22.2%)
	Area of low attenuation	1 (5.6%)	4 (22.2%)
	Abscess	11 (61.1%)	0 (0.0%)
Ring Enhancement	Yes	4 (22.2%)	0 (0.0%)
	No	7 (38.9%)	9 (50.0%)
	Unknown	7 (38.9%)	9 (50.0%)
Laterality	Left	8 (44.4%)	7 (38.9%)
	Right	3 (16.7%)	7 (38.9%)
	Both	6 (33.3%)	4 (22.2%)
	Unknown	1 (5.6%)	0 (0.0%)



Results

- 39 cases were reviewed from 34 publications
- The mean age at diagnosis was 5 years and approximately 79% were male
- 92% of patients underwent imaging at 50% between abscess and phlegmon. Only 13% had confirmed abscess in the operating room
- 52% of patients diagnosed with DNI underwent medical management

Conclusions

- KD can mimic DNI on radiology
- Radiology often non-congruent with intraoperative findings.
- This should raise suspicion for alternative diagnosis in peds patient

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Contact

Henya Sandhaus, DO
SIU SOM
720 N Bond St, Springfield IL
hsandhaus21@siumed.edu

Acute Labyrinthitis associated with COVID-19 Infection

Wen Jiang, M.D.,[1,2] Ellen Smith, Au.D.,[2]

[1] Department of Otolaryngology, University of California San Diego, School of Medicine, San Diego, CA, USA [2v]Rady Children's Hospital, San Diego, CA, USA

Introduction

Sudden sensorineural hearing loss (SSNHL) is defined as a hearing loss of at least 30 dB in at least three consecutive frequencies that has developed within 3 days[1]. It affects 5 to 27 per 100,000 people annually, with about 66,000 new cases per year in the United States[2]. The etiology is still unknown, with attributable causes including viral infection, autoimmune disease, vasculitis and tumors. Viral causes have been described, mainly from the herpesviruses. We present a pediatric case of unilateral SSNHL and labyrinthitis in a pediatric patient with an acute COVID-19 infection.

Methods

Case report with review of literature

Results

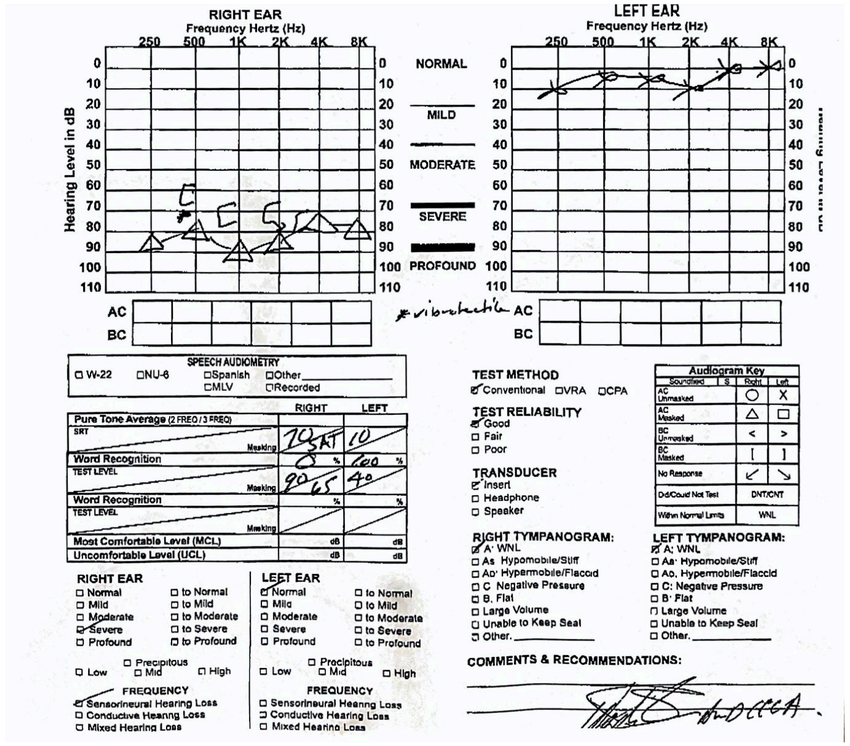
Patient is a previously healthy 12-year-old boy who presented to the emergency room with sudden onset right-sided hearing loss, tinnitus, vertigo, abdominal pain, nausea and vomiting. On admission, his SARS-CoV-2 PCR was positive, and all other viral respiratory pathogens PCRs were negative.

He had no other symptoms including fever, congestion, cough, or any respiratory symptoms. He was treated with prednisone, lorazepam and ondansetron during his 4-day hospitalization.

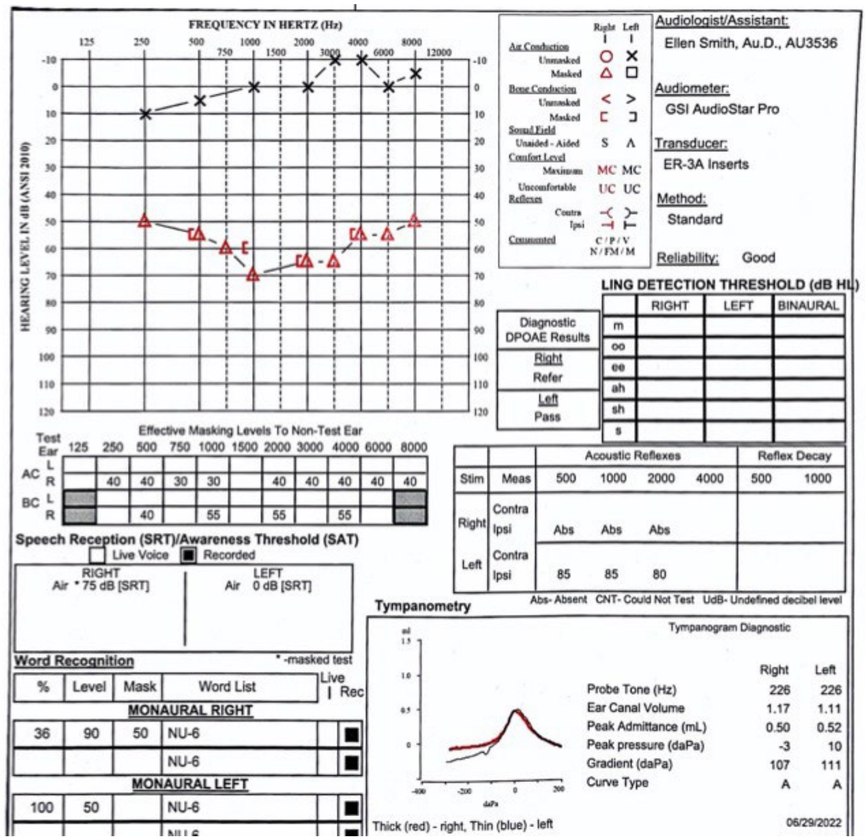
Computed tomography of the temporal bone was normal and magnetic resonance imaging with contrast of the brain and internal auditory canal was normal as well.

First audiogram following discharge showed a severe sensorineural hearing loss from 250-8000 Hz with poor word recognition score in the right ear and normal hearing in the left ear. At his two-month follow-up visit, his vertigo symptoms had resolved. However, he had persistent loud tinnitus and hearing loss.

Repeat audiogram 8 months later showed improvement in pure tone responses to moderate to moderate-severe SNHL, but the word recognition scores remained poor at 36% at multiple presentation levels, suggesting unlikely benefit from traditional amplification.



Initial Audiogram:
Normal hearing on the left, severe SNHL on the right



Recent Audiogram:
Normal hearing on the left, moderate to moderately-severe SNHL on the right

Discussion

Since the onset of the COVID-19 pandemic, there have been a few reports of SSNHL associated with either acute viral infection or as a result of the mRNA vaccine. Chern *et al.*[3] reported a case of bilateral SSNHL and intralabyrinthine hemorrhage in an 18-year-old patient who presented with similar symptoms of hearing loss, aural fullness and vertigo with documented SARS-CoV-2 infection. To our knowledge, this is the first reported case of a pediatric patient with COVID-19 labyrinthitis.

With or without vestibular symptoms, hearing loss is often overlooked during an acute COVID-19 infection especially when patients present with severe respiratory symptoms which may require intubation or acute intensive care admission. However, increased awareness is essential to realize the hearing symptoms and early initiation of systemic steroids affords the patients the best chance of hearing recovery.

Conclusion

COVID-19 may present with viral labyrinthitis with severe vestibular symptoms and SSNHL. During the current COVID-19 pandemic, there have been very few reported otologic manifestations of this viral infection. The etiology of the labyrinthitis is unclear. Short-term treatment involves supportive care and high dose steroid. Close audiologic follow-up is essential.

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Contact: Wen Jiang, MD wjiang@rchsd.org

Video demonstration vs puzzle for rigid bronchoscope assembly training

Ashley Stone BA[1]; Scott W. Gorthey, MD[2]; Angela Cao, MD[2]; Christina J. Yang, MD[1, 2, 3];
[1]Albert Einstein College of Medicine, Bronx, NY
[2]Department of Otorhinolaryngology-Head and Neck Surgery, Montefiore Medical Center, Bronx, NY
[3]Department of Pediatrics, Children's Hospital at Montefiore, Bronx, NY

Abstract

Background:
Rigid bronchoscope assembly is critical for management of airway emergencies and considered essential to otolaryngology resident education, but clinical exposure and case volumes vary. We sought to investigate whether the "see one, do one, teach one" model could be improved by technology-enhanced or low-fidelity simulation.

Methods:
Medical student novices were randomized to one of two arms - watching a videotaped demonstration (n=10) or using a printed photograph puzzle of bronchoscope parts (n=13) - and then videotaped attempting to assemble a 5.0 rigid bronchoscope. Videos were reviewed by two blinded raters. Individual steps were scored for accuracy (0- not attempted, 1-incorrect, 2-correct) and time-to-completion, and overall proficiency was rated on a 5-point Likert scale. Scores and time required for assembly were compared by the two-tailed t-test, and inter-rater reliability was assessed with Cohen's kappa.

Results:
There were no differences in years of medical training between the video demonstration and puzzle groups. The video demonstration group required less time for assembly than the puzzle group (median 69 sec vs. 133 sec, p=0.01) and had a higher overall proficiency score (median 3 vs 2, p=0.02). There was fair inter-rater agreement (kappa=0.387; p<0.001).

Conclusions:
The group watching the video demonstration had shorter assembly times and higher overall proficiency scores than the group assembling the puzzle. Further study is needed to identify potential benefits of simulation related to the concept of productive failure, where students struggle through a process to gain a deeper understanding.

Introduction

- Rigid bronchoscopy an essential part of airway management, for example foreign body removal, and used in low-volume, high-risk settings.
- The use of simulation in surgical training is emerging to improve competency, reduce surgical errors[1], and increase exposure to case volumes that can be variable for each trainee.

Acknowledgments

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- Dr. Yang is a Clinical Research Training Program scholar supported by NIH/National Center for Advancing Translational Science (NCATS) Einstein Montefiore CTSA Grant Number UL1TR001073

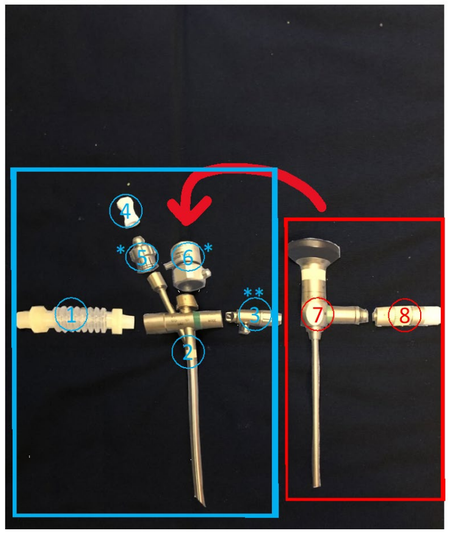
Methods

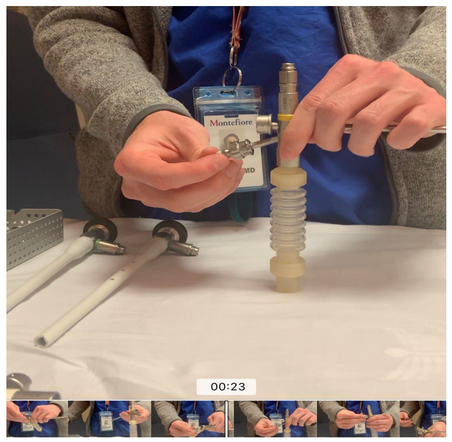
- Inclusion criteria: 1st and 2nd-year medical students with no prior experience with rigid bronchoscopy or airway interventions (N=23)
- The video group viewed a 1.5-minute video demonstration of bronchoscope assembly twice, which showed a senior resident explaining the different parts and their function as they assembled it.
- The puzzle group was given 3 minutes to view an instructions sheet (see right) and practice with the paper puzzle as many times as they wished.
- Participants were randomized by a random number generator and videotaped as they assembled the rigid bronchoscope for within the maximum time of 5 minutes.
- Videos were reviewed by 2 blinded raters and individual steps were scored for accuracy, time-to-completion, and overall proficiency scores.
- Time-to-completion and scores were compared by the two-tailed t-test.

Results

- For time to assembly, the video group required median 69 seconds vs the puzzle group required median 133 seconds (p=0.01)
- For overall proficiency scores, the video group received a median score of 3 vs the puzzle group received a median score of 2 (p=0.02)
- The Cohen's Kappa statistic for inter-rater agreement of proficiency scores was 0.387 (p<0.001)

1. Assemble 1-6 (blue) then combine 7+8 (red). Slide 7+8 onto 1-6.
*5 and 6 should be "clicked" into place.
** 3 should be fitted into the first slot





Steps in assembly:	Time (seconds)			Scores (0 to 5)		
	Puzzle Mean	Video Mean	p value	Puzzle Mean	Video Mean	p value
Attach anesthesia circuit to bronchoscope	8.3	5.1	0.09	2.00	2.00	NA
Attach suction adapter (metal) to bronchoscope	12.7	9.2	0.39	1.54	1.70	0.10
Attach suction port cover (rubber) to suction adapter (metal)	15.5	11.1	0.49	2.00	2.00	NA
Attach telescope adapter to bronchoscope	19.2	10.6	0.01	1.96	1.95	0.86
Attach light prism to bronchoscope	20.0	9.1	0.03	1.77	1.85	0.54
Attach the light cord to telescope	13.5	9.1	0.23	1.50	2.00	0.03
Slide telescope into bronchoscope	25.4	10.9	0.26	1.15	1.60	0.04
Total	162.8	74.8	0.01	2.00	2.90	0.02

Conclusions

- Faster time-to-assembly and higher overall proficiency scores were observed in the the video demonstration group than the puzzle group
- Future directions: follow-up with learners regarding skill retention and study among PGY-1 residents

Summary

- Novices learning rigid bronchoscope assembly through a video demonstration had faster and more accurate assembly than novices who learned through a low-fidelity paper puzzle model
- For novices, traditional demonstration may lead to more proficiency in bronchoscope assembly than low-fidelity models
- The utility of low-fidelity simulation warrants further study, such as possibly reinforcing skills to more experienced learners or improving learners' ability to problem-solve and retain the skill for longer periods of time

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Contact:
Ashley Stone
MS3, Albert Einstein College of Medicine
ashley.stone@einsteinmed.edu

Actionable Fingertip Data to Enhance the Safety of Children with a Tracheostomy

SENTAC 2022 Abstract #58

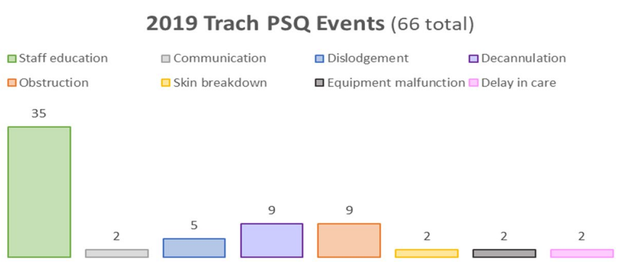
Wendy Arafiles, MD
Mark E. Gerber, MD
Vinay Vaidya, MD
Melinda Loya, MSN

Abstract

Despite a growing number of children with long term tracheostomy tubes, inconsistent event reporting obscures the true incidence of airway safety events. A multidisciplinary group was created to improve patient safety and quality of care for this vulnerable population through enhanced staff education on tracheostomy management and improved organization of data.

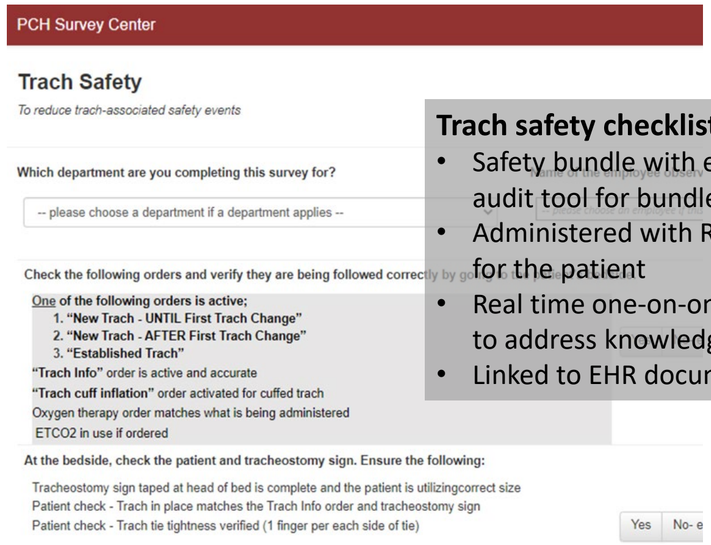
Background

- *Phoenix Children's Hospital (PCH) in Phoenix, Arizona*
  - 433 bed free-standing pediatric tertiary care center
  - 14-26 tracheostomy patients admitted each month
  - 3-7 new tracheostomy creations each month
- *PCH Tracheostomy Program:* multi-disciplinary membership, designed/clarified care standards, created safety checklist, championed EHR tools
- *Tracheostomy Resource Nurse:* Clinical Educator for nursing, respiratory therapy, and caregivers
- *Quarterly review of patient safety and quality (PSQ)* event reports involving patients with a tracheostomy
  - 2019 PSQ data revealed 66 events involving trach patients, >50% related to staff education



Methods & Results: Tracheostomy Dashboard

- *Population identification:* Tracheostomy-related ICD-10CM and/or CPT codes
- *Order set utilization:* Concurrent prompts and reminders to stakeholders, utilization tied to otolaryngology division metrics
- *Population capture:* 100% inpatient capture after multiple rounds of validation
- *Dashboard design:* (ongoing design improvements based on the needs of end-users)
  - All currently admitted patients with location in the hospital
  - Ventilator type: home, conventional, or none
  - Assigned nurse's experience with tracheostomy patients and tasks
  - Tracheostomy size/type and tube change data
  - Resident and attending provider contact information



Trach safety checklist

- Safety bundle with electronic audit tool for bundle compliance
- Administered with RN/RT caring for the patient
- Real time one-on-one education to address knowledge gaps
- Linked to EHR documentation

Conclusions

Bringing actionable data to the fingertips of the inpatient tracheostomy team enhances the ability of clinical bedside staff to efficiently maintain the safest and highest quality care for a vulnerable patient population.

New Trach (< 7 days)	Room	Patient	LOS	Service	Attending	Resident	Nurse	Assigned nurse # Trach shifts last 180 days	Assigned Nurse Total # Trach Changes	Nurse # Day Last Change Trach	#Days Trach Change	Days since ENT OP Note	Days since ENT IP Note	PULM OP Note	PULM IP Note	Surgeon	Trach Date (New)
	6142-A		1	Gener...				28	6	44	5		72	205			
	6130-A		5	Gener...				85	6	190			21	47	1		
5	6143-A		18	Critical...				33	1	275			1		2		10/21/2...
8	6136-A		26	Gener...				71	19	63	14		3	174	204		10/18/2...
	6131-A		32	Gener...							5		5		1		10/6/202
	6126-A		65	Hemat...				110	6	35	6		14		2		9/8/2022
9	1843-A		77	Neona...				6	1	225	2		6		6		10/17/2...

Cholesteatoma after Cochlear Implantation – A Rare, Devastating Complication

Steven Engebretsen, DO,[1,2] Carissa Wentland, DO[1-3]

Children's Hospital of Michigan[1], Michigan State University[2], Wayne State University[3]

Introduction

- Complications (relatively common) of cochlear implantation:
  - Device Failure/concerns
  - Vestibular concerns
  - Taste Disturbance
  - Infection
  - Facial Nerve injury
- Cholesteatoma after cochlear implantation accounts for 0.5% of complications.[1]
- Iatrogenic sources may include canal wall damage or introduction of squamous debris to mucosal cavities.

Case Presentation

- A 9-year-old Female with bilateral cochlear implants (Nucleus Freedom Contour Advance) developed left sided post-auricular erythema and pain 5 years after implantation supposedly after mild trauma to the area.
- Imaged (Figure 1A) – suspicion for cholesteatoma and acute left mastoiditis.
- Underwent cortical mastoidectomy and tympanostomy tube placement.
- Intra-operative findings:
  - Cholesteatoma in the mastoid cavity (Figure 1B) and facial recess (Figure 2) surrounding the electrode.
  - Removal of the implant to the level of the electrode in the facial recess was then performed.
- Placed on IV antibiotics
- A second procedure was performed (1 month later):
  - Remaining cholesteatoma removed
  - Electrode replacement with spacing electrode
  - With acute infection treated -- A posterior ear canal defect at the annulus (~1cm) was noted (Figure 3).
  - The defect was repaired with a tragal cartilage graft.
- A Final procedure was performed (7 months post-explantation) to restore hearing with a 512 Cochlear Nucleus Electrode.

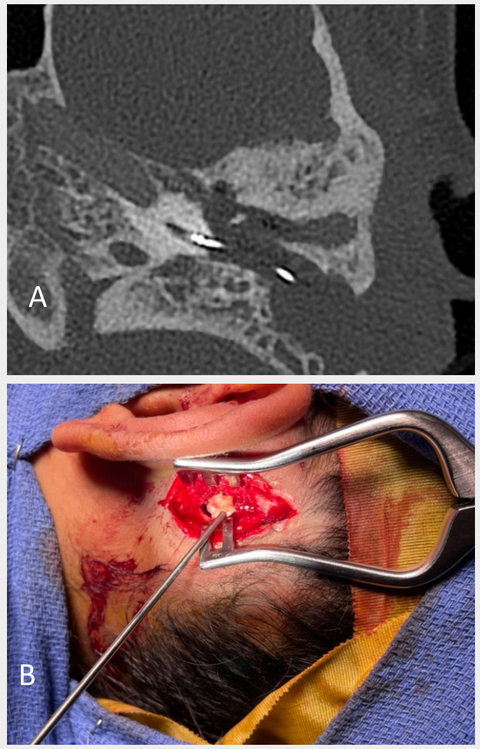


Figure 1. (A) Axial Non-contrast CT of left temporal bone showing soft tissue density and posterior canal defect with implant in situ. (B) Cholesteatoma filling the mastoid cavity in the setting of acute mastoiditis.

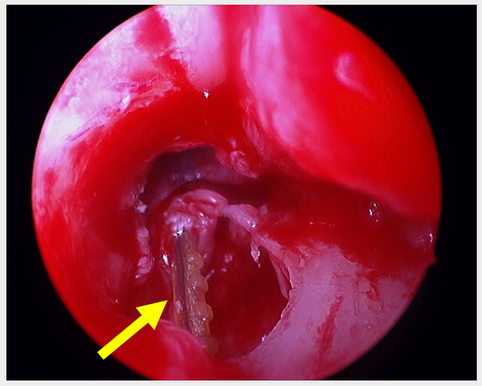


Figure 2. Facial recess showing cut electrode (arrow) in cochlea and surrounding cholesteatoma.

Discussion

- This case likely represents an iatrogenic cholesteatoma from cochlear implantation.
- Small posterior canal defects during drilling can result in sequentially larger defects with time and negative pressure.
- Reimplantation and cholesteatoma risk worse hearing outcomes.
- No published treatment algorithm exists for this devastating complication, but it seems patients can be re-implanted.

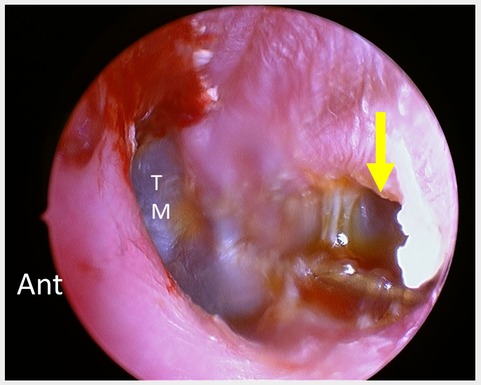


Figure 3. Endoscopic Trans-canal view of posterior canal defect.

Conclusions

- Pediatric cochlear implantation can result in a delayed iatrogenic cholesteatoma.
- It is unknown the true prevalence of this seemingly rare occurrence.
- Care should be exercised regarding the posterior canal drilling during cochlear implantation and in subsequent monitoring.
- Low threshold recommended for repair of canal defects intraoperatively at the time of implantation.

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Cost Effectiveness of Pharmacologic and Non-pharmacologic Tympanostomy Tube Prophylaxis

Josiah Brandt, BS[1] , William Clinkscales, MD[1,2]; Anthony Sheyn, MD[1,2]
[1]University of Tennessee Health Science Center, [2]Department of Otolaryngology

Abstract

Objective: Otorrhea is a common complication of tympanostomy tubes. Guidelines for prophylaxis are not firmly established. Given the comparable efficacy of topical agents and their highly variable costs, this study seeks to determine the most cost-effective intraoperative management strategy for preventing postoperative otorrhea.
Methods: An observational analysis of purchasing records at a children's hospital was performed. Methods were adapted from Yeakel et al. Using a conservative initial infection rate of 10% and observing local prices, a break-even analysis was performed. Absolute risk reduction (ARR) and final infection rates to make intraoperative prophylaxis cost-effective were calculated using pharmacologic and non-pharmacologic treatments. These included ofloxacin, ciprofloxacin-dexamethasone ophthalmic version, ciprofloxacin-dexamethasone otic version, saline, and oxymetazoline (Afrin).
Results: Break-even infection rate (BIR) of saline compared to ofloxacin outpatient treatment was 0.0982 and break-even absolute risk reduction (BARR) was 0.0018. BIR of Afrin compared to ofloxacin outpatient treatment was 0.017 and BARR was 0.083. BIR of saline compared to ophthalmic ofloxacin-dexamethasone was 0.0993 and BARR was 0.0007. BIR of Afrin compared to ophthalmic ofloxacin-dexamethasone was 0.676 and BARR was 0.324. BIR of saline compared to otic ciprofloxacin-dexamethasone was 0.0996 and BARR was 0.0004. BIR of Afrin compared to otic ciprofloxacin-dexamethasone was 0.0839 and BARR was 0.0161.
Conclusion: Routine intraoperative antibiotic prophylaxis after placing tympanostomy tubes is unlikely to be cost-effective in most scenarios. However, saline flushes and oxymetazoline should be considered for prophylaxis due to their low cost and their ability to prevent mucous plugging.

Introduction

Infectious otorrhea is the most common complication of tympanostomy tubes. The cited incidence is highly variable. In general it is relatively low but reports range from 3-75%. Historically after placing tubes, topical antibiotic drops have been applied intraoperatively and a short outpatient course prescribed as prophylaxis to prevent otorrhea and plugging of the tube with blood and drainage. There has been a trend away from this due to a lack of clear reduction in infection and plugging rates. The most recent guidelines recommend against the routine prescription of prophylaxis after placing tubes. However "regarding intraoperative management, the guideline authors leave it to clinician preference, which may involve saline washout, a single application of antibiotic ear drops (with or without a steroid), or simply no treatment at all.

Methods and Materials

Our analysis was modeled after Mckinnon et al, who borrowed a formula from the orthopedic literature. Using the local inpatient and outpatient costs of topical agents and the baseline rate of otorrhea, the final infection rate and ARR required to make the cost of prophylaxis break even with the cost of outpatient treatment was determined. Below is the formula:

$$S_{total} \times C_t \times IR_i = (S_{total} \times C_p) + (S_{total} \times C_t \times IR_f)$$

Solving for IR_f yields :

$$IR_f = \frac{(IR_i \times C_t) - C_p}{C_t}$$

Where: S_{total}= total annual surgeries; C_t= total cost outpatient treatment; C_p= cost of inpatient prophylaxis; IR_i= initial infection rate; IR_f= breakeven infection rate.

Results

Outpatient Treatment (cost)	In-Hospital Prophylaxis (cost)	Break-even Infection Rate	Break-even ARR
Ofloxacin ($15.54)	Ofloxacin ($17.27)	-1.01	1.11
	Cip-Dex Otic ($59.26)	-3.71	3.81
	Saline ($0.028)	0.0982	0.0018
	Afrin ($1.29)	0.017	0.083
Ophthalmic Ofl-Dex ($39.76)	Ofloxacin	-0.33	0.43
	Cip-Dex Otic	-1.39	1.49
	Saline	0.0993	0.0007
	Afrin	0.0676	0.0324
Otic Cip-Dex ($79.30)	Ofloxacin	-0.12	0.22
	Cip-Dex Otic	-0.64	0.74
	Saline	0.0996	0.0004
	Afrin	0.0839	0.0161

Outpatient Treatment	In-Hospital Prophylaxis	Cost of ppx	Break-even Infection Rate	Break-even ARR
Ofloxacin	Ofloxacin	$17.27	-1.01	1.11
Ofloxacin	Ciprofloxacin-dex	$59.26	-3.71	3.81
Ciprofloxacin-dex ophthalmic	Ofloxacin	$17.27	-0.33	0.43
Ciprofloxacin-dex ophthalmic	Ciprofloxacin-dex	$59.26	-1.39	1.49
Ciprofloxacin-dex otic	Ofloxacin	$17.27	-0.12	0.22
Ciprofloxacin-dex otic	Ciprofloxacin-dex	$59.26	-0.64	0.74

IRi	Treatment	IRf	Treatment	IRf	Treatment	IRf
0.05	ofloxacin	-1.082	cipro-dex ophthalmic	-0.38	cipro-dex otic	-0.17
0.1		-1.031		-0.33		-0.12
0.15		-0.98		-0.28		-0.07
0.2		-0.929		-0.23		-0.02
0.25		-0.878		-0.18		0.03
0.3		-0.827		-0.13		0.08
0.35		-0.776		-0.08		0.13
0.4		-0.725		-0.03		0.18
0.45		-0.674		0.02		0.23
0.5		-0.623		0.07		0.28
0.55		-0.572		0.12		0.33
0.6		-0.521		0.17		0.38
0.65		-0.47		0.22		0.43
0.7		-0.419		0.27		0.48
0.75		-0.368		0.32		0.53

Discussion and Conclusion

The blue table shows that with a conservatively high estimate of 10% rate of post-PE tube otorrhea without treatment, the break-even infection rate would have to be less than zero regardless of the pharmacologic agent used. Since it is impossible to reduce the infection rate even to zero, there is no combination of pharmacologic prophylaxis and treatment which is ideally cost effective.

The grey table shows the break-even infection rates using the cheapest pharmacologic prophylaxis (ofloxacin) with various outpatient treatment options while increasing the original infection rate (IRi = initial infection rate; IRf = break-even infection rate). This shows that even with an initial rate of otorrhea of 75%, ofloxacin prophylaxis is not cost effective when using ofloxacin as outpatient treatment. The initial infection rate would have to be 45% in order to be cost effective when using ophthalmic drops as treatment and it would have to be 25% percent when using otic ciprofloxacin-dexamethasone. In other words, if 1 out of every 4 patients receiving PE tubes experienced postoperative otorrhea without prophylaxis, then using ofloxacin prophylaxis and otic ciprofloxacin-dexamethasone would economically break even. Of note, this is assuming that this prophylaxis truly reduces the risk by 22%.

In conclusion, routine intraoperative antibiotic prophylaxis after placing tympanostomy tubes is unlikely to be cost-effective in most scenarios. However, saline flushes and oxymetazoline should be considered for prophylaxis due to their low cost and their ability to prevent mucous plugging.

Contact

Josiah P Brandt
University of Tennessee Health Science Center
College of Medicine
jbrandt5@uthsc.edu
(865)-806-5702

Otolaryngologic manifestations of Helsmoortel-Van Der Aa syndrome

Colleen Schindlerle, BA[1]; Carlos Sendon, MD[2]; Samantha Vergano, MD[2]; Thomas Gallagher, DO[3]

Eastern Virginia Medical School[1]; Department of Pediatrics, Children's Hospital of the King's Daughters[2]; Department of Otolaryngology, Eastern Virginia Medical School[3]

Abstract 61

Introduction

- Helsmoortel-Van Der Aa syndrome (HVDAS), also known as *ADNP*-related disorder, is a genetic disorder related to autosomal dominant inherited de novo mutations in the *ADNP* (activity-dependent neuroprotective protein) gene on chromosome 20q13.[1]
- *ADNP* gene is crucial in brain development and function. Characteristics of HVDAS include developmental and intellectual delays, speech problems, low muscle tone, and autism spectrum disorder, in the vast majority of patients. Facial features such as a prominent forehead, wide-spaced eyes, and ptosis as well as a happy demeanor resembling Angelman syndrome in younger kids with the diagnosis are also associated with the syndrome.[2]
- *ADNP* is estimated to be mutated in 0.17% of autism cases due to genetic inheritance, making it a leading association.[3] HVDAS is estimated to affect one in 20,000 people in the United States and Canada; may be underreported as it was only discovered in 2014.[4]
- The management of three new patients with HVDAS in our clinic prompted further investigation into the otolaryngologic manifestations of the disorder. Two out of the three patients had otolaryngologic problems such as aspiration, sensorineural hearing loss, otitis media and obstructive sleep apnea. To our knowledge, there are no descriptions of the otolaryngologic symptoms of HVDAS in the otolaryngologic literature. Our objective was to review these symptoms in detail and report our findings in order to help further the understanding of this syndrome and the role of an aerodigestive team in coordinating care.

Methods

- Case series with chart review of pediatric patients with ADNP syndrome who have undergone previous diagnosis based on known manifestations of the disease.
- The electronic medical record system was queried for the ICD-10 code for Helsmoortel-Van der As Syndrome (Q87.89).
- Patient charts were to be reviewed, compared, and presented in order to elaborate otolaryngologic symptoms reflected in patients with this syndrome.

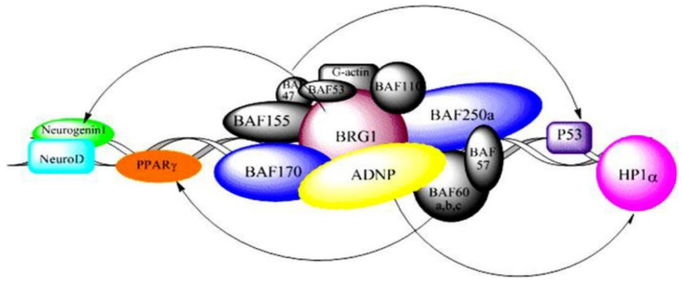

Fig. 1 is a model of the SWI/SNF chromatin remodeling complex, which ADNP regulates to impact dendritic and axonal development.[5]

Results

- Three patients were identified with all three having confirmed *ADNP* mutations consistent with Helsmoortel-Van Der Aa syndrome.
- This included two males and one female with an average age at the time of assessment of 9.3 years with a range from 2 to 18 years.
- Of the three identified patients, two expressed otolaryngologic disorders. One patient underwent adenotonsillectomy, two had aspiration of liquids, one had auricular hematoma, one had a neck abscess, two patients had either otitis media with or without effusion, two had obstructive sleep apnea, and two had hearing loss.

Presentation of Cases

Case 1

- Patient one was evaluated for bilateral sensorineural hearing loss after a failed ABR (Auditory Brainstem Response) test, and was found to have normal sloping to mild to moderate mid to high frequency sensory healing loss, bilaterally.
- This patient presented with developmental delay, speech and language delays, asthma, and autism.
- Prior history involved laryngomalacia, recurrent otitis media requiring myringotomy, aspiration on thin liquids, and an adenotonsillectomy due to mild obstructive sleep apnea.

Case 2

- Patient two was evaluated for hearing loss with an ABR and found to have mild conductive hearing loss. They underwent placement of a gastrotomy tube due to feeding difficulties and chronic lung disease due to aspiration.
- This patient presented with autism, ankyloglossia, breath-holding spells, global developmental delays, and vision impairment.
- Prior history involved mild obstructive sleep apnea, gastroesophageal reflux, aspiration and feeding difficulties, bilateral otitis media with effusion, and left neck abscess (requiring incision and drainage).

Case 3

- Patient three was seen for a self-inflicted hematoma in the left auricular region.
- This patient presented with autism, developmental delay, and self-injurious behavior.
- Prior history involved constipation, gastroesophageal reflux, rectal prolapse, eczema, excessive flatulence, poor weight gain, and standard wisdom tooth extraction.

Summary of Otolaryngologic Findings Across all Three Patients

Otolaryngologic Problem/ Procedure	Patient 1	Patient 2	Patient 3
Adenotonsillectomy	✓		
Aspiration	✓	✓	
Auricular Hematoma			✓
Neck abscess		✓	
Otitis Media	✓		
Otitis Media with Effusion		✓	
Obstructive Sleep Apnea	✓	✓	
Hearing Loss	✓	✓	

Conclusions

Although this case series is not a definitive description of all of the head and neck manifestations of HVDAS, it does indicate that awareness of the syndrome as an otolaryngologist or as part of an aerodigestive team is important.

Limitations include:

- Potential missed patients due to relatively new diagnosis of HVDAS
- Retrospective study
- Small sample size

Future directions:

- As the use of genetic testing becomes more common, HVDAS diagnosis will likely increase allowing larger numbers of patients to be identified.
- Communication with genetics and developmental pediatrics to increase awareness of head and neck manifestations of the disorder
- Multidisciplinary approach to these patients in an aerodigestive clinic

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Acute mastoiditis in children under six months of age: a case series

Emma Office BA[1], Tina Tan MD[1,2]

[1]Northwestern University Feinberg School of Medicine, Chicago, IL USA [2] Department of Pediatrics, Division of Infectious Diseases, Ann and Robert H. Lurie Children's Hospital, Chicago, IL USA

Background

- **Acute mastoiditis (AM) is a serious complication of acute otitis media (AOM)** that occurs when infected fluid spreads posteriorly from the middle ear to the mastoid.[1] AM can cause **complications such as hearing loss, meningitis, brain abscesses, and sigmoid sinus thrombosis**.[1-4]
- In the US, estimated incidence of AM was **3.5/100,000 in children under two years of age** in 2012.[5] While rare in infants, there is **limited literature on AM in this demographic**. Given the risk of serious complications, a better understanding of AM is warranted.
- Our goal is to **define the epidemiology, clinical signs and symptoms, and outcomes of AM in infants less than six months of age.**

Methods

All cases of AM occurring at Children's Memorial Hospital/Lurie Children's Hospital were identified and charts were reviewed. Inclusion criteria were: AM diagnosis based on clinical symptoms (with or without radiographic findings) and age less than six months.

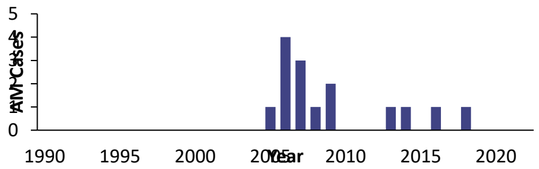


*Figure 1. AM Cases by Year. A total of **15 cases were seen over the 31-year study period**.*

Patient Demographics

Average age (months)	Range (months)
4.4	1.75 - 6

Characteristic	Number (%)
Sex	
Male	10 (67)
Female	5 (33)
Race	
Black/African American	3 (20)
White	5 (33)
Race listed as 'other'	6 (40)
No race specified	1 (7)
Ethnicity	
Hispanic	5 (33)
Non-Hispanic	7 (47)
No ethnicity specified	3 (20)
Medical History	
Previously healthy	12 (80)
Chronic condition	3 (20)
Prior history of AOM	4 (27)

Vaccination History	Number (%)
Age-appropriate PCV received	14 (93)
No PCV doses	3 (20)
PCV7 doses	
1 dose	2 (13)
2 doses	7 (47)
3 doses	1 (7)
PCV13 doses	
1 dose	0 (0)
2 doses	3 (20)
3 doses	0 (0)

Table 1. Patient Demographics. Chronic comorbidities included: chronic kidney disease, severe combined immunodeficiency, and ex-prematurity. Pneumococcal conjugate vaccination (PCV) was not available prior to 2000, but no AM cases were identified 1990-1999. The seven-valent vaccine (PCV7) was introduced in 2000, followed by the 13-valent vaccine (PCV13) in 2010.

Clinical Presentation and Evaluation

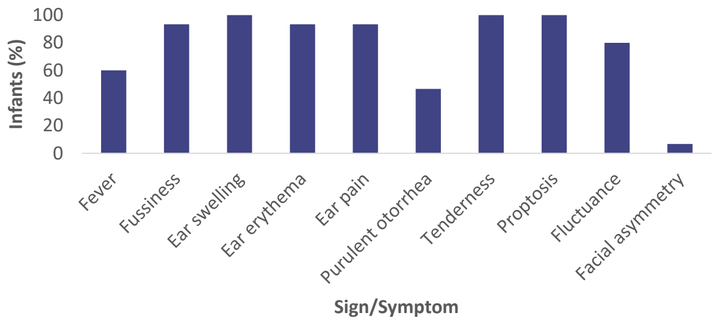
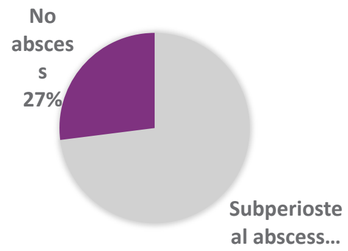


*Figure 2 (above). Clinical Presentation. Patients presented on average with **5 days of non-specific symptoms and 3 days of ear related symptoms**. Figure 3 (right). Imaging Findings. Almost all patients (93%) received CT imaging and most were found to have a subperiosteal abscess.*



Microbiology

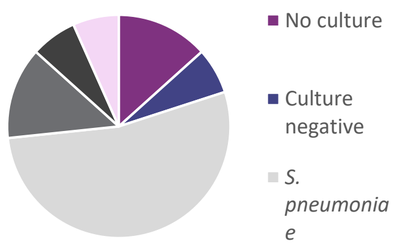
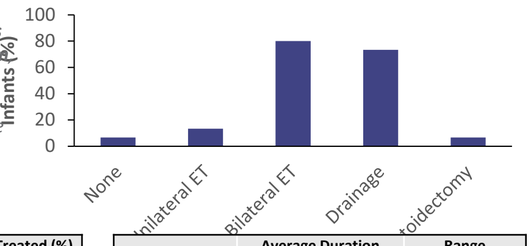


*Figure 4. Microbiology of AM. The **most isolated organism was Streptococcus pneumoniae**, followed by Coagulase-negative Staphylococci (CoNS). Group A Streptococcus (GAS) and Pseudomonas aeruginosa were each isolated once. **Four (27%) isolated organisms had some degree of antimicrobial resistance**, but all were susceptible to at least one antibiotic.*

Treatment and Outcomes

*Figure 5 (right). Surgical Treatment. **Almost all infants underwent some intervention**, most commonly ear tube (ET) placement with drainage of abscess if present (fine needle aspiration or incision and drainage).*



IV antibiotic	Number Treated (%)
IV treatment only	5 (33)
Ampicillin-Sulbactam	6 (40)
Cefuroxime	1 (7)
Ceftriaxone	8 (53)
Clindamycin	1 (7)
Levofloxacin	1 (7)
Meropenem	1 (7)

PO antibiotic	Number Treated (%)
Amoxicillin-Clavulanate	7 (47)
Amoxicillin	1 (7)
Cefdinir	2 (13)
Levofloxacin	1 (7)

Antibiotic Route	Average Duration (days)	Range (days)
IV	11	3-27
PO	10	0-30
Total	21	15-35

*Table 2 (above, left). Antibiotic Treatment. Eight children (53%) had received antibiotic treatment before AM diagnosis (not shown). After diagnosis, all received **at least 2 weeks of antibiotics.** Most (67%) were transitioned from IV to oral (PO) antibiotics at discharge. Hospitalization lasted on average 5 days after diagnosis (range 2-12 days). There were **no further intracranial complications during hospitalization.***

Conclusions

- While AM is a severe illness, it can be **managed successfully with prompt antibiotic therapy and surgical intervention.**
- The **transition from PCV7 to PCV13 in 2010 led to a significant decline in pneumococcal disease** including otitis media caused by *S. pneumoniae*.[6] This may in part account for the decrease in the number of cases of AM seen after its implementation
- Limitations to this study include small sample size and limited patient follow up.

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knowledge changing life

Nasopharyngeal Stenosis Following Adenotonsillectomy in a Pediatric Obstructive Sleep Apnea Patient: A Case Report

Mit Patel, BS, Samantha LaPrade, MD, Michael McCormick, MD
Medical College of Wisconsin, Milwaukee, WI and Children's Wisconsin, Milwaukee, WI

Children's Wisconsin
Kids deserve the best.

Nasopharyngeal Stenosis (NPS)

- NPS is restricted or complete obstruction of communication between the nasopharynx and oropharynx.
- Acquired NPS has been reported in multiple case series secondary to tonsillectomy, adenoidectomy, radiotherapy, and uvulopalatopharyngoplasty.[1,2]
- Treatment include excision of scar tissue with mucosal advancement flap, balloon dilation, corticosteroid injection, and stent placement.[3]

Case Presentation

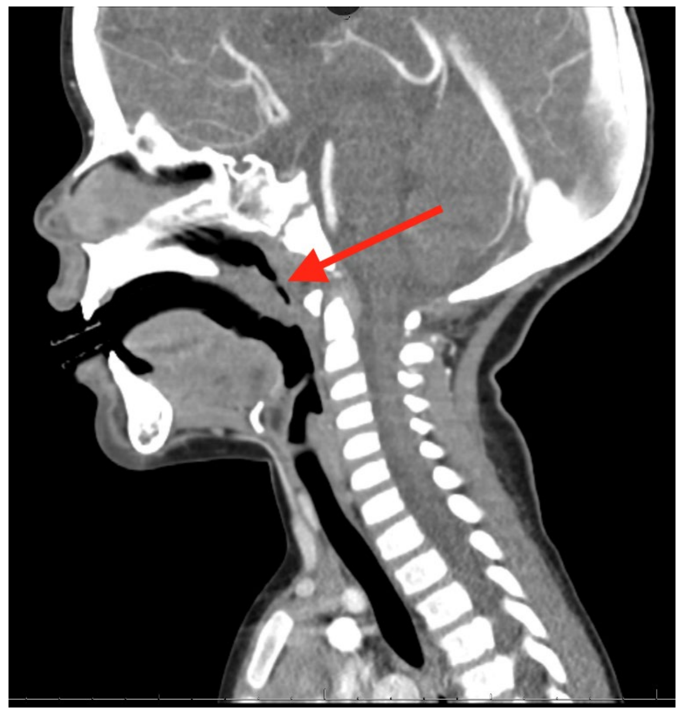
3-year-old male with history of autism spectrum disorder, recurrent acute otitis media, chronic adenoiditis, moderate obstructive sleep apnea status post adenotonsillectomy presented with persistent snoring and nasal drainage.

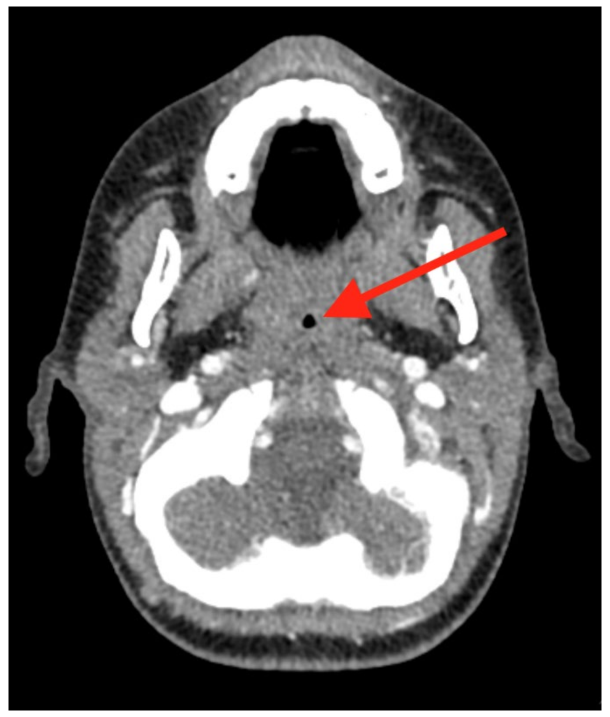
Surgical history
- 01/2021: 1st set of PE tubes
- 03/2021: 2nd set of PE tubes and adenoidectomy
- 12/2021: tonsillectomy

Initial presentation
- 04/2022
- Continues to snore loudly with some episodes of apneas, gasping
- Thick nasal drainage despite saline sprays

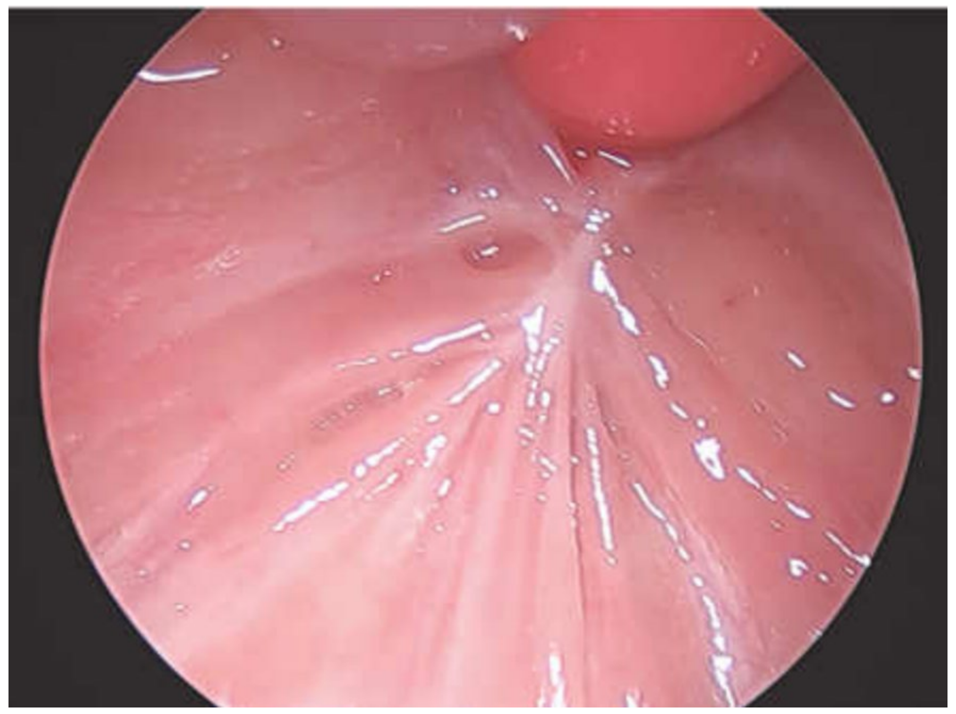
Diagnostic Workup
- 05/2022
- DISE: nasal airway - 1, nasopharynx - 2
- Flexible nasopharyngoscopy showed possible choanal narrowing
- CT neck: moderate to severe stenosis of the nasopharynx

Imaging





Preoperative CT neck showed circumferential moderate to severe stenosis of the nasopharynx. Oropharynx and hypopharynx widely patent with no enlargement of adenoids and palatine tonsils.

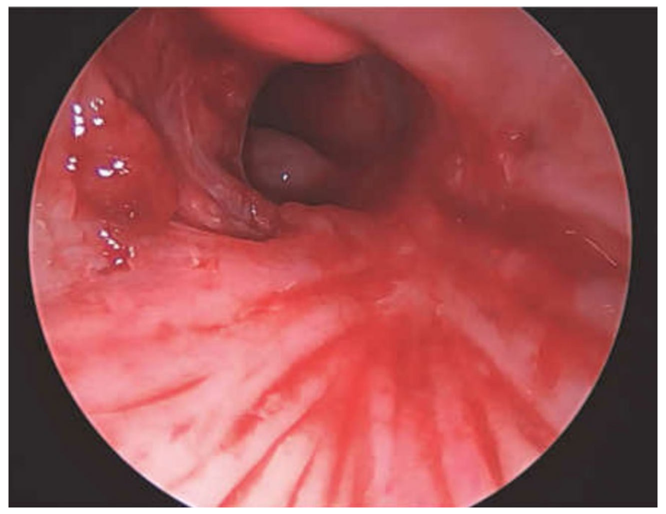


Near complete stenosis with red rubber catheter barely passing through (inferior view)

Management

Initial surgery (t) t = 07/2022

Findings: near complete obstruction of NP airway from a scar in the superior nasopharynx
Interventions: Kenalog-10 injection → scar resection under both direct visualization in the mouth and nasopharynx → sequential endoscopic balloon dilation (8-12mm)



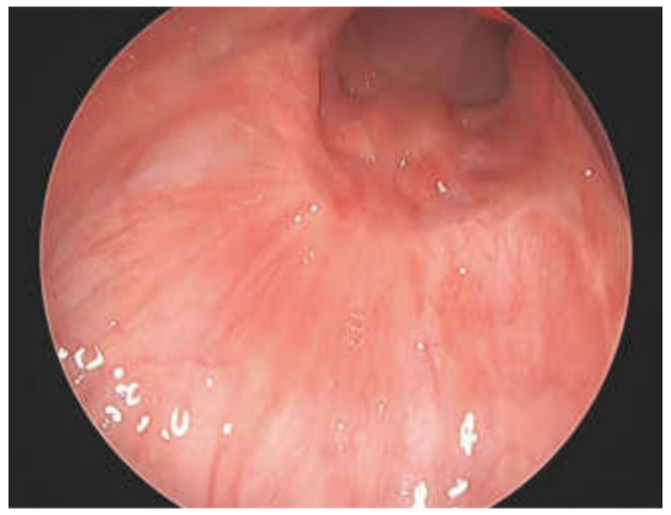
Post-op (inferior view)

Follow-up surgery (t + 12 weeks)

Findings: 90% restenosis of NP airway
Interventions: scar resection → injection of Kenalog-40 into the scar

Follow-up surgery (t + 14 weeks)

Findings: no signs of significant granulation tissue and no obvious scar
Intervention: injection of Kenalog-40 into the previously lysed scar tissue



Post-op (inferior view)

Conclusions

- NPS is an infrequent complication following adenotonsillectomy . It should be considered in patients with persistent or recurrent OSA and can be diagnosed using sleep endoscopy and imaging.
- There are multiple surgical options for correction of NPS including endoscopic resection and balloon dilation.

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Complications after Pediatric Temporal Bone Fractures by Classification System: A Systematic Review and Meta-Analysis

Nicolas S Poupore, BS; Annie F Britt, BS, Shaun A Nguyen, MD, MA; David R White, MD
Medical University of South Carolina, Department of Otolaryngology – Head and Neck Surgery, Charleston, SC, USA

ABSTRACT

Objectives: To analyze the rates of complications after pediatric temporal bone fractures (TBF) and the utility of traditional and new classification systems in predicting these complications.

Methods: Per PRISMA guidelines, PubMed, Scopus, and CINAHL were searched from the date of inception through April 27, 2022. Studies of children with TBFs were included. Meta-analyses of proportions were used to analyze complications and compare rates between the traditional and new classification systems.

Results: A total of 22 studies with 1,376 TBFs were included. Children with TBF had higher rates of CHL than SNHL (31.3% [95%CI 23.2-40.1] vs. 12.9% [8.9-17.5]). No statistical differences in both conductive hearing loss (CHL) and sensorineural hearing loss (SNHL) were seen between longitudinal and transverse TBFs; however, OCV TBFs had higher rates of SNHL than OCS TBFs (59.3% [95%CI 27.8-87.0] vs. 4.9% [95%CI 1.5-10.1]). Of all patients, 9.9% (95%CI 7.2-13.1) experienced facial nerve (FN) paresis/paralysis, and 13.4% (95%CI 5.9-23.2) experienced CSF otorrhea. Transverse TBFs had higher rates of FN paresis/paralysis than longitudinal (27.7% [95%CI 17.4-40.0] vs. 8.6% [5.2-12.8]), but rates were similar between OCS and OCV TBFs. No differences in CSF otorrhea were seen in the traditional or new classifications.

Conclusion: CHL was the most common complication seen after TBF in children; however, neither classification system was superior in identifying CHL preferentially. The traditional classification system was more effective at identifying FN injuries, and the new classification system was more robust at identifying SNHL. While these results suggest that both classification systems might have utility in evaluating pediatric TBFs, these analyses were limited by sample size. Future research on outcomes of pediatric TBFs stratified by type of fracture, mainly focusing on long-term outcomes, is needed.

METHODS

Search Criteria:

- This study followed the PRISMA guidelines, and utilized the PubMed, Scopus, and CINAHL databases.
- The initial search strategy was made for PubMed, around the concepts “temporal bone fracture” and “pediatrics”. The PubMed search strategy was then formatted for Scopus and CINAHL.

Selection Criteria:

- The inclusion criteria consisted of studies that analyzed the outcomes of children with TBF.
- Exclusion criteria consisted of non-English language, non-human studies, review articles, case reports of less than four patients, duplicates, and inaccessible articles.

Data Extraction:

- Two reviewers (N.S.P. and A.F.B.) independently extracted the data and compared for accuracy.
- Various methods of injuries were categorized as a motor vehicle accident, fall, bicycle or skateboard, assault, motor vehicle versus bicycle or pedestrian, and other.
- Types of hearing loss (conductive, sensorineural, and mixed) were collected, along with whether the hearing loss was resolved or persistent past each study follow-up period.
- Complications including hemotympanum, vertigo, bloody otorrhea, tympanic membrane perforation, otalgia or fullness, tinnitus, CSF otorrhea, and facial nerve paralysis or paresis, were documented.

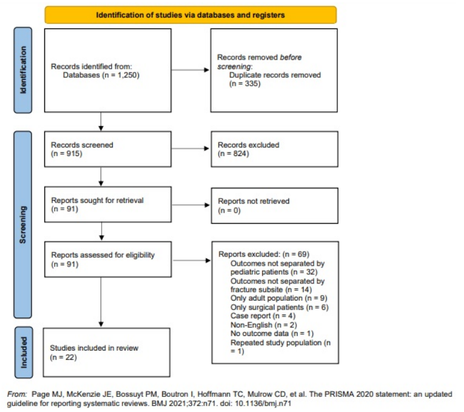
RESULTS

The literature search yielded 915 unique articles. Title and abstract screening excluded 824 articles. A full-text review of the remaining studies further excluded 69 articles, with the most common exclusion reason being “outcomes not separated by pediatric patients”. This process left **22 remaining articles for inclusion** in the final data extraction and analysis.

A total of 1,301 patients with 1,376 TBFs were included from all 22 studies. Included patients ranged from 0.0 to 18.0 years of age, with males consisting of 67.5% of the included patients. A total of sixteen studies classified their TBFs using the traditional method, with thirteen using the new classification system. **Longitudinal TBFs were the most common** (61.0% [95% CI 46.9 – 74.2]), as compared to both transverse (13.3% [95% CI 9.4 – 17.8]) and mixed (19.3% [95% CI 7.6 – 34.8]) TBFs. **There was a much higher rate of OCS TBFs compared to OCV TBFs** (91.7% [95% CI 88.3 – 94.5] vs. 8.3% [95% CI 5.5 – 11.7]) in studies utilizing the new classification system.

The two most common methods of injury included **falls** (41.6% [95% CI 35.9 – 47.5]) and **motor vehicle accidents** (27.1% [95% CI 20.3 – 34.4]). Twelve studies included clinical information on follow-up after the acute episode, with reported times ranging from 0.2 to 46.0 months.

Figure 1. PRISMA flowchart of study selection



From: Page MJ, McKenzie JE, Bossuyt PM, Boutron I, Hoffmann TC, Mulrow CD, et al. The PRISMA 2020 statement: an updated guideline for reporting systematic reviews. BMJ 2021;372:n71. doi: 10.1136/bmj.n71

Figure 2. Rates of CSF Otorrhea in Pediatric TBFs

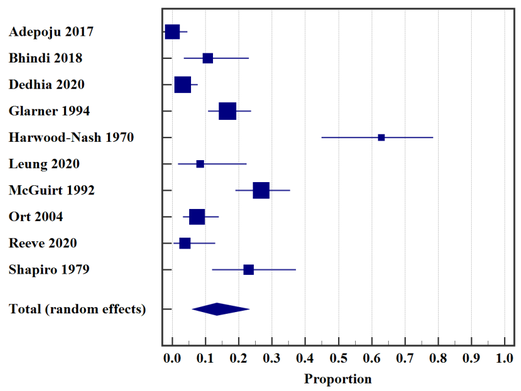


Figure 3. Rates of FN Paresis in Pediatric TBFs

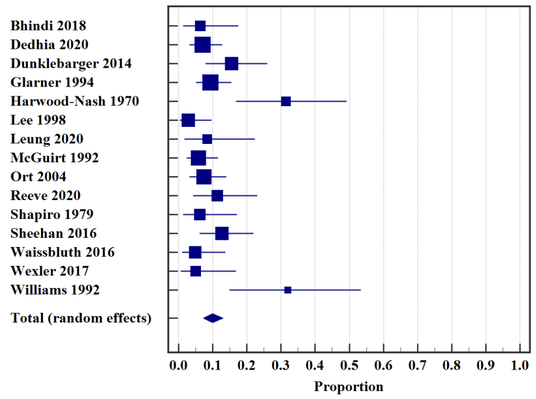
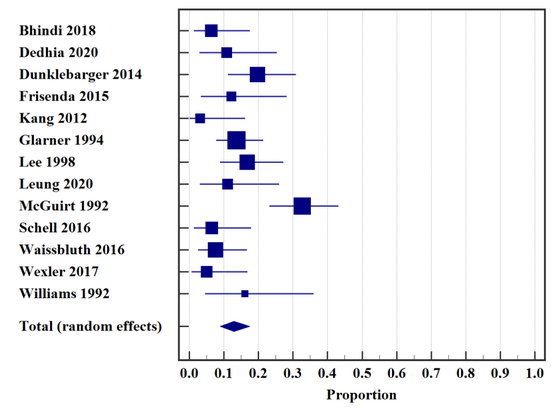


Figure 4. Rates of SNHL in Pediatric TBFs



Most children after TBF experienced **hemotympanum** (61.8% [95% CI 47.7 – 74.9]), with a smaller proportion experiencing **bloody otorrhea** (35.8% [95% CI 19.8 – 53.5]). Similar rates in pediatric TBFs were seen for **CSF otorrhea** (13.4% [95% CI 5.9 – 23.2]), **tympanic membrane perforation** (10.9% [95%CI 3.2 – 22.3]), and **facial nerve paralysis/paresis** (9.9% [95% CI 7.2 – 13.1]) . Regarding hearing loss, children had significantly higher rates of **conductive hearing loss** after TBF (31.3% [95% CI 23.2 – 40.1]) compared to SNHL (12.9% [95% CI 8.9 – 17.5]) and mixed hearing loss (5.6% [95% CI 3.0 – 8.8]).

Children with **transverse TBFs had higher rates of facial nerve paresis than children with longitudinal TBFs** (27.7% [95% CI 17.4 – 40.0] vs. 8.6% [95% CI 5.2 – 12.8]). Regarding CSF otorrhea and hearing loss, there were not statistically significant differences in rates between longitudinal, transverse, and mixed.

TBFs that were OCV resulted in significantly higher rates of SNHL than OCS TBFs (59.3% [95% CI 27.8 – 87.0] vs. 4.9% [95% CI 1.5 – 10.1]). No differences were seen between OCV and OCS TBFs regarding conductive hearing loss. OCV TBFs did not have statistically higher rates facial nerve paresis nor CSF otorrhea compared to OCS TBFs.

CONCLUSIONS

- In pediatric TBFs, **patients had slightly higher facial nerve injury rates than the reported rates in adults**; however, rates of CSF otorrhea and SNHL were marginally lower.
- The traditional classification system was more effective at identifying facial nerve paresis/paralysis, and the **new classification system was more robust at identifying SNHL**.
- Both classification systems might have utility in evaluating pediatric TBFs, but these **analyses were limited by sample size.**
- Future research on complications and outcomes of pediatric TBFs stratified by type of fracture, mainly focusing on younger patients and long-term outcomes, is warranted.

SUMMARY

- Temporal bone fractures (TBF) have been shown to occur in 14% to 22% of all patients with skull fractures and 3% of all trauma patients, with evidence supporting a **range of 8.9% to 22% occurring in pediatric patients.**
- TBFs can be classified according to **two different systems**: (1) longitudinal, transverse, oblique and mixed or (2) otic capsule sparing (OCS) and otic capsule violating (OCV).
- Complications of TBFs can have sequelae with clinically significant morbidity and mortality, such as meningitis or persistent hearing loss, so **proper diagnosis and management** of these injuries are crucial to reduce long-term negative outcomes.
- We performed a systematic review and meta-analysis of pediatric TBFs, with a **primary aim** was to classify the most frequent complications and sequelae after TBFs in children, and a **secondary aim** to analyze TBFs by fracture type to identify which classification system was robust at predicting outcomes and complications in children.
- A **prospective observational trial with consistent follow-up** using standardized definitions and protocols for labeling TBFs, particularly separating younger children, would be valuable in further clarifying the complications seen in pediatric TBFs.

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Audiometric outcomes following active transcutaneous osseointegrated implants in children

Madyson Hund, AuD; Christine Cook, MS, CCC-A; Scott Brietzke, MD; Steven Andreoli, MD | Nemours Children's Health, Jacksonville

Abstract

Background: Active transcutaneous bone conduction implant systems have been shown to decrease feedback and limit high frequency gain roll off in adults. Limited data is available for active bone implants in children, particularly with regards to speech discrimination performance.

Methods: A retrospective review was performed for patients less than 21 years of age undergoing Cochlear™ Osia® implantation at a tertiary children's hospital. Pre- and post-operative audiometry including pure tone average(PTA) and speech receptions thresholds(SRT) were compared. Aided discrimination testing in soft conversational speech, normal conversational speech and normal conversational speech in noise using the Northwestern University Auditory Test No. 6(NU-6) was performed three to six months following implantation.

Results: 23 patients underwent 26 implants from August 2020 to September 2022 with a mean age of 13.2 ± 4.4 years. Hearing loss was conductive or mixed in 18 patients and single sided sensorineural in five patients. One child did require device explantation due to infection. Pre- and post-operative PTA were 76.4 ± 16.5 dB and 24.3 ± 7.0 dB respectively (p<0.0001) for a mean functional gain of 52.1 dB. SRT improved from 72.0 ± 15.0 dB to 21.5 ± 4.3 dB after implantation (p<0.0001). Postoperative discrimination using NU-6 at 50 dB and 50 dB + 5 dB signal to noise ratio(SNR) was 94.2 ± 4.9% and 88.4 ± 6.8%.

Conclusions: Active transcutaneous osseointegrated hearing implantation in children is successful with a mean functional gain of 52.1 dB. Monosyllabic discrimination following implantation is excellent at 50dB and good and 50dB + 5 dB SNR. Future pediatric prospective studies regarding quality-of-life improvement and long term followup are required.

Introduction

Bone conduction hearing devices (BCHDs) offer an alternative to traditional hearing aids for children with unilateral or bilateral conductive, mixed loss or unilateral profound sensorineural hearing loss. BCHDs have undergone an evolution in the recent decades transitioning from devices with percutaneous abutments to modern devices deploying a transcutaneous magnet. This evolution has both decreased morbidity associated with skin and soft tissue complications and increased pediatric compliance with device use. However, transcutaneous devices were previously hindered by sound transmission across the scalp requiring significant external pressure form the amplifier for satisfactory sound results. The development of the Cochlear ™ Osia® device in 2020 offers a transcutaneous alternative and utilizes an active piezoelectric transducer minimizing skin irritation while providing adequate gain. Limited data is available related to pediatric implantation focusing on pure tone averages(PTA). This study explores our early data after Osia® implantation with follow up audiometry and discrimination testing results.

Methods

A retrospective review was performed with IRB approval to examine children undergoing Osia® implantation by two pediatric otolaryngologists at a tertiary pediatric hospital from August 2020 to September 2022. Demographic data, medical history and audiological history were extracted from the medical record. Surgical details related to prior device explantation and implant location were collected from operative records. The devices were routinely activated three to four weeks following surgery. Follow up audiometry with device was performed three to six months following implantation. Speech perception testing was conducted at soft conversational speech (35 bB HL), normal conversational speech (50 dB) and normal conversational speech in noise (50 dB + 5 dB SNR). All tonal stimuli (warble tones) and speech stimuli were presented to the ear with the Osia® processor. All speech perception testing excluding speech-in-noise testing was completed with the contralateral ear having an earplug and earmuff (plug-and-muff). During speech and noise testing, speech noise was presented to the contralateral ear to simulate real world listening environment. Two of the participants were bilateral Osia® users; therefore, speech perception testing was completed in the sound booth at 0 degree azimuth. Each processor was tested individually and bilaterally. For these participants, speech was presented to the front with noise to the back. Data was analyzed using paired t-test with significance at p<0.05.

Results

23 children underwent 26 implants during the study period. The mean age was 13.2 ± 4.4 years (range 6-19) with a female majority (56.5%). Conductive or mixed loss was present in 18 children with single sided deafness in 5 children. 13 children underwent simultaneous explantation of a Sophono® transcutaneous device. One child had previously undergone removal of a Baha Connect® abutment and skin flap for chronic skin irritation.

All children were successfully implanted with 4mm BI300 implant screws without operative complications. One child with single sided deafness did require explantation due to infection. Five weeks following surgery the implant site developed swelling and redness. After 48 hours of intravenous antibiotics without improvement a skin fistula formed and the device was surgically removed.

Prior to surgery, the PTA was 76.4 ± 16.5 dB. Following implantation, PTA improved to 24.3 ± 7.0 dB for a mean gain of 52.1 dB (p<0.0001). SRT improved from 72.0 ± 15.0 dB to 21.5 ± 4.3 dB after implantation (p<0.0001). Individual frequency testing is summarized in Table 1.

With regards to speech perception testing, discrimination at normal conversation speech (50dB) was excellent at 94.2 ± 4.9%. Discrimination in soft speech (35dB) was good at 84.7 ± 5.9%. Discrimination in noise simulating the classroom setting (50dB ± 5 dB SNR) was good at 88.4 ± 6.8%.

Frequency	Pre-Implantation (dB)	Post-Implantation (dB)	Gain (dB)	p-value
500 Hz	79.2 ± 16.1	27.2 ± 7.5	52.0	p<0.0001
1 kHz	76.2 ± 18.6	22.4 ± 12.1	53.8	p<0.0001
2 kHz	73.8 ± 20.2	23.3 ± 3.5	50.8	p<0.0001
4 kHz	72.3 ± 21.8	29.4 ± 3	42.9	p<0.0001
PTA	76.4 ± 16.5	24.3 ± 7.0	52.1	p<0.0001
SRT	72.0 ± 15.0	21.5 ± 4.3	50.5	p<0.0001

Table 1. Summary of frequency specific, puretone average(PTA) and speech reception thresholds (SRT)

Conclusion

The active transcutaneous Cochlear™ Osia® device offers children with unilateral conductive or mixed and unilateral profound sensorineural hearing loss a new option for amplification. The device carries less risk of skin irritation compared to percutaneous devices. The active component decreases the risk of skin erythema and compression related to first generation transcutaneous devices.

In our series, mean functional gain was similar to that reported in two prior pediatric studies. Discrimination testing was previously reported in one study in quiet ranging from 80-94% based on heterogenous testing sets. Our data used NU-6 for all participants identifying excellent discrimination in quiet 94.2%. Hearing in noise testing paralleling the classroom setting has not been previously reported. Our data revealed good discrimination at 50 dB + 5 dB SNR at 88%.

One patient in our cohort did require explantation due to infection. This occurred in the subacute setting weeks after activation. This complication has not been previously reported in children and no device nor surgical factors were identified to contribute. Explantation has been previously reported in adults.

Further prospective studies are required to between understand school performance, long-term compliance and use and device safety.

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Utility of Sinus CT for Diagnosis of Invasive Fungal Sinusitis in Immunocompromised Children

Park, Maria[4] ; Puccia, Ryan[1]; White, Shane[2]; Broglie, Larisa[3]; Shay, Sophie[4]; Robert Chun, MD[4]

[1]*Medical College of Wisconsin, Milwaukee*
[2]*University of Kentucky College of Medicine, Department of Otolaryngology*
[3]*Children's Wisconsin, Milwaukee, Division of Hematology/Oncology*
[4]*Children's Wisconsin, Milwaukee, Division of Pediatric Otolaryngology*

Introduction

Invasive fungal sinusitis (IFS) is a disease of the nasal passages and paranasal sinuses which can be rapidly progressive and fatal. It commonly affects neutropenic patients or those with functional neutropenia due to poorly controlled diabetes[1]. Effective treatment requires prompt diagnosis, aggressive surgical debridement, and systemic antifungal medication. The mortality rate of IFS has varied from previous reports of 50-80% to more recent values of 7-18%, likely due to earlier detection and intervention[2, 3]. The nidus of infection is presumed to be in the nasal cavity, with the middle turbinate testing positive in 14 of 21 IFS positive patients biopsied in one study[1]. Common symptoms associated with IFS include nasal congestion or discharge, facial pain or numbness, fever, and headache with more severe symptoms presenting with disease progression.

Given the importance of prompt diagnosis, imaging modalities such as CT and MRI have been used liberally in this population. Many papers in the adult literature have attempted to characterize the imaging features associated with invasive fungal disease on both CT and MRI, however, there is lack of consensus. CT scans may demonstrate mucosal thickening, bony erosion, turbinate hypertrophy, or unilateral opacification. In a review of twenty-three patients with confirmed IFS and preoperative CT imaging, DelGaudio and colleagues found that soft tissue edema of the nasal mucosa was a sensitive, though nonspecific finding. They also found that concerning signs such as bony erosion or air-fluid levels were uncommon[4]. A recent paper in the pediatric population determined that CT could play a significant diagnostic role in the appropriate clinical context, evidence of fungal specific signs on early imaging (bony destruction, extra-sinus inflammation, and nasal mucosal ulceration) was associated with a high likelihood (79%) of invasive fungal disease [5]. While CT is superior for detection of bony changes, MRI provides improved visualization of possible subtle signs of invasion into the orbit or cranial vault. For these reasons, MRI has been reported to have a greater sensitivity than CT for IFS detection, however, there is a lack of specific findings for both modalities[6]. The improved sensitivity of MRI, especially early in the disease process prior to bony erosion, has led some to favor MRI as the imaging modality of choice for IFS. Given the ease and convenience of CT, however, it continues to be widely used for evaluation.

While imaging lacks clear specific signs of invasive fungal disease, the use of bedside endoscopy provides an alternative method of evaluation. In actuality, early use of nasal endoscopy and biopsy of concerning regions are recommended for optimal patient outcomes[1]. Concerning findings on bedside nasal endoscopy include mucosal discoloration, crusting, granulation, ulceration or changes to sensation, and lack of bleeding. Recently, a retrospective review of pediatric patients who underwent bedside nasal endoscopy confirmed that a normal bedside exam was reassuring for no invasive fungal disease and that all of their patients who had invasive fungal disease also had findings on exam (Mulvey).

There is a paucity of data on the usefulness of imaging in pediatric patients for invasive fungal disease. Most of the current information and guidelines are extrapolated from the adult population. Given the importance of early diagnosis and the lack of convincing data for CT imaging, we decided to evaluate the work up of fever of unknown origin at our institution looking specifically at immunocompromised children with an ENT consult for evaluation of invasive fungal sinusitis. The goal of this study was to identify imaging trends at our institution regarding the use of CT. Secondary aims included evaluating the usefulness of CT imaging in pediatric patients with a concern for invasive fungal disease.

Methods

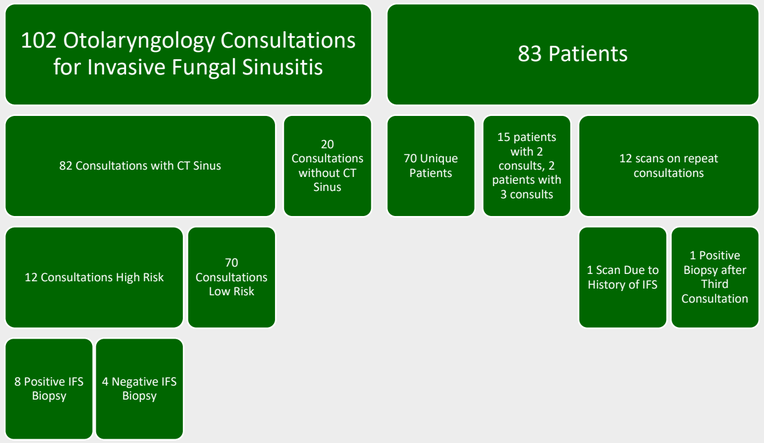
Data collection:
After approval by the institutional review board of Children's Wisconsin, a retrospective medical record review was performed of hospitalized pediatric oncology patients who had an Otolaryngology consult for concern for invasive fungal sinusitis placed between 2012-2020. Data collection included demographics, comorbidities, flexible fiberoptic examination information, CT imaging data and surgical pathology. Timing of the placement of the consult to timing of obtaining CT imaging was also obtained.

Statistical analysis:
Data analysis included Mann-Whitney tests and Fischer's exact tests. Significance was evaluated with a p-value of < 0.05.

Lund-Mackay Scoring:
A traditional Lund-Mackay (LM) score was tabulated for all children who had all sinuses developed. For children who had undeveloped sinuses, a modified Lund-Mackay (mLM) system was used which did not count the points for undeveloped sinuses, and these were tabulated as a separate group.

Nasal Endoscopy Risk Stratification:
The patients were classified as low risk or high risk based on chart review. High risk was classified as those patients who had any of the following findings on nasal endoscopy: Decreased sensation, pale patches, necrotic or dark patches, or ulceration. Low risk was classified as patients with normal nasal endoscopy, normal post-operative changes, or

Figures



Tables

	N= 102	SD
Age at Consultation	10y	6y
# Unique Patients	83	
Sex (# Male of 70 unique)	42	60%
Oncologic Diagnosis		
-Hematologic malignancy*	80	96%
-Ewing Sarcoma	2	2%
-Medulloblastoma	1	1%
Biopsy confirmed IFS	8	
Average ANC in Biopsy +	448	913
Biopsy Negative IFS	4	
Average ANC in Biopsy -	0.8	1.3
# Days FUO	5	4
ANC at time of Consultation		
>500	7	
100-500	1	
1-100	31	
0	56	
Time from Admission to Consultation, d	14	11
Consultations with CT	82	80%
CT Prior to Consultation	40	40%
CT with abnormal Radiology Read and IFS - on Biopsy (4)	3	75%
CT with abnormal Radiology Read and IFS + on Biopsy (8)	6	75%
CT with abnormal Radiology Read and Normal ENT Exam (70)	45	64%
ANC of Patients with Abnormal CT but Normal Exam	83	256

Table 1: Demographic Information of Patients at Consultation
* - Includes AML, ALL, B cell Lymphoma, DLBCL, Burkitt, NK cell
FUO = Fever Unknown Origin
ANC = Absolute Neutrophil Count
IFS = Invasive Fungal Sinusitis

		Age	ANC	FUO	LM Score
Biopsy +	Mean	13	162	7	7
N=6	SD	4	396	7	5
Biopsy – or Low Risk	Mean	13	107	5	6
N=49	SD	5	272	4	6

Table 2: Patients with All Sinuses on CT Amenable to Traditional LM Score at Time of Consultation
LM = Lund-Mackay

		Age	ANC	FUO	mLM Score
Biopsy +	Mean	2.7	1306	5	8
N=2	SD	0.5	1760	0	1.5
Biopsy – or Low Risk	Mean	3.5	34	5	7
N=25	SD	2	163	5	5

Table 3: Patients with Underdeveloped Sinuses on CT Undergoing Modified LM Score at Time of Consultation
mLM = Modified Lund-Mackay

Results

Between 2012-2020, the Otolaryngology service at Children's Wisconsin was consulted 102 separate times for persistent fevers of unknown origin in immune compromised patients with concern for IFS. The demographic of this population is shown in Table 1. Of the 102 consultations, there were 83 distinct patients. 15 patients had 2 separate consultations during different admissions. 2 patients had 3 consultations each during a different admission. This is outlined in Figure 1. Of these 102 consultations, there were 82 associated with a CT scan of the sinus (80%). Of these, 49% had been obtained prior to Otolaryngology consultation. Of the consultations with CT imaging, 85% were deemed low risk on physical exam including nasal endoscopy. 8 of 102 (7.8%) consultations and 8 of 83 (10%) patients were confirmed via biopsy to have IFS.

All 8 patients who were positive for IFS had undergone CT imaging. Of the 8 CTs, 2 (25%) were obtained prior to Otolaryngology consultation and 2 (25%) had been read as normal by Radiology. The patients with normal CT findings were taken to surgery because of concerning bedside endoscopy findings and the patients' absolute neutrophil counts (ANCs) of 0. Of the 8 biopsy positive patients, 6 (75%) demonstrated concerning findings on initial endoscopy including turbinate necrosis and dark or pale lesions. Nasal congestion was the only finding reported on one endoscopy exam, though progression to facial swelling prompted a trip to the operating room. The final case demonstrated significant bilateral congestion on endoscopy, though high index of suspicion and finding of a small septal abscess on CT was the impetus for going to the operating room.

Of the 4 patients taken to the operating room and found to be negative for IFS, three had a normal bedside endoscopy exam. Of the three normal endoscopy patients, one was biopsied due to a history of IFS and opacification on CT. The final two were taken to the operating room due to abnormalities on CT (one with bony erosion, one with fat stranding/mucosal edema) despite normal bedside endoscopy. The fourth patient is this subgroup had abnormal bedside endoscopy. This included dark debris that could not be cleared from the nasopharynx, though the middle turbinate was normal. Biopsies from the concerning area returned normal for invasive fungal disease.

Table 2 and 3 demonstrate the average age, days of fever of unknown origin, ANC and Lund-Mackay (L-M) score for the traditional and modified (mL-M) systems, respectively. There was no statistically significant difference in L-M or mL-M score in the biopsy positive and low risk/biopsy negative groups. There was also no statistically significant difference in average ANC for the traditional group.

Discussion

Our data demonstrates the high use of CT in patients with fever of unknown origin looking for invasive fungal sinusitis. 80% of consultations for evaluation for invasive fungal disease had a CT sinus and half of these scans were obtained prior to ENT consultation. The vast majority of patients in our cohort were being treated for hematologic malignancy and were high risk for invasive fungal disease with most ANCs <100, of these the majority being <1. There is no uniformity over the threshold for increased rates of invasive fungal disease. One systematic review of IFS in the pediatric population found that ANC was lower than 600 in 99% of patients [7]. Our data is consistent with this literature in terms of ANC, underlying diagnosis, and causative species except for a higher rate of Alternaria in our cohort. Interestingly, one of the main predictors of mortality in this study was the presence of pain which highlights the importance of physical exam in the work up of these patients. Presenting symptom of only fever was also not associated with overall mortality [7]. In another prospective study of invasive fungal infections in pediatric patients, the only predictors of invasive fungal disease on univariate analysis were an abnormal CXR, facial pain, and an ANC of <100 [8]. While no reported literature discusses the duration of fever in these children and IFS risk, in our cohort, the average time of fever of unknown origin was 5 days at the time of consultation. One reason for the thought that immunocompromised children would not have much mucosal inflammation on CT is that they have low ANC and cannot mount a robust immune response. One takeaway from our data is that the average LM and mLM score for our group of immunocompromised children without invasive fungal disease was 6±6 and 7±5, respectively. This indicates that even for children without invasive fungal disease with low ANC, there can be a wide range of scattered mucosal thickening. There was no association with our LM and mLM scores with ANC in our population. Three patients were taken to the operating room on CT findings alone who were negative on biopsy. Interestingly, in two of these patients, the findings on CT that prompted a trip to the operating room were bone erosion and extra sinus fat stranding which are two proposed specific findings for IFS [5]. Overall, these additional radiology findings outside of mucosal thickening/opacification were rare.

While many of these children have already been exposed to much radiation as part of their work up and surveillance, there are known risks to radiation in children including a higher lifetime incidence of solid cancers [9]. These concerns regarding radiation must be balanced with the more immediate need to diagnose IFS, as early detection is so crucial for survival. Our data indicates that there was one case in eighty-two in which CT imaging played a critical role in diagnosis. In this case, the patient had a normal physical exam with congestion on bedside nasal endoscopy. We were unable to determine if the congestion was severe enough to preclude a good exam or if it was a mild finding. In this case, CT imaging demonstrated a small septal abscess that prompted a trip to the operating room with positive biopsies of invasive fungal disease which led to early successful treatment.

Our data demonstrates that at our institution, immunocompromised children with fever of unknown origin undergo high rates of CT sinus imaging. Our data is consistent with the adult literature that there is poor sensitivity of CT imaging to identify invasive fungal disease. Given the high utilization of CT scans in this population with known risks of radiation exposure in children, we propose bedside exam and endoscopy as the best first line screening modality for these children. CT imaging may play a role in patients in which there is a high degree of suspicion with otherwise normal bedside endoscopy.

Limitations of the study include the retrospective nature and inability to understand decision making at the time through chart review alone.

Conclusions

CT imaging is a highly used modality for the evaluation of invasive fungal disease. Physical exam and bedside nasal endoscopy should guide initial screening in patients who there is a concern for invasive fungal sinusitis. For unique situations in which there remains a high index of suspicion but without remarkable nasal endoscopy, CT can be considered for guiding operative biopsy. Negative CTs alone are not a replacement for thorough evaluation and bedside nasal endoscopy.

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Is YouTube a Reliable Source for Laryngomalacia Information?

Alyssa D. Reese BA BS[1], Lauren A. DiNardo BS[1], Austin Knorz BS[1], Emilie Christie BS[1], Kristina Powers BS[1], Michele M. Carr DDS MD PhD[2]

[1]Jacobs School of Medicine and Biomedical Sciences at the University at Buffalo, Buffalo, NY
[2]Department of Otolaryngology – Head and Neck Surgery, Jacobs School of Medicine & Biomedical Sciences at the University at Buffalo, Buffalo, NY

Introduction

- The internet has become a widely used resource to learn more about medical concerns. This holds true for parents who have a child with otolaryngological complaints
- With the vast amount of information available on the internet, the accuracy and credibility of content is widely variable
- The quality of medical content on the internet and its influence on patients have not been extensively studied

Objectives

- Assess the quality of information in YouTube videos on laryngomalacia

Methods

- YouTube search of "laryngomalacia" was performed and the first 100 videos considered "relevant" were included
- Videos that did not discuss laryngomalacia and/or were not spoken in English were excluded
- Two students watched each video and evaluated each based on a variety of factors (Author, Category, Goal, Video and Audio Quality, Basic YouTube Metrics, Laryngomalacia Definition, Means of Diagnosis, Symptoms, and Treatment Options) as well as a modified DISCERN criteria (Table 1)

Table 1: Modified DISCERN Criteria

1	**Are the aims clear and achieved?**
2	**Are reliable sources of information used?**
3	**Is the information presented balanced and unbiased?**
4	**Are additional sources of information listed for patient reference?**
5	**Are areas of uncertainty mentioned?**

Results

- A total of 95 videos were analyzed
- Of the various author types, patient family or friends was the most common at 34.7% (N=33) (Figure 1)

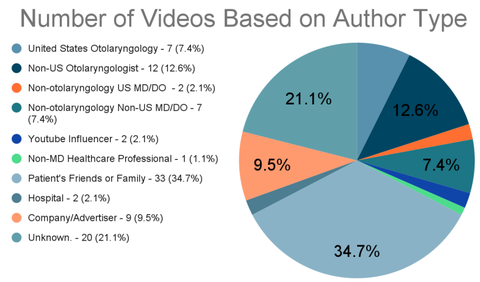

Figure 1: Distribution of videos by author types

- The goals of the videos were most frequently the recognition of symptoms of laryngomalacia (N=30, 31.6%) and providing a complete overview of laryngomalacia (N=24, 25.3%) (Figure 2)

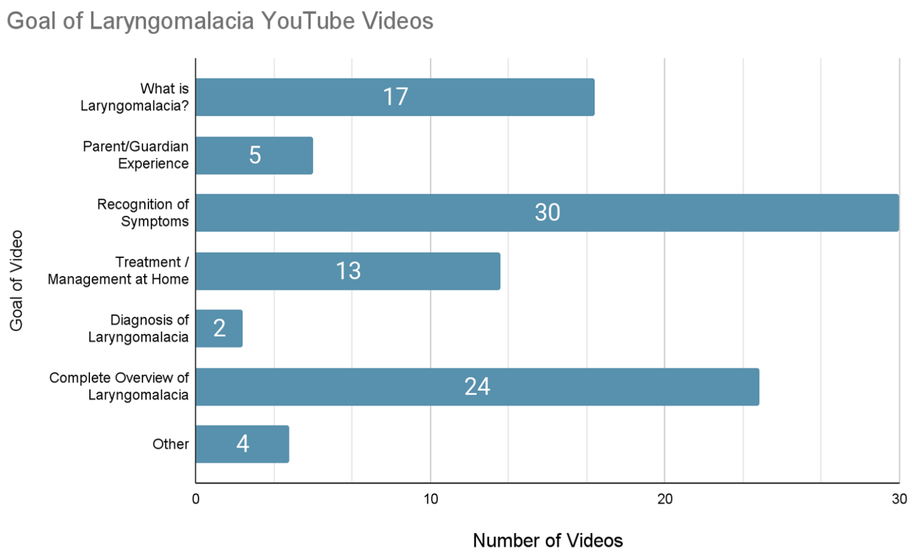

Figure 2: Number of Videos Based on Goal of Video

- Videos made for educational purposes by hospitals and providers were statistically more likely to be of better quality in terms of video clarity, text, and graphics (p=.001) when compared to videos classified as testimonial and patient caregiver experiences
- 48.3% (N=28) of the educational videos were classified as 3/3 for video quality, while 10.8% (N=4) of the testimonials and patient caregiver experience videos were rated 3/3
- The mean number of DISCERN criteria met was 1.53 (σ = .987) and the majority of the videos met only one of the five DISCERN criteria (N=56, 58.9%) and
- Only one of the videos (N=1, 1.0%) shared resources with viewers

Discussion

- A prior study that evaluated YouTube videos focusing on major procedures in pediatric otolaryngology based on a 15 point DISCERN criteria tool found that videos met a mean of 2.39 out of 15 DISCERN criteria. In our study, we used a shorter DISCERN criteria, but also found that the majority of laryngomalacia YouTube videos met a small number of the DISCERN criteria.
- An analysis of 62 videos on Facebook pertaining to otitis media found that the majority (56%) focused on complementary and alternative medication without mentioning any current guidelines [2]. The data from our study also showed that few videos included references to the information discussed.
- Videos were most frequently made by the patient's family and friends, which may explain why few videos contained unbiased information and areas of uncertainty.

Conclusion

- When parents search YouTube for information about laryngomalacia, most results will be home videos of personal testimonials and caregiver experiences
- Misconceptions about laryngomalacia may be reduced if medical professionals refer patients to high quality social media sources

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Is Preoperative Imaging Indicated Before Pharyngeal Surgery in Children with 22q11.2 Syndrome?: A Systematic Review and Meta-Analysis

Kelsey A. Duckett BS, Nicolas S. Poupore BS, William W. Carroll MD, Phayvanh P. Pecha MD

Department of Otolaryngology – Head & Neck Surgery, Medical University of South Carolina, Charleston SC

BACKGROUND

- The incidence of 22q11 Deletion Syndrome (22qDS) is 1 in 2,000 to 7,000 births.[1-3]
- Compared to the general pediatric population, there is a high prevalence of velopharyngeal dysfunction (VPD), or velopharyngeal insufficiency (VPI), in patients with 22qDS.[1-4]
- Patients with 22qDS often have more severe VPD refractory to nonsurgical management, commonly requiring pharyngeal surgery. [1-4]
- VPD surgery is challenging for patients with 22qDS due to concerns of medialization of the internal carotid artery (ICA).[2,3,5]
- While ICA medialization is a concern in pharyngeal operations, there is no consensus on whether routine preoperative imaging should be obtained in patients with 22qDS before VPD surgery.[2,6]

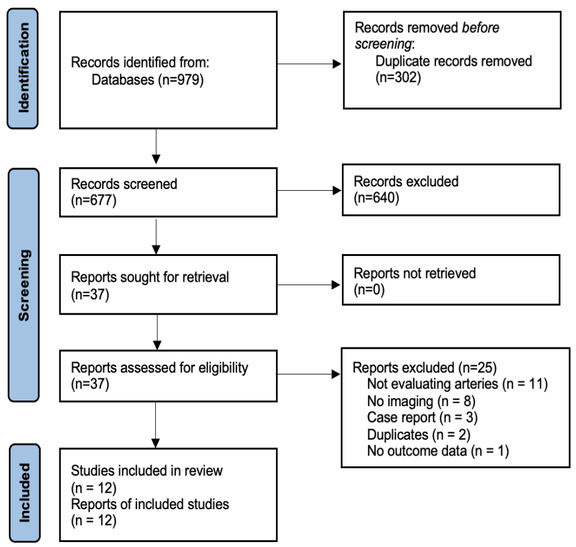
Study Aims

- **Primary:** To evaluate trends of preoperative imaging before pharyngeal surgery in patients with 22qDS in identifying internal carotid artery (ICA) medialization.
- **Secondary:** To assess reported rates of nasopharyngoscopy as a modality to predict medialization in this population.

METHODS

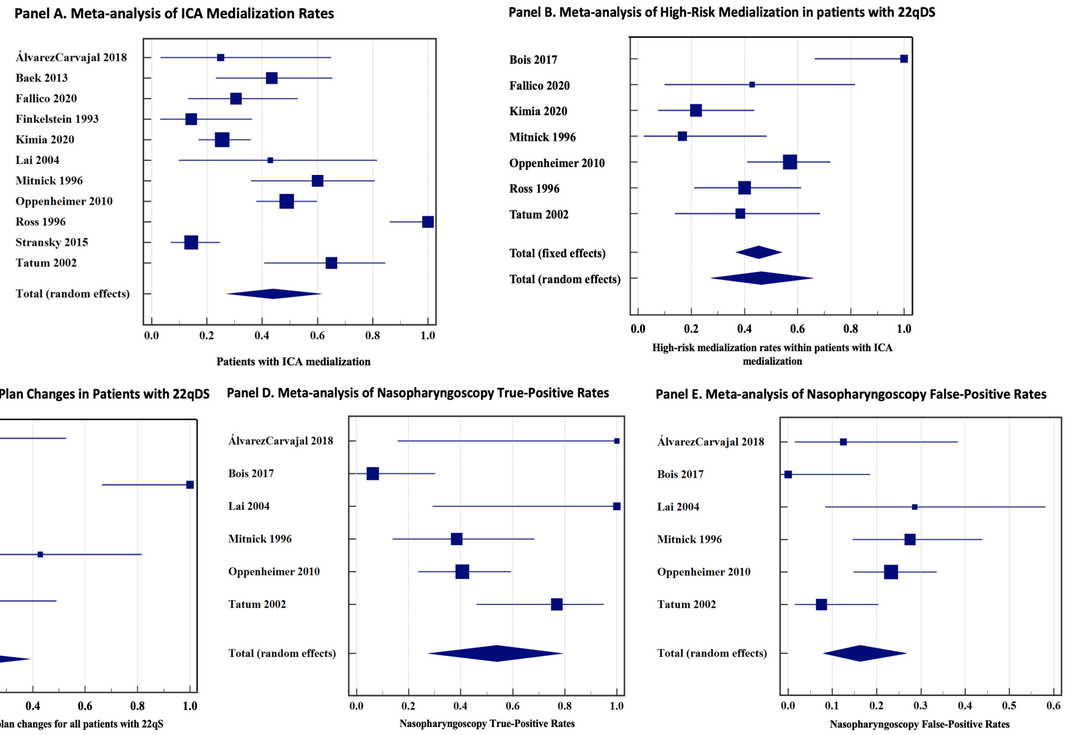
- ***Data Sources:*** PubMed, Scopus, and CINAHL.
- ***Review Methods:*** Following PRISMA guidelines, a systematic review was performed.
- ***Terms:*** "22q11 Deletion Syndrome", "imaging", "surgical procedures", and "pediatrics".
- Studies of patients with 22qDS who underwent preoperative imaging (MRA, CTA, or videofluoroscopy) to identify ICA anomalies were included.
- High-risk medialized ICAs were defined as either submucosal, retropharyngeal, Pfeiffer Grade III-IV, or <3mm from the pharyngeal mucosa.
- Meta-analyses of proportions were performed.

Figure 1. PRISMA flow diagram of study selection



RESULTS

Figure 2. Meta-analyses



- A total of 12 studies (N = 419 patients with 22qDS), all retrospective case series or case-control analyses, were included.
- Weighted mean age was 7.4 years.
- In 402 patients with 22qDS who underwent imaging, the rate of ICA medialization was 44.0% (95%CI 27.3-61.4), 46.3% (95%CI 27.4-65.8) of those being high-risk.
- Of the 254 cases that reported whether imaging altered operative plans, 19.4% (95%CI 5.7-38.8) of surgeries were modified by changing surgical method or flap size due to medialized ICA.
- Of 274 patients with reported surgical cancellation rates, 1.2% (95%CI 0.3-3.3) of patients overall and 3.6% (95%CI 0.9-9.4) of patients with medialization had their operation cancelled.
- In 6 studies that used nasopharyngoscopy pulsations to predict medialization, the true-positive rate was 53.9% (95%CI 27.5-79.2), the false-positive rate was 16.2% (95%CI 7.9-26.8), and the false-negative rate was 18.4% (95%CI 4.5-38.7).
- Ten of the twelve studies (83.3%) recommended universal preoperative imaging of the ICAs in patients with 22qDS undergoing pharyngeal surgery.
- No cases identified perioperative bleeding secondary to ICA injury.

Table 1. Description of Included Studies

Author (Year)	Total 22qDS Patients (N)	Patients with Imaging (N)	ICA Medial-ization (N)	Completed surgery (N)	Imaging modality used	Endorsed universal preoperative imaging? (Yes/No)
Álvarez Carvajal (2018)	8	8	2	8	CTA	Y
Baek (2013)	23	23	10	23	MRA*	N
Bois (2017)	9	9	9	9	MRA or CTA	Y
Fallico (2020)	40	23	7	NR	MRA	Y
Finkelstein (1993)	21	21	3	NR	VFS	Y
Kimia (2020)	90	90	23	NR	MRA	Y
Lai (2004)	7	7	3	6	MRA &/or CTA	Y
Mitnick (1996)	20	20	12	15	MRA	Y
Oppen-heimer (2010)	86	86	42	NR	MRA*	Y
Ross (1996)	25	25	25	NR	CTA	N
Stransky (2015)	70	70	10	69	MRA or CTA	Y
Tatum (2002)	20	20	13	20	MRA or CTA	Y

CTA = Computed Tomography Angiography; MRA = Magnetic Resonance Angiogram; N=number of participants; NR=not reported; VFS = Videofluoroscopy; *=MRA unless contraindicated or other CTA indication

CONCLUSIONS

- Most studies endorse routine preoperative imaging to assess for ICA medialization in patients with 22qDS undergoing pharyngeal surgery, which can lead to surgical modifications.
- Studies found that nasopharyngoscopy could not reliably predict ICA medialization as an alternative to imaging.
- Outcomes in children who do not undergo routine pre-operative imaging are unknown. Additional studies are needed to compare outcomes in children with or without preoperative imaging.

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Importance of Early Objective Auditory Testing in the Presentation of Sudden Sensorineural Hearing Loss in Children

Tanya Chen MD[1,2], Vicky Papaioannou MSc[1-3], Gillian Liberman MSW[4], Karen A Gordon PhD[1-3,5,6], Blake C Papsin MD[1,2,5,6], Sharon L Cushing MD[1,2,5,6]

[1]Department of Otolaryngology – Head & Neck Surgery, Hospital for Sick Children. Toronto, Canada. [2]Department of Otolaryngology – Head & Neck Surgery, Faculty of Medicine, University of Toronto, Toronto, Canada. [3]Department of Communication Disorders, Hospital for Sick Children. Toronto, Canada. [4]Social Work Department, Hospital for Sick Children. Toronto, Ontario. Canada. [5] Archie's Cochlear Implant Laboratory, Hospital for Sick Children, Toronto, Canada. [6] Institute of Medical Science, University of Toronto, Toronto, Canada

Introduction

Workup for sudden sensorineural hearing loss (SSHNL) routinely includes only subjective behavioural audiologic assessment which may be consistent with hearing loss. Pseudohypoacusis can be due to an auditory manifestation of conversion disorder. This is often underdiagnosed and improperly treated in children.

Definitions

Pseudohypacusis: hearing loss in the absence of an organic cause, also known as functional or non-organic hearing loss
Conversion disorder: somatoform disorder, usually triggered by psychological stressor, with physical symptoms that cannot be explained by an underlying medical pathology
Malingering: intentional production of false or grossly exaggerated physical or psychological problems for secondary gain

Figure 1. Treatment algorithm of sudden sensorineural hearing loss.

Objectives

To present our experience of three children presenting with delayed diagnoses of pseudohypacusis due to conversion disorder and highlight the considerations in evaluation of pediatric SSNHL, specifically of early objective audiometric testing.

Methods

This is a case series of 3 pediatric patients seen in a tertiary care Otolaryngology clinic with a diagnosis of idiopathic SSNHL between Sep 2019 - Jul 2022.

Case 1

ID: 10 year old male, bilateral SNHL
Past medical history: cleft lip and palate repair
Presenting symptoms: initial SSNHL in left ear in 2016, then onset of SSNHL to right ear in 2019
Triggering event: Unknown
Initial treatments: oral steroids, hearing aids
Investigations: CT, MRI, CMV testing
Audiogram: bilateral profound SNHL
Speech discrimination: did not respond to SRT, 100% SAT bilaterally at PTA
DPOAE testing: present 2-8 kHz
ABR: normal bilaterally at 500-4000 Hz
Intervention: counselling sessions
Delay in diagnosis: 3.5 years
Final outcome: Return of normal hearing, cessation of HA use

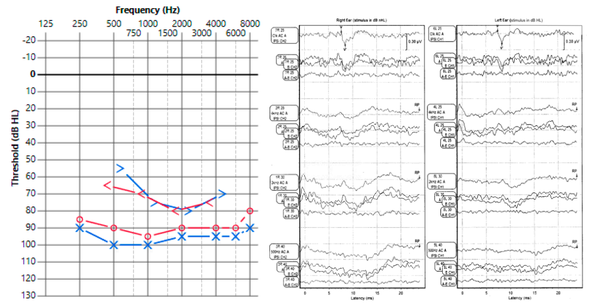

Figure 2. Initial audiogram of Case 1 (left) and ABR (right)

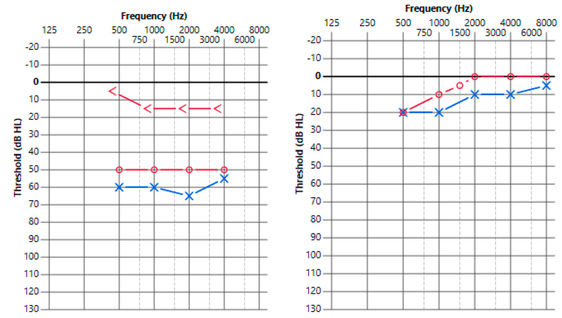

Figure 3. Audiograms of Case 1 during (left) and after psychotherapy (right)

Case 2

ID: 14 year old female with right SNHL
Reason for referral: CI consideration
Past medical history: none
Triggering event: trauma to contralateral side of head 1 week prior, anniversary of mother's death 1 year prior
Initial treatments: oral steroids, HBOT
Investigations: MRI
Speech discrimination: 100% left ear at 10 dBHL, right ear no awareness at 105 dBHL
DPOAE testing: present 2-8 kHz
ABR: normal bilaterally at 500-4000 Hz
Stenger's test: + at 1kHz
Intervention: counselling sessions
Delay in diagnosis: 2 months
Final outcome: Ongoing therapy

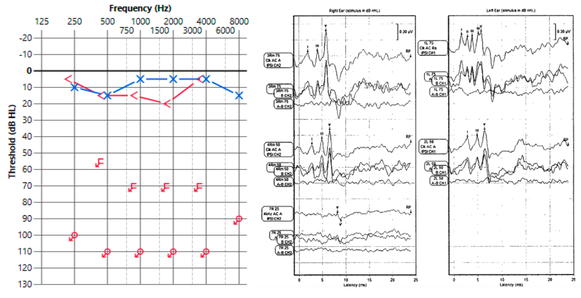

Figure 4. Initial audiogram of Case 2 (left) and ABR (right)

Case 3

ID: 14 year old female, with bilateral SNHL
Reason for referral: CI consideration
Past medical history: social anxiety
Triggering event: traumatic family events same summer of hearing loss
Initial treatments: oral steroids, hearing aids
Investigations: MRI
Audiogram: bilateral severe-profound SNHL
Speech discrimination: 80% right ear and 72% left ear at 95 dBHL
DPOAE testing: present 2-8 kHz
Stenger's test: + at 1kHz
ABR: normal bilaterally at 500-4000 Hz
Intervention: counselling sessions, antidepressant
Delay in diagnosis: 3.5 years
Final outcome: subjective hearing improvement

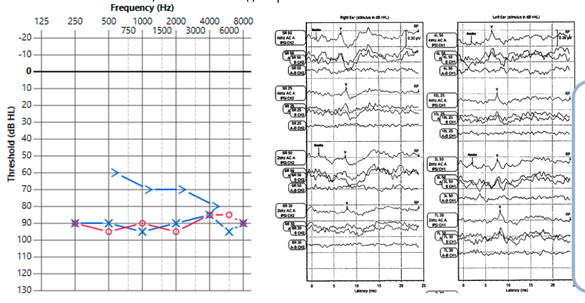

Figure 5. Initial audiogram of Case 3 (left) and ABR (right)

Conclusion

The three cases presented above were all examples of conversion disorder-related pseudohypacusis. All three patients received unnecessary investigations and treatment prior to correct diagnosis. Inconsistency between behavioural audiological testing and objective measures is important in recognizing pseudohypacusis. Early use of other objective measures in the workup of SSNHL can avoid unnecessary and potentially harmful therapies.

Air-bone gap in absence of middle ear dysfunction, lack of vibrotactile response, discrepancy in SRT/discrimination and PTA

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Safety and Outcomes of Early Cochlear Implant Activation in Pediatric Recipients

Robert Cox MD[1], Evie Landry MD, FRCSC[2], Hengameh Behzadpour MSHS[2], Tracey Ambrose AUD-CCA[3], Diego Preciado MD, PhD[2], Brian Reilly MD[2]

[1.] Division of Otolaryngology-Head and Neck Surgery, The George Washington University School of Medicine and Health Sciences, Washington, DC, USA.

[2.] Department of Otolaryngology, Children's National Hospital, Washington, DC, USA.

[3.] Division of Hearing and Speech, Children's National Hospital, Washington, DC, USA

Introduction

Cochlear implants (CI) are the standard of care for pediatric patients with severe-profound hearing loss due to substantial clinical benefits. There is limited evidence for the ideal time to activate the processor for optimal clinical outcomes.[1] Current studies demonstrate safety in activations earlier than traditional timeframes, without increased wound complications or long-term changes in impedance measurements.[1-3] We evaluated the complication rates and hearing outcomes between early activation (EA; ≤3 weeks) and standard activation (SA; >3 weeks) of CIs in pediatric patients.

Methods

A retrospective chart review of pediatric patients with cochlear implantation between 2015-2022 was performed. Data collected included demographics, clinical variables, complication rates, median time from implantation to Ling 6 sounds, and median time from implantation to target MAP. Complications were defined as any pathologic event or device failure occurring during the postoperative period. Mann-Whitney-U test, Chi-squared and Fisher's exact test were used as appropriate to determine presence of statistically significant differences between patients undergoing early versus standard activation.

Results

98 patients were included in analysis, 33% in the EA cohort. There were no significant differences between EA and SA cohorts with respect to age, gender, race/ethnicity, state of residence, laterality of implant, CI brand, or presence of comorbid syndrome. Privately insured patients made up a larger proportion of the SA cohort compared to the EA cohort (p=0.015).

No major complications were reported in either group. No significant difference was identified in complication rates between cohorts (15.2% vs. 25.0%, p=0.238). Similarly, no significant differences in median time (days) to Ling 6 at 100% or target MAP were identified (98.0 vs. 87.5, p=0.552; 58.5 vs. 64.0, p=0.411).

Outcomes	Overall (N= 98)	Standard Activation (N= 66)	Early Activation (N= 32)	P value
Complications, n (%)	18 (18.4)	10 (15.2)	8 (25.0)	0.238
Median time to Ling 6 at 100% (days) [IQR]	93.5 [42.0, 275.2]	98.0 [49.0, 249.8]	87.5 [41.5, 316.0]	0.552
Median time to target map (days) [IQR]	61.0 [35.0, 98.0]	58.5 [29.8, 87.0]	64.0 [38.8, 126.0]	0.411

*P values were obtained from Mann-Whitney-U test for continuous data and Chi-square test for binary and categorical data

Within the early activation cohort, device brand and age (≤18 or >18 months) were not found to have significant influence on complications or audiologic outcomes. However, bilateral implantation had a significantly higher complication rate compared to unilateral implantation.

Outcomes	Age Category	Univariate (Unadjusted)	
		Estimate	P value
Complications, OR (95% CI)	≤18 months >18 months	Ref 0.24 (0.03-1.6)	Ref 0.133
Time to Ling 6 at 100% (days), Coefficient (95% CI)	≤18 months >18 months	Ref 40.1 (-247.9, 328.2)	Ref 0.778
Time to target map (days), Coefficient (95% CI)	≤18 months >18 months	Ref 20.8 (-66.9, 108.5)	Ref 0.631

Outcomes	Side	Univariate (Unadjusted)	
		Estimate	P value
Complications, OR (95% CI)	Unilateral Bilateral	Ref 6.3 (1.2-41.0)	Ref **0.037**
Time to Ling 6 at 100% (days), Coefficient (95% CI)	Unilateral Bilateral	Ref -104.9 (-344.6, 134.8)	Ref 0.379
Time to target map (days), Coefficient (95% CI)	Unilateral Bilateral	Ref -28.5 (-101.9, 44.8)	Ref 0.433

*OR= odds ratio, CI= confidence interval

Conclusions

We conclude there is no significant increased risk of complications or difference in audiologic outcomes between the standard and early activation cohorts. Similarly, patient age and device brand did not significantly influence these outcome variables within the EA cohort. This highlights the similar safety and efficacy profile of early CI activation and the possibility for shorter periods between implantation and activation without complication, allowing for earlier device usage and audiological programming.

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Association between no-show rates and interpreter use in a pediatric otolaryngology clinic

Bita Naimi BA,[1] Pratima Agarwal MD,[2] Haoxi Ma MS,[3] Jessica R Levi MD[1, 2]

[1]Boston University School of Medicine, Department of Otolaryngology, Boston, MA, USA

[2]Boston Medical Center, Department of Otolaryngology, Boston, MA, USA

[3]University of Connecticut, Department of Statistics, Storrs, CT, USA

Introduction

- A patient no-show is defined as a missed appointment with no prior notification to the healthcare provider
- Higher no-show rates have been shown to be associated with poor health outcomes and increased all-cause mortality[1]
- Within pediatric otolaryngology, increased no-show rates are associated with increased referral to appointment latency, appointments occurring in the afternoon, complex psychiatric history, and complex maternal medical history[1-3]

- To date, no prior studies have investigated the relationship between interpreter use at the patient's first visit and subsequent attendance in pediatric otolaryngology
- In this study, we aim to understand the effect of interpreter utilization at the patient's first visit on no-show rates in pediatric Otolaryngology

- Retrospective cohort study
- Academic tertiary care center, safety net hospital
- 1270 new patients from 12/31/2014 to 12/31/2019 with at least one no-show to the pediatric otolaryngology clinic
- Demographic data included patient age, primary language, interpreter requirement, ethnicity, primary insurance, maternal education level, and number of recorded patient medical problems at the time of the first visit
- Attendance data included total number of no-show appointments and total number of completed appointments
- All patients who needed interpretation at their initial appointment, or who were seen by a certified Spanish bilingual provider, were categorized as receiving interpretive services

Table 1. Patient Demographics by No Show Rates

Patient Characteristic, (N)	No Show Rate, Mean ± SD (Median)	P-value
Age (1270)	0.37 ± 0.16 (0.33)	0.8173
Primary Language (1270)		**0.0474**
English (696)	**0.38 ± 0.16 (0.33)**	
Spanish (363)	**0.36 ± 0.16 (0.33)**	
Haitian Creole (67)	**0.38 ± 0.17 (0.40)**	
Other (144)	**0.39 ± 0.14 (0.38)**	
Ethnicity (1240)		0.1160
Hispanic (490)	0.36 ± 0.16 (0.33)	
Non-Hispanic (750)	0.38 ± 0.16 (0.33)	
Insurance (1264)		0.7208
BMC (528)	0.38 ± 0.16 (0.33)	
Tufts (85)	0.39 ± 0.15 (0.40)	
Medicaid (509)	0.37 ± 0.16 (0.33)	
BCBS (38)	0.33 ± 0.14 (0.33)	
Other (104)	0.37 ± 0.15 (0.33)	
# medical problems at first visit (102)	0.38 ± 0.16 (0.33)	0.2074

Table 2. No Show Rates by Language Needs and Interpreter Use at First Visit

Patient Characteristic, (N)	No Show Rate, Mean ± SD (Median)	P-value
Language Needs (1270)		0.1417
No Interpreter Needed (850)	0.38 ± 0.16 (0.33)	
Interpreter Needed (420)	0.36 ± 0.16 (0.33)	0.5584
English (9)	0.34 ± 0.13 (0.33)	
Spanish (289)	0.36 ± 0.16 (0.33)	
Haitian Creole (32)	0.39 ± 0.17 (0.41)	
Other (90)	0.38 ± 0.14 (0.33)	
For patients who need interpreter:		0.3674
No interpreter at first visit (267)	0.30 ± 0.18 (0.33)	
	0.32 ± 0.19 (0.33)	**0.0265**
Interpreter at first visit (153)	**0.31 ± 0.19 (0.29)**	
Spanish (94)	**0.42 ± 0.18 (0.43)**	
Haitian Creole (11)	**0.32 ± 0.19 (0.33)**	
Other (48)		

Demographics (Table 1)

- Primary language
  - Highest no-show rate: Haitian Creole, median 40%
  - **Lowest no-show rate: English and Spanish, both median 33%**
  - All other primary languages: median 38%
  - No difference in no-show rates by patient's age, ethnicity, insurance, number of medical problems at the time of the first visit, maternal education level, or maternal primary language

Interpreter Use (Table 2)

- Overall interpreter use was not significantly associated with no-show rates (p=0.3674)
- However, there was a statistically significant difference in average no-show rates for patients based on the language in which interpretation was provided (p = 0.0265)
  - **Lowest no-show rate: Spanish interpreter at the first visit (31% ± 19%)**
  - Highest no-show rate: Haitian Creole interpreter at the first visit (42% ± 18%)

- Overall, interpreter use at the first visit does not affect no-show rates
- Among patients that did use an interpreter at the first visit, patients receiving interpreter services in Spanish had lower average no-show rates compared to patients receiving interpreter services in other languages
- It is important to understand factors associated with no-show rates, especially in minority or underserved populations, in order to…
  - Address barriers to accessing health care
  - Reduce healthcare disparities
  - Promote health equity in these populations

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Assessing the safety of topical epinephrine in pediatric functional endoscopic sinus surgery

Natalie Quinn[1], Austin Schafer[2], Tran Bourgeois[3], Charles A. Elmaraghy[1,2]

Otolaryngology—Head and Neck Surgery, Nationwide Children's Hospital, Columbus, OH[1], The Ohio State University College of Medicine, Columbus, OH[2], Center for Surgical Outcomes Research, Nationwide Children's Hospital, Columbus, OH[3]

Abstract

Objective: To assess the safety of topical epinephrine during pediatric functional endoscopic sinus surgery (FESS).

Methods: After obtaining IRB approval, electronic medical records of patients aged 0-18 undergoing FESS in 2021 were retrospectively reviewed and divided into two cohorts based on the topical vasoconstrictive agent utilized during the case: oxymetazoline or epinephrine. Outcome variables consisted of pre-operative (PO) and maximum intra-operative (IO) heart rate (HR), systolic blood pressure (SBP), and diastolic blood pressure (DBP), as well as any administration of intraoperative antihypertensive medications. Patients who received both or neither vasoconstrictive agent were excluded.

Results: Among a total cohort of 96 FESS patients, 75 met inclusion criteria and were primarily male (59%), with median age of 13 (IQR 8,16). Oxymetazoline was administered to 60% (n = 45) of the cohort, while 30% (n = 30) received epinephrine. After adjusting for all confounders, mean differences between both agents were -0.04 (95% CL: -12.01,11.92, $P = 1$) for PO HR, -3.90 (95% CI: -15.75, 7.95, $P = 1$) for IO HR, 6.88 (95% CI: -5.89, 19.65, $P = 0.8813$) for PO DBP, 5.04 (95% CI: -7.26, 17.35, $P = 1$) for IO DBP, 6.64 (95% CI: -8.68, 21.95, $P = 1$) for PO SBP and 4.94 (95% CI: -9.90, 19.79, $P = 1$) for IO SBP. No intraoperative antihypertensives were administered to patients in either cohort.

Conclusion: Topical epinephrine and oxymetazoline have similar risk profiles at each individual time point as well as over time. A prospective study should be developed to evaluate the efficacy of topical epinephrine and to further evaluate its safety profile.

Background

- Functional endoscopic sinus surgery (FESS) is commonly performed in pediatric otolaryngology, with the most common indication being chronic rhinosinusitis (CRS)[1]
- Topical vasoconstrictors are critical to improve intraoperative visualization and reduce the risk of complications[2-5]
- Topical epinephrine is of growing interest for its powerful arteriolar and venous-sinusoidal constriction[6,7]
- However, its use is not without the potential risk of cardiovascular side effects such as tachycardia and hypertensive crisis[4,6]
- Thus, the aim of this study is to evaluate the safety profile of topical epinephrine during pediatric FESS

Methods

Study Population

- All patients undergoing FESS at Nationwide Children's Hospital in 2021
- Divided into 2 cohorts based on the vasoconstrictive agent: those with pledgets soaked with 0.05% oxymetazoline or those with epinephrine 0.1mg/ml (1:10,000)
- Patients outside of the age range 0-18 and those who received both or neither vasoconstrictive agent were excluded

Outcome and Independent variables

- Administration of intra-operative antihypertensive medications
- Pre-operative and maximum intra-operative heart rates (HR), systolic blood pressures (SBP), diastolic blood pressures (DBP)
- Demographic features and clinical characteristics such as indication for surgery, history of cystic fibrosis (CF), primary ciliary dyskinesia (PCD) or preexisting cardiovascular conditions

Statistical Analysis

- Chi square / Fisher's exact test and Wilcoxon Mann Whitney for bivariate analyses
- Univariate and multivariable linear mixed models used to assess differences in outcomes over time

Results

- No differences between epinephrine and oxymetazoline were observed at pre-operative and intra-operative time periods for all outcome variables ($P > 0.5$)
- Interaction between topical vasoconstrictor and time were also not significant ($P > 0.05$) indicating no difference between both agents over time

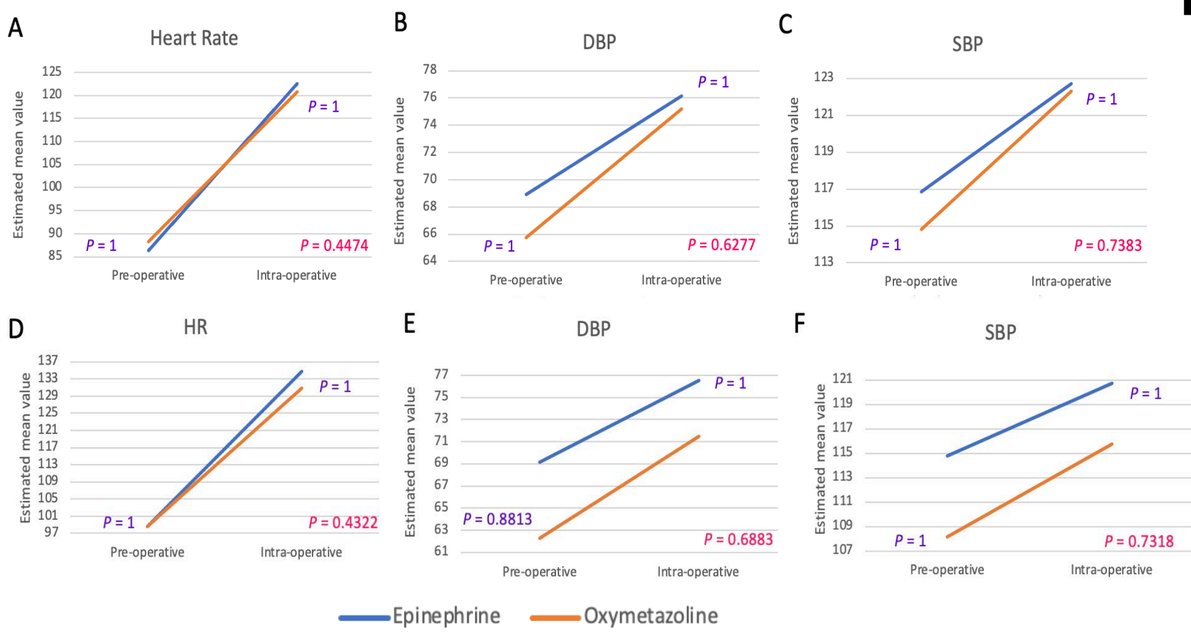


Figure 1: Outcome comparisons between epinephrine and oxymetazoline over time.

Predicted values for heart rate (HR), diastolic blood pressure (DBP), and systolic blood pressure (SBP) at pre-operative (time (0)) and intra-operative (time (1)) timepoints for both epinephrine (blue) and oxymetazoline (orange). Both univariable linear mixed models (A – C) and multivariable linear mixed models adjusting for confounders (D-F) are shown. *P* values (purple) at each time point indicate Bonferroni adjusted *p* value comparing change in HR, DBP, and SBP between epinephrine and oxymetazoline at pre-operative and intra-operative time points. Non-significant *p* values (red) at lower right corner look at the interaction of topical vasoconstrictor and time and indicates no difference between topical vasoconstrictor over time.

Discussion

- To our knowledge, this is the first study to evaluate the safety of topical epinephrine exclusively in children undergoing FESS
- We found no significant differences in intraoperative hemodynamic parameters between oxymetazoline and epinephrine
- No antihypertensive administration was required in either cohort
- Topical epinephrine does not appear to significantly alter intraoperative hemodynamic parameters
- Results are concordant with multiple adult studies
- Based on these findings, it is reasonable to develop a prospective study to evaluate the efficacy of topical epinephrine and to further evaluate its safety profile

References and Contact Information

Chronic Post-Tympanostomy Tube Otorrhea: Stepwise Management and Review of the Literature

Drew Gottman, BS; Dee Dee Gilbert, CPNP; Bethany Thomas, CPNP; Christian Francom, MD; and Sarah A. Gitomer, MD
Children's Hospital Colorado and the University of Colorado School of Medicine

Children's Hospital Colorado

Affiliated with University of Colorado Anschutz Medical Campus

BACKGROUND

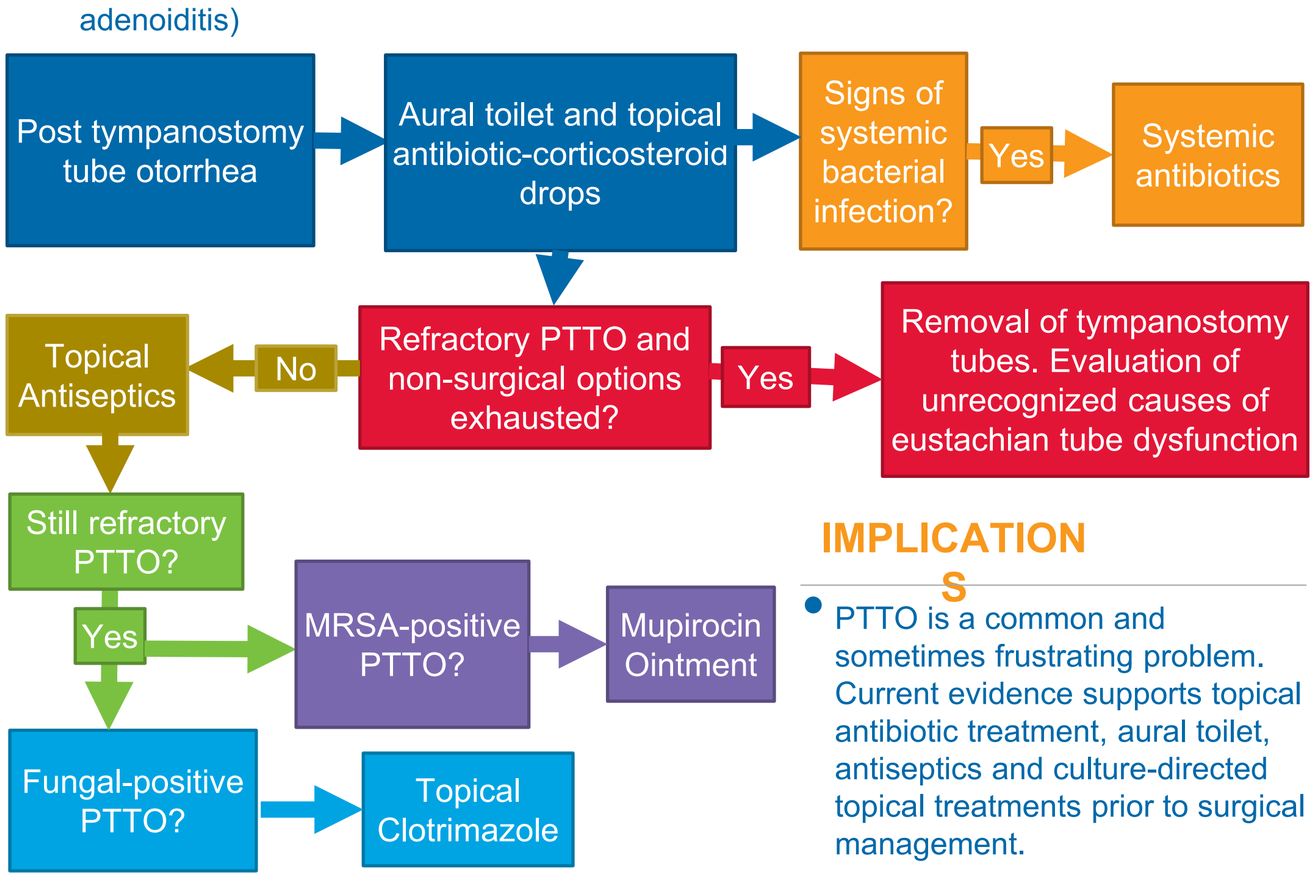
- Post-tympanostomy tube otorrhea (PTTO) is the most common complication of tympanostomy tube placement
- It is often difficult to manage with limited evidence-based guidance on treatment
- Academy of Otolaryngology–Head and Neck Surgery Foundation (AAO-HNSF) guidelines updated in 2022 recommend topical antibiotic drops for treating uncomplicated acute PTTO
- Other treatment options are not considered in the updated AAO-HNSF guidelines for more complicated cases of PTTO
- **Objective:** To provide an updated review of the literature and evidence-based algorithm for management of PTTO in children

METHODS

- Review of the English-language literature
- Articles from 2004-2022 were identified using PubMed and Google Scholar databases
- 23 studies are included

RESULTS

- First line: aural toileting + antibiotic-corticosteroid drops
- Adjuvant therapy for refractory PTTO includes:
  - Antiseptics: Acetic acid, hydrogen peroxide, alcohol, and aluminum acetate
  - Antibiotics/Antifungals: Mupirocin for MRSA-positive TPPO. Clotrimazole for fungal-positive TPPO
- Removal of tubes for persistent TPPO
- Concurrent evaluation of eustachian tube dysfunction and underlying syndromes (SMCP, adenoiditis)



CONCLUSIONS

- Review of the literature supports a stepwise for treatment of chronic PTTO
- Antiseptics are a reasonable second-line option for PTTO
- Otolaryngologists should consider other causes of eustachian tube dysfunction in cases of chronic PTTO

IMPLICATIONS

- PTTO is a common and sometimes frustrating problem. Current evidence supports topical antibiotic treatment, aural toilet, antiseptics and culture-directed topical treatments prior to surgical management.

DISCLOSURES

- No financial disclosures or conflicts of interest

University of Colorado Anschutz Medical Campus

Children's Hospital Colorado

A Silent Burden of the COVID-19 Pandemic: The Undocumented Surge in Facial Dog Bites

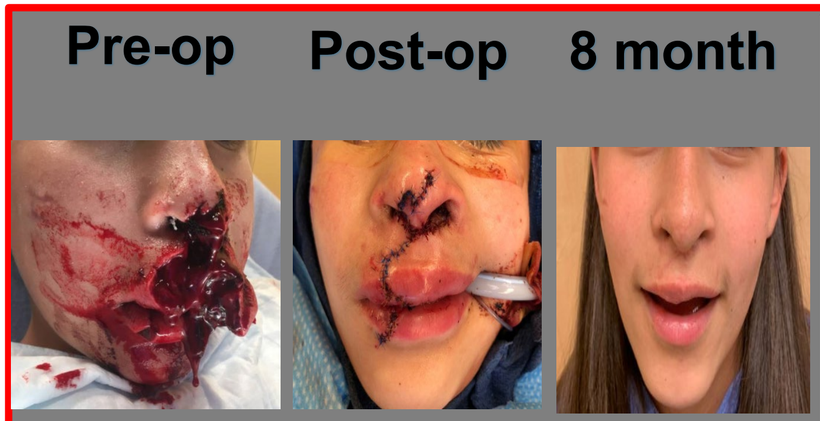
PURPOSE

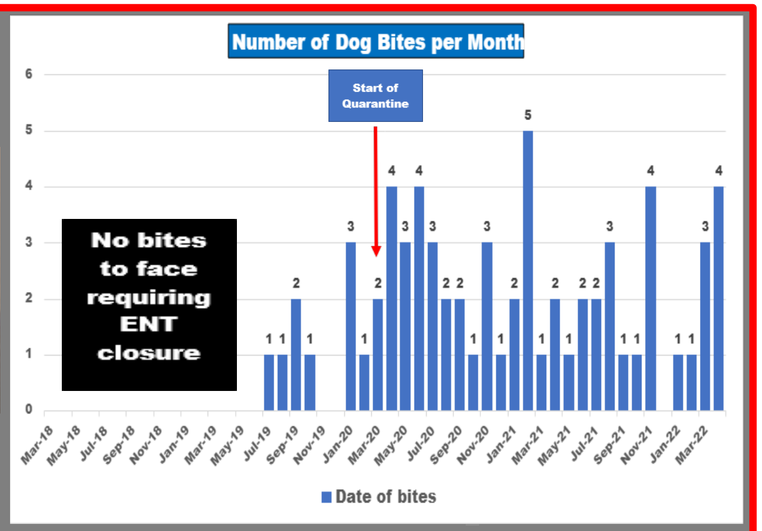
- During the 2020 COVID-19 pandemic lockdown, a presumed increase in pediatric facial dog bites requiring ENT surgical intervention was noted.
- The aim of this study is to document and compare the number of pediatric facial dog bite cases that occurred before and during the lockdown at Children's Hospital Colorado – Colorado Springs & UCHealth Memorial Hospital Central.

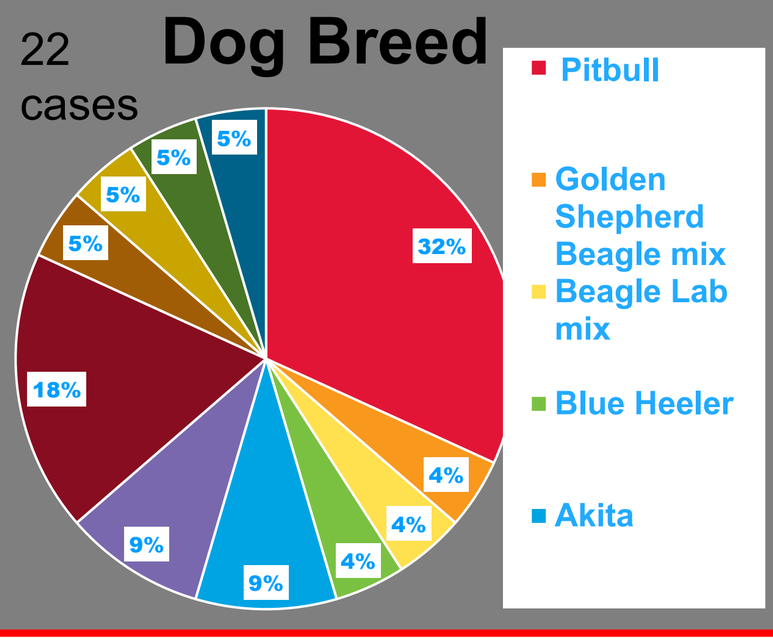
METHODS

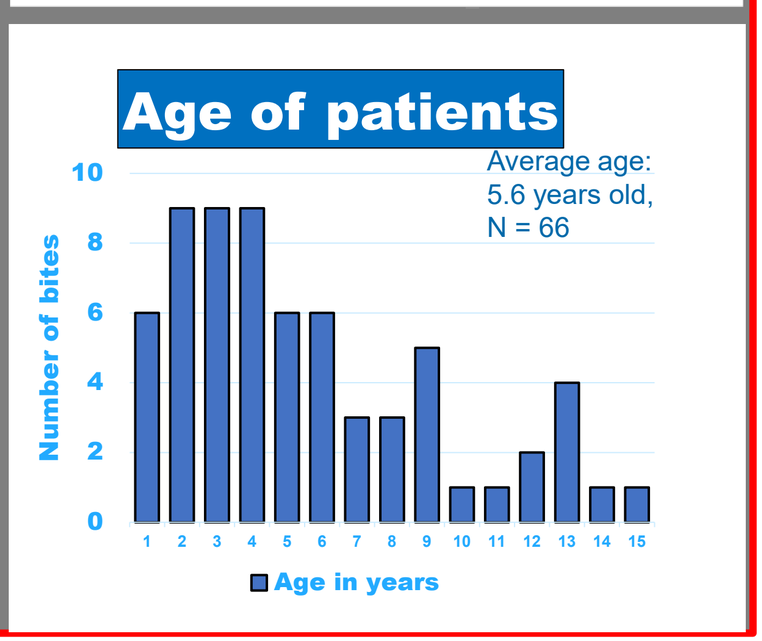
- COMIRB approval
- Chart review of pediatric patients with facial dog bites that required operative closure by ENT surgeons between March 2018 and March 2022.

RESULTS









RESULTS

- 1 dog bite repair/month 9 months prior to quarantine
- 2.5 dog bite repairs/month 9 months during and after quarantine
- 52% male
- Average age 5.6 years
- Nearly 75% of patients were Caucasian with over 20% Hispanic

Conclusions/Future Directions

- Significant increase in cases of dog bites at our institution during the COVID19 lockdown.
- Continued elevation in # of bites despite lift of quarantine. Unclear reason.
- Improved reporting to animal control and local pediatricians.
- Community education and policy changes.

A Rare Case of Retained Foreign Body in Maxillary Sinus after Penetrating Globe Injury

Mason Soeder BA, Lara Reichert MD
Department of Otolaryngology, Albany Medical College, Albany, NY 12208.

Introduction

Advancements in spring and gas-powered BB and pellet guns have led to increased projectile velocity and power of these "toy" weapons[1].

13,000 gunshots of children with non-powder guns occur annually in the US, resulting in 6500 retained foreign bodies every year[2,3].

Retained foreign bodies within the sinus can lead to complications including sinusitis, bleeding, and lead poisoning[4, 5, 6].

We report a unique case of a retained BB, which entered the maxillary sinus by perforating the left globe and crossing the orbital floor; the first documented case of a retained maxillary BB with this route of entry.

Hospital Course and Follow Up

Emergency Department

13-year-old male presented to ED after being shot in the left eye with a BB gun. On examination by the ophthalmology service, the left eye was swollen, and the patient was unable to open the left eyelid, and the left globe was ruptured.

Otolaryngology service: no palpable bony step-offs, and the face sustained no lacerations or abrasions. The nose was clear anteriorly and negative for a septal hematoma.

CT orbital and facial scans: acute displaced fracture of the left orbital floor posteriorly. CT scan also revealed a BB at the floor of the left maxillary sinus 5 mm in diameter.

Surgical Intervention

Emergent ruptured globe repair was performed.
Post-operatively, patient reported no sinus pressure or pain. The patient was discharged with oral Augmentin.

Follow Up

One-week post-op patient reports no anterior rhinorrhea but often spits out bloody mucous. No facial pain or pressure.

Elective maxillary antrostomy and foreign body removal was recommended. However, the patient was eventually lost to follow up.

Imaging and Figures

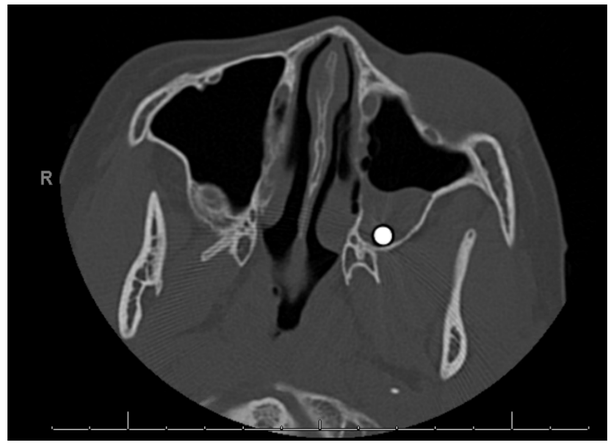


Figure 1: Axial Non-contrast Head CT. No evidence of an acute intracranial injury was seen. In the left maxillary sinus, a 5 mm BB can be seen with surrounding fluid. There are additional linear radiopaque fractured fragments with hemorrhage in the left maxillary sinus.

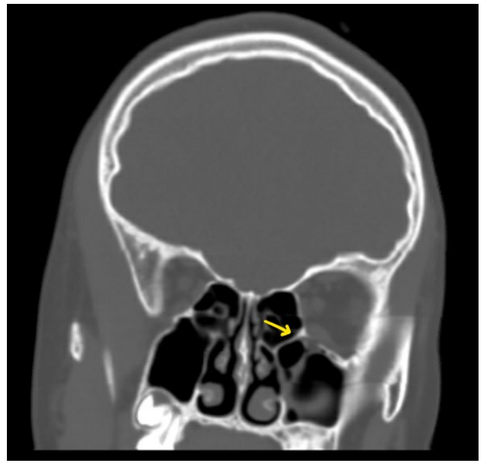


Figure 2: Coronal Non-contrast Facial CT. Constellation of findings is most suggestive of left globe rupture secondary to penetrating BB injury with associated displaced fracture involving the left orbital floor. This is associated with focal herniation of intraorbital fat into the adjacent sinus.

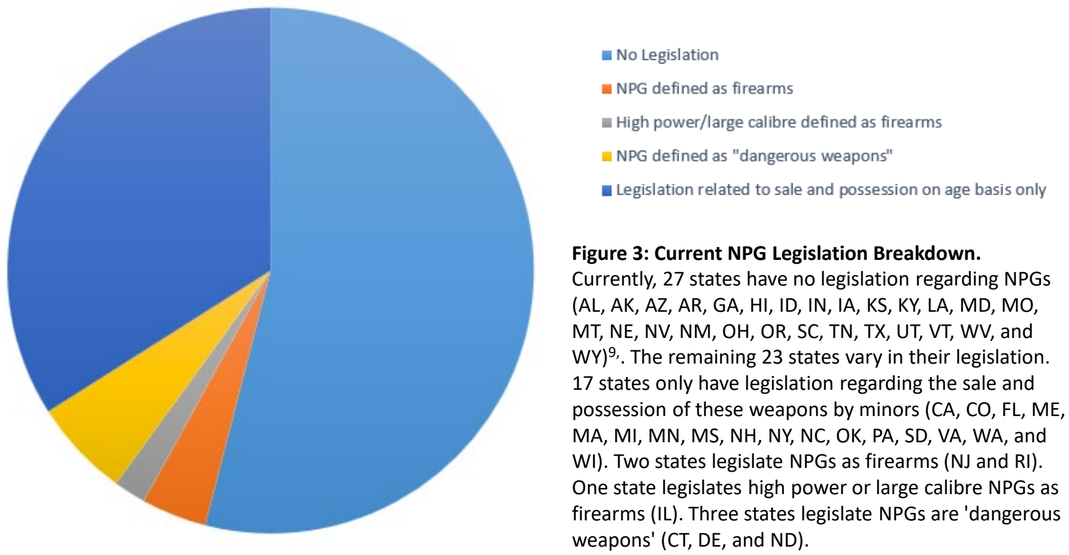


Figure 3: Current NPG Legislation Breakdown. Currently, 27 states have no legislation regarding NPGs (AL, AK, AZ, AR, GA, HI, ID, IN, IA, KS, KY, LA, MD, MO, MT, NE, NV, NM, OH, OR, SC, TN, TX, UT, VT, WV, and WY)[9]. The remaining 23 states vary in their legislation. 17 states only have legislation regarding the sale and possession of these weapons by minors (CA, CO, FL, ME, MA, MI, MN, MS, NH, NY, NC, OK, PA, SD, VA, WA, and WI). Two states legislate NPGs as firearms (NJ and RI). One state legislates high power or large calibre NPGs as firearms (IL). Three states legislate NPGs are 'dangerous weapons' (CT, DE, and ND).

Discussion

Overall rates of BB gun injuries have declined since 1990, rates of *eye* injury and severity of injury have steadily risen[3].

Most common route of entry into the maxillary sinus is through the skin and thin anterior bone covering the sinus[3].

Retained foreign bodies within the sinus can trigger acute local inflammation, leading to sinusitis, bleeding, disruption of mucocilliary clearance, and potentially meningitis[4].

Odd Findings and Complications

Brinson et. al: 8-year-old female patient also reported hypoesthesia of the infraorbital region. It has been proposed that lead BBs may increase lead concentrations in children[7].

Edetanlen et. al: Patients with retained lead shrapnel from gunshot wounds in the craniofacial region showed a 366% increase in lead blood concentrations at 3 months compared to control subjects. Risk unknown, as the concentrations remained lower than the threshold values published by the CDC[5].

Kuehnel et. al described a patient with a retained lead BB in the maxillary sinus for over 50 years. After removal by endoscopic antrostomy, the diagnosis of squamous cell carcinoma was confirmed by histopathological examination of the maxillary sinus[8].

Future Considerations

Recommend elective maxillary antrostomy.

Does this case warrant a CPS call?

Analyze BB composition for the presence of lead. If the BB is lead, a combination of chelation and surgical removal will be recommended.

Consider updating antiquated legislation that defines BB guns as 'weapons which shoot BBs' while not considering the force and velocity of the projectile.

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ASA Physical Status Classification as a Predictor of Adenotonsillectomy Complications

Abstract #98

Leyn Shakhtour BS[1] , Ishwarya Shradha Mamidi BS[2] , Ryan Lee MBA[1] , Lilun Li MD[3] , Joel W. Jones MD[2] , Andrew Mattisoff MD[4] , Brian K. Reilly MD[5]

[1]George Washington University School of Medicine and Health Sciences [2]Department of Otolaryngology; Louisiana State University [3]Division of Otolaryngology; The George Washington University Hospital [4]Division of Cardiac Anesthesia, Children's National Health System [5]Division of Otolaryngology, Children's National Medical Center

ABSTRACT

- ***Objective:*** To identify the association of pre-operative American Society of Anesthesiologists Physical status classification (ASA-PS) with 30-day complication rates and adverse events following tonsillectomy with or without adenoidectomy (T±A).
- ***Methods:*** A retrospective analysis was performed using data from the American College of Surgeons' National Surgical Quality Improvement Program database (ACS-NSQIP) of patients who underwent T±A between 2005 and 2016. Patients were stratified into ASA-PS Classes I/II and III/IV. Postoperative outcomes in the 30-day period following surgery were compared between the two subsets of ASA-PS groups.
- ***Results:*** On multivariate analysis, patients with ASA class III and IV were more likely to experience an unplanned readmission (OR 1.39, 95% CI 1.09-1.76; p=0.007), overall complications (OR 1.49, 95% CI 1.28-1.72; p<0.001), major complications (OR 1.52, 95% CI 1.31- 1.77, p= <0.001), reoperation (OR 1.33, 95% CI 1.04-1.69; p=0.022), and extended length of stay >1 day (OR 1.78, 95% CI 1.41-2.25; p<0.001).
- ***Conclusion:*** Higher ASA-PS classification is an independent predictor of complications following T±A. Surgeons should aim to optimize the systemic medical conditions of ASA-PS classes III and IV patients prior to T±A and implement post-operative management protocols specific to these patients to decrease morbidity, complications, and overall health care cost.

METHODS

- The American College of Surgeons (ACS) National Surgical Improvement Program (NSQIP) database was queried to identify all patients who underwent concurrent tonsillectomy and adenoidectomy from 2005 to 2016.
- Patients were isolated using the Current Procedural Terminology (CPT) code 42821 (tonsillectomy and adenoidectomy) and CPT code 42826 (isolated tonsillectomy).
- Radical resections of tonsils (CPT codes: 42842, 42844, and 42845), primary adenoidectomies (CPT code: 42831), and secondary adenoidectomies (CPT code: 42836) were excluded from this retrospective study in an effort to provide guidance specifically for candidates of simple removal of tonsils simultaneously with or without adenoids.
- All 5,936 selected patients were ≥ 16 years old and had complete data regarding their ASA classification.
- All patients were stratified into two cohorts: The first cohort included patients in ASA Classes I and II. The second cohort included patients in ASA Classes III and IV.
- NSQIP database variables can widely be categorized into five categories: demographics, preoperative comorbidities, laboratory values, administrative/operation-specific variables, and postoperative outcomes in the 30-day period following surgery.
- Pearson's chi-squared tests and Fischer's exact tests (expected cell sizes <5) were used to compare the prevalence rates of categorical variables, expressed as number and rates of occurrence.
- An unpaired t-test was used to compare differences in mean values and respective standard deviations of continuous variables.
- The multivariate logistic regression models yielded odds ratios (OR) with 95% confidence intervals (CI) as measures of increased levels of risk for each respective complication following tonsillectomy with or without adenoidectomy.
- All statistical findings with p-values less than or equal to 0.05 were considered significant for this retrospective study. All statistical tests were two-sided and performed at the 0.05 level of significance. Statistical analysis was performed using R statistical software, version 4.0.014.

RESULTS

Table 1a. Patient Demographics between ASA-PS Classes I/II vs. ASA-PS Classes III/IV

	Level	Total	Classes I/II	Classes III/IV	p-value
n		29163	25861	3302	
Age (mean (SD))		30.0 (11.9)	28.6 (10.6)	40.5 (15.5)	*<0.001
Age by Category (%)	<30 years	17807 (61.1)	16831 (65.1)	976 (29.6)	*<0.001
	>30 years	11356 (38.9)	9030 (34.9)	2326 (70.4)	
Sex (%)	Male	9855 (33.8)	8508 (32.9)	1347 (40.8)	*<0.001
	Female	19295 (66.2)	17342 (67.1)	1953 (59.2)	
Race (%)	White	19817 (68.1)	17633 (68.4)	2184 (66.2)	*<0.001
	Black or African American	2355 (8.1)	1851 (7.2)	504 (15.3)	
	Other	1698 (5.8)	1459 (5.7)	239 (7.2)	
	Unknown	5226 (18.0)	4855 (18.8)	371 (11.2)	
Hispanic (%)	No	21564 (76.5)	18937 (75.9)	2627 (81.4)	*<0.001
	Yes	2424 (8.6)	2147 (8.6)	277 (8.6)	
	Unknown	4199 (14.9)	3874 (15.5)	325 (10.1)	
BMI (mean (SD))		29.1 (7.7)	28.0 (6.6)	37.3 (10.2)	*<0.001
BMI Class (%)	1	10050 (35.0)	9687 (38.1)	363 (11.1)	*<0.001
	2	8152 (28.4)	7675 (30.2)	477 (14.6)	
	3	5060 (17.6)	4451 (17.5)	609 (18.7)	
	4	2761 (9.6)	2202 (8.7)	559 (17.1)	
	5	2670 (9.3)	1418 (5.6)	1252 (38.4)	
Obesity (%)	No	18202 (63.4)	17362 (68.3)	840 (25.8)	*<0.001
	Yes	10491 (36.6)	8071 (31.7)	2420 (74.2)	

Table 2. Administrative and Intraoperative Variables between ASA-PS Classes I/II vs. ASA-PS Classes III/IV

	Level	Total	Classes I/II	Classes III/IV	p-value
n		29163	25861	3302	
Total Operative Time (mean (SD))		29.6 (30.6)	28.3 (27.0)	40.3 (49.3)	*<0.001
Days from Admission to Operation (mean (SD))		0.1 (4.1)	0.1 (4.3)	0.1 (0.8)	0.931
Days from Operation to Discharge (mean (SD))		0.2 (1.9)	0.2 (1.8)	0.6 (2.5)	*<0.001
Total LOS (mean (SD))		0.3 (2.1)	0.2 (2.0)	0.7 (2.7)	*<0.001
Primary Anesthesia Modality (%)	General	29119 (99.9)	25826 (99.9)	3293 (99.8)	0.07
	Other	39 (0.1)	31 (0.1)	8 (0.2)	
Wound classification (%)	1	706 (2.4)	614 (2.4)	92 (2.8)	*<0.001
	2	26384 (90.5)	23458 (90.7)	2926 (88.6)	
	3	1132 (3.9)	994 (3.8)	138 (4.2)	
	4	941 (3.2)	795 (3.1)	146 (4.4)	

Table 1b. Preoperative Comorbidities between ASA-PS Classes I/II vs. ASA-PS Classes III/IV

	Total	Classes I/II	Classes III/IV	p-value
n	29163	25861	3302	
Diabetes = Yes (%)	899 (3.1)	390 (1.5)	509 (15.4)	*<0.001
Smoking history = Yes (%)	5277 (18.1)	4451 (17.2)	826 (25.0)	*<0.001
Dyspnea = Yes (%)	489 (1.7)	249 (1.0)	240 (7.3)	*<0.001
COPD = Yes (%)	141 (0.5)	30 (0.1)	111 (3.4)	*<0.001
Ventilator dependence= Yes (%)	13 (0.0)	3 (0.0)	10 (0.3)	*<0.001
Ascites = Yes (%)	3 (0.0)	2 (0.0)	1 (0.0)	0.229
CHF = Yes (%)	14 (0.0)	3 (0.0)	11 (0.3)	*<0.001
HTN = Yes (%)	2605 (8.9)	1430 (5.5)	1175 (35.6)	*<0.001
Acute renal failure = Yes (%)	2 (0.0)	1 (0.0)	1 (0.0)	0.084
Dialysis dependence = Yes (%)	13 (0.0)	2 (0.0)	11 (0.3)	*<0.001
Open wounds and/or wound infection = Yes (%)	59 (0.2)	36 (0.1)	23 (0.7)	*<0.001
Steroid use for chronic conditions= Yes (%)	327 (1.1)	210 (0.8)	117 (3.5)	*<0.001
Significant weight loss[a] = Yes (%)	47 (0.2)	26 (0.1)	21 (0.6)	*<0.001
Hematologic disorder = Yes (%)	119 (0.4)	72 (0.3)	47 (1.4)	*<0.001
Preoperative blood transfusions[b] = Yes (%)	5 (0.0)	2 (0.0)	3 (0.1)	*0.001
Systemic sepsis = Yes (%)	273 (0.9)	206 (0.8)	67 (2.0)	*<0.001
Functional status (%)				*<0.001
Independent	28758 (98.6)	25525 (98.7)	3233 (97.9)	
Dependent	43 (0.1)	16 (0.1)	27 (0.8)	
Unknown	361 (1.2)	319 (1.2)	42 (1.3)	

Table 3b. Major Postoperative Complications between ASA-PS Classes I/II vs. ASA-PS Classes III/IV

	Total	Classes I/II	Classes III/IV	p-value
n	29163	25861	3302	
Deep Incisional SSI = Yes (%)	14 (0.0)	12 (0.0)	2 (0.1)	0.726
Organ/space SSI = Yes (%)	41 (0.1)	32 (0.1)	9 (0.3)	*0.032
Unplanned intubation = Yes (%)	45 (0.2)	27 (0.1)	18 (0.5)	*<0.001
Pulmonary Embolism = Yes (%)	7 (0.0)	5 (0.0)	2 (0.1)	0.15
Ventilator Dependence[b]= Yes (%)	19 (0.1)	5 (0.0)	14 (0.4)	*<0.001
ARF = Yes (%)	1 (0.0)	0 (0.0)	1 (0.0)	*0.005
CVA/stroke = Yes (%)	2 (0.0)	1 (0.0)	1 (0.0)	0.084
Cardiac Arrest requiring CPR = Yes (%)	3 (0.0)	3 (0.0)	0 (0.0)	0.536
MI = Yes (%)	2 (0.0)	1 (0.0)	1 (0.0)	0.084
DVT = Yes (%)	10 (0.0)	7 (0.0)	3 (0.1)	0.062
Systemic sepsis = Yes (%)	47 (0.2)	30 (0.1)	17 (0.5)	*<0.001
Septic shock = Yes (%)	6 (0.0)	3 (0.0)	3 (0.1)	*0.003
Unplanned Readmission = Yes (%)	835 (2.9)	692 (2.7)	143 (4.3)	*<0.001
Unplanned Reoperation = Yes (%)	943 (4.1)	824 (4.1)	119 (4.6)	0.231
Extended LOS[c]= Yes (%)	644 (2.2)	425 (1.6)	219 (6.6)	*<0.001
Death = Yes (%)	6 (0.0)	2 (0.0)	4 (0.1)	*<0.001

Table 3c. Total Postoperative Complications between ASA-PS Classes I/II vs. ASA-PS Classes III/IV

	Total	Classes I/II	Classes III/IV	p-value
n	29163	25861	3302	
Minor Complication = Yes (%)	293 (1.0)	233 (0.9)	60 (1.8)	*<0.001
Major Complication = Yes (%)	2034 (7.0)	1618 (6.3)	416 (12.6)	*<0.001
Overall Complication = Yes (%)	2203 (7.6)	1764 (6.8)	439 (13.3)	*<0.001

Table 4. Role of ASA Classes III/IV as an Independent Risk Factor for Complications

Complication Type	OR (95% CI)	p-value
Overall complications	1.49 (1.28, 1.72)	*<0.001
Major complications	1.52 (1.31, 1.77)	*<0.001
Minor complications	1.27 (0.86, 1.85)	0.219
Unplanned readmission	1.39 (1.09, 1.76)	*0.007
Reoperation	1.33 (1.04, 1.69)	*0.022
Extended LOS (>1 day)	1.78 (1.41, 2.25)	*<0.001

DISCUSSION

- This study found that higher ASA-PS classification is associated with increased risk for post-operative complications. Specifically, this analysis demonstrated that patients classified as ASA Class III and IV have a significantly increased risk of readmission, reoperation, estimated length of stay, and a risk for minor and major complications within 30 days in comparison to patients denoted as ASA Class I and II.
- The main limitation of this study was retrospective design. The NSQIP database does not specify the cause of the readmission; thus, the paper hypothesized the main causes of readmission after a tonsillectomy with or without adenoidectomy based on the consensus in the literature of the primary causes.
- Additionally, any long-term complications were not studied because the database only captures complications and post-operative outcomes 30 days after the surgery. Finally, the NSQIP database only reports outcomes on patients aged 16 years or older, thus limiting the generalizability of these results to the pediatric population less than 16 years of age.
- However, given the results of this study, future studies may aim to assess tonsillectomy and adenoidectomy outcome in children younger than 16 years old in relation to their ASA classification.

CONCLUSIONS

- This study identified a positive and independent correlation between higher ASA-PS classification and the risk of unplanned readmission following T ± A.
- These findings highlight the need for surgeons to risk stratify patients and appropriately counsel patients on the risks of surgery preoperatively.
- Additionally, surgeons should ensure pre-operative optimization of underlying systemic diseases and implement targeted post-operative measures for ASA-PS classes III and IV patients to decrease morbidity, unplanned re-admissions and reoperations, and overall health care cost.

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Email lshakhtour@gwu.edu

Complications of Pediatric Cochlear Implant Surgery

Andrew Stefan BSE[1], Kareem Elhage, BS[1], Sabrina Bernado BS[1], William Azkoul MD[2], Jordan Grauer MD[2], Bianca Siegel MD[3]

[1]Wayne State University School of Medicine; [2]Department of Otolaryngology-Wayne State University School of Medicine; [3]Division of Pediatric Otolaryngology, Children's Hospital of Michigan

INTRODUCTION

Cochlear implantation surgery is the standard of care treatment for pediatric patients diagnosed with severe to profound sensorineural hearing loss. There is strong data that supports cochlear implantation can greatly improve hearing, language comprehension, and speech development in children of all ages. While the operation is considered relatively safe, any invasive procedure involving the cranium puts patients at risk for adverse events. However, there are few recent studies in the United States that have analyzed the postoperative complication rates among children that have received cochlear implants. Furthermore, existing studies fail to investigate specific risk that led to complications, nor offer solutions to reduce their incidence.

Study aims:

1) To determine the incidence of specific postoperative complications in pediatric cochlear implant patients
2) To identify evidence-based risk factors for each complication so pediatric otolaryngologists can better identify high risk patients prior to surgery

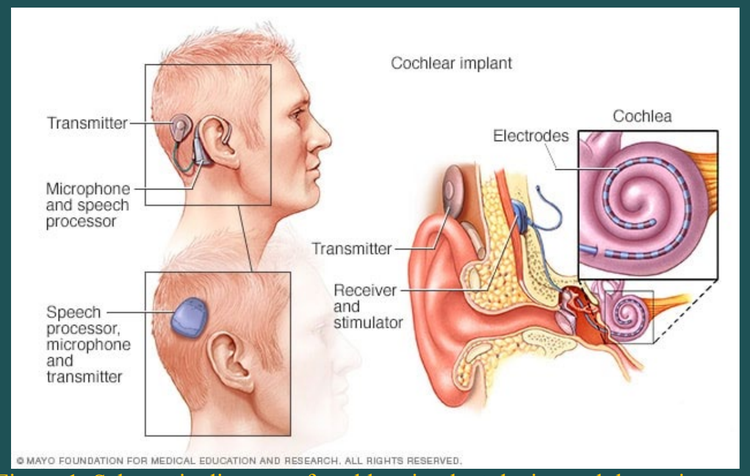


Figure 1: Schematic diagram of cochlear implant device and the various external (left) and internal (right) parts. *Image courtesy of mayoclinic.org*

METHODS

This is a single institution retrospective cohort study analyzing the pre-, intra-, and post-operative data from 72 pediatric patients who underwent cochlear implant surgery from 2016 to 2021.

Preoperative data: patient age and demographic information, comorbidities, history of ear infections, previous ear or skull-based surgeries, and CT scan abnormalities of inner ear.

Intraoperative data: unilateral vs bilateral surgery, simultaneous vs sequential surgery, total operative time, and antibiotics used

Postoperative data: complications, infection types, device removal

Statistical analysis completed using SPSS statistical software.

RESULTS

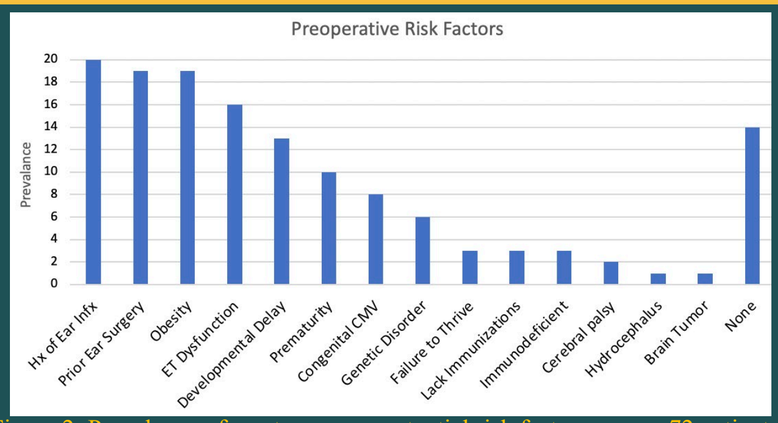
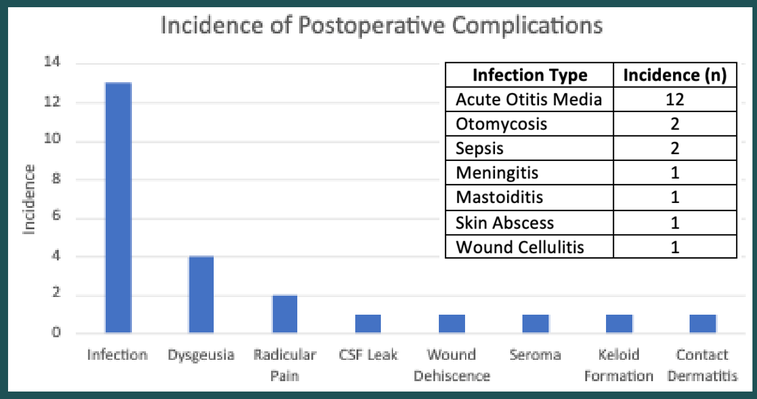


Figure 2: Prevalence of most common potential risk factors among 72 patients.

Among 72 patients, there were 101 ears operated on. There were 27 males and 45 females with age ranging from 8.9 months to 25 y/o. 52 patients (72.2%) had ≥ 1 preoperative comorbidity. 41 patients (56.9%) had a preoperative inner ear abnormality on imaging. Obesity was the most common preoperative comorbidity occurring in 26.4% of patients.



Infection Type	Incidence (n)
Acute Otitis Media	12
Otomycosis	2
Sepsis	2
Meningitis	1
Mastoiditis	1
Skin Abscess	1
Wound Cellulitis	1

Figure 3: Prevalence of all postoperative complications among 101 ears. The table shows the breakdown of the different types of infection.

24 of 101 ears had ≥ 1 postoperative complication yielding a complication rate of 23.8%. Infection and dysgeusia were the two most common complications occurring in 12.9% and 3.96% of ears, respectively. AOM was the most common type of infection. There were 4 ears with >1 type of infection. 2 of 101 ears (1.98%) required cochlear implant device removal.

CONCLUSIONS

Cochlear implantation is a relatively safe procedure that can greatly improve hearing and overall quality of life in patients. There was an overall complication rate of 23.8% with infection and dysgeusia being the most common complications. Pending further statistical analysis, further conclusions can made regarding the the correlation between preoperative risk factors and complication rates.

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Assessing trends in intervention for epistaxis during the COVID-19 pandemic

Megan McNutt, BS[1], Amy Fulmer, BS[1], Austin Schafer, BA[1,2], Natalie Quinn, BS[1], Tran Bourgeois, BS, MPH[3], Michael Boutros, BS[1], Charles Elmaraghy, MD, FACS, FAAP[1,2]

1. Otolaryngology—Head and Neck Surgery, Nationwide Children's Hospital, Columbus, OH
2. The Ohio State University College of Medicine, Columbus, OH
3. Center for Surgical Outcomes Research, Nationwide Children's Hospital, Columbus, OH

abstract

Background:
The current treatment algorithm for recurrent epistaxis in children is well understood. However, it is unclear how the COVID-19 pandemic has influenced trends in management. This study aims to determine if the COVID-19 pandemic affected intervention for children with recurrent epistaxis.
Methods:
After obtaining IRB approval, electronic medical records were retrospectively reviewed for patients aged 0 to 18 seen in the ENT clinic for epistaxis (R04.0) from 2018-2021. Clinical characteristics such as laterality of epistaxis, history of nasal trauma, digital trauma and bleeding disorders were recorded. Intervention included conservative, silver nitrate (SN), electrocautery, or both SN and electrocautery. Patients with epistaxis secondary to surgery or those who underwent previous cautery were excluded. Pre-pandemic was defined as 2018-2019 while the pandemic was defined as 2020-2021.
Results:
Among a cohort of 2424 patients, 50% (n = 1222) were evaluated for epistaxis pre-pandemic and 50% (n = 1202) were evaluated during the pandemic. After adjusting for all confounders, conservative management increased during the pandemic (+6.92% (95% CI: 2.94%, 10.91%, *P*= 0.0007) while electrocautery decreased (-5.15% (95% CI: -8.21%, -2.09%, *P*= 0.001). Treatments with SN and both remained similar (-0.57%, 95% CI: -4.15%, 3.01% *P*= 0.7552, -1.20%, -2.44%, -0.04% *P*= 0.0572, respectively).
Conclusions:
The pandemic altered intervention patterns for epistaxis. Future studies should evaluate recurrence rates among each treatment group to determine the clinical implications of these changes.

background

- Epistaxis is a common childhood disorder, with 75% of children being diagnosed with at least one episode and up to 9% of children experience recurrent epistaxis[1,2]
- Treatment options include conservative methods like moisture therapy, as well as silver nitrate cautery and bipolar electrocautery[1,2]
- Type of intervention is determined by patient preference and disease severity[3]
- COVID-19 made following treatment algorithms challenging
- Objective is to determine the pandemic's effect on the clinical management of epistaxis

methods

Study population
- All patients aged 0 to 18 who presented in the clinic for epistaxis between 2018-2021

Exclusion criteria
- Patients who underwent cautery prior to 2018 or who had epistaxis secondary to a surgical intervention

Outcome variables
- Clinical characteristics, such as epistaxis laterality, history of nasal and digital trauma, bleeding disorders, and treatment with either conservative management, silver nitrate cautery, or electrocautery
- Intervention rates were compared between pre-pandemic years (2018-2019) and post pandemic years (2020-2021)

Statistical analysis
- Bivariate analyses conducted with Chi square / Fisher's exact tests
- Univariable and multivariable general linear models conducted to determine differences in management between pre-pandemic years and post pandemic years

results

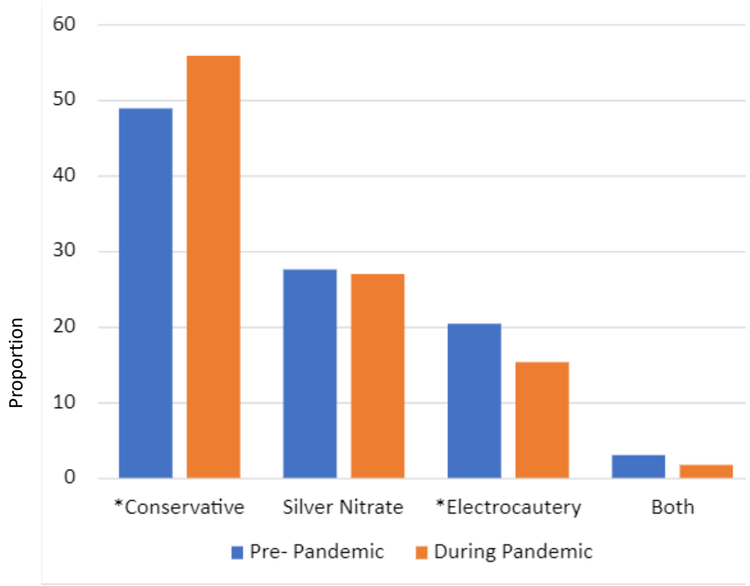

Figure 1: Multivariable general linear regression modeling change in treatment type for epistaxis pre-COVID-19 pandemic compared to during the COVID-19 pandemic. Models adjusted for digital nasal trauma, previous nasal trauma, and previous history of easy bleeding.

discussion

- There were no differences in demographic characteristics between patients evaluated pre pandemic and those evaluated during the pandemic
- Compared to pre-pandemic years, encounters for patients with a history of digital nasal trauma, previous nasal trauma, and previous history of easy bleeding all significantly increased during the pandemic
- In line with surgical procedures being shut down during the pandemic, conservative treatment for epistaxis increased significantly during the pandemic while electrocautery procedures significantly decreased during the pandemic
- Treatment rates with SN remained similar
- The pandemic did have a significant impact on type of intervention for recurrent epistaxis
- Implications of these changes should be investigated in future studies by evaluating recurrence rates among different treatment groups

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110 - Sclerosing extramedullary hematopoietic tumor in the masticator space of a child

University of Colorado Anschutz Medical Campus

Scott Hirsch, MD[1]; Christian Francom, MD[2]

[1]University of Colorado, Department of Otolaryngology – Head and Neck Surgery
[2]Children's Hospital Colorado, Department of Pediatric Otolaryngology

Abstract

Background: Sclerosing extramedullary hematopoietic tumor (SEMHT) is a rare tumor-like condition usually presenting in adult patients with history of chronic myeloproliferative disorders (CMPD). It is known to involve a variety of different organs including lung, skin, gastrointestinal tract, breast, kidney, lymph node, and thyroid gland. We present a unique case of SEMHT not only found in an unreported location, but also in a pediatric patient.

Case Description: 16-year-old male with history of myelofibrosis found to have a facial mass in the right masticator/retro-maxillozygomatic space. It measured 2.0 x 1.5 x 2.5cm and was T1 isointense and slightly T2 hyperintense on contrasted MRI. He had been asymptomatic and unaware of the mass. Physical exam was notable for a non-tender, right-sided mass palpable just anterior to the retromolar trigone. Removed was via a trans-oral approach and final pathology returned with SEMHT. Molecular genetic testing was significant for a type I variant p.Leu367ThrfsTer46 in *CALR* and a variant p.Gln1381ThrfsTer77 in *SAMD9L*.

Discussion: SEMHT is a rare tumor, described almost exclusively in adults, and occurs in patients with a CMPD (most commonly myelofibrosis). Characteristic features include prominent atypical megakaryocytes and dense collagen fibrosis, helping to distinguish it from extramedullary hematopoiesis. Morphologically, SEMHT has a sclerotic to myxoid background with thick collagen strands and trapped fat. Although mutations in *JAK2* and *CALR* type 2 are more common, there is increased survival with the *CALR* type I mutation. Both the *CALR* and *SAMD9L* mutations predispose to myeloid neoplasms. The prognosis of SEMHT is unknown.

Introduction

- SEMHT is an extremely rare lesion that is most often diagnosed in elderly patients
- Often described in patients with advanced-staged myeloproliferative disorders
- Characterized by mature hematopoietic elements that form fibrotic extramedullary tumors
- Features including prominent atypical megakaryocytes, dense collagen fibrosis, and positive staining for factor VIII, CD15, CD30, and S100.

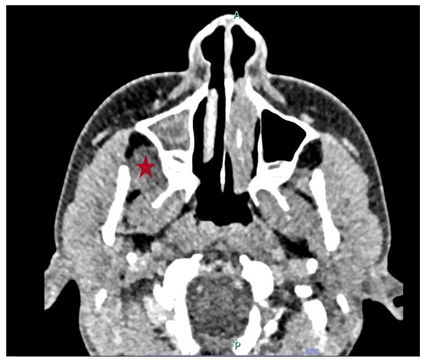

Figure 1. Axial contrasted CT

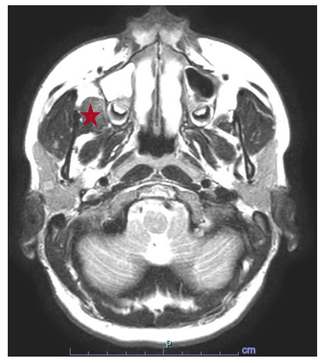

Figure 2. Axial T2 MRI

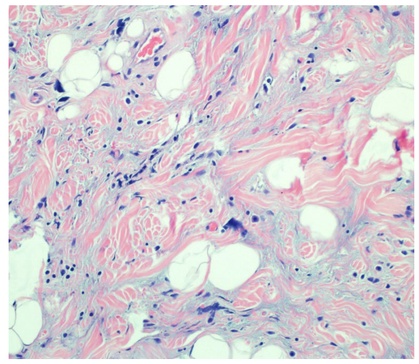

Figure 3. H&E Stain 20x with fibrosis

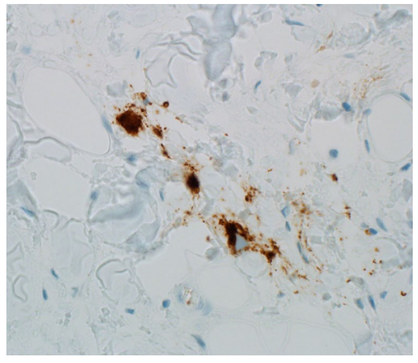

Figure 4. CD61 Immunostain

Case Presentation

- 16yo male with a medical history significant for myelofibrosis (with the following WHO criteria: megakaryocytic hypoplasia with atopia and reticulin fibrosis grade II, absence of diagnostic criteria for other myeloproliferative disorders, and a CALR mutation), who on pre-bone marrow transplantation workup was found to have a right retromaxillozygomatic space mass on CT sinus with contrast (**Figure 1**) and verified on an MRI face w/wo contrast **(Figure 3**)**.**
- He had had no symptoms related to the mass and but did complain of intermittent facial pressure, nasal drainage, and nasal congestion. On exam, the mass was palpable between the maxilla and mandible near the retromolar trigone without any overlying skin changes
- He underwent endoscopic right maxillary antrostomy with removal of polypoid tissue and mucopurulance and transoral approach to the oral mass.
- Specimen was 3 x 2.8 x 1.6cm; poorly circumscribed, relatively paucicellular spindle cell lesion consisting primarily of dense, focally keloidal, collagen bundles admixed with adipose and trabeculated myxoid stroma **(Figure 3**)**.** Megakaryocytes present by positive immunostaining for CD31 and CD61 **(Figure 4**)**,** Associated with islands of extramedullary hematopoiesis.
- STAT-6 negative, diffusely CD34 positivity, pan-TRK negative. Mutations found in the *CALR* and *SAMD9L* genes

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Discussion

- Sclerosing extramedullary hematopoietic tumor is a rare lesion that is most often diagnosed in association with chronic myeloproliferative disorders
- Extramedullary hematopoiesis usually takes place in the spleen, liver, or lymph nodes, but can manifest itself in the formation of discrete masses. However, to date, none have been described in the facial soft tissues
- Differential diagnosis includes sclerosing liposarcoma, malignant fibrous histiocytoma/pleomorphic sarcoma, sarcomatoid/anaplastic carcinoma, and Hodgkin lymphoma
- It has only been described in the adult population, but here, we present a case in a pediatric patient
- Calreticulin (CALR) mutations are seen in myeloproliferative disorders and has been described with SEMHT
- SAMD9L mutations are also found in myelodysplastic syndromes

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Research in Outcomes for Children's Surgery

Children's Hospital Colorado

Extranodal NK/T-cell Lymphoma Nasal Type in an Adolescent Male

Melissa Raines, MSN, CPNP-PC; Steven Leoniak, MD

Department of Otolaryngology, University of Colorado Anschutz Medical Campus, Aurora, CO | Pediatric Otolaryngology, Children's Hospital Colorado, Colorado Springs, CO

Background

- Lymphoma is the 3rd most common sinonasal malignancy.
- Extranodal NK/T-cell lymphoma nasal type (ENKTL-NT) is a rare, aggressive variant related to Epstein-Barr Various (EBV).
- Known for geographic predominance in Asian and Latin American countries.
- Majority of cases reported in adult males, with rare presentation in children or adolescents.
- EBV may play a key role as it is a common factor across all affected groups.
- 5 year survival ranges from 40-80%

Methods

- Case report

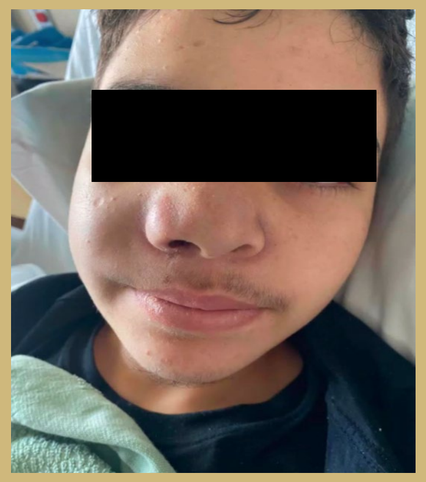
Objective

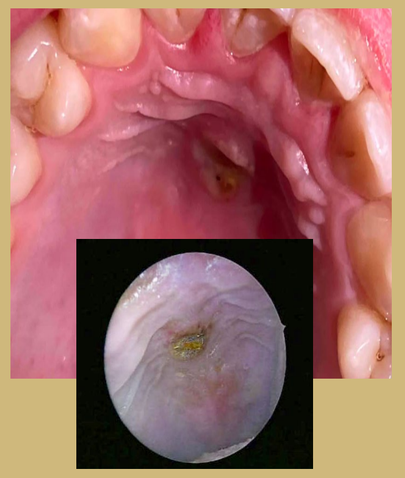
- Understand the importance of differential diagnosis in children with prolonged, complicated cases of chronic nasal congestion and sinusitis unresponsive to common medical therapies.

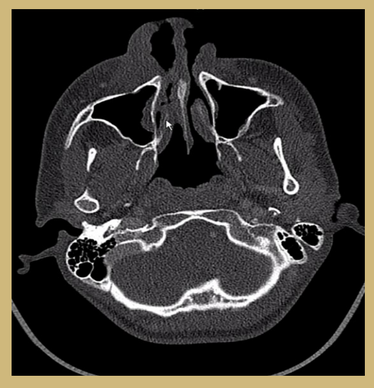
Case Report

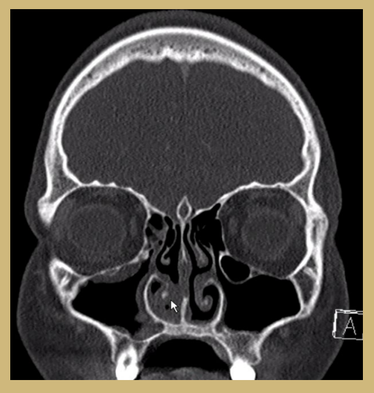
- 17 year old Hispanic male
- Presented to emergency room with severe right-sided nasal congestion.
- Unresponsive to 1 month of medical treatment.
- Flexible nasal endoscopy revealed an extensive ulcerative and exophytic mass of the right nasal cavity extending through the hard palate.
- CT sinus showed an abnormal density & asymmetric soft tissue thickening of the right nasal ala extending to the nasal apex and the soft tissues of the face overlying the right maxillary sinus.
- Endoscopic biopsy demonstrated fragments of mucosa & deep tissue with extensive involvement by an atypical lymphoid cell proliferation positive for EBV-encoding RNA.

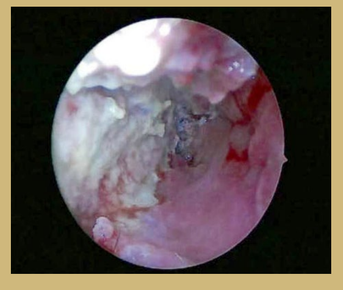
Clinical Presentation

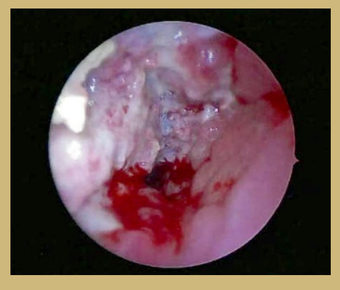












Case Report cont.

- Pre-treatment PET with right nasal tumor SUV 15, right neck levels 1b, 2, 3 lymphadenopathy SUV 16.4, 3.3, 2.5 respectively.
- MRI with ill-defined T2 hyperintense right facial and nasal mass with restricted diffusion centrally and several abnormally enlarged right neck lymph nodes.
- EBV PCR 8457 copies/mL
- 2 cycles mSMILE chemotherapy
- Post-treatment PET with persistent FDG activity right nasal tumor SUV 11.3, resolution of FDG activity in neck
- 3 cycles DEVIC chemotherapy
- Concurrent IMRT 56Gy (50 tumor bed/neck + tumor bed boost)
- PET with mild FDG activity of right inferior turbinate without definable lesion, SUV 3.4
- MRI decreased thickness and extent of right facial and nasal mass but with residual T2 hyperintensity around the premaxilla and hard palate and resolution of central diffusion restriction
- EBV PCR undetectable
- Exam with resolution of palatal ulceration and flexible nasal endoscopy with moderate to severe mucositis without evidence of tumor
- MRI q3 months x1 year, EBV PCR q1 month x6 months
- Head and neck exam & flexible nasal endoscopy q6 weeks x1 year
- Post-treatment audiogram

Conclusion

- High clinical suspicion is key for diagnosis of ENKTL-NT in children.
- Otolaryngologists should be particularly wary in cases of prolonged, complicated nasal congestion and sinusitis unresponsive to typical medical management.
- Prompt diagnosis and subsequent initiation of treatment is key to ensuring the best chance of survival.

Comparing Loss to Follow-up Among Rural and Non-rural Children with Tracheostomy

Adam Van Horn, MD[1], Raquel Good, MS[2], Javan Nation, MD[2], Erin Kirkham, MD MPH[3], Patricia L Purcell, MD MPH[3]
[1]Otolaryngology, Marshall University, Huntington, WV; [2]Pediatric Otolaryngology, Rady Children's Hospital, San Diego, CA; [3]Pediatric Otolaryngology, University of Michigan, Ann Arbor, MI

OTOLARYNGOLOGY-HEAD AND NECK SURGERY
UNIVERSITY OF MICHIGAN HEALTH SYSTEM

Background

- Most pediatric tracheostomy patients require frequent follow-up with high rates of healthcare utilization given medical complexity and reliance on medical technology and equipment.[1-6]
- Social determinants of health may affect pediatric tracheostomy outcomes and long-term care.[7-10]
- Rural patients experience barriers to otolaryngologic care, however this has not been examined in pediatric tracheostomy populations.[11,12] Rural pediatric tracheostomy patients may experience different long-term outcomes compared to nonrural patients.

Objectives

- We hypothesize rural pediatric tracheostomy patients will have lower rates of follow-up after receiving a tracheostomy compared to nonrural patients.
- This study aims to examine follow-up rates among pediatric tracheostomy patients in rural and nonrural cohorts using nationally representative data.

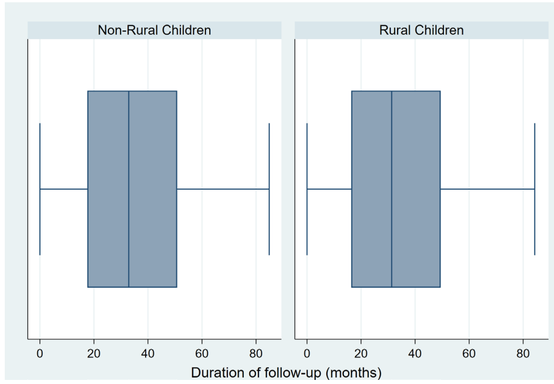
Methods

- Retrospective cohort study of patients who underwent tracheostomy from 2013-2017 in the pediatric health information system (PHIS) database
- Rural and nonrural cohorts defined by rural-urban commuting area codes[13]
- Demographic and medical characteristics for patients collected
- Follow-up information collected
  - Length of follow-up and lost to follow-up
- Univariate analysis performed using chi-squared and Mann-Whitney U test
- Multivariate logistic regression analysis performed to control for covariates

	Nonrural (n=4,614)	Rural (n=890)	p
Demographics			
Age (years)	3.2 ± 5	3.6 ± 5.3	0.04
Female sex	1,996 (43)	377 (42)	NS
Non-white race	2,295 (50)	233 (26)	<0.001
Comorbidities			
Neuromuscular disorder	1,957 (42)	355 (40)	NS
Ventilator dependence	2,228 (48)	380 (43)	<0.001
Follow-Up			
Length of follow-up (months)	35 ± 22	34 ± 22	NS
Lost to follow-up*	634 (12)	147 (14)	0.03

Table 1. Characteristics of nonrural and rural pediatric tracheostomy patients. Data are presented as frequency (percentage) or mean (± standard deviation). NS=not significant. *Patients not included in preceding rows or in column heading count.

Figure 1. Box plot demonstrates median length of follow-up. Shaded box represents single standard deviation and bracketed line represents range. There was not a significant difference in follow-up duration between rural and non-rural children.



	Follow-up (n=890)	Lost to follow-up (n=147)	p
Demographics			
Age (years)	3.5 ± 5.3	4.8 ± 6	<0.001
Female sex	377 (42)	64 (44)	NS
Non-white race	188 (22)	42 (32)	0.02
Comorbidities			
Neuromuscular disorder	355 (40)	71(48)	0.06
Ventilator dependence	380 (46)	77 (55)	0.06

Table 2. Demographic and medical characteristics of rural patients based on whether they had follow-up at PHIS hospital after tracheostomy. Data are presented as frequency (percentage) or mean (± standard deviation). NS=not significant.

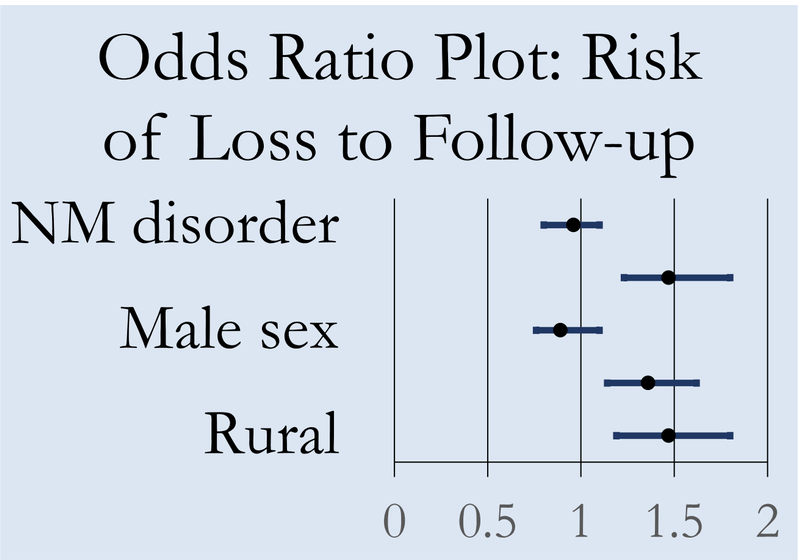


Figure 2. Odds ratios demonstrating the association between demographic and medical characteristics and loss to follow-up from multivariate logistic regression model. Error bars represent 95% confidence interval. Rural status, non-white race and ventilator dependence were associated with a significantly increased risk of loss to follow-up. NM = neuromuscular.

Conclusions & Discussion

- Rural pediatric tracheostomy patients have lower rates of follow-up at PHIS reporting hospitals than nonrural counterparts following tracheostomy hospitalization.
- Rural patients who maintained follow-up had similar length of follow-up as compared to nonrural patients.
- Patients who are less likely to follow-up at PHIS reporting hospital may be more likely to receive care outside of tertiary care centers, which can have implications on outcomes.[14]

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Does Parental Postop Decisional Regret Correlate with Persisting Symptoms in Children Who Undergo Tonsillectomy?

Beatrice R. Bacon BS, Nicole M. Favre BA, Katherine A. Foote BA MA, Michele M. Carr DDS MD PhD

Jacobs School of Medicine and Biomedical Sciences at the University at Buffalo, Department of Otolaryngology- Head and Neck Surgery

Introduction

As one of the four pillars of medical ethics, patient autonomy, and by extension, informed consent, is key when making decisions regarding medical procedures. After explaining any risks, benefits, and alternative options[1], the patient or patient's guardian must come to a shared decision with the health care provider before moving forward with a given procedure. In pediatric populations, decisional regret (DR) in parents who elect to have their children undergo tonsillectomy is related to preoperative decisional conflict (DC), how comfortable they are with their decision to do surgery[2-4]. Intuitively, it would seem that decisional regret should be inversely related to how successful the surgery is to resolve preoperative complaints. However, this has not yet been shown in this population. Despite proper preoperative counseling[3,5],there are numerous knowledge gaps, unrealistic expectations, unclear values, social pressures[6], and persisting symptoms that can cause parental decisional regret.

Objective

- Our goal is to examine the relationship between parental postop DR and persisting symptoms in children who undergo tonsillectomy for sleep disordered breathing (SDB).

Methods

- Parents with children aged 2-14 years who were booked for tonsillectomy +/- adenoidectomy for SDB were recruited.
- OSA-18 (a quality-of-life scale for children with SDB) scores were collected preop and one month postop.
- Preop DC and postop DR scores were collected.
- Analysis was undertaken with correlations and linear regression.

Results

- N = 31
- Demographics included: 18 (58%) females and 13 (42%) males with mean age of 5.4 years
- Mean preop OSA-18 score was 72.6 **[FIGURE 1].**

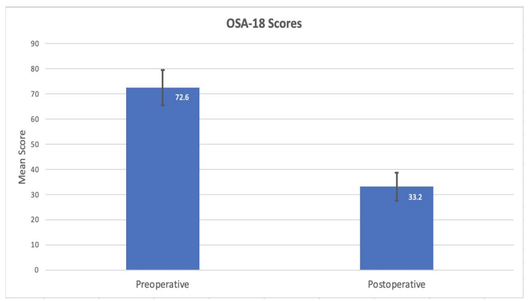

Figure 1. Comparing mean preoperative OSA-18 score to mean postoperative OSA-18 score, with associated 95% CI

- Mean preop DC score was 4.3 (95% CI 1.4-7.3), consistent with low DC.
- Mean postop OSA-18 score was 33.2 which was significantly improved **[FIGURE 1]**.
- Mean improvement: 42.6 (95% CI 34.7-50.4 (p<.001)
- Mean postop DR score was 6.0 (95% CI 2.8-9.3), indicating low DR **[FIGURE 2]** .
- 19 parents had DR scores of 0, indicating no postop regret.

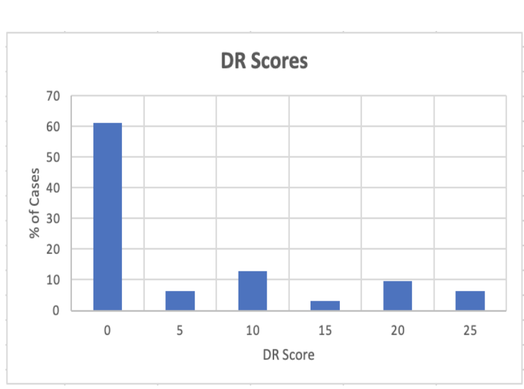

Figure 2. Percentage of cases per DR score

- There was no significant contribution to DR scores from preop or postop OSA-18 scores, change in scores or preop DC when analyzed with linear regression and correlation.

Discussion

- Preoperative DC has frequently been associated with postoperative DR[3], and may also be related to canceled surgeries and follow up visits.[7]
- DR has specifically been found to be associated with parental education level and ongoing symptoms.[5]
- There have been mixed results surrounding the association between postoperative DR and postoperative complications.
- This suggests that other external factors may play a role in decisional regret.

Conclusion

In this group, we were not able to demonstrate a correlation between parental postop DR and persisting SDB symptoms in children. This may reflect the low overall postop regret in these parents.

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Cell-Signaling Pathways Involved in BMP2-Treatment Response Heterogeneity in Pediatric Osteoblast-Like Cells

Priya Arya, BS[1], Jane Jang, BS[2], Archana Kamalakar, PhD, MS[2], Shelly Abramowicz, DMD, MPH, FACS[2,3], Steven Goudy, MD, MBA[2,3]

[1]Mercer University School of Medicine, Savannah, GA 31404
[2]Emory University School of Medicine, Atlanta, GA 30322
[3]Children's Healthcare of Atlanta, Atlanta, GA 30322

Background

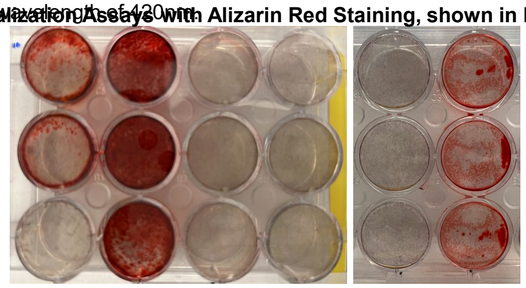
Craniofacial bone loss as seen in pediatric patients occurs due to congenital and traumatic bone loss. These defects include conditions such as maxillary hypoplasia and craniosynostosis, and are complex to manage, requiring multimodal treatment regimens. These range from short-term bone stabilization and soft tissue repair to newer regenerative therapies; one such therapy that is used in adult patients is the local administration of bone morphogenetic protein (BMP2), which is physiologically involved in the development of bone and cartilage. Treatment with BMP2, however, is not FDA-approved for use in pediatric patients due to inflammatory and oncogenic sequelae (James *et al.* 2016). In addition to the inflammation, BMP2 has a heterogenous response in patients, suggesting that bone pathways have a differential response during BMP2-induce bone formation that could be exploited to improve bone regener[illegible]

Patient with maxillary hypoplasia, before and after surgical reconstruction (Wang *et al.* 2019).

Methods

In this study, heterogenous patient responses following treatment with BMP2 were studied by utilizing human bone-derived osteoblast-like (HBO) cell lines isolated from pediatric bones (n=6). Following identification of responder versus non-responder cell lines via Western blots, as well as Luminex-based assays to identify what pathways were undergoing phosphorylation, mineralization assays were used to validate the findings. These were conducted by treating cells for 21 days with the following treatments: growth media (control), osteogenic media, TGF-β inhibitor (TI), AKT inhibitor (AI), TI+AI, BMP2, BMP2+TI, BMP2+AI, and BMP2+TI+AI. Cells were given half feeds at a time point of every 5 days. On day 21, cells were harvested and fixed. They were then stained with Alizarin Red to evaluate for calcium-rich deposits by the cells (Gregory *et al*. 2004), and absorbance of each well was read[illegible] at a wavelength of 420nm.

Mineralization Assays with Alizarin Red Staining, shown in HBO 16



Osteogenic Media | BMP2 | AKT Inhibitor (AI) | BMP2 + AI | TGF-beta Inhibitor (TI) + AI | BMP2 + TI

Results

Mineralization assays validated the responder and non-responder designations previously identified in the lab. HBO 21 had not been characterized previously and produced responder-like results, with uniquely high mineralization results shown.

Responders

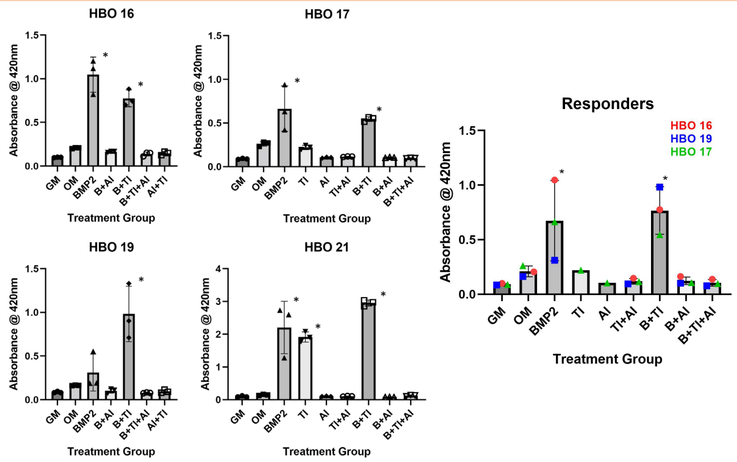


Figure 1. HBO 16, 17, 19, and 21 Responses to Treatment. Increases in mineralization levels were observed in these cell lines. In addition to BMP2, the PI3K/AKT pathway was shown to be responsible for mineralization amongst the treatments tested, as demonstrated by significant increases in mineralization when the TGF-beta pathway was inhibited. These responses were noted in each cell line, designating them as "responders" to treatment. Of note, HBO 21 only demonstrated significant mineralization potential in the presence of TGF-beta inhibitor, without BMP2.

Key take-away: BMP2-mediated phosphorylation of the PI3K/AKT pathway is essential for mineralization observed in cell lines designated as responders.

Non-Responders

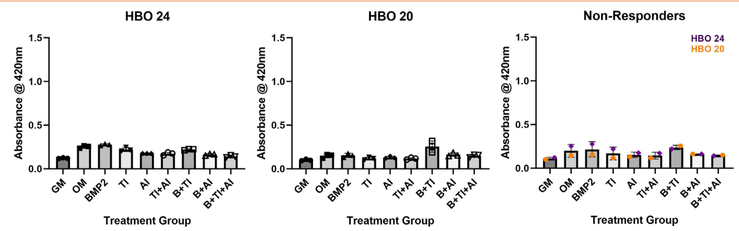


Figure 2. HBO 20 and 24 Responses to Treatment. These cell lines did not produce as significant of an increase in mineralization potential following treatment schemes. These cell lines, therefore, were designated as "non-responders" to treatment.

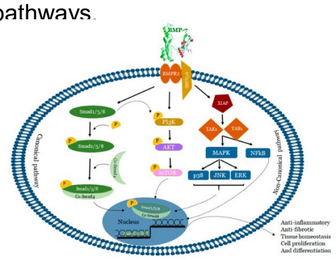
Key take-away: Slight mineralization capability was seen in non-responder cell lines in the presence of BMP2, and even more so with BMP2+TI, suggesting TGF-beta may play an inhibitory role in mineralization.

Conclusion

- The initial findings from this experiment indicate that non-canonical signaling pathways (particularly the PI3K/AKT pathway) are potential targets of interest for bone regenerative therapies.
- These therapies could be applied towards pediatric patients with craniofacial defects as an alternative to direct BMP2 administration, which produces undesirable side effects that are not seen in adult patients.

Future Avenues of Study

- Elucidating additional cell-signaling pathways that may be involved in the heterogeneity of HBO cell responses, not only between responders and non-responders, but also those that may be responsible for variation seen within responder cell lines themselves (i.e., HBO 21).
  - Utilizing RNA sequencing to see other genes/pathways being activated in cellular responses.
  - Luminex-based assays to see what other proteins may be involved, as well as inflammatory responses.
  - Understanding to what degree cross-talk may occur between these pathways.



BMP canonical and non-canonical cellular signaling pathways (Narasimhulu *et al.* 2020).

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Single-Staged Laryngotracheal Reconstruction for a Type IV Laryngeal Web in a Neonate: A Case Report and Literature Review

Éolie Delisle [1], Mathieu Bergeron BPharm, MD, FRCSC [1,2], Carol Nhan MSc, MD, FRCSC [1,2,3,4,5]

[1]Faculté de médecine, Université de Montréal; [2] Division d'Oto-rhino-laryngologie, Centre Hospitalier Universitaire Mère-Enfant, CHU Sainte-Justine;
[3] McGill University Faculty of Medicine; [4]Division of Otolaryngology, Montreal Children's Hospital; [5]Division of Otolaryngology, Jewish General Hospital

CHU Sainte-Justine
Le centre hospitalier universitaire mère-enfant

Faculté de médecine
Université de Montréal

Abstract

Background: Congenital laryngeal webs are rare anomalies. Type III and IV laryngeal webs are not commonly repaired in a single-staged procedure at birth.

Methods: Case report and literature review.

Results: A two day-old 36.5-week preterm infant with 22q11.2 deletion syndrome presented with respiratory distress accompanied by biphasic stridor, aphonia and CO2 of 69 mmHg despite CPAP. Naso-pharyngo-laryngoscopy confirmed a type IV laryngotracheal web. He was stabilized by adding heliox to his CPAP and operated the next day at 2700g. The patient was intubated with a 2.0 endotracheal tube over a telescope. The subglottis was over 70% stenosed. A single-staged laryngotracheal reconstruction (LTR) with thyroid alar graft anteriorly and posterior cricoid split was performed. The patient was extubated one week post-operatively. He was on room air within two days and full PO by nine days post-extubation. Apart from prolonged sedation withdrawal syndrome, there were no major post-surgical complications and he was discharged 59 days after laryngotracheoplasty. Review of the literature found eleven papers with sufficiently detailed reports of type III and/or IV congenital laryngeal webs. Only 6/50 reported cases were managed with a single-staged laryngotracheal reconstruction at ages three months to 4 years at the time of surgery.

Conclusion: This is the youngest case in the literature of a type IV laryngeal web successfully treated with a single-staged laryngotracheal reconstruction. In select patients a single-staged repair may avoid burdens associated with tracheostomy.

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Introduction

Laryngeal webs may be acquired or congenital. Acquired webs are most often the result of trauma to the vocal folds, for example, traumatic or prolonged intubation, previous endo-laryngeal surgery, caustic ingestion, as well as laryngeal inflammatory processes such as vasculitis or infections[1]. Congenital webs originate from an incomplete recanalization of the primitive laryngotracheal airway between weeks 8 and 10 of embryonic life.These are rare abnormalities of the larynx as they are seen in 1/10,000 to 1/50,000 births and represent only 5% of all laryngeal anomalies[5,6].

The Cohen classification describes four types of laryngeal webs[1]. Type I has a web involvement of less than 35% with little to no subglottic involvement. Type II glottic webs involve 35-50% of the glottis and can be thin or thick with possible low grade subglottic stenosis. Type III glottic webs involve 50-75% of the laryngeal lumen and are thick webs with cartilaginous subglottic involvement. Finally, type IV glottic webs consist of 75-90% involvement of the glottic airway and are thick with significant cartilaginous subglottic stenosis. Degree of respiratory distress and dysphonia or aphonia varies by severity of the web[2,7]

Congenital laryngeal webs may be approached endoscopically or with an open surgical procedure depending on severity[6]. In severe type III or IV webs, patients present with respiratory distress and the airway is generally secured by tracheostomy. A laryngotracheal reconstruction is preferred to augment the airway in children with more severe webs that significantly involve the subglottis[8]. In the current literature, it is suggested to delay definitive surgery until 6 to 12 months of life[16]. This paper presents a literature review of the treatment of congenital type III and IV laryngeal webs, and reports a case of congenital type IV web successfully treated with a single-staged laryngotracheal reconstruction on day three of life.

Methods

Pubmed was searched for publications that described the treatment of congenital laryngeal web cases. A total of 28 articles were found in a Pubmed search on June 27, 2022 (see Figure 1). The titles and abstracts were screened by all authors for the following inclusion criteria: (1) a primary research study including case series or case reports; (2) study included data on the pediatric population (0-18 years old); (3) study reported on the treatment of case(s) of congenital laryngeal webs meeting the criteria for type III and/or IV webs; (4) study documented surgical intervention(s) and outcomes; (5) English or French language study; and (6) not a duplicate study or a study on the same data set. Eleven articles that did not meet inclusion criteria were removed. The same reviewers then screened he full texts of all chosen citations; studies that did not meet the selection criteria were excluded. All discrepancies were resolved by consensus. Another five articles were removed due to incomplete information. Fifty patients from these eleven articles met inclusion criteria and information on web type, age at operation, type of surgery, and time to decannulation was recorded (see Table 1).

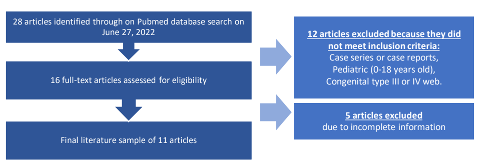


Figure 1. Flow chart of studies found, excluded and included in our literature review.

Articles	Patient number in the articles	Type of laryngeal web (Cohen)	Grade of subglottic stenosis (Cotton-Meyer)	Age at operation	Trached prior to definitive management	DS/SS	Time to decannulation after definitive surgery
Amir et al.	1	3(60%)	SGS, grade non-specified	24 months	No	Endoscopic laser failure	Time unspecified for all patients
	2	4(90%)	SGS, grade non-specified	3 months	Yes trached at 24h of life	Endoscopic laser with tracheostomy	
	4	3(74%)	SGS, grade non-specified	5 months	No	DS	
	5	4(90%)	SGS, grade non-specified	6 months	No	DS	
	6	3(70%)	SGS, grade non-specified	12 months	No	DS	
Alkan and al.	1	4	SGS, grade non-specified	N/A	No	DS	Time unspecified
	2	4	SGS, grade non-specified	N/A		DS	No, permanent tracheostomy
	3	4	SGS, grade non-specified	N/A		DS	Time unspecified
	4	3	SGS, grade non-specified	N/A		Endoscopic with tracheostomy	Time unspecified
	5	3	SGS, grade non-specified	N/A		Endoscopic with tracheostomy	Time unspecified
Avelino and al.	4	3	SGS, grade non-specified	36 months	Yes	DS	90 days or more
	5	3	SGS, grade non-specified	18 months	Yes	DS	90 days or more
	6	4	SGS, grade non-specified	19 months	Yes	DS	60 days or more
	7	4	SGS, grade non-specified	18 months	Yes	DS	85 days or more
Baburajan and al.	1	3	SGS, grade non-specified	3 years	No	DS	6 months
	2	4	SGS, grade non-specified	2 years	No	DS	3 months
Bowe and al.	1	3 (50%)	SGS, grade non-specified	4 years	No	DS	1 month
Cheng and al.	1	3 (66%)	SGS, grade non-specified	3 months	No	SS	N/A
	2	4 (75%)	SGS, grade non-specified	4 years	Yes, at 27 days	SS	N/A
	3	3	SGS, grade non-specified	5 months	No	SS	N/A
	4	4	SGS, grade non-specified	3 months	No	SS	N/A
DeTrey and al.	1	4	3	1,2 years	All patients were trached but time unspecified for each case (before or during the surgery)	DS	Time unspecified for all patients
	2	4	3	2,8 years		DS	
	3	4	3	1,9 years		DS	
	4	3	2	3,2 years		DS	
	5	4	3	0,4 years		DS	
	6	4	3	0,5 years		DS	
	7	4	3	1,3 years		DS	
	8	3	3	4,4 years		DS (after failed endoscopic surgery)	
	9	4	3	2,6 years		DS	
	10	4	3	0,2 years		DS	
	11	4	2	4,6 years		DS	
	12	4	3	1,3 years		DS	
	13	4	3	3,1 years		DS	
	14	4	2	6,2 years		DS	
LeRiger and al.	1	4	SGS, grade non-specified	2 days	Yes	DS	Time unspecified for all patients
Milcuk and al.	4	3	SGS, grade non-specified	17 months	Yes, at 2 months and 2 days	DS	22 months
Sorichetti and al.	1	4 (95%)	2	5 days	No	DS	76 days
Wyatt and al.	8	3 (60%)	2	11 months	No	SS	N/A
	9	3 (60%)	2	16 months	Yes, at 14 months	DS	5 months
	10	3 or 4	2	13 years	Yes, at 5 months	DS	For the present surgery: 2 years and 5 months Total in life-time: 5 years
	12	4 (75%)	3	22 months	Yes, at 4 months	DS	24 months
	13	4 (75%)	3	4 years 8 months	Yes, at 10 days	DS	4 years 11 months 21 days
	14	4 (80%)	3	15 months	Yes, at 12 days	DS	19 months 19 days
	15	4 (80%)	3	17 months	Yes, at 16 months	DS	9 months
	16	4 (90%)	3	3 years and 6 months	Yes, at 11 months	DS	4 years 1 months
	17	3 or 4	3	13 months	No	SS	N/A
	18	3 or 4	3	23 months	Yes, at 7 months	DS	20 months
	19	3 or 4	3	24 months	Yes, at 1 month	DS	26 months
	20	3 or 4	3	27 months	Yes, at 1 month	DS	3 months3

Table 1. Summary of cases

Case Report

A 36.5-week preterm infant with 22q11.2 deletion syndrome presented with biphasic stridor, respiratory distress and aphonia at birth and was stabilized on CPAP. On day two of life, OTL was consulted since his PC02 remained at 69 mmHg despite CPAP. A nasopharyngolaryngoscopy confirmed a type IV web and he was temporized by the addition of heliox to his CPAP until OR availability the next day. At day three of life and 2700g, he underwent an MLB which showed a type IV laryngeal web with a subglottic stenosis of at least 70%(see Figure 2). His airway was secured with a 2.0 endotracheal tube and a single-staged laryngotracheoplasty with a thyroid alar graft anteriorly and a posterior cricoid split was performed. He was extubated on post-operative day seven. In less than 48h, he was on room air, and nine days later he was feeding fully by bottle. Planned MLB and balloon dilatation on post-operative days 21 and 58 were performed. He remained hospitalized during this time for prolonged sedation withdrawal syndrome. A small neck incision dehiscence at the site for the previous penrose drain was revised concomitant with the first dilatation. He has been followed in outpatient clinic and at nine months old continues to have no respiratory issues and a good voice.

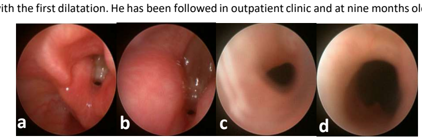


Figure 2. MLB of the larynx; a) Supraglottic view, b) Vocal fold view showing a type IV laryngeal web, c)Subglottis stenosis of more than 70%, d) Deep subglottic view

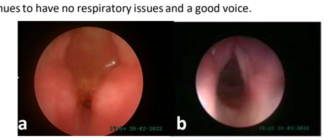


Figure 3. Pre-operative and post-operative view of the larynx; a)Pre-operative b) Post-operative

Results

Of the 50 patients, 19 were clearly reported as having a tracheostomy for some time prior to definitive surgery. The median and mean age at definitive surgery was 18 and 26 months, respectively .

Forty patients had a double-staged LTR, six patients had single-staged LTR, and four had an endoscopic surgery (three while with tracheostomy, one patient without tracheostomy but surgery failed).

For the double-staged surgery (n = 40), the median and mean age of the children at the time of double-staged surgery was 19 and 27 months, respectively. The age range at time of surgery was from two days of life to six years-three months.

The median and mean age at the time of singled-staged surgery (n = 6) was 8 and 14 months, respectively. The age range at the time of single staged surgery was from three months to 4 years.

Abbreviations

CPAP: Continuous positive air pressure
DS: Double staged LTR
LTR: Laryngotracheal reconstruction
MLB: Microlaryngobroncoscopie
OTL: Oto-rhino-laryngology
PO: Per-os
SGS: Subglottic stenosis
SS: Single staged LTR

Discussion

Patients with type III and IV laryngeal webs typically present at birth with respiratory distress and may require immediate surgical management perinatally to establish an airway.[5] Regardless, they are generally treated for some duration with a tracheostomy whether before definitive surgery or after a double-staged LTR[1,3]. Tracheostomy in infants and children is associated with significant morbidity and even mortality, as well as cost to the healthcare system and burden to the family [10]. Forty-four out of 50 infants were tracheostomized either before or after definitive surgery in this review.

When determining the surgical approach, the severity of the web and subglottic stenosis should be considered. According to *Lawler et al.'s* study, the web type is predictive of symptomatology, the number of surgeries the patient will require, and if a tracheostomy will be necessary. Although the surgical techniques used to address the various webs are often surgeon-dependent and challenging, open surgeries tend to be performed in type III and IV webs with significant subglottic extension or when a previous endoscopic technique has failed[1]. In the case series of *de Trey et al.,* case 8 was a type III laryngeal web that failed endoscopic surgery and required revision LTR.[8] *Amir et al.* also reported a failed attempt at endoscopic laser and mitomycin C repair of a type III web[7]. Their second case was treated with endoscopic repair after emergent tracheostomy. This patient however had minimal subglottic extension and the treatment was successful after two iterations.

The present case report refers to an infant with a type IV laryngeal web and significant subglottic cartilaginous extension and therefore an LTR with thyroid alar graft for augmentation and posterior cricoid split offered the best chance for success.

Since laryngeal webs are associated with 22q11.2 microdeletions, it is important to complete genetic, cardiac, endocrinologic and immunologic workup[3,17,18]. The patient in this report was found to have a 22q11.2 microdeletion and early detection allowed management of the various systems involved.

The primary limitation to this study is that it is a review of very heterogeneously reported cases studies. Information available was limited, and if it was present, it was not always classified according to recognized classifications. Also, times data was reported for a group of patients and did not allow consideration of individual cases. Laryngeal web being rare, there is a dearth of data reported.

Conclusion

In the literature, patients with type III and IV laryngeal webs are tracheostomized for an average of 29 months. The case reported is that of a 2700 g three-day-old infant with a type IV laryngeal web successfully treated with a single-staged thyroid alar graft. This approach avoids the morbidity and use of scarce resources involved in caring for infants with tracheostomies. In select patients, a single-staged repair may avoid burdens associated with tracheostomy.

Children with Previous COVID-19 Infection Are More Likely to Present with Recurrent AOM or Tube Otorrhea

Beatrice R. Bacon BS, Michele M. Carr DDS MD PhD

Jacobs School of Medicine and Biomedical Sciences at the University at Buffalo, Department of Otolaryngology- Head and Neck Surgery

Introduction

Since December 2021, the number of children with COVID-19 infections has increased. Sequelae in children have not been well-described.

Objective

- Determine if children with a history of COVID-19 infection (C19 group) were more likely to present with recurrent acute otitis media (rAOM) or post-ventilation tube otorrhea (VTO) than children who had no history of COVID-19 infection (NoC19 group).

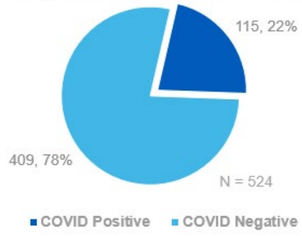
Methods

- Charts of consecutive children presenting at a pediatric otolaryngology clinic from March-May 2022 were reviewed.
- Demographics, COVID test history, co-morbidities, ultimate diagnosis, physical exam findings, and management plan were included.
- No children had a known COVID-19 infection at the time of visit.

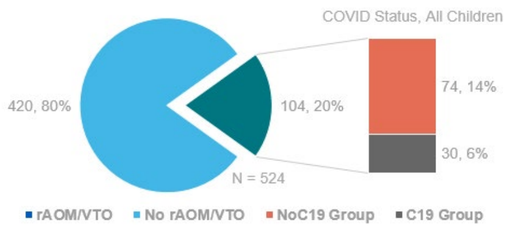
Results

- 524 children
- 228 (43.5%) girls and 296 (56.5%) boys
- Mean age 5 years (95% CI: 4.6-5.4), no difference between C19 and noC19 (p=.50)
- There was no difference in incidence of otitis media with effusion, tonsil/adenoid hypertrophy, sleep-disordered breathing, or epistaxis between the C19 and NoC19 groups.

History of Positive COVID-19 Test

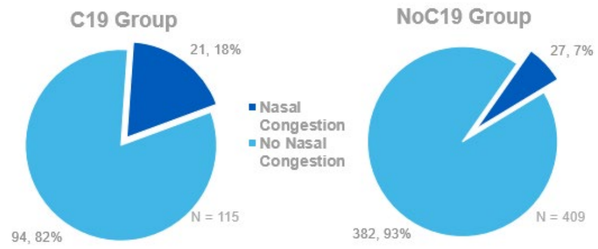


Diagnosis of rAOM or VTO



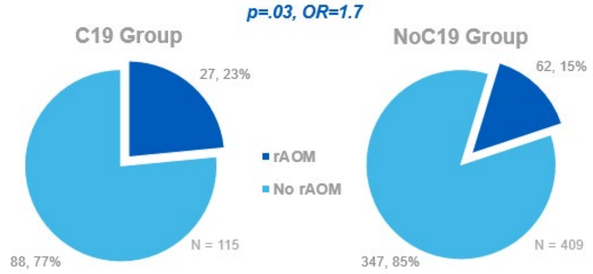
Diagnosis of Nasal Congestion

p<.001, OR=3.2



Diagnosis of rAOM in Children Without Ventilation Tubes

p=.03, OR=1.7



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Discussion

Without undermining the devastating personal and societal upsets caused by the COVID-19 pandemic, a positive, unexpected result of social isolation was a decrease in the prevalence of communicable diseases[1]. Within the field of pediatric otolaryngology, the COVID-19 pandemic initially reduced rates of rAOM and subsequent medical and surgical interventions[1]. This was likely related to daycare and school closures. However, when investigating the clinical presentations of patients who have been infected in the recent past, most studies to date have substantial limitations or do not find a meaningful difference between children who have or have not been infected by COVID-19[2]. Whether COVID-19 infection presages rAOM or whether co-varying factors such as daycare attendance and COVID-19 infection are responsible remains to be seen.

Conclusion

Infection with recent strains of COVID-19 may be associated with an increased risk of rAOM and VTO in children. This may affect healthcare utilization by increasing the need for pediatric and otolaryngologic care.

Outcomes of Pediatric Canal Wall-Down Mastoidectomy with Ossicular Chain Reconstruction

Daniel Juno Lee MD[1], Tyler G. Chan BS[2], Kristan Alfonso MD[1,3], Nandini Govil MD MPH[1,3]

[1]Department of Otolaryngology-Head & Neck Surgery, Emory University, Atlanta, Georgia, U.S.A.; [2]Emory University School of Medicine, Atlanta, Georgia, U.S.A.;
[3]Children's Healthcare of Atlanta, Atlanta, Georgia, U.S.A.

Contact: d.juno.lee@emory.edu

Introduction

- Canal wall-down (CWD) mastoidectomy is usually used as a last resort surgical option to address recalcitrant cholesteatoma in children, since conductive hearing loss is an inevitable sequela.
- Functional outcomes and post-operative complications of ossicular chain reconstruction (OCR) in pediatric CWD patients are not well described.[1,2,3]
- We report our experience with OCR in six pediatric patients who underwent CWD mastoidectomies for recidivistic cholesteatoma.

Methods

- **Study Design:** Case series retrospective single center chart review of patients at a pediatric tertiary referral hospital.
- **Inclusion Criteria:** All pediatric patients (<18yo at first mastoidectomy) who underwent CWD for recalcitrant cholesteatoma from January 2015 to April 2022 (n=37). Operative reports were reviewed for inclusion if CWD mastoidectomy was performed.
- **Data collected:** Past surgical history, indications for CWD mastoidectomy, number of mastoidectomies prior to CWD, if and when OCR was performed
- **Outcome measures:** pre and post-operative audiometric hearing results, and implant complications such as OCR displacement and extrusion.

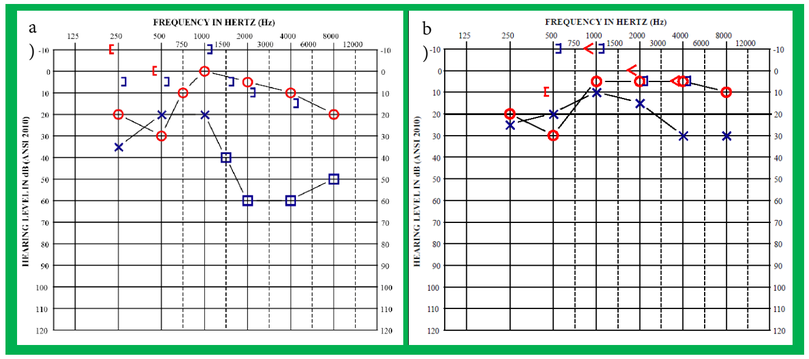

Figure 1. Audiograms of patient DH show significant improvement of hearing in L ear after OCR implantation. a) Pre-operative; b) Post-operative, 4 months.

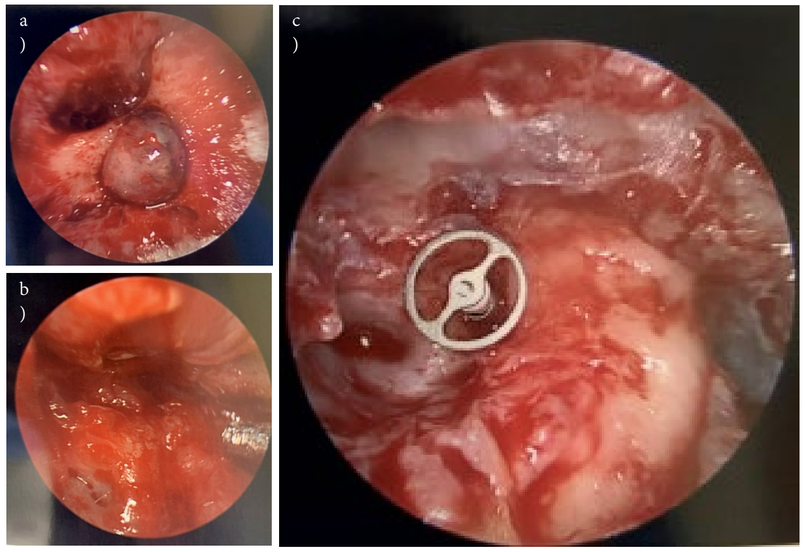

Figure 2. Intraoperative photos of patient AC's left middle ear taken with endoscope. a) Mastoid filled with thickened mucosa and pocket of old blood contents, debris, and cholesteatoma; b) Middle ear cavity, cholesteatoma removed; c) TORP in place with relation to facial ridge.

Results

- 37 patients received CWD mastoidectomy with a mean age of 12.05 years old (range 3-19). The mean number of prior mastoidectomies before CWD mastoidectomy was 1.83 (range 0-6). All cases were completed for cholesteatoma and all but 1 ear had partial or complete erosion of the ossicular chain.
- Of the 37 eligible CWD patients, 6 received OCR in ipsilateral ears. The mean number of prior mastoidectomies performed before CWD in patients who received OCR is 3.3 (range 2-5). Out of these 6 OCR recipients, 4 patients had implant complications including prosthesis displacement or extrusion.
- These 4 OCR failure patients were subsequently referred for bone conduction device (BCD) placement. One of these 4 patients (TD) showed mild audiogram improvement at 3-month post-operative follow-up. However, after an episode of otitis media, he developed displaced OCR and worsened hearing on audiogram.
- Another OCR recipient (DH) had hearing improvement on audiogram at 4-month post-operative follow up.

Patients (n=6)	# of sx before CWD	Timing of OCR Placement	Pre-op PTA (dB)	Post-op PTA (dB) [time after OCR placement]	OCR Failure	BCD Placement
LF	3	During CWD	28.3	35 [2 months]	Extrusion	Planned
MG	2	39 months after CWD	51.6	58.3 [3 months]	Displaced	Planned
CP	2	34 months after CWD	51.6	46.6 [4 months]	Extrusion	Yes
DH	3	During CWD	35	15 [4 months]	N/A	No
TD	5	During CWD	51.25	40 [3 months] 60.3 [24 months]	Displaced	Yes
AC	5	20 months after CWD	80	80 [2 months]	N/A	No

Table 1. CWD pediatric patients who underwent OCR.

Conclusions

- In our study, pediatric patients with CWD who had OCR had variable outcomes, with 67% (4/6) of patients ultimately requiring or planning on bone anchored hearing aids for hearing rehabilitation.
- Ossicular chain reconstruction after CWD can have favorable hearing outcomes in a pediatric patient, but it should be carefully considered due to potential post-operative prosthesis displacement and poor hearing outcomes.
- Determinants for successful CWD mastoidectomies with OCR and patterns of post-operative hearing loss warrant further investigation.

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Intraoperative Ergonomic Risk Assessment Among Otolaryngologists Performing Tympanostomy Tube Insertions

Adam G.F. Smith, B.S., Priyanka Reddy, M.D., Natalie Kelly, B.S., Faye Anderson, Ph.D., M.S., Natalie Quinn, Catherine Russo, Megan McNutt, Prasanth Pattisapu, M.D., MPH,
Tendy Chiang, M.D.

Introduction

- Up to 85% of surgeons regularly experience musculoskeletal (MSK) symptoms in the workplace.[1]
- Among otolaryngologists, 97% reported experiencing MSK pain from a work-related injury, 74% of which had their work affected by these symptoms.[2]
- Our previous work found that common otolaryngologic procedures carried substantial ergonomic risk as measured by Rapid Upper Limb Assessment (RULA) score and by the Craniovertebral Angle (CA) risk assessment tool during adenotonsillectomy.

Figure 1: Select Body Positions Included in the Rapid Upper Limb Assessment. (A) Upper Arm, (B) Lower Arm, (C) Neck, and (D) Trunk Positions.[3]

Objectives

- The purpose of this study was to evaluate ergonomic risk during tympanostomy tube insertion (TTI), a common, short-duration otolaryngology procedure. We hypothesized that while performing TTI, surgeons experience non-negligible ergonomic risk as measured by both RULA and CA.

Find Me

Our Ergonomics Research

Methods

- 11 attending pediatric otolaryngologists participated in this single institution prospective study.
- As participants performed TTI, a video recording that included head, neck, and upper back was captured.
- CA was quantified each minute, including the start of the procedure. A mean CA per TTI was calculated.
- RULA was scored at time of tympanostomy tube insertion.
- RULA scores >2 and CA measurements <50º were considered at-risk.
- Scores were summarized using means and standard deviations, and ranges were then utilized to stratify intraoperative ergonomic risk.

RULA Score	Level of Musculoskeletal Risk
1 - 2	acceptable posture
3 - 4	further investigation, change may be needed
5 - 6	further investigation, change soon
7+	investigate and implement change

Figure 2: Level of Musculoskeletal Risk Associated with RULA Score.

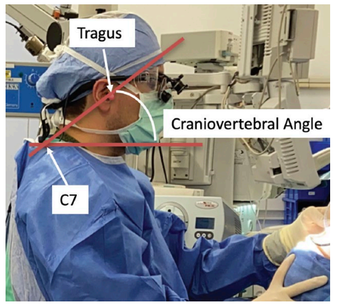

Figure 3: Measurement of Craniovertebral Angle.[4]

Results

- **Elevated ergonomic risk was found in most of the 232 TTI cases according to RULA score (n=206, 89%) and CA (n=199, 86%) measurements**.
- A median of 22 evaluations per surgeon were conducted with a range from 8 to 37 evaluations.
- Mean procedure duration was 1.7 minutes (SD 0.6 minutes).
- RULA neck/trunk/leg score moderately correlated with CA **(r = -0.57, p < 0.0001)** while RULA lower arm score was found to inversely correlate with CA **(r = 0.22, p < 0.0009)**.

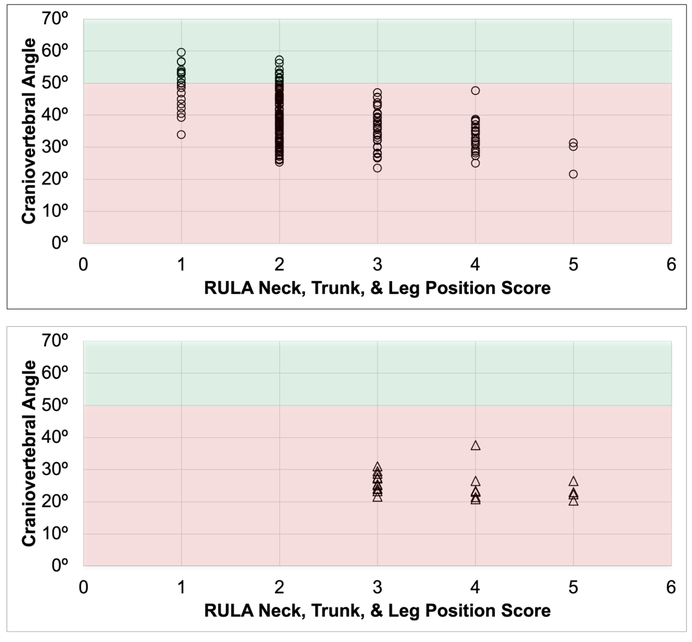

Figure 4: Scatterplots of craniovertebral angle and Rapid Upper Limb Assessment Neck, Trunk, and Leg Score. (Top) Procedures Performed While Seated, (Bottom) Procedures Performed While Standing.

Table 1: Select Ergonomic Risk Assessment Measurements of Otolaryngologists While Performing TTI

	TTI Performed While Seated	TTI Performed While Standing
Number of Observations	211	21
Mean CA (SD)	**39.1° (8.1°)**	**25.4° (4.0°)**
Mean Overall RULA (SD)	3.0 (0.5)	3.5 (0.5)
Mean RULA Wrist/Arm Score (SD)	3.1 (0.7)	2.9 (0.6)
Mean RULA Neck/Trunk/Leg Score (SD)	**2.3 (0.8)**	**3.7 (0.8)**

Discussion

- Otolaryngologists are exposed to ergonomic risk during TTI as measured by both RULA and CA.
- Opposing risk assessment trends between CA and RULA lower arm score were observed. This suggests that improvement of some aspects of posture may negatively impact others during TTI.
- Performing TTI while seated carried less ergonomic risk compared to performing while standing, particularly based on CA measurement.
- Further research is needed to identify interventions that might reduce symptoms and risks of injury, especially related to cervical angle in otolaryngology procedures.

Limitations

- Due to the relatively small sample size of this current study, analyses are underpowered at the α of 0.05 considered.
- Participants in this study were aware that they were being recorded. As a result, observational bias was potentially introduced.

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Clinical-epidemiological study of children referred for evaluation of swallowing disorder

Débora Bressan Pazinatto[1], Myrian Maria Andreotti Favaro[1], Flavia Peixoto[1], Luciahelena Prata Trevisan[1], Maria Angela Bellomo Brandão[2], Rebecca Maunsell[1]

[1]UNICAMP, Department of Otolaryngology, Campinas, SP, BRAZIL

[2]UNICAMP, Department of Gastroenterology, Campinas, SP, BRAZIL

Introduction

- Swallowing dysfunction in children can result in aspiration, chronic lung disease and poor weight gain/malnutrition, therefore its early diagnosis can prevent negative impacts[1].

Objectives

- To describe the clinical-epidemiological characteristics of children referred for swallowing evaluation at a Brazilian public university hospital.

Methods

- Prospective cohort study of 60 patients referred for swallowing evaluation by otolaryngologist and speech therapist between June 2019 and June 2021.
- For the assessment of swallowing, each child was submitted to a clinical feeding/swallowing evaluation which followed a standard protocol (PAD-PED)[2] and to instrumental FEES examination. Dysphagia was then classified in normal swallowing/no dysphagia, mild, moderate or severe.

Results

- Study with evaluation of 60 children and mean age of 4,25 years old.
- Mean duration of complaints until the first swallowing evaluation was 2 years, although 46 (76%) patients had at least one hospitalization for a respiratory cause in the previous year and are in follow-up with the average of 3 different pediatric specialties despite of otolaryngology.
- Of all the patients, swallowing was considered normal only in 7. Of the remaining patients, 25 (47%) had severe dysphagia with evidence of aspiration disease on fiberoptic endoscopic evaluation of swallowing (FEES), 17 of them were feeding orally before and had the feeding route changed due to the risk of aspiration.
- The main complaint across all patients was choking, nevertheless drooling was most experienced by the group of severe dysphagic patients. The average number of hospitalizations for respiratory causes in the last year was higher in children with severe dysphagia.
- There was a statistically significant association between the group of neuropathic children and dysphagia diagnosis ($p<0,05$).
- 96% of referred children with cerebral palsy and seizures had confirmed suspicion of dysphagia, of which 46% were severe cases with evidence of aspiration.
- Table 1 shows the classification of the degree of dysphagia and clinical features.

Swallowing	Mean age (years)	Feeding route	Comorbidities	Clinical complaints	Number of hospitalizations in the last year*	SPL suspected aspiration	Aspiration evidenced on FEES	Total number of patients
Normal	2,44	Oral (6) G-tube (1)	COPD (3) GERD (3) Heart disease (2) Tracheostomy (1) Tracheomalacia (1) Laryngomalacia (1) Esophageal atresia (1) Neuropathy (0)	Choking (4) Vomiting (1) Recurrent pneumonia (1) Cough (1)	0,85	Yes (1) No (6)	Yes (0) No (7)	7
Mild dysphagia	4,37	Oral (9) NET (3) G-tube (2) G-tube + oral (1) TFs/ Modified diet (1)	Neuropathy (12)/ Cerebral palsy (2) Genetic syndrome (10) COPD (8) Heart disease (6) GERD (5) Tracheostomy (1)	Choking (7) Recurrent pneumonia (3) Difficulty gaining weight (3) Drooling (3)	1,18	Yes (4) No (12)	Yes (0) No (16)	16
Moderate dysphagia	2,95	Oral (5) NET (2) G-tube (2) G-tube + oral (1) TFs/ Modified diet (1)	Neuropathy (11)/ Cerebral palsy (2) COPD (9) Genetic syndrome (6) Tracheostomy (5) GERD (3) Heart disease (2)	Choking (6) Recurrent pneumonia (4) Drooling (1)	1,72	Yes (9) No (2)	Yes (3) No (8)	11
Severe dysphagia	4,69	G-tube (8) G-tube + oral (6) Oral (5) NET (4) TFs/ Modified diet (2)	Neuropathy (25)/ Cerebral palsy (9) COPD (15) Tracheostomy (12) Genetic syndrome (9) GERD (8) Heart disease (4)	Drooling (10) Choking (9) Recurrent pneumonia (3) Difficulty gaining weight (2) Vomiting (1)	2,56	Yes (20) No (5)	Yes (23) No (2)	25

Table 1: Overview of evaluated patients. *Mean number of hospitalizations in the last year for respiratory causes. NET= nasoenteral tube; TFs= Thickened fluids; COPD= Chronic obstructive pulmonary disease; GERD= Gastroesophageal reflux disease; SPL= Speech Language Pathologist; FEES= Fiberoptic endoscopic evaluation of swallowing

Conclusion

- The diagnosis of swallowing disorders allows introduction of the safest feeding route with potential impact on the prevention of lung damage.
- The long time between the onset of symptoms and diagnosis shows the importance of the multidisciplinary approach for clinical suspicion and early referral for evaluation.

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Novel Application of Onyx Intralesional Embolization in the Treatment of Pediatric Lingual Venous Malformations

Ramya Bharathi MD[1], Holly Sprow BS[2], Maharshi Panchal MD[3], Mark Vecchiotti MD[1,2]

[1]Department of Otolaryngology, Tufts Medical Center, Boston, MA, USA. [2]Tufts University School of Medicine, Boston, MA, USA. [3]Department of Radiology, Tufts Medical Center, Boston, MA, USA

Abstract

Background: The treatment of head and neck venous malformations with preoperative embolization minimizes recurrence, cosmetic and functional morbidity. N-butyl cyanoacrylate (NBCA) has been the embolic agent of choice, however, Onyx is a new agent with a potential of being more effective based on the success on intracranial application.

Methods: We describe a patient with a lingual venous malformation treated with preoperative intralesional embolization using Onyx. We also present a review of the literature on current management with embolization agents and explore the mechanism of action of Onyx.

Results: Onyx has been used successfully in intracranial malformations. In our experience, Onyx delineates the borders of the lesion effectively, minimizes removal of uninvolved tissue, and reduces blood loss. We theorize that it is likely related to its mechanism of action versus other embolic agents. NBCA is an adhesive polymer that causes immediate solidification, therefore, there is less control over injection time, penetration, and extent of solidification. Onyx allows the user to determine the rate of solidification by controlling the amount of dimethylsulfoxide (DMSO) in the solution. This allows for better penetration, and an accurate solidification of the vascular malformation. Thus, increasing the chances of complete surgical resection.

Conclusions: The Onyx embolization system is a new alternative to traditional agents used in preoperative embolization of lingual venous malformations. Its unique mechanism of action favors more effective and accurate embolization and solidification, as we outlined in our case presentation. More research is necessary to effectively compare the onyx system with more traditional embolization agents.

Background

Pre-operative embolization agents: NBCA vs Onyx

NBCA

- Used since 1980s to embolize a lesion prior to surgical excision of high-flow intracranial and extracranial arteriovenous malformations.[1]
- Reduces operative blood loss and allows for complete or partial resection of the lesion.[1,2]
- NBCA is an adhesive polymer, and it works by forming casts and thrombus.[3]
- It also causes adhesion to the vascular wall, and causes an acute inflammatory reaction with vascular endothelial damage.[3]
- This results in fibrosis, and enhances the embolizing effects of NBCA.[3]

Onyx

- FDA approved in 2001[4]
- Pre-mixed, radiopaque, injectable embolic fluid consisting of the following components: EVOH (ethylene vinyl-alcohol copolymer), DMSO (dimethyl-sulfoxide) and TA (micronized tantalum powder).[4]
- The substance works by precipitating instead of polymerizing, to occupy the target area slowly without solidifying immediately like glue.[5]
- Can control how much solidifies by changing amount of DMSO.[5]

Case Presentation

11-year-old female with a 2 cm VM of the superficial aspect of the right tongue (figure 1). The VM was delineated with the use of Onyx embolization injected by Interventional Radiology (Figures 2). The patient was brought to the operating room immediately and underwent a successful and complete resection of the malformation with minimal blood loss, and minimal resection of the tongue musculature (Figure 3). We were able to achieve primary closure of the wound with preservation of surrounding tissue for optimal functional results. The patient had excellent wound healing post-op and no issues with oral intake or speech (Figure 4).

Figures

Table 1: Comparative features between Glue and Onyx

	NBCA (Glue)	Onyx
Time to solidification	Seconds	Minutes/variable
Injection time	Few seconds	Several minutes
Adhesiveness	High	Low
Final Consistency	Rocky/Hard	Foam-like/spongy
Solidification process	Polymerization	Copolymerization
Color	Colorless	Black

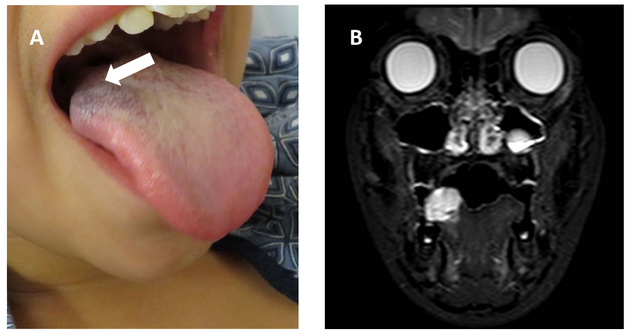

Figure 1: (a) Preoperative, post embolization of the tongue venous malformation (White arrow). (b) MRI, coronal, STIR image of the same lesion.

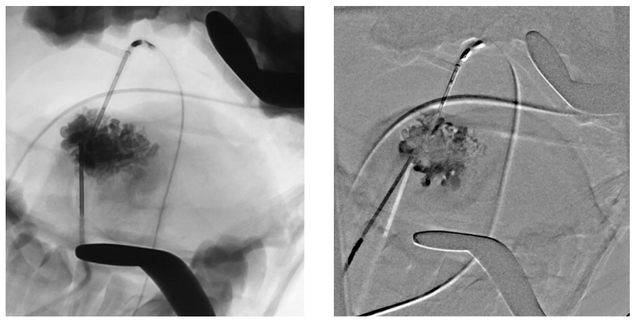

Figure 2: Sagittal, fluoroscopic images of percutaneous embolization using Onyx of the right tongue vascular malformation done by Interventional Radiology.

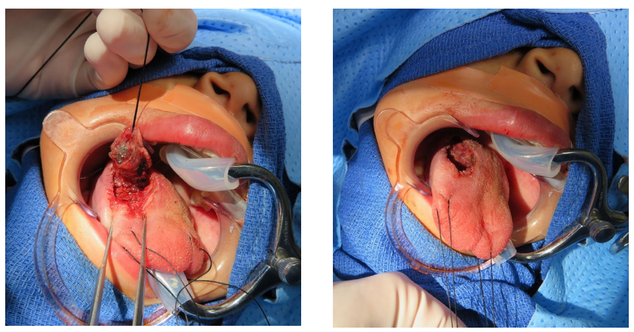

Figure 3: Intraoperative resection of the venous malformation after Onyx embolization. There was minimal blood loss and minimal resection of tongue musculature

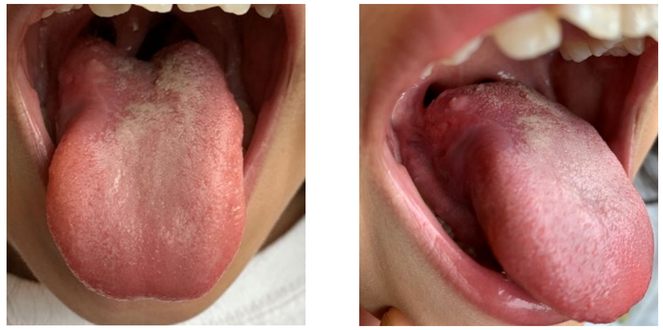

Figure 4: Post operative view of the tongue demonstrating well healed surgical incisions and preserved tongue architecture in the right posterolateral region.

Discussion

Traditionally, NBCA was widely used for pre-operative embolization of VMs. But with the advent of the Onyx liquid system, a new option was presented. In our experience the Onyx liquid system performs better than NBCA for surgical excision and it is likely related to their differences in their mechanisms of action (Table 1). NBCA is an adhesive polymer that causes immediate solidification.[3] There is less control over extent and penetration of the glue. This may cause incomplete penetration of the vascular malformation, which can lead to inadequate resection of the lesion. While Onyx is a non-adhesive, copolymer, that solidifies only after it contacts ionized fluid.[6,7] The usage of DMSO allows for greater control and precision during embolization. If there is unintended solidification of some areas, then more DMSO can be injected to reduce the solidification.[7] Then the catheter can be redirected to the desired areas. This process can be done slowly, carefully avoiding draining veins by pausing, allowing the Onyx to solidify, and then continuing the injection thus ensuring that the entire nidus of the malformation is covered.[7] Another advantage of Onyx is its color. The tantalum powder is black in color, allowing for easy visualization of the malformation extent during surgical excision.

Loh and Duckwiler performed a multicenter, randomized trial comparing NBCA to the Onyx system in embolizing AVMs. Their primary outcome was technical success in reducing the AVM volume by >50%, other outcomes were operative blood loss and procedure time. They found the Onyx system is equivalent to the NBCA as a preoperative embolic agent, Onyx achieved volume reduction in 96% or cases compared to 85% for NBCA, however, not statistically significant.[8] In our experience, Onyx has allowed us to minimize the surgical excision in cosmetically an/or functionally sensitive areas.

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Corresponding Author: Ramya Bharathi, MD
Department of Otolaryngology – Head and Neck Surgery, Tufts Medical Center
Email: rbharathi@tuftsmedicalcenter.org

Post-operative Admission Lengths After Adenotonsillectomy for Children with Trisomy 21

Alexis Lopez MD MPH[1,2] Raquel Good MA,[1] Javan Nation,[1,2]

[1]Rady Children's Hospital, San Diego, USA A [2]University of California, San Diego, San Diego, USA

Abstract

Background: Obstructive sleep apnea (OSA) is very common in children with Trisomy 21. Adenotonsillectomy (AT) is a common procedure in this patient population as it is the first line therapy for OSA. Some studies have suggested these children may have a higher risk of prolonged hospital stay, and few studies have examined this. The objective of this study is to assess the length of stays and factors associated with prolonged hospitalization among children with Trisomy 21 following AT.

Methods: A retrospective review was completed using the Pediatric Health Information System (PHIS) database. The patients included Trisomy 21 children <18 years of age who underwent an AT. The variables examined included age, gender, hospital length of stay (LOS), and presence of various comorbidities including cardiovascular, neurologic, respiratory or prematurity.

Results: A total of 1,822 children with Trisomy 21 aged 0-17 years (mean age of 6) were evaluated from 2017 to 2021. The LOS ranged from 0-17 days with 92.5% being admitted, and 83.2%, 6.8%, and 1.4% requiring a 1, 2, and 3 day admission, respectively. A Pearson correlation found a negative relationship (r=-.08), between age and LOS (p< 0.001). An independent samples t-test did not find a significant association between having a comorbidity (cardiac, neurologic, respiratory, prematurity) and LOS.

Conclusions: Children with Trisomy 21 have a high rate of admission following AT. A longer length of stay was associated with younger age, but not with having additional comorbidities.

Introduction

Obstructive sleep apnea (OSA) is very common in children with Trisomy 21, described as high as 60-80%.[1-2] This population is predisposed to OSA due to numerous anatomic factors, including macroglossia and a narrow nasopharyngeal airway.[2] Thus, adenotonsillectomy (AT) is a common procedure among patients with Trisomy 21.[1-2] Studies have demonstrated a moderate improvement in polysomnographic parameters and a mean decrease of the apnea-hypopnea index, but in many cases is non-curative.[2]

Patients with Trisomy 21 are at increased risk of post-operative complications requiring medical intervention, including respiratory issues, decreased oral intake and post-operative hemorrhage.[3-4] Respiratory complications range from atelectasis to requirement of continuous positive airway pressure.[4] In addition, this population has been found to have an increased length of stay and associated cost following AT.[3] The objective of this study is to assess the length of stays and factors associated with prolonged hospitalization among children with Trisomy 21 following AT.

Materials and Methods

A retrospective review was completed using the Pediatric Health Information System (PHIS) database. PHIS is a comparative database with clinical and resource utilization data for inpatient, ambulatory surgery, emergency department (ED) and observation unit encounters for more than 49 children's hospitals. It is managed by the Children's Hospital Association, an organization consisting of more than 220 children's hospitals.

The review included patients with Trisomy 21 aged <18 years of age who underwent an AT from 2017 to 2021. The variables examined included age, gender, hospital length of stay (LOS), and presence of various comorbidities including cardiovascular, neurologic, respiratory or prematurity.

The comorbidity was designated to patient if an additional ICD code fell within certain subcategories within each flag subset. For example, the cardiovascular flag included ICD codes with heart and great vessel malformations, endocardium diseases, cardiomyopathies, conduction disorders and dysrhythmias, or transplantation.

Results

Figure 1. Number of Admissions by Age

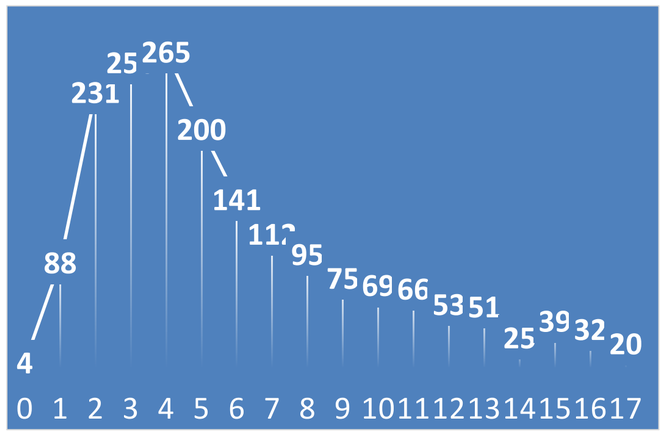
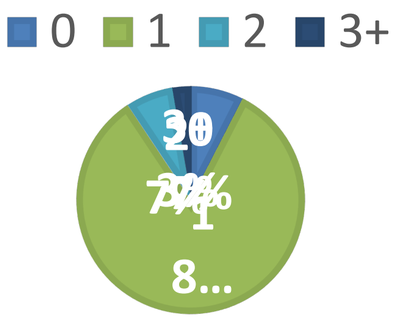


Figure 2. Length of Stay in Days by Percent



Demographic	
Age (mean)	6 years
Gender	
Female	876 (48.1%)
Male	946 (51.9%)
Comorbidity	
Respiratory	107 (5.9%)
Cardiovascular	163 (9.0%)
Neurologic	24 (1.3%)
Prematurity	3 (0.2%)

Conclusions

The majority of children within this review were admitted for at least one night following AT. Just over 50% of this admitted group were aged 2-5 years of age. The most common comorbidity associated with this population was cardiovascular, although this was not significantly associated with length of stay. Children below 6 years of age more commonly were admitted beyond 2 days.

Children with Trisomy 21 have a high rate of admission following AT. A longer length of stay was associated with younger age, but not with having additional comorbidities. This study is limited by its retrospective nature and lack of comparison group. In addition, the ICD for the diagnosis prior to surgery (ie, OSA) was not available.

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Skull Base Osteomyelitis and Retropharyngeal Abscess Following Injection Pharyngoplasty with Calcium Hydroxyapatite for Velopharyngeal Insufficiency

Michaele Francesco Corbisiero MSc, MPH[1]; Steven Leoniak MD[1,2]; Melissa Raines CPNP[1,2]
Brandi Axford CPNP[1,2]; Allison M. Dobbie MD[1,2]
[1]University of Colorado School of Medicine
[2]Children's Hospital Colorado, Colorado Springs, CO, USA

Children's Hospital Colorado

Affiliated with University of Colorado Anschutz Medical Campus

BACKGROUND

- Velopharyngeal insufficiency (VPI) is a disorder that manifests in patients as hypernasal resonance due to incomplete velopharyngeal closure during phonation.
- Depending on the patient's individual VPI characteristics, a variety of palatal and pharyngeal surgeries can be performed.
- Surgeons may elect for injection pharyngoplasty to augment the posterior pharyngeal wall.
- We herein report a rare presentation of a 9-year-old boy who suffered from osteomyelitis following calcium hydroxyapatite injection for VPI.

METHODS

- Case Report.

CASE SYNOPSIS

- A 9-year-old boy with cleft lip and palate underwent pharyngoplasty with calcium hydroxyapatite injection for VPI.
- Over the next two weeks, the patient developed significant neck pain and torticollis.
- CT neck showed a 20x24x44mm retropharyngeal abscess.
- Subsequent MRI of the neck following incision and drainage demonstrated skull base osteomyelitis involving the clivus, occipital condyles, and basisphenoid region.

IMAGING

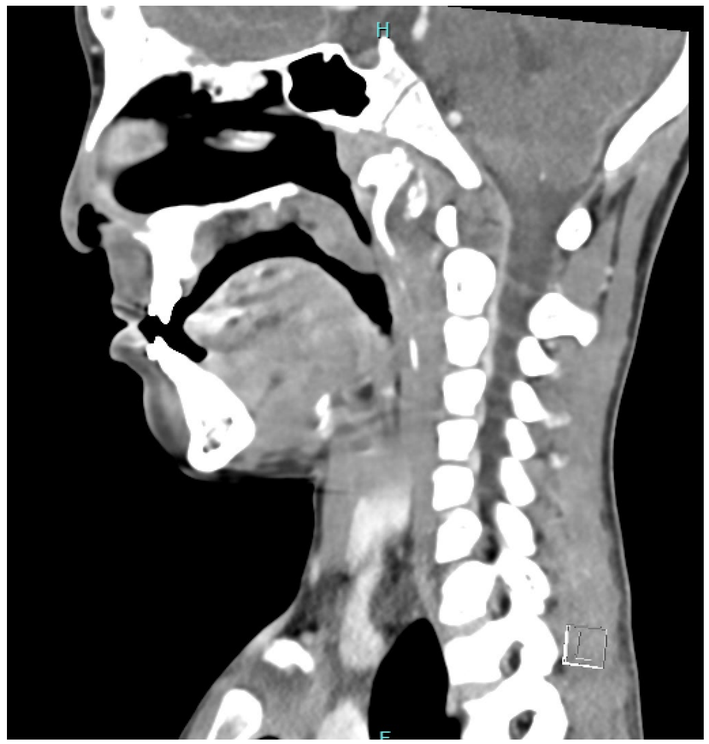


Figure 1. CT neck showed a 20x24x44mm retropharyngeal abscess.

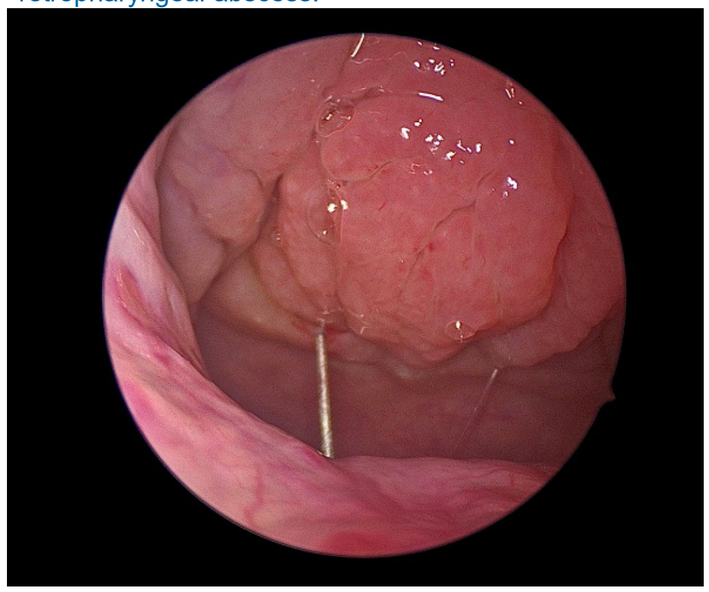


Figure 2. Injection of calcium hydroxyapatite.

CLINICAL COURSE

- POD#9 – ED visit for fever, headache, and neck pain with decreased ROM. IV Vancomycin and Ceftriaxone initiated.
- POD #11 – I&D completed with aspiration of 10 mL of purulent fluid, which grew *Strep intermedius* and *Staph aureus*.
- POD#14 – Vancomycin switched to Clindamycin per ID recommendations.
- POD#15 – Ceftriaxone stopped as CSF cultures were negative for 48 hours.
- Discharged on Oral Clindamycin 75 mg for 6 weeks. Repeated X-Rays after 3 months were normal – no osteomyelitis.

CONCLUSIONS

- In certain clinical circumstances, injection augmentation pharyngoplasty is an effective intervention for VP.
- However, risk of contamination and subsequent infection has not been explored in the current literature.

IMPLICATIONS

- This case highlights the serious infectious risk that accompanies this procedure.
- Preventive methods, such as a 0.5% povidone iodine irrigation, should be considered to mitigate this life-threatening, morbid complication.

DISCLOSURES

- None.

Surveying Dysgeusia/Anosmia in the Pediatric Population Infected with COVID-19

Bilal S Siddiq, BS; Chad Nieri, BS; Walter J Humann, MD; Anthony Sheyn, MD

Department of Otolaryngology – Head and Neck Surgery, College of Medicine, The University of Tennessee Health Science Center

Introduction

COVID-19 infection carries significant morbidity and mortality risks, ranging in severity from pneumonia and thrombus formation to anosmia and dysgeusia.

The chemosensory deficits seen are not only typically the earliest signs of asymptomatic carriers, but they can often be the only signs of infection. COVID-induced anosmia has affected ≈ 53% of patients. While these relationships with COVID have been elucidated in adults, a significant correlation between anosmia and dysgeusia, and COVID-19 in the pediatric population has not yet been cemented.

As of now, it is known that children infected with COVID tend to be mostly asymptomatic compared to adults. Possible etiologies for this difference include decreased expression of angiotensin converting enzyme 2 (which functions as the receptor for SARS-CoV-2), an overall less intense immune response, and/or viral interference due to the presence of other viruses also infecting the host.

Recognizing early anosmia and dysgeusia is important as it allows for the identification of otherwise asymptomatic carriers early on in the disease course, thus enabling intervention and limiting the spread of the virus. Additionally, earlier interventions in the pediatric population could increase odds of recovery of smell and taste

This study aims to clarify the relationship between anosmia and dysgeusia in diagnosed COVID-19 pediatric patients as well as its resolution in that population.

Methods

- Study participants were children who had tested positive for COVID-19 within the UTHSC and Le Bonheur Children's Hospital patient databases
- Only COVID-19 positive patients were contacted
- The children's caretakers were contacted via a text messaging surveying system through Questionpro
- The survey was direct and consisted of a total of 6 questions. Questions inquired as to
  - The patient's age at time of COVID-19 diagnosis
  - Their anosmia and dysgeusia status at the time of diagnosis and
  - Whether both of those symptoms resolved after resolution of their COVID-19 infection

Results

- A total of 279 people viewed the survey. A total of 46 patients actually participated in the survey, with 23 participants finishing the survey (8.2% response rate).
- 1 participant was over the age of 18, so they were excluded from the study results
- At the time of COVID-19 testing:
  - 50% of patients had at least some alteration in their ability to smell
  - 59.09% of patients had an alteration in taste
- At the time of resolution of their infection:
  - 9.09% of patients still had an alteration in smell
  - 9.09% of patients still had an alteration in taste
- Conversely, 90.91% of patients had no long-term changes in smell and 90.91% of patients had no long-term changes in taste

Figure 1. Participant Age

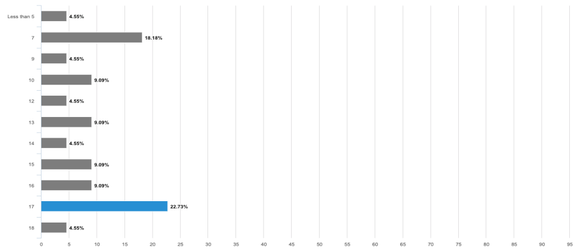


Figure 2. How long ago was the positive test?

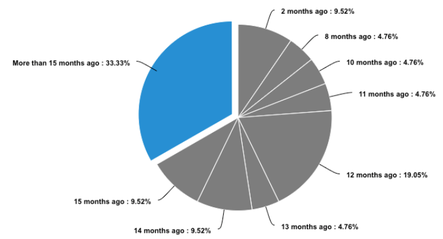


Figure 3. Smell alteration at time of positive COVID-19 test

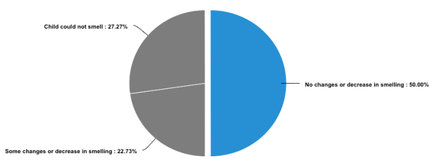


Figure 4. Taste alteration at time of positive COVID-19 test

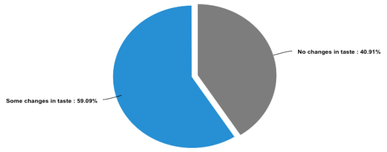


Figure 5. Anosmia status post infection resolution

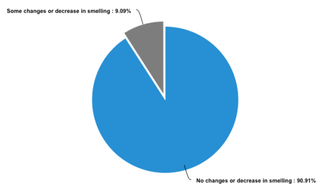
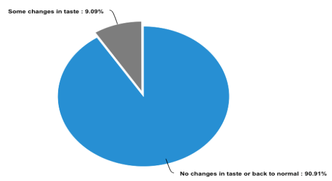


Figure 6. Dysgeusia status post infection resolution



Conclusions

This study demonstrates that the majority of the pediatric population affected with COVID-19 did not have ongoing anosmia/dysgeusia.

However, 2 patients did have long term sequelae from infection. Further research should seek to delineate the degree of loss in those patients with sequelae, as well as track them over a longer time period to assess for natural recovery in smell and taste function.

Additionally, attempts to elucidate the exact mechanism of recovery/dysgeusia in kids should be assessed for therapeutic interventions.

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CI Patient Adherence to Post-Implant Care Recommendations

Shannon Doolittle, AuD [1], Daniela Carvalho, MD [1,2], Julie Purdy, PhD[1], Megan Ellmaker, AuD[1], Keri Colio, AuD [1]
Rady Children's Hospital [1] and University of California San Diego [2]- San Diego, CA

BACKGROUND

Continued utilization rates of full-time cochlear implant use by young adults who were implanted as children has been reported by several authors in the literature [1,2]. A common report in these studies is that only a percentage of those implanted were queried to determine utilization rates. Many subjects are reported as being lost to follow up. Consequently, percentage of continued use is only available for subjects who can be successfully tracked. The percentage of and factors contributing to patients who are lost to follow-up are not widely reported. Additionally, strategies to reduce lost to follow-up rates have not been provided. It was the purpose of this study to examine cochlear implant patient adherence to post-implant care recommendations to include annual follow-up appointments for both Otolaryngology and Audiology.

METHODS

We received IRB approval through UCSD and RCHSD to investigate reasons for lost to follow up at our center. We retrospectively examined patient appointment data for patients implanted between the years of 1998-July 2021 for a total of 503 patients. Our center counsels patients preoperatively about the lifetime commitment of cochlear implantation and recommends annual appointments for otolaryngology and audiology once a child has reached stability in their implant process. Any child who had not been seen within the past 12 months was designated lost to follow-up. Contact was initiated via MyChart (email from our medical record system) and by phone for all the patients regardless of the duration of the loss to follow up.

RESULTS

Of the 503 patients seen, they were identified as being in one of nine broad categories:

Category	# / % of Patients
Current Patient Otolaryngology & Audiology	137 (27.4%)
Current Patient Audiology/Lost to FU Otolaryngology	117 (23.3%)
Lost to FU by both Otolaryngology & Audiology	89 (17.7%)
Moved	59 (11.7%)
Insurance change	58 (11.5%)
Followed elsewhere	29 (5.8%)
Current Patient Otolaryngology/Lost to FU Audiology	9 (1.8%)
Deceased	4 (0.8%)

For the 89 patients who were lost to follow up for both Otolaryngology and Audiology, the duration of loss ranged from 7 months to 24 years for Audiology and from 1 year to 19 years for Otolaryngology.

Lost to Follow-up by both Otolaryngology and Audiology	# / % of Patients
Non-User/Inconsistent User	21 (23.6%)
Insurance/Finance	5 (5.6%)
Seen Elsewhere	3 (3.4%)
Moved	5 (5.6%)
Did not believe they needed a return visit	6 (6.7%)
COVID Precautions	2 (2.2%)
Could Not Reach	47 (52.8%)

For the nine patients who had been seen by Otolaryngology but not followed up with Audiology, the loss to follow up for Audiology ranged from 14 months to 13 years.

Current Patient Otolaryngology/Lost to Follow-up Audiology	# / % of Patients
Non-User/Inconsistent User	2 (22.2%)
Insurance/Finance	1 (11.1%)
Did not believe they needed a return visit	3 (33.3%)
Could Not Reach	3 (33.3%)

DISCUSSION

For two of our lost to follow-up groups, of the patients for whom we were able to initiate response, 23/98 or 4.6% of the total were lost to follow-up due to being a non or inconsistent user of their cochlear implants. Consequently, when longitudinal studies report utilization rates but have patients for whom they do not have data, it is possible that the reported rates are higher than is actually taking place. Additionally, the fact that a patient has moved or has different insurance does not preclude that they might also be non-users.

CONCLUSION

When looking at loss to follow-up rates, it is important to identify potential barriers to returning care. Established cochlear implant users still may need assistance with adherence to follow-up recommendations.

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101 - SARS-CoV-2: A New Cause of Pediatric Hearing Loss Due to Labyrinthitis Ossificans

Tal Honigman[a], Fahad El-Dahari[a], Melissa Hazen[a,b], Vicky Papaioannou[a,b], Karen A. Gordon[a,b], Helen Branson[C], Blake C. Papsin[a,b], Sharon L. Cushing[a,b]

[a] Department of Otolaryngology – Head and Neck surgery and Institute of Medical Sciences, Faculty of Medicine, University of Toronto, ON
[b] Archie's Cochlear Implant Laboratory, Department of Otolaryngology, The Hospital for Sick Children, Toronto, ON, [C] Department of Diagnostic Imaging, The Hospital for Sick Children, Toronto, ON

ABSTRACT

Background: Otolaryngologic presentations of SARS-CoV-2 (COVID-19) primarily involved upper respiratory tract with few reports of otologic manifestations. There are a few reports of sudden onset sensorineural hearing loss (SNHL) in and around the time of acute COVID-19. Most were treated with corticosteroids, some were treated with hyperbaric oxygen and a few patients required and were treated with cochlear implantation (CI). Outcomes were variable.

Methods: Report of a pediatric patient treated with CI following COVID-19 related bilateral SNHL complicated by labyrinthitis ossificans with a review of hearing outcomes following reported cases of COVID-19 related sudden SNHL.

Results: 4-year-old was referred with new sudden onset sudden bilateral cochleo-vestibular loss following COVID-19 infection. He had regression of milestones and disequilibrium which could represent vestibular compromise. Audiogram showed bilateral profound SNHL that was confirmed by ABR. MRI findings suggested labyrinthitis bilaterally and assessment of his cerebrospinal fluid demonstrated abnormal white blood cell count without bacteria and normal protein as well as glucose levels.
Given the MRI findings consistent with inflammation that is known to lead to labyrinthitis ossificans, the child underwent urgent bilateral CI within a week. Intraoperative findings suggested intracochlear inflammation with fibrosis of the lumen. Despite this full insertion bilaterally of the electrode array was achieved via cochleostomy.

Conclusion: Sudden SNHL may be a sequelae of a recent infection with SARS-CoV-2.In patients with profound SNHL, urgent imaging is warranted to detect signs of inner ear inflammation and define the time-course of hearing loss evaluation and treatment given the risk of labyrinthitis ossificans.

BACKGROUND

SARS-CoV-2 (COVID-19) and its heterogenous symptoms and presentations have been reported with great interest in the scientific literature since late 2019. Reported presentations in the head and neck region primarily involve the upper respiratory tract symptoms. Reports of SARS-CoV-2 related otologic manifestations have been scarce, although there are a number of reports of sudden sensorineural hearing loss implicating SARS-CoV-2 as the potential underlying etiology [1].

Most reported patients with suspected SARS-CoV-2 related SNHL were treated with corticosteroids (enteral and/or intratympanic), some received hyperbaric oxygen therapy [2]. The SNHL in some adults were severe enough to require cochlear implantation (CI). Reported outcomes following these therapies are short term and variable.

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CASE SUMMARY:

4-year-old previously healthy child presented with sudden bilateral cochleo-vestibular loss following SARS-CoV-2 infection. A month prior his mother had confirmed COVID-19 infection. Timeline from presentation to diagnosis and treatment over a total of less than 6 weeks is shown in Figure 1.

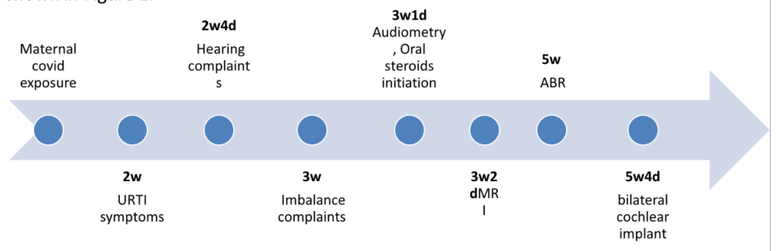

Figure 1: Timeline from possible exposure to hearing loss and cochlear implant

IMAGING:

MRI reported leptomeningeal enhancement suggesting an element of acute meningitis, bilateral avid inner ear enhancement including both the cochlea and vestibular end organs with loss of the fluid signal, especially in the basal turn of the cochlea suggesting labyrinthitis. Assessment of his cerebrospinal fluid did demonstrate abnormal white blood cell count, but no bacteria and normal protein and glucose levels were present.

Given the MRI finding, the child was deemed to be at high risk for early ossification of the labyrinth, and therefore a plan for urgent cochlear implantation was made. ABR was conducted under anesthesia confirming bilateral profound sensorineural hearing loss.

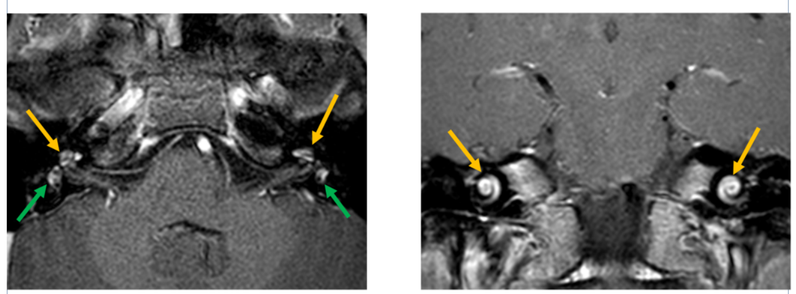

Figure 2: Axial and coronal T1 FS post-Gadolinium images demonstrate abnormal enhancement of both cochlea (orange arrows) and semicircular canals (green arrows)

TREATMENT:

The patient received bilateral cochlear implantation within the week of confirming bilateral deafness.. The round window was difficult to visualize bilaterally as is typical in the setting of ossification and raising further suspicion beyond the imaging concerns for intracochlear inflammation with fibrosis of the lumen. Upon entry into the cochlea, there was bleeding which again is typically seen in the setting of early ossificans. The Cochlear 612 counter advanace implant was inserted via cochleostomy with full insertion. Intra-operative evoked stapedial reflexes were present and the neural response telemetry confirmed responsiveness to implant stimulation.

Vestibular assessment was performed post implantation as he had significant imbalance and impaired ambulation at the time of the acute event. Gradual improvement occurred over time and subsequent to implantation with progression to ambulation on a broad base, running and return to participation in sport activities. Cervical VEMP were absent bilaterally.

At last follow up, 4 months after the procedure, He was doing well with implants responding to sound and planning return to school.

DISCUSSION & CONCLUSION

To our knowledge, this is the first reported case of bilateral profound hearing loss with labyrinthitis ossificans requiring cochlear implantation in a pediatric patient potentially due to SARS-CoV-2 infections.

In the case of SARS-CoV-2 infections induced SNHL three major entities remain unclear, 1) configuration of hearing loss (laterality, severity), 2) natural history of SNHL (recovery, response to conservative and nonconservative therapy) and 3) the pathophysiology of the underlying cochlear/neurologic insult. In addition, it is unclear whether profound SSNHL in the setting of SARS-CoV-2 is an isolated injury at the level of the inner ear, or part of a more general neurological insult (i.e viral meningitis/encephalitis) or some combination of the two.

The greatest concern was the presence of cochlear inflammation and clinical signs of ossificans which In its extreme form can lead to complete ossification of the cochlea can become fully ossified precluding implantation and leading to poor functional outcomes. Unfortunately, most cases of SSNHL that were reported in the context of a SARS-CoV-2 infection did not undergo any formal imaging of the brain and inner ear at the time of presentation or subsequently making it challenging to assess the frequency of cochlear ossification [1].

This case underlines the importance of an urgent audiological assessment in patients presenting with hearing changes in the context of a SARS-CoV-2 infection. In keeping with this, when profound SNHL is identified, expeditious evaluation of the inner ear with MRI can assess for early signs of inner ear inflammation, a risk for future ossification. This would aid in defining whether an accelerated time course to implant evaluation and ultimately implantation where appropriate is required prior to the development of irreversible cochlear ossification.