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# ABSTRACTS

SENTAC ANNUAL MEETING

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# MANAGEMENT OF LARYNGEAL CLEFT IN CHILDREN: EXPERIENCE OF A MULTI-STATE CHILDREN'S HEALTH SYSTEM

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Objective: Review presentation, diagnosis, management, and outcomes in children with laryngeal cleft(LC) seen in a single pediatric health system across three children's hospitals.

Methods: Retrospective summary of all cases of laryngeal cleft treated between August 2014 to December 2017. Data collected included demographics, comorbidities, presentation, preoperative and postoperative modified barium swallow, type of LC and intervention (observation, feeding therapy, injection laryngoplasty, or endoscopic repair.

Results: Twenty-one patients (11 males) presented at median age of 2 years (range 0.01-12.6), with 62% non-Hispanic, 52% commercial insurance. Fifty-two percent were treated in Central Florida, 28.6% in Northeast Florida, and 14% in Delaware. Type I LC was most common, followed by types II and III (86%, 10%, 5%, respectively). Most common symptom was cough (74%), followed by choking on feedings (47%), stridor (42%), aspiration (26%), and recurrent pneumonia (21%). Gastrointestinal symptoms occurred in 93%; 40% had diagnosis of congenital syndrome; and 40% had neurologic diagnosis. Intervention of any type occurred in 71%. Feeding therapy was performed in 19% (median 12 interventions; 8-27 IQR; range 8-31); single injection performed in 52%; surgical repair in 29% (median 1, range1-2). Eleven patients (52%) underwent injection; 3/11 required surgical repair due to persistent symptoms. Three (16%) underwent initial endoscopic repair with 1 requiring revision. Three patients did not demonstrate resolution of symptoms, all of whom required multiple interventions.

Conclusion: Variation in LC management is influenced by clinical condition, severity of aspiration, age, and comorbidity status.

# A COMPARISON OF THE PARAMETERS OF CHILDREN WHO FAIL AND SUCCEED TYPE ONE LARYNGEAL CLEFT REPAIR: A CASE CONTROL DESIGN

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Objective: Laryngeal cleft type 1 (LC1) repair has gained increased popularity in managing children with swallowing dysfunction (SD). Several investigations reported neurological diagnoses as the independent predictor of failure of the surgery, based on multivariable analysis of different independent variables. We aimed to assess other variables related to the patient through a case control design.

Method: in a retrospective study based on a tertiary referral practice we examined a prospectively kept surgical database for all the patients who had undergone a LC1 repair. Only patients who had undergone the procedure for SD, had a minimum of 3 months follow-up, and had complete information on outcome were included. A sex and age matched group was selected for comparison of outcome. Failure was defined as persistence of clinical symptoms and or pre-operative swallowing test abnormalities (functional endoscopic evaluation of swallowing - FEES & videofluoroscopic swallowing study - VFSS). Variables related to presentation, VFSS findings, anatomical findings and neuro-developmental findings were compared using McNemar's test then multivariable analysis was performed

Results: over 5.5 years, seventy-two repairs were performed. Seven were excluded. The mean age was 1.97±1.49 years (0.23-6.97), and there was 22 females. Twenty-two failed. There were no statistically significant differences between the variables of the two groups on comparison of proportion. The only significant predictor of success (coeff 0.56, p = 0.017) was the absence of an anterior larynx (grade 2-3 view).

Conclusion: The presence of an anterior larynx appears to be the most significant predictor for the outcome of LC1 repair.

# SCREENING FOR VOCAL CORD PARALYSIS IN HIGH RISK PREMATURE INFANTS AFTER PATENT DUCTUS ARTERIOSUS (PDA) LIGATION

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Objective: To determine clinical compliance with postoperative screening for vocal cord paralysis and associated comorbidities in infants after Patent Ductus Arteriosus (PDA) ligation. Given that vocal cord paralysis is a known complication of PDA ligation, an understanding of the current screening methodology is needed for improvement of care.

Methods: A three year retrospective chart review of all premature infants who underwent PDA ligation at a single tertiary pediatric hospital was performed. Vocal cord palsy, Otolaryngology postoperative consultations, and comorbidities (voice abnormalities, feeding difficulties, gastrostomy tube placement, and supplemental oxygen at discharge) were extracted from the charts. Standard statistical methodology was applied.

Results: 74 patients were identified (33M/41F; mean [SD] gestational age 25.6 [3.5] weeks). 17 patients (23%) were diagnosed with vocal cord paralysis; (14 patients (19%) left-sided, 2 patients bilateral, and 1 right-sided). Otolaryngology evaluated 27 patients (36%), despite having 33 (45%) with post-operative voice abnormalities and 48 patients (65%) with post-operative feeding difficulties. Patients with known vocal cord paralysis had a higher rate of gastrostomy tube placement (53% vs. 12%, p= 0.0003), supplemental oxygen (64% vs. 36%, p= 0.0417), and need for chronic ventilation at discharge (18% vs. 0.09%, p=0.0013) compared to those who did not.

Conclusions: The prevalence of vocal cord paralysis may be underestimated due to the lack of Otolaryngology evaluation after PDA ligation, highlighting the need for a better screening protocol. Premature infants with known vocal cord paralysis after PDA ligation have greater feeding and respiratory comorbidities. Timely diagnosis may improve discharge planning and patient/parent counseling.

# ASSESSING PREVALENCE OF OBSTRUCTIVE SLEEP APNEA IN PEDIATRIC PATIENTS WITH LARYNGOMALACIA

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Purpose: Congenital laryngomalacia is the most common cause of newborn inspiratory stridor. Fortunately, laryngomalacia often improves with age and most patients do not need surgery. The link between laryngomalacia and obstructive sleep apnea (OSA) is well known in children being treated for OSA - 3.9% of children with residual OSA after adenotonsillectomy have sleep-dependent laryngomalacia. However, it is unclear how laryngomalacia and OSA in infancy are related. We seek to examine OSA prevalence among primary laryngomalacia patients without obstructive symptoms, identify predictors for patients with laryngomalacia subsequently diagnosed with OSA, and track the clinical course of OSA versus laryngomalacia.

Methods: Retrospective chart review of pediatric patients with a history of laryngomalacia diagnosed over the last 5.5 years.

Results: Of 108 patients with laryngomalacia (median age <1 year old; 41.1% female; 58.9% male), 56 (52%) had sleep studies as per routine care of laryngomalacia patients by our pediatric otolaryngologist. Of patients with sleep studies, 44 (79%) had mild (73%), moderate (11%), or severe OSA (16%). There was no correlation between severities of laryngomalacia and OSA. 11 patients (25%) underwent surgery, after which the apnea-hypopnea index (AHI) decreased by a mean of 14.0 points (95% CI, 4.9-23.1; $P<0.05$). Of all patients (surgical and non-surgical) with laryngomalacia and OSA, 38 (86%; $P<0.001$) outgrew both conditions.

Conclusion: Laryngomalacia in infants is associated with OSA. Although in most cases both laryngomalacia and OSA resolve with age and seem to improve after supraglottoplasty, OSA may be an important indication for earlier treatment.

# INJECTION OF BOTULINUM TOXIN INTO EXTERNAL LARYNGEAL MUSCLES IN PEDIATRIC LARYNGEAL PARALYSIS: AN UPDATED EXPERIENCE

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Objective: describe an uncontrolled case series of neonates and children with bilateral idiopathic laryngeal paralysis (BILP) treated with botulinum toxin type A (BTA) injections into the external laryngeal muscles. Method: this is a retrospective study at a tertiary referral center. All patients diagnosed with bilateral laryngeal paralysis will be identified from a prospective surgical database. Only children diagnosed with BILP, based on flexible , rigid endoscopy with laryngeal electromyography (LEMG) and treated with BTA were included. Demographics, etiology, LEMG data, follow up, tracheostomy and outcome at last follow up presented descriptively.Results:over a 14 years period, fourteen patients in total were diagnosed with bilateral paralysis and treated with BTA.Eight females, median age at diagnosis 0.22 months (0.06-3 months).Five had tracheostomy performed; one had concomitant subglottic stenosis (died before repair or reassessment), one had microcepahly and laryngeal web, and one had diaphragmatic paralysis. Median of 10 units of BTA were used (5-16 units). Four were decannulated. Median follow up was 14 months (3-66 months).Median age at recovery 8 months (1-36 months) No morbidity or mortality resulted from the treatment.

Conclusion: BTA injection of external laryngeal muscles is associated with a significant rate of recovery of laryngeal function. This is likely due to neuromodulatory effect of the drug.

# UTILITY OF POLYSOMNOGRAPHY IN THE MANAGEMENT OF PEDIATRIC PATIENTS WITH CONGENITAL BILATERAL VOCAL FOLD DYSFUNCTION

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Introduction: Congenital bilateral vocal fold dysfunction(BVFD) presents in 0.75 cases per 1,000,000 neonates annually and historically requires tracheostomy in approximately 50% of patients. However, in neonates and infants with milder presentations of BVFD, one is often faced with the dilemma of recommending tracheostomy until BVFD resolution occurs or observing and expectantly managing the patient's BVFD. This study sought to review our institution's experience with polysomnography(PSG) in the management of BVFD patients.

Methods: Retrospective case-series

Results: From 2009-2018, 10/45 patients with BVFD were identified at our institution with PSGs performed postnatally and prior to tracheostomy. Average patient age at BVFD diagnosis and polysomnography was 7.4 days and 22.9 days, respectively. Bilateral paramedian paralysis and mixed vocal fold paralysis/paresis was found in 7/10 and 3/10 BVFD patients, respectively. Severe OSA was found in 2/10 patients with mean non-REM/REM AHI, O2 Nadir, Peak CO2 of 11.5/17.5, 81%, 69 torr - both of whom underwent subsequent tracheostomy. Mild-moderate OSA was found in the remaining 8/10 patients with mean non-REM/REM AHI, O2 Nadir, Peak CO2 of 3.6/6.4, 84%, 53 torr - none of whom underwent tracheostomy. 8/10 patients' PSGs were performed on room air while 2/10 were performed on minimal supplemental oxygen. Aspiration was noted in 2/10 patients pre-PSG and chronic tube feeding required in 1 patient pre-PSG.

Conclusions: Mild-moderate OSA vs. severe OSA was found in 8/10 vs. 2/10 BVFD patients, respectively, with the latter patients ultimately undergoing tracheostomy. PSG findings in BVFD patients with a mild respiratory/feeding burden may be helpful in determining surgical vs. non-surgical management.

# IMPROVING LOSS TO FOLLOW-UP - A COLLABORATIVE CITYWIDE INITIATIVE

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BACKGROUND AND OBJECTIVE:

With 95% of babies screened prior to discharge nationally, the implementation of newborn hearing screens has been very successful; however, loss to follow-up and loss to documentation (LTF/LTD) continues to be a significant hurdle. A baby is considered LTF/LTD when s/he does not receive timely and appropriate diagnostic testing and early intervention. Nationally, LTF/LTD rates are approximately 33%. In some states, LTF/LTD rates are higher. The focus of the current research is to reduce LTF/LTD rates in a geographical area prone to high LTF/LTD rates.

METHODS:

Our institution partnered with other institutions to coordinate efforts to improve communication among providers and between providers/institutions and families. In 2017, the birth hospitals began conducting both the inpatient and outpatient hearing screens. If the patient failed the outpatient screen, the screener and the parent together, at the time of the outpatient screen, immediately contacted the audiology provider institution to schedule the diagnostic testing. Scheduled weekly communications between the impatient/outpatient screen providers and diagnostic audiology providers were implemented to reduce the likelihood of LTF/LTD. Data was collected from the state newborn hearing screen database.

RESULTS:

Data from 2015-2017 was reviewed in the state newborn hearing screen database. LTF/LTD rates were significantly higher than the national average in 2015 (84%) and 2016 (64%). Following our implementation of the citywide collaboration, LTF/LTD rates decreased to 14% in 2017.

CONCLUSION:

We implemented a city-wide collaboration of providers/institutions and successfully reduced LTF/LTD in a geographical area with a history of high LTF/LTD rates.

# OUTCOMES OF BAHA CONNECT VS BAHA ATTRACT IN PEDIATRIC PATIENTS

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The two most commonly implanted bone-anchored hearing aid (BAHA) systems are the BAHA Connect and BAHA Attract. The BAHA Connect uses a skin-penetrating titanium abutment. The BAHA Attract uses an implanted magnet, leaving the overlying skin intact. Limited data is available on the difference in complication rates between the two systems. Our hypothesis was that there would be no difference in complications and audiologic data.

Retrospective chart review was performed of patients who had BAHA Connect vs. Attract at our children's hospital from 2006-2018. Pre- and post-operative information, such as demographics, related diagnoses, outcomes and complications were compared between the systems using Mann-Whitney U tests for 1) one year post-implant and 2) until the last follow-up. Audiology data was analyzed with a two-way mixed ANOVA.

24 Attract and 18 Connect BAHA surgeries were identified from 37 patients. Eleven Connect patients had the surgery completed in two stages. Connect patients followed up an average of 6 years post-implant and 9 months for Attract. 58.8% of patients with Connect surgeries had complications within a year and 82.4% had a complication by their last follow-up. There were no complications with Attract surgery. Patients with Connect surgeries had significantly more skin overgrowths, cultured infections, times on antibiotics, nursing phone calls, and ENT visits within the first year and for all records, $p<.05$.

The pure-tone average was significantly lower for both Connect (unaided-M=61.41,SD=5.32; aided-M=25.31,SD=5.62) and Attract (unaided-M=61.00,SD=5.11; aided-M=18.70,SD=16.43) after implant, $p<.001$.

Implantation of both systems lead to improved hearing outcomes with profoundly different complication rates.

# ASSESSING LOSS TO FOLLOW-UP AFTER NEWBORN HEARING SCREENING: ANALYSIS OF RISK FACTORS AT AN URBAN SAFETY-NET HOSPITAL

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Introduction: In 2014, the Massachusetts Department of Public Health estimated that approximately 1,300 newborns were referred on their newborn hearing screen, with 8.3% receiving a status of completely lost to follow-up or final diagnosis pending (referred to as LTF). This study seeks to analyze recent data to assess risk factors for loss to follow-up at Boston Medical Center (BMC), an urban safety-net hospital.

Methods: A retrospective chart review was conducted of patients born from January 2015 - March 2018 who received a referral status on their newborn hearing screen at BMC.

Results: 187 patients received a referral status; of those, 35 (19%) were completely lost to follow-up or received a status of final diagnosis pending. Of those who attended an initial audiology evaluation, the median time between screening and appointment was 29 days. Preliminary analysis indicates that maternal smoking status is associated with loss to follow-up ($p=0.01$). 43% of current smokers, compared to 14% of never smokers, are LTF. Further, an extended NICU stay was significantly associated with LTF ($p=0.02$; 28% of NICU babies, compared to 15% of non-NICU babies, were LTF). There is also a significant association between children of mothers with multiple prior deliveries and subsequent complete loss to follow-up ($p=0.04$).

Conclusion: Our results indicate that patients whose mothers are current smokers and those who had an extended NICU stay require additional support to increase compliance with follow-up after newborn hearing screening. In addition, strategies to address families who have multiple children can decrease the rate of LTF.

# OVERLAPPING DIAGNOSES WHICH COEXIST WITH THE DIAGNOSTIC DYAD OF CONGENITAL CYTOMEGALOVIRUS AND HEARING LOSS

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Objectives: To define the prevalence of congenital cytomegalovirus infection (cCMV) in children presenting with sensorineural hearing loss (SNHL) and the prevalence of co-existent diagnoses associated with congenital cytomegalovirus infection (cCMV) and hearing loss.

Materials and Methods: Retrospective review of children presenting with a diagnosis of SNHL between 2011 and July 2018 whose birth dried blood spots were tested for CMV by polymerase chain reaction (PCR).

Results: Testing revealed that 56/660 (8.5%) of children were confirmed to have cCMV. All were otherwise asymptomatic at presentation. Additional diagnoses not considered to be cCMV-related were noted in 5/56 (9%) of the children with cCMV including: achondroplasia, aural atresia, autism and genetic variants associated with Ushers type 1. Additionally, one child with cCMV had a family history of hearing loss due to branchio-oto-renal syndrome.

Conclusions: The prevalence of congenital CMV is high in children presenting with SNHL and can overlap with other conditions that affect hearing. These findings support both universal screening for cCMV, which can have long term consequences in development, as well as a multimodal approach to diagnostic evaluation of SNHL in children. In addition, these data suggest that the use of iterative algorithms that reach a stop upon a first and single diagnosis could lead to failure to identify co-existing causes of SNHL.

# COMPREHENSIVE GENETIC TESTING FOR CONGENITAL NONSYNDROMIC SENSORINEURAL HEARING LOSS

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Background

Guidelines for working up congenital nonsyndromic sensorineural hearing loss (NSSNHL) emphasize the use of comprehensive genetic testing (CGT) with next generation sequencing (NGS), yet these tests have limited accessibility and may not be covered by insurance providers. Our objective was to analyze CGT results for pediatric patients (<18 years) with NSSNHL.

Methods

A retrospective review was performed between 2014-2017 at a tertiary pediatric hospital. Demographics, clinical data and CGT results were analyzed. Logistic regression models identified characteristics associated with pathogenic variants (PV) and variants of unknown significance (VOUS).

Results

Fifty-five of 430 patients reviewed had CGT results. A mutation was discovered for 95% (n= 52). A definitive genetic etiology was found for 24% (n=13), over half of which were not of connexin mutations. At least one PV was identified for 33% (n=18), while at least one VOUS for 93% (n = 51). Three patients without a definitive genetic etiology had one PV and VOUS of the same gene. There were no significant differences in PV presence or number of VOUS across any characteristic except race. African Americans had significantly higher rates of VOUS with a rate ratio and 95% CI of 1.61 [1.11 - 2.34], p=0.01. Asians trended towards higher rates (1.96 [0.95 - 4.05], p = 0.06).

Conclusion

CGT is of high utility in the identification of relevant mutations and definitive genetic etiologies for pediatric patients with NSSNHL, especially for those who are non-Caucasians. Future studies should evaluate the efficacy of formulating race specific genetic panels.

# IDENTIFYING DIFFERENCES IN EARLY HEARING DEVELOPMENT USING THE LITTLEARS AUDITORY QUESTIONNAIRE

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This study asks whether the LittlEARs Auditory Questionnaire (LEAQ), a caregiver measure, can differentiate between the early auditory development of children with bilateral cochlear implants (CI), bilateral hearing aids (HA), and Auditory Neuropathy Spectrum Disorder (ANSD). The LEAQ is sensitive to impaired auditory development but has not previously been used to distinguish developmental changes between groups. We collected retrospective longitudinal LEAQ results from 43 children with HAs, 43 with CIs, and 18 with ANSD. The children with ANSD wore hearing technology. They were a similar age to the children without ANSD (23 mths, SD=15), while the CI group (14 mths, SD=8) was younger than the HA group (24 mths, SD=18) ($F(2,98.48)=3.4$, $p=.04$). The CI group often participated in their first LEAQ pre-treatment. Participants completed between one and seven LEAQs. Scores ranged between zero and thirty-five (Mean: 18.36). We conducted a linear mixed effects analysis of the relationship between LEAQ score and age. Results confirmed that children with HAs progressed faster than those with ANSD, who progressed faster than children with cochlear implants ($x^2(2)=17.25$, $p<.001$). Additionally, children with developmental delays demonstrated significantly slower growth in all three groups ($x^2(1)=11.64$, $p<.001$). Although we observed the expected pattern of responses in the CI and HA groups, there was more variability in the responses of the ANSD group. There was no connection between LEAQ growth and speech perception outcomes in a sub-sample ($r(6)=.42$, $p=.30$). The LEAQ is a useful tool for monitoring auditory development in very young children and can inform early treatment decisions.

# A PILOT STUDY VALIDATING A PHONE TRIAGE EVIDENCE BASED CLINICAL PATHWAY FOR TYMPANOSTOMY TUBE OTORRHEA IN CHILDREN

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Introduction: As healthcare moves away from volume-based to value based delivery models, evidence based clinical pathways detail essential steps in patient care to reduce the costs and utilization of health care resources. Ideal pathways lead towards standardized, patient-centered care through an algorithm that is evidence-based, interventions with criteria-based progression, and measurable endpoints or quality indicators. Using these standards, a clinical pathway for managing tympanostomy tube otorrhea beginning with phone triage was developed in accordance with AAO-HNSF Guidelines.

Methods: Retrospective case series of all consecutive patients calling the otolaryngology nurse's line at a tertiary pediatric hospital 3/2018-6/2018 regarding otorrhea. Nurses completed a standardized and evidence based form based on parent responses regarding purulence, tympanostomy tubes/perforation, fever>102 degrees, ear redness, bacterial rhinosinusitis, sore throat, and immunodeficiency, which was sent to the APPs to assess for antibiotic drops. Otorrhea form information and tympanostomy tube history, subsequent phone calls, clinic visits, and antibiotic prescriptions for otorrhea were extracted.

Results: Fifty-two patients were included. Median child age at phone call was 2.3 years (range 0.3-19.7 years), and 46.2% were female. All patients had prior tubes and active purulent otorrhea. No parents reported bacterial rhinosinusitis, sore throat, cellulitis, or immunodeficiency. All were prescribed antibiotic drops by an APP [ofloxacin (57.7%), ciprofloxacin (30.8%), or ciprofloxacin with dexamethasone (7.7%)] or already had drops (3.8%). Nine (17.3%) received another antibiotic prescription and 15.4% had a subsequent clinic or urgent care visit for otorrhea.

Conclusions: This pathway obviated clinic visits in 84.6% of patients with a 82.7% treatment cure.

# USE OF OTIPRIO TO TREAT CHRONIC OTORRHEA: A NOVEL, MINIMALLY INVASIVE SOLUTION FOR A DIFFICULT PROBLEM.

Pameal Hanson

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Introduction: Chronic suppurative otitis media (CSOM) and persistent tympanostomy tube otorrhea (TTO) are often relentless problems, severely impacting families. If initial topic treatment fails, other options may be increasingly invasive: oral antibiotics, intravenous antibiotics, ear washout, mastoidectomy. We propose a novel use of Otiprio (high dose gelatinizing ciprofloxacin drop) to treat TTO and CSOM. This is a review of our experience after 20 patients were considered for this protocol.

Methods: Children who have failed topical and/or systemic treatment for TTO or CSOM were offered Otiprio placement under sedation. This project was exempted through IRB. Patient charts were reviewed for prior treatment, cultures, comorbidities, resolution/partial resolution of otorrhea.

Results: While 20 patients were scheduled for Otiprio placement, 13 had it placed in 25 ears. Cultures were obtained in 7/13 patients with the most common bacteria being MRSA (42%). Patients had used an average of 1.8 (range 1-3) courses of topical drop and 1.5 (range 0-4) courses of systemic antibiotics prior to Otiprio. 3/13 patients had craniofacial or genetic comorbidities and 1 had immunodeficiency. We had complete or partial resolution in 12/13 (92%)patients or 23/25 (92%) ears.

Conclusions: We have had success using Otiprio as an adjuvant treatment in persistent and recalcitrant otorrhea cases. In reviewing our off label use of Otiprio, we demonstrate 92% success in resolving ear drainage. This is an important tool for all pediatric Otolaryngolgists to consider.

# PARENTAL HEALTH LITERACY IN PEDIATRIC OTOLARYNGOLOGY: A PILOT STUDY

Nicole Leigh Aaronson (M.D.)

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Objective: Although pediatric otolaryngology encompasses the highest frequency of elective surgical cases in children, little is known regarding health literacy of parents making health decisions for these children. This is a pilot study to assess parental health literacy in the pediatric otolaryngology clinic and evaluate relation to personal demographics.

Methods: Parents in a pediatric otolaryngology clinic completed the Short Assessment of Health Literacy-English (SAHL-E). Parents were defined as low (0-14) or high (>14) literacy based on mean scores. Two-tailed t tests and the Mann-Whitney U test were used to evaluate relationships between parent demographics and literacy. Postoperative and diagnosis-based leaflets were evaluated for readability using the Flesch Reading Ease Score (FRES) and the Flesch Kincaid Grade Level (FKGL) to provide context for literacy results.

Results: Fifty parents, average age 36 years, participated (80% female, 76% white, 80% privately-insured, 90% some post-high-school education). Seventy-four percent had no experience working in healthcare. Mean SAHL-E score was 17.6 (range: 16-18); all 50 parents scored >14, indicative of proficient health literacy. Continued education after high school was associated with higher scores (mean 16.8 vs 17.69, P=0.003) and was the only significant variable in multivariable linear regression. Educational leaflets ranged 5.8-8 on FKGL and 62.5-77.5 on FRES.

Conclusion: Parents in this clinic had high health literacy, inclusive of word recognition and association. Educational materials are written at appropriate levels for public health information. This pilot study is intended to highlight issue of literacy and advance methods to person-oriented research.

# CLINICAL AND RADIOLOGIC CHARACTERIZATION OF FRONTAL SINUSITIS IN THE PEDIATRIC POPULATION

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Introduction

Frontal sinusitis in the pediatric population is a condition that hasn't been fully studied and characterized.

Objectives

To characterize the clinical presentation, radiologic variables, treatment modalities, complications and prognosis associated with acute and chronic frontal sinus disease in the pediatric population.

Methods

IRB-approved retrospective cohort study of pediatric patients who were diagnosed with acute(AFS) or chronic frontal sinusitis(CFS) and underwent frontal sinus surgery at a tertiary level children's hospital 2006-2016. Patients with AFS were compared to patients with CFS. Statistical analysis completed using chi-square test or Fisher's exact test, statistical significance set at $p<0.05$.

Results

Nineteen patients with AFS and fifteen patients with CFS were analyzed. There was male predominance in AFS and female predominance in CFS($p<0.05$). AFS patients were less likely to have seasonal allergies, prior sinus disease and significant comorbidities($p<0.05$). Additionally, AFS patients presented with constitutional, neurologic and ocular symptoms. The CFS group had predominant sinonasal symptoms. CT-scan analysis showed that AFS patients had higher prevalence of complex frontal anatomy (Type-II cells,concha bullosa and Haller-cells) compared with CFS patients($p<0.05$). Culture results were positive in 78% in AFS group with S.Millieri(53%),Anaerobes(20%) and normal flora(17%). In the CFS group cultures were positive in 60%:normal flora(56%),H.Influenzae(13%),Pseudomonas(6,5%) and others(24.5%). CFS patients were more likely to have persistent sinus disease and require repeat sinus procedures($p<0.05$).

Conclusion

There are two distinct presentations of frontal sinus disease in the pediatric population. Patients with AFS vary significantly from the patients with CFS. Correct identification and understanding is crucial for the appropriate management of this disease process.

# THE IMPACT OF BILATERAL TURBINOPLASTY WITH OR WITHOUT ADENOIDECTOMY ON THE QUALITY OF LIFE OF CHILDREN: RETROSPECT

Hamdy El-Hakim (M.D.)

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Objectives: To quantify the improvement in quality of life of children with chronic rhinitis (CR) who had undergone inferior turbinoplasty (IT) with or without adenoidectomy (Ad).

Methods: a retrospective cohort study was undertaken at a tertiary pediatric center. Eligible patients were those diagnosed with CR refractory to medical treatment and had undergone IT with or without Ads. We excluded those with polyposis, septal deviation, symptoms of recurrent acute or chronic sinusitis, had other concomitant sino-nasal procedure, dysmorphic features, syndromes, or other congenital nasal pathology. Patients were identified from a prospectively kept surgical database. The outcome measure used was the Glasgow Children's Benefit Inventory (GCBI),

Results: 89 patients treated between January 2009 and December 2016 were eligible. 60 patients were included (mean age 14.03 years, 31 males, 29 females). The mean follow-up duration was 39.61.87 months (range 8.7-93.6). The overall median GCBI score was 22.9, 25th-75th percentile 6.25-39.6). 42 had IT and 18 had both IT and Ad. The median GCBI score of patients who had IT only was 22.9, (25th and 75th percentiles of 6.25 and 41.67, respectively). The patients who underwent IT and Ad showed a median GCBI score of 23.9, with 25th and 75th percentiles of 8.3 and 35.4, respectively. There was no statistically significant difference between them ($p = 0.891$). One patient developed toxic shock syndrome.

Conclusion: This study provides evidence on long term maintenance of improved quality of life after IT with or without Ad in children with CR.

# TESTING FOR TRACHEOSTOMY ALLERGY AS A CAUSE OF RECALCITRANT CHRONICALLY INFLAMED LARYNX

Lyndy Wilcox (M.D.)

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PURPOSE/INTRODUCTION: Pediatric patients with laryngotracheal stenosis often require surgical intervention to achieve decannulation. An increasingly recognized subset of these children have chronic laryngeal inflammation. While gastroesosphageal reflux and eosinophilic esophagitis are known potential contributors, some children have negative testing or persistent inflammation despite treatment of these conditions. Azithromycin has previously been tried with success in a portion of these patients, but some have persistent inflammation that delays surgical intervention or greatly reduces its success rate. This study presents patch testing for a delayed hypersensitivity reaction to components of tracheostomy tubes or care products to evaluate for this a cause of ongoing laryngeal inflammation.

METHODS: With IRB approval, a retrospective case series was performed on all children who underwent patch testing to tracheostomy components from January 2016 to May 2018 due to chronic laryngeotracheal inflammation. Data regarding endoscopic findings, diagnostic evaluation, prior interventions, and allergy testing results were collected. If allergy testing was positive and intervention was instituted, the post-intervention endoscopic findings were also evaluated.

RESULTS: Over the study timeframe, eight children ages 1 to 9 years underwent patch testing for chronically inflamed larynx. Other symptoms prompting testing included severe skin irritation and peristomal granulation. Four of the children demonstrated positive patch testing to various tracheostomy components. Post-testing interventions resulted in improvement in the laryngeal inflammation with two of the children being considered candidates to undergo laryngotracheal reconstruction.

CONCLUSION: Testing for tracheostomy allergy should be considered in patients with recalcitrant laryngeal inflammation preventing proceeding with surgical intervention for laryngotracheal stenosis.

# DIAGNOSIS AND MANAGEMENT OF IDIOPATHIC TRACHEAL STENOSIS IN CHILDREN

Philip D Knollman (M.D.)

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Objectives: To describe the evaluation, diagnosis, and management of idiopathic tracheal stenosis in children.

Methods: Retrospective review of 10 consecutive pediatric patients with idiopathic tracheal stenosis at a tertiary care pediatric medical center.

Results: Ten patients (6 males) with a median age of 10.5 years old (range 19 months to 15 years) were reviewed. The median time to diagnosis was 8.5 months (range 3 to 48 months), and the median duration of followup was 58.5 months (range 12 to 145 months). No patients had significant medical co-morbidities, preceding episodes of airway trauma, or prior intubations. All patients were diagnosed by endoscopic airway evaluation, revealing isolated focal tracheal stenosis (n=7) or multilevel airway stenosis, including the trachea (n=3). Biopsies of stenotic lesions were performed in 8 patients, revealing evidence of fibrosis with chronic inflammation. Overall, patients underwent a mean of 2.6 (±1.0) endoscopic procedures and 1.7 (±0.8) balloon dilations per year. Common adjuvant treatments included steroid injection (n=7), lysis of stenosis (n=6), and systemic immunosuppression (n=4). Three patients underwent open airway reconstruction; two patients underwent cervical slide tracheoplasty, and one underwent laryngotracheoplasty with subsequent cricotracheal resection. At most recent followup, 5 patients had significant clinical improvement and were asymptomatic. Four patients experienced recurrent symptoms after periodic endoscopic intervention. One patient experienced progressive disease.

Conclusions: This study represents the largest series of pediatric patients with idiopathic tracheal stenosis, a clinical entity almost exclusively described in adults, illustrating the evaluation and management of this rare and difficult to treat condition.

# PATIENT-CENTERED VALUE PROPOSITION OF AN AIRWAY FOREIGN BODY PROTOCOL THAT INCLUDES CT AIRWAY

Courtney A. Hill (M.D.)

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Purpose: The decision to perform bronchoscopy in children with suspected airway

foreign body (AFB) can be challenging. We present patient-centered quality outcomes of an AFB protocol that includes a computed tomography (CT) airway for children who were deemed at intermediate risk of having an AFB (only one of: witnessed choking event, unilateral wheezing, or unilateral air trapping/foreign body on CXR).

Methods: Retrospective review of children presenting to a tertiary pediatric hospital for a year-long period before and after institution of the airway foreign body protocol. Chi-squared, T-test, and Mann-Whitney test were used for statistical analysis.

Results: Forty-eight visits to the emergency department (ED) with suspicion of AFB were compared to 77 visits after institution of the protocol. The protocol significantly increased the use of CT airway (p=0.15); once stratified by

risk, this was evident only for the intermediate cases (p=0.045). Rate of bronchoscopies significantly decreased for intermediate risk cases (p=0.027) but were unchanged overall. Overall negative bronchoscopy rates decreased

significantly from 50% to 29.6% (p=0.02). Median ED and total visit time tended to decrease across all risk stratifications but were not statistically significant. Median hospital charges tended to decrease for intermediate risk cases.

Conclusions: Institution of this protocol including CT airway led to improved care for patients who were not obviously with or without an AFB. We rely on quality outcomes that matter to patients and providers rather than contrasting the risk-benefit analysis of anesthetics (bronchoscopy) vs radiation (CT airway) exposure in intermediate risk cases.

# TRACHEO-INNOMINATE FISTULA IN CHILDREN: A SYSTEMATIC REVIEW OF THE LITERATURE

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Background/Objective:

Tracheo-innominate fistula (TIF) is a rare but fatal complication of tracheotomy, traditionally described as occurring in the first three weeks post-tracheotomy. To date, there has been a paucity of literature in pediatric TIFs. The objective of this study was to conduct a systematic review of observational studies documenting the development and management of TIF in pediatric patients.

Methods:

We conducted a systematic review using MEDLINE, Embase, Cochrane Database of Systematic Reviews, Web of Science, CINAHL. We included all studies with pediatric patients (under 18 years of age) who developed TIF following tracheostomy.

Results:

Fifty-four publications met inclusion criteria, reporting on 77 cases. The most common indication for tracheostomy was prolonged intubation and the need for ventilatory support (38.6%). Patients with neurological co-morbidities were the most common (72.7%). The mean time to develop TIF was 395.7 days (95%CI, 225.9-565.5). Fifty-four patients (70.1%) presented with massive hemorrhage while 18 patients (23.3%) presented with a sentinel bleeding event. The most common diagnostic modalities were diagnostic imaging and bronchoscopy (55.8%). A substantial number of patients did not have any investigations (41.6%). 70.1% of patients underwent surgical management. Mortality was 38.6% in reported cases with variable follow-up periods.

Conclusion:

TIF may occur in long-term tracheostomy-dependent children beyond the conventionally described 3 week period and the mortality may not be as high as previously reported with timely intervention. Our results are limited by inherent risks of bias. Further research including well-designed cohort studies are needed to guide an evidence-based approach to TIF.

# BEDSIDE PEDIATRIC TRACHEOSTOMY: A RECIPE FOR SUCCESS

Heena Narsi Prasla

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Prolonged intubation may necessitate tracheostomy placement. Although adult tracheostomies are often performed at the bedside, pediatric tracheostomies are typically performed in the operating room. There are times the risks of transport are too great. In this case series we will present our institutional experience with pediatric bedside tracheostomies.

This is a retrospective review of bedside tracheotomies performed at a tertiary care hospital from December 2011 to January 2018. Data collected included age and weight at the time of surgery, reason for bedside procedure, perioperative complications, and admission mortality.

Ten bedside tracheostomies were performed during the review period. The median age was 317.50 (range 92-4,364) days. Deciding factors for bedside tracheostomy included patients with pulmonary hypertension (PHTN) requiring nitric oxide (iNO), extracorporeal membrane oxygenation (EMCO), or continuous dialysis. The mean surgical time was 44.4 (range 32-67) minutes; mean total procedure time was 85.5 (range 73-114) minutes. There were no perioperative complications within the first 2 weeks; however 5 (50%) patients died during that hospitalization. Mortality was unrelated to the tracheostomy.

The decision to perform bedside tracheostomy requires a collaborative effort by the otolaryngologists, anesthesiologists, and intensivists. Factors to be considered are the patient’s airway status; comorbidities preventing safe transport; logistical accomodation for the surgical staff; and surgeon’s comfort level. In our institutional experience it is possible to safely perform bedside tracheostomies in patients who are unable to transport to the operating room. However, given the high 30-day mortality, the indications and desired outcomes of tracheostomy need to be clearly delineated.

# EFFECT OF INPATIENT MULTIDISCIPLINARY TRACHEOSTOMY TEAM ROUNDS ON TIMELY OUTPATIENT FOLLOW-UP

Jessie Marcet-Gonzalez

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Objectives: To assess the effect of inpatient tracheostomy team rounds on timely outpatient follow-up in patients with tracheostomies.

Design: Retrospective review of outpatient follow up rates for patients before and after initiation of weekly tracheostomy rounds.

SETTING: Tertiary children's hospital in Houston, Texas.

PARTICIPANTS: Patients with tracheostomies discharged between 2012 and 2017

Methods: A retrospective review was performed of all patients with tracheostomies discharged from the hospital during the study period. The ideal follow up period for patients with newly placed tracheostomies was defined as within 6 weeks of discharge, and within 6 months for those with existing tracheostomies. The rate of appropriate follow up in the Otolaryngology clinic was determined for both populations before and after the initiation of inpatient multidisciplinary tracheostomy team rounds (begun 12/1/2015).

MAIN OUTCOME MEASURE: Compliance with clinic follow-up after hospital discharge.

RESULTS: From July 2012 to December 2017, a total of 942 patients were hospitalized. Hospitalizations for any reason (not only respiratory/tracheostomy-related) were included. Prior to inpatient trach rounds, the follow up rate was 16% within 6 months of discharge. After initiation of rounds, the follow up rate increased to 22%. In the subset of patients with newly placed tracheostomies, follow up within 6 weeks of discharge rose from 38% in 2015 to 63% in 2017.

CONCLUSIONS: The introduction of regular inpatient tracheostomy team rounds significantly improved follow up rates, specially in patients with newly placed tracheostomies. This study highlights opportunities for improvement in appropriate follow up after discharge for patients with existing tracheostomies.

# IMPACT OF TRACHEOSTOMY ON LANGUAGE AND COGNITIVE DEVELOPMENT IN INFANTS WITH SEVERE BRONCHOPULMONARY DYSPLASIA

Betsy Cammack (M.D.)

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Background

More and more infants with severe bronchopulmonary dysplasia (BPD) who are ventilator-dependent with tracheostomy are now being successfully managed at home. The impact of tracheostomy on the cognitive and language development in these infants is not known.

Objective

To compare the cognitive and language outcomes among infants with severe BPD with and without tracheostomy for home assisted ventilation. We hypothesize that tracheostomy has an independent negative impact on the cognitive and language development in infants with severe BPD.

Design/Methods

This is a retrospective cohort study of de-identified data of infants born from Jan 1, 2010 to Dec 31, 2015, who received tracheostomy and assisted ventilation at <2 years of age. The cohort was compared to infants hospitalized within the same time frame with severe BPD but did not require a tracheostomy. The outcomes measured were cognitive, receptive, and expressive language scores at 2 to 3 years of age as determined by Bayley II or III scale of development.

Results

33 patients with tracheostomies and 29 patients without tracheostomies were analyzed. There was no significant difference in total language development between patients with trachs and those without (trachs 1.28, range 0.32-2.24, non-trachs 1.30, range 0.06-2.54). The analysis of cognitive development also showed no significant difference (trachs 1.30, range 0.06-2.54, non-trachs 1.91, range 0.84-2.98).

Conclusion

These data suggest that tracheostomy does not independently impact the language and cognitive development of infants with severe BPD. A multicenter collaboration with larger cohort is needed to further validate our findings.

# TONSILLECTOMY IN PATIENTS LESS THAN 3 YEARS-OLD: A CLOSER LOOK AT EPIDEMIOLOGY AND POST-OPERATIVE ADMISSION PRACTICES

Sallie Cataldo

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OBJECTIVE The current clinical practice guidelines from the American Academy of Otolaryngology released in 2011 recommends post-operative admission of patients less than 3 years-old undergoing tonsillectomy. This recommendation is based on both increased risk of post-operative respiratory compromise and cost-effectiveness of inpatient versus outpatient management. The purpose of this study is to evaluate how children's hospitals nationwide are adhering to the guidelines with the goal of identifying measures to further improve care.

METHODS The Pediatric-Health-Information-System provides data on 48 participating United States children's hospitals. Pediatric patients younger than 3 years-old who underwent tonsillectomy from 2010 to 2016 were identified by query.

RESULTS Between 2010 and 2016, 30257 tonsillectomies were performed in patients under 3 years-old (mean age 1.74±0.46 years). 12478 (41%) were inpatient and 17779 (59%) were outpatient. 62% were male, 60.3% white, 19.8% black, and 19.9% other. Outpatient tonsillectomy increased with age: 32.8% (0-11 months-old), 44.2% (1 year-old) and 63.6% (2 years-old). Comparing 2010 to 2016, there was a 76% decrease in outpatient tonsillectomy and a 60% decrease in total tonsillectomies. There was wide variability between institutions in total tonsillectomies performed as well as inpatient versus outpatient post-operative management. There were 9 (0.02%) mortalities, all of which were inpatient admissions.

CONCLUSION After the release of the 2011 guidelines, the incidence of outpatient tonsillectomy and overall tonsillectomy has decreased in children under 3 years-old. There is still variability in admission practices among institutions nationwide. Further study of individual institutional and practitioner admission guidelines would help to understand the identified variabilities.

# DIET AFTER TONSILLECTOMY: ARE RESTRICTED DIETS NECESSARY IN PEDIATRIC PATIENTS

Maxwell Bergman

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Background: Evidence for the necessity of diet restriction after pediatric tonsillectomy is sparse in the literature despite many institutions routinely recommend a diet restricted to soft foods after surgery

Methods: Randomized controlled trial of patients aged 3-12 years undergoing tonsillectomy with or without adenoidectomy. Patients were randomized to either follow a postoperative diet consisting of soft foods only for 10 days or a non-restricted diet. Outcomes were assessed at the first postoperative visit. Primary outcomes were pain and bleeding. Secondary outcomes included return of normal diet and activity, pain medication usage, weight change, and days missed from school.

Results: (Note: patients are currently being enrolled and data will be updated accordingly for any presentation) Seventeen patients were enrolled over approximately 3 months. Ten patients were randomized to follow the standard soft diet and 7 patients were randomized to follow a non-restricted diet. The average age was 7.6 years. Mean pain score at the first postoperative visit was less than 1. There were more nighttime awakenings and more days of opioid use reported in the standard soft diet group compared with the non-restricted group (6 vs. 2.2 nights and 4.3 vs. 2 days, respectively). Patients on average missed 1 week of school, though the mean number of days missed was slightly higher in the standard soft diet group (7.5 vs. 6.6 days).

Conclusions: The preliminary results of this study suggest that postoperative outcomes are not influenced by diet in any clinically significant way.

# THE USE OF DRUG-INDUCED SLEEP ENDOSCOPY OR CINE MRI TO GUIDE ADENOTONSILLECTOMY IN PEDIATRIC OBSTRUCTIVE SLEEP APNEA.

David R. Lee (M.D.)

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Objective: For children with obstructive sleep apnea (OSA) without obvious tonsillar hypertrophy, drug induced sleep endoscopy (DISE) and/or cine MRI are increasingly used to identify treatment options. The aim of our study was to assess polysomnographic (PSG) outcomes for patients who underwent adenotonsillar surgery after a DISE or cine MRI.

Design: Single-institution retrospective case series.

Methods: Medical records were reviewed of children under the age of 18 who underwent a DISE or cine MRI at a pediatric children's hospital and underwent either adenoidectomy, tonsillectomy, or adenotonsillectomy. We excluded those without a preoperative and postoperative PSG.

Results: We assessed 132 children (33% female, 72% white race) with a mean age of 10.1 /-4.8 years (range 1-18); 59 (45%) had severe OSA, 40 (30%) had moderate, 33 (25%) had mild. Comorbidities were common with 58% having craniofacial disorders including 35% with Down syndrome. Adenoidectomy (primary or revision) was performed for 77 (58%), tonsillectomy for 8 (6%), and adenotonsillectomy for 47 (36%). There was no difference in resolution rates between techniques with oAHI<1 at 21%,25%, and 13%, respectively(P=0.47); oAHI,5 was 61%, 50%, and 3%, respectively. There was no difference by age, race, or gender in surgical technique selected (P=0.18-0.54).

Conclusion: When DISE or cine MRI suggest that adenoidectomy, tonsillectomy, or both should be carried out in children without tonsillar hypertrophy, resolution rates were 19% for an oAHI<1 and 58% for an oAHI<5. The use of these studies can be helpful to identify patients who will have a successful response to adenotonsillar surgery alone.

# THE ECONOMIC IMPACT AND RESOURCE UTILIZATION OF SURGICAL INTERVENTION IN REFRACTORY OBSTRUCTIVE SLEEP APNEA

Nicholas Kuhl

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Introduction: There have been no publications regarding the economic impact of adenotonsillectomy (T&A) used to treat refractory obstructive sleep apnea (ROSA). This study investigates total costs and resource utilization associated with success and failure of T&A.

Methods: Pediatric patients under 18 years who were treated for ROSA between 2008-2017 were included. Variables assessed were clinical characteristics and co-morbid diagnoses. Cost data included hospital and professional costs associated with T&A, drug-induced sleep endoscopy (DISE), hospitalization post-T&A, outpatient clinic visits, and polysomnograms. Pearson chi-squared tests assessed categorical variables; Wilcoxon rank-sum, Kruskal-Wallis, and Spearman assessed continuous variables. Statistical significance was set a priori at $p<0.05$.

Results: One hundred and forty-eight patients met inclusion criteria. Ninety-five (64%) underwent re-intervention via DISE. Younger age at T&A ($p=0.032$), greater OSA severity ($p=0.005$), decreased body mass index (BMI) ($p=0.025$), higher inpatient care level ($p<0.001$), and longer length of stay ($p<0.001$) were associated with increased total cost. Gender, race, ethnicity, and co-morbid medical conditions did not have a significant impact on total cost. Number of outpatient ENT clinic visits were increased by lower BMI ($p=0.049$) and younger age ($p=0.016$). The number of polysomnograms increased as the severity of OSA increased ($p=0.034$), and longer LOS was associated with decreased BMI ($p=0.008$) and age ($p<0.001$). Higher level of hospital care was associated with greater severity of OSA ($p<0.001$), younger age ($p<0.001$), and male gender ($p=0.041$).

Conclusion: These findings provide additional justification for the increased total cost and resource utilization incurred by hospitals wherein more complex ROSA patients are seen.

# INCIDENCE OF FISTULA FORMATION AND VELOPHARYNGEAL INSUFFICIENCY IN EARLY VERSUS STANDARD CLEFT PALATE REPAIR

Stephen Hadford

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Purpose:

To compare the rates of fistula formation and velopharyngeal insufficiency (VPI) in early versus standard cleft palate repair.

Methods:

A retrospective chart review from a single institution identified 668 patients treated for cleft palate from 2000-2017. 219 patients met the inclusion criteria of having initial palatoplasty at the institution and sufficient medical record data to permit assessment for post-operative complications. Data collected included age at repair, patient-specific factors, and incidence of both post-operative fistula formation and VPI. “Early repair” is defined as palatoplasty before 6-months of age. “Standard repair” is palatoplasty at or beyond 6-months old.

Summary of Results:

Of the patients reviewed, 110 had early repair and 109 had standard repair. Rates of fistula formation were found to be significantly higher in early repairs (chi-square statistic 9.0536, p-value = 0.0026). Development of VPI was not significantly different between the two groups (chi-square statistic 1.2068, p-value = 0.27196). Incidence of post-palatoplasty VPI was significantly higher in patients who had a post-operative fistula when compared to those who healed without fistula formation (chi-square statistic 4.3627, p-value = 0.0367).

Conclusions:

There is significant debate in the literature regarding the optimal timing of cleft repair to maximize speech outcomes and minimize risks. Our data shows that post-operative fistula formation occurs at a significantly greater incidence when performed on patients prior to six months old. Furthermore, while the rate of VPI was not significantly affected by age at time of surgery, it was significantly higher in those who experienced a post-operative fistula.

# IMPLEMENTATION OF AURICULAR MALFORMATION SCREENINGS IN THE NEWBORN POPULATION

Barbra Novak (PhD)

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BACKGROUND AND OBJECTIVE:

Research has shown that it is important to initiate ear molding early for children with auricular malformation in order to achieve the best result. Currently our institute relies on the traditional primary care physician (PCP) referral system which does not recognize the time sensitivity of the visit in patients with auricular malformation. The purpose of the current research is to implement a new screening protocol for identifying auricular malformations in the newborn population and thus expedite the clinic visit.

METHODS:

The hearing screen technicians (HSTs) were trained to identify some of the most common auricular malformations. A picture guide of 11 types of auricular malformations were given to the HSTs to use as a reference. At the time of the newborn hearing screen, the HSTs examined the pinnas of each baby. When an auricular malformation was identified, the auricular malformation team was immediately alerted and a bedside consultation with ENT occurred.

RESULTS: Comparison was made of the referral rate between pre- and post-implementation of the protocol which showed an increased rate of identification (five referrals in the 12-month period pre-implementation versus eight referrals in the 3-month period post-implementation).

CONCLUSION:

We successfully implemented an auricular malformation screening protocol that was linked to newborn hearing screenings. The frequency of identification has increased with the implementation of the new screening protocol and has resulted in earlier initial ENT consultations for ear molding with the goal of improving patient satisfaction and results.

# RENAL ANOMALIES IN MICROTIA AND CONGENITAL AURAL ATRESIA PATIENTS AT A TERTIARY PEDIATRIC CENTER - AN UPDATE

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INTRODUCTION: Microtia and congenital aural atresia (CAA) are severe pediatric conditions that hinder quality of life. Microtia is associated with renal abnormalities and several genetic syndromes involving the kidneys. Current guidelines suggest performing a renal ultrasound with the presence of preauricular pits and ear anomalies associated with dysmorphic features but not isolated microtia and atresia.

OBJECTIVE: To characterize the prevalence of renal anomalies among microtia and CAA patients at a tertiary pediatric center.

METHODS: An updated retrospective review of 237 children with microtia and/or CAA was conducted from 2001 through 2018. Patients were identified as syndromic or non-syndromic. Data included renal ultrasounds performed, anomalies found, and follow-up.

RESULTS: In the 237 patients, 221 (93%) had microtia and CAA, 14 (6%) had isolated microtia, and 2 (1%) had isolated CAA. 28 (8.5%) were syndromic, the most common being Goldenhar. 98 patients (41%) had documented renal ultrasounds, of which 14 (14%) were syndromic. Anomalies were found in 24/98 (24.4%) of patients screened and included disorders such as renal agenesis, pelviectasis, renal ectopia, hydronephrosis, vesico-ureteral reflux, and duplicated collecting systems. 18/24 (75%) of the patients with abnormal results were non-syndromic. 8/24 (33%) required nephrology follow-up for chronic kidney disease or renal failure. Of the syndromic patients, 14/28 (50%) had not received an ultrasound.

CONCLUSION: Children with microtia and CAA are at a significant risk of structural renal abnormalities, even when isolated outside of a genetic syndrome. We recommend the consideration of performing a screening renal ultrasound in all patients with microtia and CAA.

# SURGICAL MANAGEMENT OF CONGENITALLY ABSENT TRACHEAL RINGS

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Introduction - Congenital tracheal stenosis secondary to absent tracheal rings is a rare congenital anomaly that is difficult to manage both clinically and surgically. This typically manifests as severe segmental tracheomalacia and only isolated cases with short-term follow-up have been previously described.1-5 Our aim is to describe our case series of children with absent tracheal rings who underwent surgical management and had a long-term follow-up

Methods - This is a retrospective case series, examining all subjects who were identified with absent tracheal rings from 2002-2016. Electronic and paper medical records were queried to obtain demographics, age at diagnosis and surgery, pre and postoperative symptoms, pre and post airway size, procedure performed, length of follow up and any adjunctive procedures performed.

Results - Nine subjects were identified who underwent slide tracheoplasty for correction of congenital absent tracheobronchial rings. Age at diagnosis ranged from 10 days to 5 years of age (median - 4 weeks). Age at surgery ranged from 3 weeks to 5 years of age (median - 5 weeks). Six out of 9 subjects were extubated on postoperative day 1. Only one subject required additional intervention for treatment which included balloon dilation, tracheobronchial stenting and aortopexy to alleviate the obstruction. Mean follow up time was 5.89 years.

Discussion - This is the largest series of children with absent tracheal rings who underwent slide tracheoplasty with long-term follow-up presented to date. Slide tracheoplasty is an effective surgical intervention for the treatment of absent tracheal rings in infants and young children.

# MANAGEMENT OUTCOMES OF CRICOPHARYNGEAL ACHALASIA IN CHILDREN: A SYSTEMATIC REVIEW

Sama Alohali (M.D.)

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Purpose

To date, there is no consensus as to the optimal treatment of Cricopharyngeal Achalasia (CPA) in children. The objective of this systematic review is to study the reported outcomes of various treatment modalities such as myotomy, dilatation, and botulinum toxin injection.

Methods

A comprehensive search of Web of Science, Medline, Global Health, Embase, Cochrane, and Biosis databases was performed in June 2018 following the PRISMA protocol to identify all previously published articles reporting results of CPA management in children. Two authors independently selected eligible studies, assessed the methodological quality, and extracted the data.

Results

Of 734 potential articles, 35 studies met the inclusion criteria. A total of 84 patients aged less than 18-year-old who underwent myotomy (48.8%), dilatation (56%), or botulinum toxin injection (16.7%) were included in the analysis. A symptom-free period of at least 24 weeks was reported in 63.4% of cases after myotomy, 8.5% after dilatation, and 21.4% after botulinum toxin injection. Death was reported in 6.4% of cases after dilatation, 4.9% after myotomy, but none after botulinum toxin injection. Other complications included perforation, fistula formation, aspiration pneumonia, and respiratory failure.

Conclusion

This is the first review to systematically compare the effectiveness of myotomy, dilatation, and botulinum toxin injection for the treatment of CPA in children. The most effective treatment seems to be myotomy, while botulinum toxin injection appears to be the safest. Most studies failed to report long-term outcomes. The authors recommend botulinum toxin injection as first-line treatment and myotomy in case of failure.

# THE IMPACT OF AGE AND TYPE OF FRENULECTOMY IN INFANTS WITH ANKYLOGLOSSIA ON PERCEIVED MATERNAL BREASTFEEDING BENEFITS

Lyuba Gitman (M.D.)

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Ankyloglossia is a common condition that can cause difficulty breastfeeding, yet optimal timing for intervention remains unclear. The purpose of this study was to investigate whether an immediate or delayed correction of ankyloglossia correlated with improved breastfeeding. The secondary outcome was to observe whether results varied based on type of frenulectomy performed. This was a prospective case series from a tertiary care center, which examined maternal feedback on infants who received frenulectomy for ankyloglossia and/or upper lip frenulum. The participants' mothers were given a pre-procedure survey to assess breastfeeding difficulty (notably pain, efficiency, skin/nipple breakdown). Frenulectomy was performed in clinic with sharp scissor dissection. Mothers filled out an immediate post-procedure questionnaire and final follow-up survey a few weeks to months later. A linear mixed model was used to compare mean pain and efficiency scores. Fifty subjects participated from 9/2017 to 6/2018. Four participants were excluded due to inability to continue breastfeeding and failure to complete surveys. Mean pain scores (scale 1-10) reduced by 3.14 and 3.61 points from pre-procedure to immediately post-procedure, and to 3rd survey, respectively; both values were statistically significant (P values <0.001). Mean efficiency score (scale of 1-10) increased by 1.44 points (from 4.64 to 6.08) and by 2.56 points when comparing pre-procedure survey to 2nd survey and 3rd survey, respectively. Both values of improvement were statistically significant (P values <.001). However, age at frenulectomy (<4 weeks vs >4 weeks) and type of frenulectomy, were not significantly related with pre- and post-pain scores or efficiency score improvements.

# PEDIATRIC MODIFIED BARIUM SWALLOW: IMPACT OF STANDARDIZED FLUOROSCOPIC VISUALIZATION PROCEDURES ON RADIATION EXPOSURE

Kathryn H. Bradburn

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Objective: To compare ionizing radiation exposure in the pediatric population undergoing modified barium swallow studies (MBSS) before and after the implementation of a standardized fluoroscopic visualization procedure.

Methods: Videofluoroscopic swallow studies were retrospectively reviewed between 2011-2017 for fluoroscopy time and dose. Ionizing radiation exposure during non-standardized clinician determined procedural approach was compared to exposure after the initiation of a standardized procedural protocol. The standardized protocol included the fluoroscopic examination of five sequential swallows at four standardized timepoints (min:sec) 00:00, 00:30, 01:30, 02:30 using thin liquids, followed by the provision of compensatory interventions as clinically indicated by the examining Speech-Language Pathologist.

Results: 270 patients were identified and included in the investigation. The median total radiation dose that patients received during the standardized MBSS protocol was 4.42 mGy (IQR: 3.02 - 7.59 mGy) and the median study time was 1.70 minutes (IQR: 1.20 - 2.22 minutes). Nonparametric t-tests showed a significant decrease in radiation dose ($P < 0.05$) and total fluoroscopy time ($P < 0.05$) compared to MBSS performed prior to protocol implementation.

Conclusion: The new MBSS protocol aims to minimize radiation exposure and maximize diagnostic yield during modified barium swallow studies. We found that capturing swallows with fluoroscopy at four standardized time points decreases study duration and effective dose.

# EFFECT OF FLUID RESTRICTING BOTTLE NIPPLES ON INFANT AIRWAY PROTECTION

Katlyn Elizabeth McGrattan (PhD)

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Purpose: Infant dysphagia is commonly treated through the use of fluid restricting bottle nipples. Despite the use of this intervention in the clinical arena, limited evidence exists to support its effectiveness as a dysphagia treatment modality. The aim of the current investigation was to test the difference in rates of bolus airway entry (penetration/aspiration) between Dr. Brown's® Preemie and Level 1 bottle nipples.

Methods: 23 infants (39 0/7-50 2/7 PMA) referred for videofluoroscopic swallow studies were provided thin Varibar® barium contrast from a Dr. Brown's® Preemie and Level 1 bottle nipple. Exams were completed using continuous fluoroscopy recorded at 30 frames/second on a TIMS high resolution recording system. Swallows were reviewed frame-by-frame for the presence of penetration or aspiration. Differences in rates of penetration and aspiration were compared using a paired t-test with results reported as mean ± standard deviation.

Results: Although no significant differences were observed in the occurrences of penetration between Preemie and Level 1 nipples (preemie 0.83 ± 1.03, level 1 0.57 ± 0.84, p= 0.299), infants exhibited significantly fewer occurrences of aspiration on preemie nipples (0 ± 0) when compared to level 1 (0.61 ± 0.78) (p=0.00012).

Conclusions: Infants exhibited improved airway protection while drinking from the preemie nipple when compared to a level 1. Future randomized investigations are necessary to better understand this intervention's full treatment effect.

# A QUALITATIVE STUDY TO EXPLORE BARRIERS OF EARLY SWALLOWING DYSFUNCTION (SD) DIAGNOSIS ON OTHERWISE HEALTHY INFANTS.

Abdulsalam Baqays (M.D.)

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Introduction:

SD is an under-reported condition among otherwise healthy infants. Nonetheless, parents of these infants still struggle until the diagnosis is made. Identification of parental perception of factors that prevent early diagnosis of SD may help. This study captures patients' reports about barriers of early SD diagnosis and related consequences.

Method:

An inductive qualitative study was done using detailed semi-structured in-person interviews with four experienced families. Audiotaping and verbatim transcription were performed. A thematic analysis approach was utilized.

Result:

Infants with SD presented with continuous coughing, choking, cold-like symptoms, and being constantly sick; or even feeding difficulties. They had been managed with antibiotics multiple times or diagnosed with the allergy, asthma, or recurrent viral infections before thinking about SD. From the parents' insight, we built a framework that composed of five themes and 23 subthemes specific to the barriers, and two related consequences. These themes are difficult access to proper healthcare resources, parent-related barriers, physician-related barriers, prioritizing differences between physicians and parents, and the presence of several false beliefs about SD among families and physicians. These barriers had severely impacted on the parents causing significant reduction in work productivity, exposure to work-related reprimands, and changes in the family dynamics. Regular hospital attendance and development of an advanced condition that warrant emergency interventions or admission were the primary consequences on the infants.

Conclusion:

Disregard to parental observations, and lack of knowledge and appropriate assessment skills for SD were the commonly cited barriers for early diagnosis of SD in infants.

# SWALLOWING FUNCTION IN CHILDREN STATUS POST ESOPHAGEAL REPLACEMENT

Claire Kane Miller (PhD)

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## Purpose

Dysphagia is a prominent issue for children with esophageal dysfunction secondary to esophageal anomalies such as congenital long gap esophageal atresia/tracheoesophageal fistula, or caustic ingestion. Esophageal replacement techniques may be used when preservation of the native esophagus is not possible. The purpose of this study was to explore swallowing characteristics viewed via videofluoroscopy in a small cohort of patients following esophageal replacement at Cincinnati Children’s Hospital Medical Center.

## Methods

The videofluoroscopic swallowing studies of 7 pediatric patients status post esophageal replacement were retrospectively reviewed. The Modified Barium Swallow Impairment Scale© standardized ratings (per speech pathology and radiology) were used for analysis of swallowing parameters viewed via videofluoroscopy.

## Results

The majority of patients were accepting at least a partial amount of oral intake at the time of their post-operative video swallow studies. Of note, oral transfer and swallowing onset was within the normal range. Pharyngeal clearance was incomplete in 50% of the patients following the first swallow as judged via the MBSImP standardized rating. In the majority, the amount of amount of bolus remaining in the pharynx was minimal, with the exception of one patient who had severe pharyngeal retention despite repeated swallowing efforts. Compensatory swallowing strategies such as effortful swallow, and alternating solid and liquid boluses were effective in improving pharyngeal and esophageal clearance.

## Conclusions

Videofluoroscopic analysis of swallowing parameters has utility in differentiating the functional parameters of the swallow following esophageal replacement, as well as the effects of compensatory strategies on the physiology of the swallow.

# THE EFFECT OF SOCIOECONOMIC STATUS ON PEDIATRIC COCHLEAR IMPLANT USAGE

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Consistent cochlear implant usage is critical for optimizing speech and language development. This study evaluated the associations between proxy measures of socioeconomic status and usage of cochlear implants.

Usage data (hours/day) was extracted from the audiology records of patients, aged 0-26 years, seen at our children's hospital from 2002-2017. Associations between usage and demographic factors such as race, housing data, insurance type, poverty level and median household income for zip code were assessed. Logistic and linear regression was performed to determine the effects of demographic variables on the duration of cochlear implant usage.

There were 143 total patients with 73 having usage data for both ears. Average implantation age was 6 years old with an average last visit age of 12 years old. Patients living in a zip code with a greater percentage of residents below the poverty level received their implants at younger ages (B=-.29,t(138)=-2.00,p=.047), and those with a lower median household income spent more time listening to quiet each day (B=-1527.95,t(138)=-2.19, p=.030). Children living in school districts that had more students qualifying for reduced-price meals were more likely to spend time listening to quiet (B=6.362 t(133)=2.31,p=.022), and spend time listening to speech in noise (B=7.911,t(133)=2.10,p=.037). Older children had greater total usage, time spent listening to speech in noise, time spent listening to quiet, and less time with the coil off, $p<.05$.

In conclusion, patients with lower socioeconomic status receive their cochlear implants at younger ages but spend more time using their implant each day, particularly in quiet conditions.

# VESTIBULAR AND BALANCE FUNCTION IN CHILDREN WITH SINGLE SIDED DEAFNESS DUE TO ABNORMAL DEVELOPMENT OF THE COCHLEAR NERVE

Patricia L. Purcell (M.D.)

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Objective: Abnormal development of the cochlear nerve has been well-characterized as a common etiology of single-sided deafness; this study is among the first to evaluate vestibular and balance function in these children.

Methods: Thirteen children with unilateral cochlear nerve aplasia/hypoplasia underwent standardized balance testing at a pediatric tertiary care center. Testing was performed using the balance subtest of Bruininks Oseretsky Test of Motor Proficiency (BOT-2). One sample t-test compared results from sample population to mean age and gender standardized score of 15. Any additional vestibular testing results were also reviewed.

Results: All children had single-sided deafness. Age at time of testing ranged from 4 to 19 years. Ten of the children (77%) performed at or below the mean of 15 on BOT-2 testing; however, 3 children were high-performing with scores of 22 or higher. Overall mean BOT-2 score was 12.8 (SD=6.6); while lower than standardized mean of 15, this difference was not statistically significant, p-value 0.25. Median BOT-2 score was 13, (range 4-24). Of 4 children with vestibular testing results, 2 did not demonstrate vestibular dysfunction, 1 had mildly reduced caloric testing in affected ear, and 1 had multiple abnormal findings.

Conclusion: Children with cochlear nerve aplasia/hypoplasia may be at greater risk of vestibular and balance dysfunction, although some are able to demonstrate a high level of performance. Complete assessment of the sensory deficits in children with unilateral hearing loss is important as we continue to define outcomes and measure response to interventions in this group.

# ENDOSCOPIC MYRINGOPLASTY WITH BIODESIGN AND TISSEEL GLUE IN CHILDREN- A MINIMALLY INVASIVE INCISIONLESS TECHNIQUE

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A novel and innovative technique of incisionless myringoplasty in children is described. Tympanic perforation is common amongst children, causing otorrhea, hearing loss and avoidance of swimming. Routinely children need to wait until they are older and then may decline surgery if phobic. The technique described offers an alternative to traditional 'open' myringoplasty and can be performed bilaterally

Results

From July 2016 to July 2018 a total of 36 procedures have been performed, age range 7 to 16 years old. To date 28 patients have been followed up.

Successful closure was achieved in 80% of patients with a perforation less than 50% area of tympanic membrane, but not successful in larger perforations, However, in the failures an average reduction of over 70% of the original perforation size was achieved; and children’s symptoms improved.

Without packing the children reported a subjective hearing gain on awakening post-anaesthesia.

No significant morbidity and no complications were reported, all children able to return to school within 1 to 2 days.

Conclusion

Endoscopic myringoplasty with biodesign and tisseel is a safe minimally invasive technique, which can be offered to younger or phobic children.

# "FAST TRACK" EAR TUBES: A QUALITY IMPROVEMENT PILOT PROGRAM TO IMPROVE ACCESS TO CARE

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Purpose: Tympanostomy tube (TT) placement is the most common ambulatory surgery performed on children in the United States, with approximately 667,000 children younger than 15 years of age receiving tympanostomy tubes each year. Often times, patients who have had frequent ear infections want to have ear tubes placed in a timely manner to avoid further antibiotic therapy. The purpose of this quality improvement pilot program was to initiate a "fast track" ear tube program for patients who meet criteria for TT placement for recurrent ear infections, improving access to care and care efficiency.

Methods: Two large pediatric primary care offices were chosen to participate in the program. Patients who were referred for "fast track" ear tubes were seen by one designated nurse practitioner; these patients were then scheduled for TT placement with one designated surgeon 48 hours later. A parent satisfaction survey was administered at the postoperative visit, and a referring provider survey was sent after conclusion of the program.

Results: Survey responses from parents were overwhelmingly positive regarding this program. Parents noted that they were happy with the process, did not feel rushed into surgery, and were satisfied with the care and education received from both the nurse practitioner and the surgeon. Referring providers also were highly satisfied with this program.

Conclusion: "Fast track" ear tubes for patients who meet surgical criteria for TT placement is a model of care that is feasible to administer and can improve efficiency of care for patients with otitis media.

# HEARING OUTCOMES IN PEDIATRIC OSSICULOPLASTY: COMPARING POSTAURICULAR APPROACH TO TOTAL ENDOSCOPIC EAR SURGERY

Patricia L. Purcell

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Objective: This study compares hearing outcomes after ossiculoplasty in children who underwent postauricular approach with a microscope to those who underwent transcanal total endoscopic ear surgery (TEES). When performing TEES, surgeons are limited to one-handed techniques for manipulation of instruments. It is important to determine whether this restriction impacts placement of ossicular prosthesis and affects hearing outcome.

Methods: Hearing outcomes were reviewed from children who underwent ossiculoplasty with titanium total ossicular replacement prosthesis (TORP) after previous cholesteatoma surgery. Data were acquired prospectively by a single surgeon at a pediatric tertiary care center. The primary outcome measure was the number of ears with normal hearing 2 months postoperatively, defined as air bone gap (ABG) 4-frequency pure-tone average ≤25dB.

Results: Complete data were available for 41 children: 22 underwent postauricular approach (mean age 12 years; preoperative ABG 42dB); 19 underwent TEES (mean age 13 years; preoperative ABG 43 dB). There were no significant differences in age or preoperative hearing thresholds. Ten (45%) of the 22 ears receiving microscopic surgery achieved normal hearing threshold, compared with 12 (60%) of the 19 ears treated with TEES. This difference was not significant (X2 test, $p = 0.3$). Mean postoperative ABG for children after microscopic surgery and TEES were 26dB and 23dB, respectively (not significant: Student's t-test, $p = 0.4$).

Conclusion: Hearing outcomes for pediatric ossiculoplasty were similar when comparing microscopic surgery to TEES. For a surgeon familiar with TEES, one-handed technique does not appear to negatively impact TORP placement. This lower morbidity approach appears justifiable.

# BENEFITS AND CHALLENGES TO WORKING MEMORY AND ACADEMICS IN CHILDREN RECEIVING EARLY SIMULTANEOUS BILATERAL COCHLEAR IMPLANTS

Claire McSweeny

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Purpose: To determine whether children who receive bilateral cochlear implants simultaneously exhibit deficits in working memory and to examine potential academic implications.

Background: The overarching goal of bilateral implantation in children is to provide access to sound and reduce listening effort. Children with unilateral cochlear implants develop age-appropriate oral speech and language but they have decreased phonological awareness and working memory capacity. These impairments have implications for academic achievement. We hypothesize that providing children with bilateral cochlear implants simultaneously reduces risks of working memory deficits with benefits for mathematics and reading.

Method: Twenty-eight children who received bilateral cochlear implants at a mean age of 2.21(SD=1.37) were tested at 8.40(SD=2.35) years of age. Measures included tests of working memory (digit span, the Corsi block tapping test and dot matrix test) and academic performance (four WIAT-III subtests). The same measures were tested in a group of 11 normal hearing peers at 8.64(SD=1.56) years of age.

Results: Accounting for improvements with age (linear regression), children with bilateral cochlear implants achieved similar results as their normal hearing peers on the digit span and dot matrix tests ($p>0.05$) but had poorer scores on the Corsi block test ($p<0.05$). Academic measures were similar to normal ($p>0.05$) with the exception of pseudo-word decoding and mathematic ($p<0.05$).

Conclusions: The impact of bilateral deafness on working memory in children may be reduced but not entirely eliminated by providing early access to bilateral cochlear implants. Reading appears to be on par with normal hearing peers but mathematics remains slightly delayed.

# TYMPANOSTOMY TUBE PLACEMENT IN CHILDREN WITHOUT GENERAL ANESTHESIA

Franklin Rimell (M.D.)

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Background: Tympanostomy tube placement (TT) is the most common surgical procedure in the U.S.. Given recent concerns regarding exposure of general anesthesia to children and in attempt to reduce cost we have begun the process of evaluating other approaches by using our pediatric sedation unit and a single pass delivery device to aid in TT placement.

Methods: Sedation procedures occurred in a dedicated pediatric sedation unit where nitrous oxide and intranasal fentanyl were delivered. Following cerumen removal, phenol was placed topically on the tympanic membrane and then a device was used to deliver a TT into both ears. Toward the end of the study some children were offered enrollment in a second approved study where the TT was delivered in the office with phenol only.

Results: There were 54 cases (108 ears) in one sedation unit and 10 cases (20 ears )in the office. All 64 cases were successful on postoperative follow up at 3 months with functioning TT. Total length of time for the procedure in the sedation unit was 9.8 minutes with an average length of stay of 79 minutes vs. 3.5 hours in the surgical unit.

Conclusion: TT placement can be successfully placed in children under two years of age with the avoidance of general anesthetic agents listed in the recent FDA warning and with reduced cost. Future experience will show that TT can be placed in younger children in the office and older children in the sedation unit with markedly reduced cost and greater parental convenience.

# USE OF A SMARTPHONE OTOSCOPE FOR TYMPANOSTOMY TUBE SURVEILLANCE

Lia Jacobson (M.D.)

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Background: Advances in technology have led to the development of a simple system that converts a smartphone into a digital otoscope for remote assessment of the tympanic membrane (TM) via HIPAA-compliant videos. This device may improve physician-patient communication and assist with diagnosis and management of ear disease.

Objectives: To evaluate the efficacy, feasibility and physician/parental satisfaction using the Cellscope® smartphone attachment (Oto) for at-home tympanostomy tube (TT) monitoring.

Methods: Videos of each TM were recorded by parents monthly and at any acute event post-TT. Comparisons were made between home-recorded videos and in-office otoscopy findings. Two independent physicians reviewed the videos and inter-rater concordance was calculated with a kappa coefficient. Satisfaction and usability questionnaires were administered to physicians and parents.

Results: 19 children (69% female) participated (mean age, 2.2 years). The average duration of device use was 16 months. A total of 206 videos were reviewed. Intra-rater agreement demonstrated excellent agreement (95.2% concordance). There was good inter-rater agreement for most otoscopy findings. Tube placement demonstrated substantial agreement (Cohen’s kappa 0.65); tube patency, extrusion, blockage and presence of otorrhea demonstrated moderate agreement (Cohen’s kappa 0.48, 0.59, 0.49 and 0.58 respectively). For acute events, over 50% of parents used the device in place of a primary care or urgent care visit. Physicians and parents expressed satisfaction and ease of use with the Oto device.

Conclusions: Oto is effective and feasible for use in TT surveillance. Smartphone otoscopy may allow otolaryngologists to follow a child’s TT remotely and reduce in-office and acute care visits.

# DOES VIDEO PATIENT EDUCATION HAVE AN EFFECT ON EAR TUBE SURGERY FOLLOW THROUGH?

Andrew Peace

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INTRODUCTION: Patients who are recommended ear tube placement do not always follow through with surgery. Timing is of essence in these patients since hearing deficits caused by persistent middle ear effusions (MEE) can have a long-lasting impact on speech and language development. A lack of follow through with surgery maybe partially due to a lack of understanding parents may have concerning the severity of the consequences of otitis media (OM) and MEE and the ease of its resolution. In this study we explore the effects of additional patient education in the form of animated videos.

OBJECTIVE: To determine if perioperative video education via text messaging can increase the number of patients following through with recommended surgery.

METHODS: After consent, parents are sent a text message with an educational video on ear tube surgery after their clinic visit for patients of four attending physicians. After a study period of 6 months, a retrospective chart review is done to assess how many patients were recommended surgery versus how many actually follow through. This is then compared to the non-study period and to non-study attending physicians. Video usage analytics for the study participants are also reviewed.

RESULTS: Preliminary data shows an increase of 16% in ear tube surgeries for the study physicians. 87% of study participants who watched the video proceeded with surgery compared to 83% of those who received but did not watch the video.

CONCLUSION: Additional perioperative video education delivered via text message preliminarily shows an increase ear tube surgery.

# DEVELOPMENT OF AN EMERGENCY ROOM AND OTOLARYNGOLOGY CLINIC MANAGEMENT ALGORITHM FOR EAR FOREIGN BODY REMOVAL

Linda Brock (RN)

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Objective: Ear foreign body (FB) removal is a commonly performed procedure in the emergency center (EC). In the Otolaryngolgoy clinic we encounter multiple instances related to complications from adhesive usage for attempted FB removal in the EC. We therefore sought to improve the process at our institution by developing an ear FB management algorithm between the EC and Otolaryngology clinic to decrease complications.

Method: A literature review and survey study performed in a tertiary referral center resulting in the development of a management algorithm and educational material for the EC physicians.

Result: Fifty-two surveys to the EC care providers were sent out with an 84% return rate (n=44). The result showed 70% (n=31) of the providers are familiar with the adhesive method while 52% (n=27) had tried it despite no supportive evidence and 90% reported learning this method through colleagues. Literature review showed a discrepancy in the recommended method for FB removal and success rate depending on the origin of published articles (Emergency Medicine (EM) vs. Otolaryngology journals). The EC and Otolaryngology departments collaborated to develop a new management algorithm with subsequent improved efficiency and decreased complications for ear FB removal. In addition, an educational course and training session was developed for the EC.

Conclusion: There are inconsistent validated recommendations of instruments and techniques in the emergency literature for ear FB removal. We identified appropriate instruments and safe techniques for ear FB removal within the emergency setting resulting in decreasing complications.

# SURVEY ANALYSIS OF QUALITY OF LIFE AFTER RECONSTRUCTIVE SURGERY IN PEDIATRIC MICROTIA PATIENTS

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Introduction: Microtia is an uncommon congenital abnormality involving the incomplete development or complete absence of the pinna. Due to the cosmetic nature of microtia, reconstructive surgery is often recommended and completed between 6 to 10 years of age. However, repair of microtia is complex and outcomes vary widely. This study aims to review the complication rate and patient satisfaction using a validated survey in children with microtia reconstruction.

Methods: Patients were identified by CPT codes for microtia and reconstruction surgery at a tertiary referral center over a 7-year period. Charts were reviewed for demographic information, post-operative complications, and other surgical outcomes. A validated quality of life survey assessed physical, emotional, social, and school functioning within the past 1 month. A score of 0 suggested no problems while 92 implied the patient almost always has problems. The data was analyzed for a correlation between patient satisfaction via a high quality of life report and surgery complications.

Results: 7 of 109 patients, 4 boys and 3 girls, were identified via CPT codes for microtia with repair surgery, with an average age during surgery of 9 years. Median length of post-operative stay was 6 days with no readmissions within 30 days and no major post-surgical complications noted. Of the responses received from the quality of life survey, the average score was a 10.6 with 60.4% of problems occurring in school functioning.

Conclusion: Microtia reconstruction has had minimal surgical complications and correlates with a high quality of life in pediatric patients.

# A CASE OF SUPRAGLOTTIC INVOLVEMENT IN CROHN'S DISEASE

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Introduction: Upper airway manifestations of Crohn's disease are rare with the most common being tracheobronchitis. Supraglottic manifestations of Crohn's disease have not previously been described.

Case description: A 12-year-old male with suspected Crohn's disease with primarily extraintestinal features, currently on mercaptopurine, was referred to Otolaryngology for evaluation of dyspnea on exertion and intermittent audible breathing. In-office nasopharyngolaryngoscopy was notable for bulky, pale arytenoids and thickened epiglottis. He underwent microlaryngoscopy and bronchoscopy (MLB) with biopsies of abnormally thickened and pale aryepiglottic folds. Pathology revealed squamous mucosa with mild inflammatory infiltrate and subjacent fibrovascular tissue with dense chronic inflammatory infiltrate, numerous plasma cells, and microcalcifications with clustered macrophages resembling granuloma. This pathology strongly supported the diagnosis of extra-intestinal Crohn's disease, and he was started on systemic mesalamine. Airway symptoms progressed and he was taken for MLB with injection of 20mg (0.5mL) Kenalog into fibrotic supraglottic tissue. Postoperatively, he reported improvement in symptoms, and exam revealed improvement in supraglottic fullness at one month. He underwent MLB with injection of 40mg (1mL) Kenalog at a three-month interval, again with postoperative improvement in symptoms and exam. Intestinal biopsy confirmed the diagnosis of Crohn's disease one year later. He has had no return of airway symptoms in a five-year follow up period.

Discussion: Here we present a case of symptomatic upper airway obstruction secondary to isolated supraglottic involvement of Crohn's disease. Inflammation resolved with intralesional steroid injection, obviating the need for systemic steroid treatment, which has been described in cases of Crohn's disease involving the airway.

# UTILIZATION OF ANCILLARY SERVICES FOR VOICE AND SWALLOWING OUTCOMES FOLLOWING CARDIOTHORACIC SURGERY

Ari G Mandler

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Objective: We sought to determine the utilization of otolaryngology and speech-language pathology services among infants following open cardiothoracic procedures. Given their susceptibility to iatrogenic-induced vocal cord dysfunction, the proper use of ancillary services is critical in limiting long-term swallowing and speech complications.

Methods: A fourteen-month retrospective review of the Society of Thoracic Surgeons database at a single tertiary children's hospital was performed to include all infants undergoing open cardiothoracic surgery. Demographic, operative, and outcome variables were identified.

Results: 67 patients were identified (36M/31F; mean [SD] gestational age 36.5 [4.5] weeks) whose surgeries included patent ductus arteriosus ligation (n=20), aortic operation (n=10), Tetralogy of Fallot repair (n=8), and Norwood procedure (n=6). Post-operatively, 20/67 patients (30%) had documented weak cry or weak cough; of these, 10 were evaluated by otolaryngology and 7 were found to have vocal cord paralysis. 30/67 patients (45%) had post-operative feeding difficulties and 13 had documented aspiration. Of the 13 with aspiration, 5 underwent a modified barium swallow with speech-language pathology and 5 had otolaryngology consultations. On discharge, 13/67 patients (19%) necessitated gastrostomy tube placement and 8/67 patients (12%) required supplemental oxygen or continued ventilation. Following discharge; 8/67 patients (12%) were followed by otolaryngology for vocal cord paralysis (n=7), persistent dysphagia (n=3), or stridor (n=2).

Conclusion: Open cardiothoracic surgery in infancy is associated with high rates of voice and swallowing dysfunction. Inpatient ancillary services are under-utilized in managing this population, and outpatient followup is poor suggesting the need for prospective protocols to ensure adequate care.

# REAL-TIME ALERT TRACKING FOR TRACHEOSTOMY DEPENDENT PEDIATRIC PATIENTS

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Introduction: Tracheostomy patients have high morbidity and mortality rates with frequent doctor and hospital visits. Methods: A novel method was developed to prospectively follow tracheostomy patients' inpatient and outpatient encounters using real-time email alerts at a tertiary children's hospital. All patients with a diagnosis of tracheostomy dependence from ages 0-26 were included. Data collected included reason for tracheostomy, comorbidities, inpatient and outpatient information, and tracheostomy-related complications, analyzed with linear and logistic regression.

Results: There were 141 patients with tracheostomies for 72 days of inpatient data and 51 days of outpatient data and a total of 304 alerts. 57(18.8%) alerts were encounters in the ED and 216(71.1%) alerts were outpatient clinics. Patients were an average of 9 years old with an average tracheostomy placement age of 4 years. Chronic respiratory failure, 58(41.4%), and ventilator dependence, 68(47.1%), were the most prevalent reasons for tracheostomy with a congenital syndrome present in 65(46.4%) patients. Patients most frequently visited the ENT, 50(18.5%), and pulmonology, 33(15.3%), outpatient departments. While controlling for age at encounter, patients who had a tracheostomy for a shorter period of time reported increased secretions more times, (B=-2.99,t(28)=-2.29,p=.030). 18(31.6%) ED visits were from patients who went to the ED for a tracheostomy-related problem and 31(54.4%) patients went to the ED with their tracheostomy as a secondary problem. Patients with tracheitis were more likely to be seen in the ED than in ENT outpatient, (OR:.09,95%CI:.03-.30,p<.001). Conclusion: Following tracheostomy patients will identify complications to better prepare both health professionals and caregivers, leading to decreased costs.

# LARYNGOTRACHEOESOPHAGEAL CLEFTS IN CHILDREN WITH DOWN SYNDROME: AN IMPORTANT CONSIDERATION FOR SWALLOWING DYSFUNCTION

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Introduction: Swallowing dysfunction is common in children with Down syndrome (DS), but often goes unrecognized. Aspiration or penetration has been reported in 50-60% of patients with DS. Laryngotracheoesophageal clefts (LTCs) can be associated with swallowing dysfunction, but their occurrence in children with DS has not been well studied. Our primary objective was to describe children with DS who underwent repair of LTC.

Methods: Retrospective review of medical records of patients with DS who underwent repair of LTC at a quaternary pediatric hospital from 2000-2018. Demographics, cleft grade, presence of bronchiectasis, aspiration/penetration status from either Video Swallow Study (VSS) or Fiberoptic Endoscopic Evaluation of Swallowing (FEES), and number of lipid laden macrophages(LLM) in broncheoalveolar lavage (BAL) were recorded.

Results: 10 patients were included; 60% male. Type of cleft: 5 (50%) deep interarytenoid notch, 1 (10%) type I, and 4 (40%) type III; no type II or IV were seen. Most common history was recurrent pneumonia (PNA) in 6 (60%). Five (50%) patients had bronchiectasis on computed tomography (CT). Of 8 patients with pre-operative FEES or VSS, 7 (87.5%) demonstrated aspiration or penetration. One (10%) patient had >20 LLM on BAL.

Conclusion: Given the high occurrence of swallowing dysfunction in children with Down syndrome, it is important to consider LTCs, particularly deep interarytenoid notch, as a potential etiology. Approximately 5% of LTC repairs performed at our institution occurred in children with DS, 60% of whom presented with recurrent PNA. These data suggest a need for further study in this population.

# DOES INJECTION LARYNGOPLASTY IMPROVE OUTCOMES ON PATIENTS WHO HAVE TYPE 1 LARYNGEAL CLEFT REPAIR

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Purpose:

To evaluate if patients with type 1 laryngeal clefts (T1LC) treated initially with injection laryngoplasty (IL) followed by surgical repair (SR) have better swallowing outcomes compared to patients who immediately undergo SR.

Methods:

A retrospective chart analysis was performed examining pediatric patients with T1LC treated at a tertiary academic center. Patients who underwent both IL and SR (IL-SR) were compared to patients who underwent SR only. Data was collected regarding patient comorbidities, conservative therapy, and swallowing outcomes and analyzed using Fisher's exact and Mann-Whitney tests. Subjective and objective swallowing outcomes before and after SR were compared between the two groups.

Results:

Thirty-three patients were identified with 18 (55%) of IL-SR patients compared to 15 (45%) SR patients. Average age at diagnosis was 2.5 years. Patient comorbidities and use of conservative therapy were similar between the two groups ($p>0.05$). The most common presenting symptom was coughing and choking with feeds. IL-SR had increased rates of aspiration on pre-surgical swallow evaluation (56% vs 33%), but this was not significantly different ($p=0.128$). Patients in both groups improved in swallow symptoms (67% vs. 73%, $p=1$), and aspiration on post-surgical swallowing evaluation (0% vs. 20%, $p=0.058$). Average follow-up was 4 years, and 42% of patients had normal swallowing.

Conclusion

Patients identified to benefit from SR of their T1LC repair after IL had improved swallowing outcomes compared to those who underwent SR only, but this was not statistically significant. Further evaluation of which patients would benefit from immediate T1LC SR without initial IL is needed.

# A GEOMETRIC MODEL TO EXPLAIN THE BENEFICIAL IMPACT OF LINGUAL FRENOTOMY FOR ANKYLOGLOSSIA IN BREASTFEEDING WOMEN

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Purpose: Frenotomy for ankyloglossia in neonates is a well-tolerated intervention that improves breastfeeding quality. This investigation aimed to describe the breastfeeding experience and outcomes of the mother-infant dyad and propose a 2-vector analysis, based on a quadrilateral pyramid model, which could explain the improved genioglossus tongue muscle function after frenotomy

Methods: A 3-year retrospective analysis of mother-infant dyads referred for ankyloglossia, poor weight gain, or maternal breast pain while breastfeeding and underwent lingual frenotomy. Data in the medical records from office visits and telephone call follow-up with parents, as approved by the IRB, were collected. Data included demographics, primary symptom, prior breastfeeding experience, degree of ankyloglossia, complications, improvement (major vs. minor), continuation of breastfeeding, and parental recommendation of the procedure.

Results: We reviewed 460 infants. Sixty-two percent were males and the median age for frenotomy was 2 weeks of age. Four out of five (83%) of the mothers were breastfeeding their firstborn child. Most mothers (92%) reported improvement or resolution of the painful feeding around one week following the procedure. There were no complications. We postulate a geometric model to describe changes in the anterior projection and lift of the tongue following lingual frenotomy.

Conclusion: Lingual frenotomy is an intervention associated with high satisfaction for the mother and infant and can be anticipated when genioglossus muscle function is optimized.

# EOSINOPHILIC ESOPHAGITIS WITH AND WITHOUT AIRWAY INVOLVEMENT -A COMPARATIVE ANALYSIS

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Background: Eosinophilic esophagitis (EoE) is an allergic esophageal disease characterized by marked eosinophilic infiltration and inflammation eventually leading to esophageal dysfunction. EoE may or may not present with airway involvement, leading to breathing difficulties.

Objective: To compare the course of disease in patients with EoE with or without airway involvement

Methods: A retrospective chart review was done on patients with a diagnosis of Eosiniphilic Esophagitis and were managed in our EoE specialty clinic from 2012- 2018. A total of 123 EoE patients were included in the study. Each patient's disease course was examined for pertinent information including - but not limited to - age at presentation, allergies, endoscopic and pathology results, treatments prescribed, and time to resolution. T-tests were used to analyze for any differences between the airway and non-airway groups for each of these variables.

Results: Of the variables analyzed, none showed any significant difference between patients suffering from EoE with and without airway involvement. However, patients with airway disease did have a younger age at presentation as compared to those without airway symptoms (6.68 years vs. 9.69 years, $p = 0.69$). Analysis of endoscopic and pathology findings revealed no difference. Similarly, no differences in treatments prescribed were found. Finally, Kaplan-Meyer analysis of the time to resolution of symptoms indicated no discernible differences.

Conclusions: Our findings indicate that the disease course of patients with EoE does not vary depending on the presence of airway symptoms. Thus, patients with airway symptoms should not be diagnosed or treated any different than those without airway symptoms.

# UPPER AIRWAY DETERGENT POD INJURIES IN CHILDREN: CLINICIAN AND INDUSTRY SUGGESTIONS

Preethi Venkat

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Objective: Accidental injuries involving laundry detergent pods (LDPs) are well-documented and common in children. This paper suggests industry improvements to reduce accidental LDP injury incidence and severity. Additionally, we provide an algorithm for otolaryngologists (ENT) to follow in the treatment of LDP exposure.

Methods: Data was taken from the National Electronic Injury Surveillance System (NEISS) database, 2012-2016. We utilized product code 949, which identified cases of laundry detergent-related ED visits.

Results: Of the LDP-related injuries identified (n=558), the most commonly affected groups were children 1-2 (46.1%) and 2-3 years of age (23.1%). The most common ENT-related symptoms were drooling (31%), difficulty breathing (14%) and wheezing (11%).

Conclusion: Educating parents about ways to limit LDP exposures has been the main approach to reducing incidence thus far. Given that exposures are still occurring, otolaryngologists should be aware of common presenting injury treatments, and industry consideration should be given to redesigning LDP packaging to reduce the risk of injury upon ingestion.

# A STERNOCLAVICULAR ANOMALY: CASE SERIES AND A REVIEW OF LITERATURE

Kimberly Luu (M.D.)

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PURPOSE:

To describe the clinical and pathologic features of a rare congenital neck mass, a sternoclavicular sinus. A sternoclavicular sinus presents similarly but distinctly from the more common branchial cleft anomaly, highlighting the importance of identifying differentiating features.

METHODS:

Patients with a congenital neck anomaly, distinct from common branchial cleft anomalies, were identified through a 10-year retrospective chart review of a tertiary care pediatric otolaryngology practice. Records including consulting evaluations, imaging studies, operative reports, and histopathology were included.

RESULTS:

Details of three patients with a congenital neck anomaly consistent with a sternoclavicular anomaly are reported. All patients presented in the same way: with a neck mass and dermal pit with imaging confirmation of a tract ending at the sternoclavicular joint. Complications of an untreated anomaly included infection and abscess formation. Surgical excision, through an external approach, was successfully performed with subsequent resolution of symptoms. The literature review compiles all the cases previously presented. Theories on embryologic origin include incomplete fusion of sternum and clavicle or unusual remnant of a fourth branchial cleft. Commonalities include left sided predominance, squamous epithelium lined sinus tract ending at the sternoclavicular junction, and successful surgical excision in almost all cases.

CONCLUSION:

A sternoclavicular anomaly is a rarely described congenital neck abnormality. Presentation and management is similar to a branchial cleft anomaly but with a distinct anatomical pathway that is important for surgeons to recognize.

# COMPARATIVE MANAGEMENT OF OTOGENIC LATERAL SINUS THROMBOSIS IN CHILDREN

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Objective:

The aim of this study is to (1) report the incidence, clinical presentation, management, and complications of children treated for otogenic lateral sinus thrombosis (LST) and (2) compare with patients treated from 1996 to 2012 at the same hospital.

Methods:

Retrospective case review of children treated for otogenic lateral sinus thrombosis from July 2012 to June 2018. Comparison of management from patients treated July 1996 to June 2012.

Results:

Six patients were identified with mastoiditis and LST (2012-2018), compared with five patients in our prior review (1996-2012). All except one patient had symptoms for greater than one week and were previously treated with antibiotics prior to presentation. All underwent mastoidectomy with or without ear tubes, and incision and drainage when indicated. All were, or are, currently being treated with anticoagulation including direct oral anticoagulants (DOAC). Only one patient has had resolution of thrombus, but none have neurological sequelae.

Discussion:

LST is a rare, yet serious complication of otitis media for which management controversies persist. This study compares recent and past management of 11 patients from our institution, and includes a preliminary report of safe use of DOAC in the pediatric population. The increased rate of otogenic LST diagnosis and treatment - 6 cases in 6 years compared to 5 cases in 16 years - is likely multifactorial and warrants further study. As only one patient has had thrombus resolution in our more recent series, yet none have any neurological sequelae, this study also challenges utility of anticoagulation for LST.

# IS THERE UTILITY IN ROUTINE PREOPERATIVE HEMOGLOBIN TESTING IN CLEFT LIP AND PALATE SURGERY - A QUALITY IMPROVEMENT STUDY

Alexa Robbins

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Objective

Since the 1960's, a minimum hemoglobin of 10 has been considered a marker of positive outcomes after cheiloplasty and palatoplasty. At our institution, we have noted consistently low intraoperative blood loss (BL) and postoperative complications (PC), and therefore undertook to examine whether patients with day-of-surgery hemoglobin results (DHR) less than 10 showed a higher PC rate.

Methods

Through current procedural terminology (CPT) code queries of the electronic medical record, we identified primary cheiloplasties and palatoplasties performed by a single surgeon between 2012 and 2017. We then completed a retrospective chart review.

Results

We identified 137 cleft lips and 188 cleft palates in 248 patients meeting inclusion criteria. The average HGB for cheiloplasties and palatoplasties was 11.8 (R 8.4-14.2) and 12.1 (R 9.4-19.7), respectively. BL was recorded as minimal (10cc) in 98% (N=123) of cheiloplasties and 79% of palatoplasties (N=132). 3% (N=8) patients were identified with a DHR below 10, and 1 (13%) of this subset (intraoperative BL 20cc, DHR 9.9) experienced a PC (tet spell). In the DHR above 10 population, there were 9 (4%) total PC's, namely dehydration, dehiscence, prolonged admission, and hemorrhage.

Conclusions

Based on this study, the PC rate in primary cheiloplasty and palatoplasty is indeed low. The one patient with low DHR who experienced a complication had significant cardiac disease. It is unlikely that the complication was related to DHR. Given these results, our institution now limits day-of-surgery hemoglobin testing to patients with comorbidities, who are at higher baseline risk.

# OTOLARYNGOLOGIC MANIFESTATIONS OF CORNELIA DE LANGE SYNDROME: A SYSTEMATIC REVIEW

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OBJECTIVES. Cornelia de Lange Syndrome (CDLS) is a rare genetic disorder estimated to affect 1 in 30.000 live births. Our goal was to systematically review the otolaryngology literature pertaining to CDLS.

METHODS. We systematically reviewed MEDLINE/PubMed, Embase and Google Scholar for original articles of otolaryngology manifestations for patients with CDLS. These articles were analyzed and pooled prevalences were calculated.

RESULTS. We analyzed 1343 patients included in 36 case series and 33 case reports. Hearing loss was present for most of the reported patients (27 studies), with sensorineural hearing loss affecting 40.1%, conductive 28.5% and mixed or unspecified 19.1%. Recurrent acute otitis media was the most frequent infectious manifestation (57.9% in 7 studies) followed by recurrent upper/lower airway infections (53.1%,5 studies). Sixty-four (64.7%, 9 studies) of patients have dysphagia and 69.8% (4 studies) have some degree of dysphonia or dysarthria. Cranio-facial anomalies were reported in 30 studies with micrognathia (73.1%) and cleft palate (59.3%) being the more common. Other physical exam abnormalities included: lips (91.3%), dentition (92.2%), mouths (88.7%), low-set ears (49.1%), low hairline (96.6%), and eyelashes (96.6%). Sleep-disordered breathing or obstructive sleep apnea affect 14.1% of patients (10 studies). Infrequently mentioned manifestations included difficult intubation (55.6%, 9 studies) and choanal atresia (37.5%,2 studies).

CONCLUSIONS. This is the first comprehensive evaluation of the CDLS literature regarding otolaryngologic manifestations. ENT manifestations are numerous, mostly regarding hearing loss, and cranio-facial anomalies. Airway disorders were reported mostly in case reports, but are of significance given the potential life-treatment condition.

# PEDIATRIC EWING SARCOMA OF THE HEAD AND NECK

Elaine Martin

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Objective: To determine the demographics, treatment modalities, and overall survival of pediatric Ewing sarcoma of the head and neck.

Methods: The National Cancer Institute's Surveillance, Epidemiology, and End Results database was used to identify cases of pediatric Ewing sarcoma of the head and neck between 1973 and 2015. Additional variables collected included age, gender, ethnicity, tumor grade, staging, treatment modality, and follow-up time. Kaplan-Meier survival curves were generated and overall survival was calculated.

Results: One hundred and twenty seven cases of pediatric Ewing sarcoma were identified. The majority of patients were male (53%), white (88%), and non-Hispanic (76%), and the mean age at diagnosis was 10.5 years. The most common tumor sites were bones and joints of the skull and face (46%), followed by soft tissue of the head, face, and neck (25%), followed by the mandible (13%). Most patients (71%) had some type of surgery, 70% received radiation, and 92% received chemotherapy as part of their treatment. Overall 1 and 5 year survival was 91% and 72%, respectively. There was no significant difference in survival based on patient gender, age, tumor location, radiation, or chemotherapy treatment.

Conclusions: Pediatric Ewing sarcoma of the head and neck is a rare malignancy which has a good prognosis with overall 1 year survival of 91% and 5 year survival of 72%. This study is the first to report in detail surgical interventions as well as characteristics about tumor extension, lymph node spread, and treatment outcomes since the advent of radiation and chemotherapy.

# ACUITY OF TONSILLITIS DOES NOT INCREASE RISK OF POST-TONSILLECTOMY HEMORRHAGE

Noah Worobetz

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Background: Patients undergoing tonsillectomy for chronic or recurrent tonsillitis have increased risk of post-tonsillectomy hemorrhage than patients with an indication of obstructive sleep apnea or sleep disordered breathing. Among patients with tonsillitis, various degrees of acuity are observed, but the extent to which acuity relates to hemorrhage risk is not well understood.

Methods: A retrospective cohort study at a single tertiary pediatric hospital was conducted. All patients who underwent tonsillectomy in 2017 for an indication of chronic or recurrent tonsillitis were included. Comorbidities, postoperative bleeding, procedure technique, and tonsillitis history were ascertained from patient medical records. Patients with high acuity had documentation of at least 7 infections in the year prior to surgery. We compared the distribution of risk factors for patients with post-tonsillectomy hemorrhage and patients without using Chi-squared and Wilcoxon-Mann-Whitney tests.

Results: Among the 456 in our study, the prevalence of high acuity was 22%. The incidence of bleeding in the total cohort was 10%, compared to 14% in the high acuity group. This difference did not reach statistical significance ($p=0.17$). Patients with a bleed had a more recent infections (median: 67 days vs 90 days), but this too did not reach statistical significance ($p=0.10$). The only risk factor significantly associated with increased risk of hemorrhage in our study was neuromuscular comorbidity ($p=0.01$).

Conclusion: High acuity of tonsillitis, nor timing of most recent infection, are associated with post-tonsillectomy hemorrhage in this retrospective study. Future research should include a prospective study which can reliably capture all events of tonsillitis.

# COMPARING THE EFFICACY OF INTRAORAL AND TRANSCERVICAL APPROACHES FOR THE TREATMENT OF RANULAS

Jeffrey K Than

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Objective: This study aimed to compare the treatment outcomes of intraoral and transcervical approaches for the treatment of simple and plunging ranulas.

Methods: A review was performed on patients treated for a ranula from February 2013 to May 2018. Clinical data was collected from medical records including patient demographics, intervention used, and recurrence rate.

Results: This study included 52 patients treated surgically for a ranula (10 plunging, 42 simple). The patients - 21 male and 31 female - ranged in age from 1 month to 17 years (mean = 9.68 years). The most common symptoms were intraoral swelling (39 cases) and pain (14 cases) with a symptom duration ranging from 1 day to 3 years. Simple ranulas were treated with intraoral excision of the mass and the SLG (27 cases), marsupialization (7 cases), intraoral excision of the mass alone (7 cases), and intraoral excision of the ranula and subsequent marsupialization after recurrence (1 case); intraoral excision of the SLG was not associated with any recurrence. Plunging ranulas were treated with intraoral excision of the mass and/or SLG (7 cases) or with a transcervical approach (3 cases). One patient was initially treated with sclerotherapy before undergoing an intraoral excision of the SLG. Two patients treated with transcervical excision of the ranula experienced recurrence compared to no recurrence with intraoral excision of the SLG.

Conclusion: Given the effectiveness of the intraoral excision of the SLG, careful consideration must be used prior to using a transcervical approach to treat simple and plunging ranulas.

# SLEEP ARCHITECTURE AND OSA SEVERITY IN A PEDIATRIC POPULATION

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Background: Previous research has demonstrated that children with obstructive sleep apnea (OSA) are more likely to suffer behavioral and neurocognitive problems. The correlation between OSA and daytime impairment is thought to be due to sleep disruption. Consequently there is a significant need to assess measures of sleep quality in this population. However, studies regarding sleep architecture in children with OSA have thus far been inconclusive.

Objective: To determine polysomnographic characteristics of sleep architecture in children with OSA and investigate effects relative to OSA severity.

Methods: Overnight polysomnograms (PSG) of children referred for suspected OSA were reviewed. Subjects were classified by apnea hypopnea index (AHI) into those with mild, moderate and severe OSA. PSG parameters of sleep architecture were recorded and analyzed according to OSA severity.

Results: A total of 211 PSGs were included (mean age 7.9 years  4.2). Children with severe OSA differed from those with moderate or mild OSA, exhibiting a greater percentage of time spent in N1 stage sleep (14.5 vs. 8.2 and 4.8, $p<0.01$), a lower percentage in N2 stage sleep (38.0 vs. 40.0 vs. 47.9, $p<0.01$), a lower percentage in REM sleep (13.6 vs. 14.5 vs. 15, $p<0.05$), and lower sleep efficiency (79.4 vs. 81.1 vs. 85.4, $p<0.01$). No significant differences were found in N3 sleep stage.

Conclusion: Our results confirm that children with OSA have altered sleep architecture. Similar to adults, children with more severe OSA appear to spend greater amounts of time in N1 stage sleep and less time in deeper sleep stages.

# GENERAL ANESTHESIA RISK ACROSS PEDIATRIC SURGICAL SPECIALTIES: LOW IN OTOLARYNGOLOGY

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Purpose: To determine the relevance of the Food and Drug Administration (FDA) warning regarding general anesthesia in children under 3 years of age for procedures lasting longer than 3 hours, by surgical specialty and for otolaryngology specifically.

Methods: A one-year retrospective review was conducted at a tertiary-care medical center for all children younger than 3 years undergoing surgical procedures with duration greater than 3 hours. De-identified data related to age, surgical service, procedure type, and general anesthesia time were collected and examined.

Results: During 2017, 430 of 11,757 patients (3.6%) met the age and duration of anesthesia criteria. Procedures performed by the Cardiothoracic Surgery service were most likely to result in duration of surgery greater than 3 hours (24.2%), followed by General Surgery (21.6%), Urology (16.5%), Plastic Surgery (8.1%), Orthopedic Surgery (7.2%), Neurosurgery (7.2%) and Cardiology (6.5%). Less than 5% of patients undergoing Otolaryngology (4.4%), Ophthalmology (1.9%), Transplant Surgery (1.2%), Dental (0.5%), Pulmonology (0.2%), and Gastroenterology (0.2%) procedures required anesthesia greater than 3 hours. In Otolaryngology, the most common procedures were otologic. In Plastic Surgery, Cleft palate procedures predominated.

Conclusion: Less than 4% of patients undergoing surgery under age 3 required general anesthesia for longer than 3 hours. The theoretical risks of general anesthesia per the FDA warning are discussed, and must be balanced against the known functional and neurodevelopmental consequences of not performing critical and time-sensitive surgery on children in this age group. Strategies for addressing parental and provider concerns are reviewed.

# PEDIATRIC CERVICAL LYMPHADENOPATHY: THE UTILITY OF WATCHFUL WAITING.

Edward R. Lee (M.D.)

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Isolated cervical lymphadenopathy is a common reason for referral to a pediatric Otolaryngologist but is a rare initial presentation of lymphoma. Often times, careful observation is all that is necessary and spares children the risks of ionizing radiation and/or invasive surgical intervention. We sought to identify patient characteristics that are associated with benign lymphadenopathy and to differentiate this group from those who warrant further work-up. We identified 77 pediatric patients with cervical lymphadenopathy who presented to our tertiary care pediatric otolaryngology practice over a 10 year period. The mean lymph node size at presentation was 1.69 cm. The mean duration of symptoms was 9.19 months. Patients, on average, were observed for 9.44 months. The factors most highly correlated with lymphoma were lymph node size and age. 9/77 children (11.6%) went on to have a FNA. The mean size of lymph nodes proceeding to FNA was 2.06 cm. 8/9 (89%) of FNA's showed benign pathology. Only 1/9 (11%) of children who underwent FNA had concerning signs of malignancy. 12/77 (15.6%) of children underwent excisional biopsy. The mean lymph node size was 2.23 cm. 1/12 children (8.3%; 1.3% of total study group) had a final pathology of lymphoma with a lymph node size of 3.7 cm. Up to 90% of children under the age of 8 will have palpable cervical adenopathy. Our study suggests that most children do well with conservative management with or without serial ultrasounds. Non-mobile, >3 cm lymph nodes should undergo consideration for work-up including FNA or excisional biopsy.

# EOSINOPHILIC ESOPHAGITIS WITH AND WITHOUT AIRWAY INVOLVEMENT -A COMPARATIVE ANALYSIS

Varun Bora

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Background: Eosinophilic esophagitis (EoE) is an allergic esophageal disease characterized by marked eosinophilic infiltration and inflammation eventually leading to esophageal dysfunction. EoE may or may not present with airway involvement, leading to breathing difficulties.

Objective: To compare the course of disease in patients with EoE with or without airway involvement

Methods: A retrospective chart review was done on patients with a diagnosis of Eosiniphilic Esophagitis and were managed in our EoE specialty clinic from 2012- 2018. A total of 123 EoE patients were included in the study. Each patient's disease course was examined for pertinent information including - but not limited to - age at presentation, allergies, endoscopic and pathology results, treatments prescribed, and time to resolution. T-tests were used to analyze for any differences between the airway and non-airway groups for each of these variables.

Results: Of the variables analyzed, none showed any significant difference between patients suffering from EoE with and without airway involvement. However, patients with airway disease did have a younger age at presentation as compared to those without airway symptoms (6.68 years vs. 9.69 years, $p = 0.69$). Analysis of endoscopic and pathology findings revealed no difference. Similarly, no differences in treatments prescribed were found. Finally, Kaplan-Meyer analysis of the time to resolution of symptoms indicated no discernible differences.

Conclusions: Our findings indicate that the disease course of patients with EoE does not vary depending on the presence of airway symptoms. Thus, patients with airway symptoms should not be diagnosed or treated any different than those without airway symptoms.

# MAXIMIZING OPERATIONAL EFFICIENCY VIA REDESIGN OF AERODIGESTIVE CLINIC PRE-PLANNING WORKFLOW AND RETURN ON INVESTMENT

Aarti Chandawarkar (M.D.)

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Background: The lead physician in our multi-disciplinary Aerodigestive Clinic used a time-consuming manual process of pre-planning for patient visits. For each session, this necessitated approximately six hours in manual data extraction from the electronic patient chart into a spreadsheet. This process was time consuming and resulted in risk, including transferring protected health information out of the medical record and data transposition errors. The Aerodigestive Team approached Physician Informatics to see how pre-planning could be automated within the Electronic Medical Record (EMR).

Methods: The Physician Informatics team and the clinic's lead physician performed a needs assessment. A semi-automated EMR-embedded "Snapshot" was created with real-time data points, which can be utilized by all clinic providers.

Results: Using EMR functionality a pre-visit report was created that can be viewed by all providers. 80% of the data included is automated. Since this report is embedded in the EMR, there is no potential for loss of PHI. Additionally, semi-automation, the most recent patient data points are populated in the report without having to do a chart search. This cut down the time spent by our lead physician by 50% prior to each clinic and resulted in substantial cost savings, specifically about $ 25,000 annually (based on the 50th percentile for AAMC salary survey for ENT physicians).

Conclusions: The use of a semi-automated EMR embedded pre-visit "snapshot" in a multidisciplinary high complexity aerodigestive clinic improved physician efficiency and satisfaction, facilitated cohesive team management, reduced risk of errors and PHI exposure and had substantial cost savings.

# GRANULAR CELL TUMOR: A RARE PEDIATRIC PAROTID TUMOR

Anthony Sheyn (M.D.)

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Introduction: Granular cell tumors are uncommon soft tissue tumors thought to arise from Schwann cells of peripheral nerves. Granular cell tumors of the parotid are particularly rare and to our knowledge less than 15 cases have been reported, few of those in the pediatric population. We present a case of a 17 year old female who presented with a left parotid granular cell tumor.

Case: A 17 year old presented to our facility with lower extremity weakness. Evaluation of her symptoms resulted in a new diagnosis of multiple sclerosis. A parotid mass was noted incidentally on one of her MRI images. A fine needle aspiration was performed during her initial hospitalization and revealed no evidence of neoplasia but presence of a cystic structure. Due to newly diagnosed multiple sclerosis, surgery was delayed until her neurologic symptoms were under control. Patient subsequently underwent a left superficial parotidectomy. Intra-operatively the mass appeared to be very inflamed and attached to surrounding tissues with an intimate relationship to the buccal branch of the facial nerve. The mass was able to be removed with preservation of all facial nerve branches. Post-operatively no facial weakness was noted. Final pathology was returned as a granular cell tumor.

Conclusion: Granular cell tumors of the parotid are extremely rare tumors in children and adults. Fine needle aspiration may not be able to identify the pathology. Treatment is generally limited to gross total excision with facial nerve preservation with good pathologic evaluation to rule out malignancy.

# ACUTE LYMPHOBLASTIC LEUKEMIA PRESENTING WITH PERIORBITAL EDEMA: A CASE REPORT AND LITERATURE REVIEW

Andrew P. Stein (M.D.)

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Objective - To describe a rare presentation of acute lymphoblastic leukemia (ALL) that is important to the field of pediatric otolaryngology.

Methods - The case of a nine-year old female who presented with left periorbital edema as her initial symptom of ALL was reviewed along with pertinent literature.

Results - At the time of patient's initial presentation, a computed tomography scan of the sinuses showed sphenoid sinus opacification and left lateral rectus muscle enlargement. She was started on antibiotics without improvement. Subsequently, magnetic resonance imaging of the brain demonstrated an infiltrative process involving the sphenoid sinus marrow space, lateral rectus muscles and dura. Her peripheral blood count revealed 40% blasts, and a bone marrow biopsy demonstrated B-cell ALL. She immediately underwent induction chemotherapy, and her post-induction bone marrow biopsy showed no evidence of residual disease.

Conclusion - This case highlights the importance of understanding and considering rare, aggressive diseases that can masquerade as simple periorbital edema.

# A MULTIDISCIPLINARY APPROACH TO RAPID SUCCESSION OF SPINE AND TRACHEAL RECONSTRUCTION IN A PATIENT WITH MORQUIO SYNDROME

Patrick Kiessling

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Morquio syndrome (Mucopolysaccharidosis IVA, MPS IVA) is an autosomal recessive lysosomal storage disease caused by a deficiency in N-acetylgalactosamine-6-sulfate sulfatase (GALNS). The disease may cause a wide variety of symptoms and manifestations, ranging from mild to severe phenotype. Issues of the spinal cord and airway are a major source of morbidity and mortality. Given the medical and surgical complexity of these patients, we believe that a multidisciplinary approach is preferable. Here we describe a 17-year-old male patient with severe Morquio syndrome, presenting with thoracolumbar kyphosis, cervical and upper thoracic spinal stenosis, and progressive severe tracheal stenosis. Coordinated care among otolaryngology, orthopedic surgery, neurosurgery, anesthesiology, cardiovascular surgery, and pulmonology teams facilitated the successful planning and execution of two major surgical interventions in rapid succession. While reconstruction of both airway and spine in a single patient has been attempted previously, this is the first such successful procedure.

# DESMOID FIBROMATOSIS: A RARE PARANASAL SINUS TUMOR OF CHILDREN

Anthony Sheyn (M.D.)

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Introduction: Desmoid tumors of the paranasal sinuses are extremely rare in the pediatric population. They are considered to be low grade tumors and can very locally destructive. To our knowledge only 8 previous cases have been reported in the literature.

Case: An 18 month old female presented to our facility after undergoing a biopsy of a rapidly enlarging right maxillary sinus mass. Parents noted that enlargement occurred in less than a month. Initial pathology was concerning for a rhabdomyosarcoma. Further review by pathology prompted a repeat biopsy which was positive for desmoid fibromatosis. Symptoms at the time of presentation included cosmetic deformity, nasal obstruction, proptosis and decreased vision. After multi-disciplinary discussion the decision was made to proceed with resection and post-operative chemotherapy. A combined endoscopic and sublabial approach was employed to perform a medial maxillectomy with resection of the mass. Part of the orbital floor was resected in order to obtain negative margins Oculoplastic surgeons then evaluated the orbital floor but no reconstruction was needed. Following surgery vision and cosmesis returned to normal and the patient was discharged on post-operative day 1. Surveillance MRI obtained at 3 and 6 months after resection noted stable to decreased post-operative changes or residual disease.

Conclusion: Desmoid tumors of the paranasal sinuses are rare tumors in the pediatric population. Surgical excision is the preferred treatment when possible. Residual tumor can be monitored with imaging and cosmesis should be maintained when possible. Post-operative chemotherapy is an option for residual disease.

# RHINOSCLEROMA WITH SINONASAL AND LARYNGEAL INVOLVEMENT IN A PEDIATRIC PATIENT

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Outcome Objective:

We describe a rare case of rhinoscleroma presenting as diffuse nasal and laryngeal polyps in United States. The case demonstrates a diagnostic challenge due to lack of awareness in non-endemic areas and biopsy is essential to obtain diagnosis.

Methods:

Case Report

Results:

A 17-year-old Hispanic male presented with a 2-year history of chronic progressive nasal obstruction, hoarseness and rhinorrhea. Flexible nasal endoscopy and laryngoscopy revealed polypoid nodules along the septum and bilateral turbinates and lesions involving bilateral arytenoid mucosa. The lesions were suspicious for granulomatosis with polyangiitis (Wegener's); therefore, formal endoscopy in the OR was recommended. Intraoperatively, nasal endoscopy showed bilateral numerous mucosal nodules that were round, smooth and pedunculated. Direct laryngoscopy showed diffuse nodular lesions involving the larynx, the epiglottis, false cords, laryngeal ventricles and arytenoid mucosa. The subglottis had circumferential small nodular lesions and fibrinous exudate. Distal airway was clear. Biopsy revealed severe mixed inflammatory cell infiltrate consisting of predominately lymphocytes, foamy histocytes with pale blue cytoplasm (Mikulicz cells). Warthin-Starry stain revealed silver-positive rods confirming the pathologic diagnosis of rhinoscleroma. The patient was referred to infectious disease specialist for treatment. At his 8 months follow-up, patient reported complete resolution of symptoms. Flexible endoscopy revealed minimal nodules in the nasal cavity, and the larynx was free of lesions.

Conclusion:

Rhinoscleroma is a chronic, slowly progressive, inflammatory disease that affects the upper and lower respiratory tracts with K rhinoscleromatis as the causative agent. Treatment involves prolonged courses of antibiotics. Despite excellent initial response, recurrence is common.

# AGGRESSIVE PARATHYROID ADENOMA WITH CONCOMITANT BILATERAL SCFE IN AN ADOLESCENT

Jordan Luttrell

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Introduction: Primary hyperparathyroidism (PHPT) is uncommon in the pediatric population. Pediatric PHPT typically presents with symptomatic hypercalcemia or complications including kidney stones, abdominal pain, and skeletal fragility, although 10-15% of cases are discovered incidentally. We present a case of a 10 year-old non-obese male who had an even more unusual presentation of slipped capital femoral epiphysis (SCFE) later found to have a single parathyroid adenoma. The number of cases of PHPT in the pediatric population is greater than 300 currently, whereas only 11 cases of concomitant PHPT and SCFE in an adolescent have been reported. Generally, the clinical course is more severe in children, requiring early surgical intervention and frequent postoperative monitoring.

Case: A 10-year-old non-obese African American male presented to an orthopedic clinic with pain in the hip and limp for 3 weeks. He was noted to have bilateral SCFE. Work-up after his presentation demonstrated a calcium of 12.6 and a parathyroid hormone of 1,862. Sestamibi scan was performed, as well as CT and ultrasound, which demonstrated a left inferior parathyroid adenoma. His SCFE was repaired acutely and he underwent excision of his parathyroid adenoma shortly after that with PTH dropping to 96.4. He had postoperative hypocalcemia that corrected with oral calcium measures.

Conclusion: Based on our research, this is the 12th case worldwide reporting an association of SCFE and PHPT. We agree that patients presenting with bilateral SCFE and hypercalcemia should be worked up for underlying endocrine pathology and if found, prompt surgical management should be instituted.

# POSTERIOR CONGENITAL CHOLESTEATOMA: UNDERSTANDING AN ATYPICAL ORIGIN OF A RELATIVELY COMMON DISEASE

Krishna Bommakanti

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Objectives: To evaluate and understand the clinical course, treatment, and embryological origin of posterior congenital cholesteatomas (PCC)

Background: Congenital cholesteatomas (CC) originate from epithelial remnants within the middle ear concurrent with an intact tympanic membrane. They typically originate in the anterior mesotympanum near the peri-Eustachian tube; however, a small subset originates in the posterosuperior quadrant. It is important to recognize PCC as a distinct entity from the more common anterosuperior cholesteatomas given their distinctive and more challenging clinical presentation, diagnosis, and treatment.

Methods: This is a retrospective review of PCC cases treated between 2014-2018 at a tertiary pediatric hospital. The outcome measures were demographics, site of origin, embryologic basis, clinical features, surgical findings, and hearing results.

Results: Three cases were found during this period. All patients presented with moderate unilateral conductive hearing loss and normal otoscopy without previous otologic history. The delay in diagnosis ranged from four months to several years and the mean preoperative speech reception threshold (SRT) was 58 dB. CT scan of the temporal bones showed possible stapes superstructure absence, but no noticeable masses. All three patients were diagnosed with cholesteatoma intraoperatively. One possible explanation for the PCC origin is that the mesenchymal to epithelial transition (MET) that occurs during middle ear development produces a stretch of abnormal epithelium that develops into a cholesteatoma.

Conclusion: PCC is a rare disease that is often diagnosed at a later age. A better understanding of this disease can expedite the diagnosis and improve treatment.

# IMPROVEMENT OF CENTRAL SLEEP APNEA AFTER SUPRAGLOTTOPLASTY FOR LARYNGOMALACIA

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Laryngomalacia is commonly associated with respiratory comorbidities such as obstructive sleep apnea. Numerous studies report on improvements in polysomnographic outcomes following supraglottoplasty but largely focus on obstructive or overall respiratory indices. Although the prevalence of central sleep apnea (CSA) is reported to be as high as 43.6% in infants with laryngomalacia, existing literature lacks information on the effect of surgical intervention on central apnea. This case series describes two patients with laryngomalacia and sleep apnea who underwent supraglottoplasty. The first patient underwent preoperative polysomnography, which showed overall apnea hypopnea index (AHI) of 202.0 events/hour. Obstructive AHI was 182.2 and central apnea index (CAI) was 17.3 events/hour. Repeat polysomnography 17 days after supraglottoplasty showed an improvement with AHI of 11.7 events/hour. Obstructive AHI was 6.2 and CAI was 4.6 events/hour. The second patient underwent preoperatively polysomnography which showed an overall AHI of 182.5 events/hour. Obstructive AHI was 50 and CAI was 132.7 events/hour. Postoperative polysomnography showed an overall AHI of 12.3 events/hour. Obstructive AHI was 9.3 and CAI decreased to 3.0 events/hour. We hypothesize that sustained hypoxia due to airway obstruction may result in decreased respiratory drive and higher occurrence of central apneas. An alternative hypothesis is that these patients, as a result of their airway obstruction, may have dwindling respiratory reserve and are therefore more prone to desaturation with physiologic central apneas, which are known to occur in infants. This is the first description of two infants with laryngomalacia who exhibit a significant decline in central apnea events after supraglottoplasty.

# SURNAMES OF CHILDREN WITH ISOLATED UNILATERAL ATRESIA: AURAL VERSUS CHOANAL

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Background and Purpose: The reported overall incidence of both aural and choanal atresia is about 1/7,000. The etiologic explanation is unknown for isolated cases (i.e., those lacking additional unrelated major defects). The quest to describe surname distributions arouse from the serendipitous simultaneous inspection of the databases of the two populations from a single metropolitan area.

Methods: Review of existent databases of children in one metropolitan area who had computed tomography (after 2001) to characterize these atresias.

Results: No choanal atresia case had a Hispanic surname (0/25), but 21 of 70 aural atresia cases had Hispanic surnames. P<.005.

Discussion: Though an increased Hispanic representation in children with aural atresia and a decreased representation in choanal atresia have been suggested, this is the first report of cases from one metropolitan area.

Conclusions: Isolated non-syndromic unilateral choanal atresia and aural atresia may be associated with genetic differences as manifested by ethnicity.

# ACUTE B-CELL LYMPHOBLASTIC LYMPHOMA AND SCALP LEUKEMIA CUTIS IN A PEDIATRIC PATIENT

Betty Yang (M.D.)

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We present the case of a 7 year-old girl who presented with an asymptomatic soft tissue mass of her scalp. Several months later, she developed concerning right cervical lymphadenopathy that was unresponsive to antibiotic therapy. She was taken to the operating room for cervical lymph node biopsy and scalp punch biopsy, which were found to be consistent with acute B-cell lymphoblastic lymphoma and leukemia cutis. Leukemia cutis is defined as cutaneous infiltration of leukemic cells which results in clinically significant skin lesions. This phenomenon is seen in patients with acute myeloid leukemia and chronic lymphocytic leukemia, however is rare in patients with acute B-cell lymphoblastic lymphoma with an estimated prevalence of 1-3%. These lesions most commonly occur on extremities, but may also occur on the face and scalp, emphasizing the important role that otolaryngologists may play in timely diagnosis. While leukemia cutis is rare, it should be included in the differential diagnosis for patients presenting with asymptomatic, rapidly growing cutaneous nodules.

# FLOOR OF MOUTH THYROGLOSSAL DUCT CYST: A CASE REPORT OF A RARE EMBRYOLOGIC COURSE WITH REVIEW OF THE LITERATURE

John Kerr

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Introduction: Thyroglossal duct cysts (TGDC) are one of the most common pediatric midline neck masses. These occur with failure of involution of the thyroglossal duct after migration of the thyroid from the tongue base to its final position in the neck. Most thyroglossal duct cysts occur in the region of the hyoid bone, with 85% occurring in the infrahyoid region. Definitive surgical management involves the Sistrunk procedure which removes the cyst and tract in its entirety, as well as the midportion of the hyoid bone.

Case Report: We present a case of an infant presenting with a midline neck mass. MRI demonstrated a concurrent hyperintense lesion in the floor of mouth. A transcervical approach revealed a cystic lesion adjacent to the hyoid bone. The central hyoid was excised and the termination of the mass was found in the central floor of mouth, representative of the mass seen on MRI. There was no connection with the base of tongue. Pathology confirmed the diagnosis of a thyroglossal duct cyst.

Discussion: Thyroglossal duct cysts typically present in the midline neck. We review all atypical presentations of a TGDC such as within the floor of mouth, base of tongue, larynx, and intrathyroid, as well as double cysts. We describe a presentation of TGDC that was found to originate in the anterior floor of mouth. The entire mass was successfully removed through a transcervical approach (Sistrunk procedure.

# PROSPECTIVE STUDY OF TIME-INDUCED SLEEP ENDOSCOPY IN THE DIAGNOSIS OF AIRWAY OBSTRUCTION AND OBSTRUCTIVE SLEEP APNEA

Nicholas R. Oberlies

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Introduction

Polysomnography (PSG) and drug-induced sleep endoscopy (DISE) are the gold standards for diagnosing obstructive sleep apnea (OSA) and identifying sites of airway obstruction, respectively, with the latter requiring general anesthesia (GA). In 2016, the FDA warned of potential adverse effects of anesthesia exposure on neurodevelopment in children under three years of age. In a subset of pediatric patients, it may be possible to observe sleep airway dynamics without GA using time-induced sleep endoscopy (TISE).

Objectives

To evaluate TISE's ability to observe sleep airway dynamics in children under three months of age without using GA.

Methods

Ten patients with suspected OSA underwent TISE and had their charts reviewed for demographic information, comorbidities, and maternal risk factors. A flexible laryngoscope was inserted through the nose, positioned above the larynx, and held for five minutes or until the patient fell asleep.

Results

Ten patients underwent TISE at a median age of 17 days (range 1-80 days). Mean gestational age, weight, and height at birth were 38 weeks (SD 2 weeks), 2.9 kg (SD 0.7 kg), and 48.4 cm (SD 4.7 cm). Laryngomalacia (50%), cleft palate (50%), and in-utero tobacco (40%) and methadone (20%) exposure were common. Five (50%) patients fell asleep during the procedure and had observable sleep airway dynamics, with an average time to falling asleep of 192 seconds (SD 72 seconds). No patients desaturated during the procedure.

Conclusions

Using TISE, it is possible to observe sleep airway dynamics in some children under three months of age without using GA.

# POST-TRANSPLANT LYMPHOPROLIFERATIVE DISEASE OF THE LARYNX

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Purpose: To describe the presentation, workup, diagnosis, and management of children with airway post-transplant lymphoproliferative disease (PTLD).

Methods: Case series and literature review at a tertiary care pediatric medical center.

Results: Two cases of endolaryngeal PTLD are described. The first was a 15-month old who underwent liver transplant at 5 weeks of age and presented with airway distress. Airway evaluation identified epiglottic and arytenoid infiltrate. Biopsy was completed and pathology was consistent with polymorphic PTLD. The second case was a 23-month old who underwent liver transplant at 13 months of age and presented with airway distress. Airway evaluation revealed sub-mucosal infiltrate of the epiglottis, arytenoids, post cricoid region, and uvula. Biopsy was consistent with monomorphic PTLD. In both cases immunosuppression was reduced and the airway symptoms resolved. A literature review of patients presenting with airway distress and found to have PTLD was performed.

Conclusion: Airway PTLD is rare. A high index of suspicion in post-transplant patients presenting with airway obstruction is required to make this critical diagnosis. There should be a low threshold for airway evaluation and biopsy for the workup of PTLD.

# COMPLEX CLOSURE OF WIDE LOCAL EXCISION OF COMPLICATED PREAURICULAR CONGENITAL CYSTS UTILIZING A CERVICAL PAROTID FLAP

Kaylee Luck

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Purpose:

To describe innovative surgical technique for closure of large defect following complicated preauricular cyst excision secondary to prior treatments and drainage. Preauricular cysts must be widely excised in order to reduce recurrence rates; however, the resultant large local excision poses cosmetic challenges. The existing literature of various preauricular cyst repair approaches is reviewed.

Methods:

Retrospective chart review of 3 patient cases who underwent excision of recurrent preauricular lesions involving cervical parotid flap closure. Review includes intra-operative and post-operative photographs for two of the three patients documenting size, shape and outcome of our technique. All three cases demonstrate preauricular congenital cysts which were infected and had prior drainage or sclerotherapy. Two patients preauricular lesions were infected congenital cysts while one patient had an infected brachial cyst. A cervicoparotid flap was used to close all defects cosmetically with no facial nerve weakness and without distortion to the oral commissure or lateral canthus.

Results:

There is a high recurrence rate seen with wide local congenital cyst excisions; as well as, cosmetically unfavorable outcomes utilizing traditional repair. Utilizing our closure technique which involves reconstructive local regional flap with cervicoparotid approach our 3 patients have had no reoccurrence of cyst or infection. Our approach also maximizes cosmetic outcomes, with reduced scar visibility. Pre and postoperative photos will be shown.

# PURE EXPIRATORY WHEEZING CAUSED BY SUBGLOTTIC CYSTS: A CASE SERIES

Dominic J. Catalano (M.D.)

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Subglottic cysts are acquired abnormalities of the airway primarily observed in pediatric patients with a history of prematurity and early intubation or airway instrumentation. The acquired lesions most commonly present with stridor or respiratory distress given their obstructive nature, and are diagnosed following complete airway evaluation for anatomic abnormalities. Herein we present a case series of three patients presenting with four total episodes of pure expiratory wheezing in the setting of subglottic cysts who experienced complete resolution of wheezing following cyst removal.

# OCULOAURICULOVERTEBRAL SPECTRUM (GOLDENHAR) WITH IPSILATERAL CAVERNOUS HEMANGIOMA: CASE REPORT AND LITERATURE REVIEW

Romy Megahed

Romy Megahed, BS (1) Jack Hachem, BA (1) Kenya Parks, MD (1)

1) McGovern Medical School

OBJECTIVE

To present a rare case of Goldenhar syndrome and the importance of a multidisplinary approach in its management.

METHODS

We examine a case of a 6-year old male with an unusual presentation of Goldenhar syndrome.

CASE REPORT:

Our patient presented to the office with newly diagnosed Goldenhar syndrome. On physical exam, the patient presented with scoliosis, right facial hemiplasia, right-sided sensorineural hearing loss, jaw asymmetry, and right preauricular skin tag. Past medical history was significant for right-sided facial paralysis at birth. At 21 months of age, genetic testing was found to be inconclusive. At 3 years of age, he arrived at the pediatric emergency room already intubated from an outside hospital. CT scan had showed a right intraparenchymal hemorrhage in his insular lobe. Surgical evacuation was scheduled and a right sided craniotomy helped to completely remove the hemangioma.

There is has only been one other case described of a cerebral cavernous hemangioma with Goldenhar syndrome. It was uncertain whether the hemangioma in this case was ipsilateral or contralateral.

Surgical management to correct his jaw asymmetry was delayed as it was thought that continuing growth would require more definitive surgeries. No other reconstructive surgeries have been done.

CONCLUSION:

As Goldenhar syndrome has no known etiology and thus no cure, mainstay treatment is on a symptomatic basis. We recommend a multidisciplinary team to best treat patients with this disease. More cases will need to be investigated to see if cavernous hemanigomas are linked to this disorder.

# SUPERNUMERARY NOSTRIL WITH OTHER ABNORMALITIES

Romy Megahed

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Objective: To present the management of the first case of a supernumerary nostril with left-handed syndactyly, bifid uvula, and cleft palate, ankyloglossia, and upper lip tie.

Methods: Case Report, Review of literature.

Case Report: A female term 2907g neonate was born to a 25-year-old white woman. The mother of the child had an episode of strep pharyngitis treated with penicillin during the pregnancy. Otherwise, the pregnancy was normal and uncomplicated.

Several services were consulted upon presentation including genetics, ophthalmology, plastic surgery, and otolaryngology.

Physical Exam showed a congenital supernumerary nostril on the left that ended in a blind pouch, hemiplastic nails on the 2nd and 5th digits, a submucosal cleft, and syndactyly on the left hand. CT demonstrated 3 nostrils with the third most left nostril filled with soft tissue ending in a blind pouch. There was a rightward deviation of the nasal septum with narrowing of the right nasal passage and no evidence of aperture stenosis or choanal atresia.

Surgery to correct the supernumerary nostril was delayed until 1 year of age. The extra nostril was excised and closed primarily without cartilage excision. During the surgery, the left nasal lacrimal duct was found to be in an aberrant location and draining into the extra nostril.

Conclusion: The patient has healed well and will still need close follow up including additional reconstruction and a possible dacryocystorhinostomy. The literature has only shown 35 cases of a supernumerary nostril with this being the first with multiple system involvement without a genetic basis.

# TIMING OF AUDIOLOGIC TESTING AFTER PE TUBES: DOES IT MATTER?

Samantha J Lemelle

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Background and Objective:

Post-operative audiologic evaluations are generally recommended for children who undergo pressure equalization (PE) tube placement. At our institution, audiologic testing is recommended 4-6 weeks after surgery; however, no consensus was found in the literature regarding the recommended timing of post-operative hearing evaluations. Factors precluding the timing of post-operative testing can include: a lapse in health insurance, missed/cancelled appointments, parents declining testing, and limited appointment availability. The study aimed to determine if hearing outcomes or PE tube patency varied with the timing of post-operative audiologic testing.

Methods:

A retrospective chart review was completed for children undergoing PE tube surgery at a tertiary care pediatric hospital. The patient population included children birth to 6 years old who had at least one documented post-operative appointment with an audiologist. Patients were divided into two cohorts. Group A included children who were evaluated within the 6-week post-operative period, and Group B consisted of those who were evaluated beyond the 6-week post-operative period. A total of 184 patients were included in the study with 78 and 106 patients in Groups A and B, respectively. Charts were reviewed to determine hearing outcomes and PE tube patency.

Results:

Results indicated no significant difference in PE tube patency or hearing status between the two cohorts. Of those who completed audiologic testing, one child had a new diagnosis of sensorineural hearing loss.

Conclusion:

The timing of post-operative audiologic testing did not affect hearing outcomes or PE tube patency status.

# CONGENITAL PSEUDOARTHROSIS OF THE FIRST CERVICAL RIB PRESENTING AS A SUPRACLAVICULAR MASS

Wen Jiang (M.D.)

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Outcome Objective:

We describe a patient with a congenital first rib anomaly that presented as a left supraclavicular mass concerning for malignancy.

Methods:

Case Report

Results:

A 12-year-old female presented to the pediatric otolaryngology clinic with 6-month history of a left supraclavicular mass. There was no antecedent trauma and patient complained of pain while carrying her backpack. On exam, there was a definite fullness in the left supraclavicular fossa with a very firm, slightly tender, and fixed mass of 2.5cm noted along the anterior border of the trapezius. Prior to referral, pediatrician obtained both ultrasound and magnetic resonance imaging (MRI). Ultrasound did not reveal any significant abnormality and MRI showed mildly prominent left supraclavicular lymph nodes that appeared morphologically normal. However, due to her persistent symptom, unusual exam and parental concern for malignancy, an exploration of the left supraclavicular fossa with possible excisional biopsy was recommended. Intra-operatively, there were no abnormal lymph nodes identified. We encountered a bony prominence posterior to the left sternocleidomastoid muscle, between to the anterior and posterior scalene muscles, and just deep to the brachial plexus. Subsequent computed tomography demonstrated an incompletely developed left cervical rib with a pseudoarthrosis at its terminus with the second rib.

Conclusion:

Pseudoarthrosis is an uncommon congenital rib anomaly with the development of an irregular joint-like fusion between the first and second ribs. No immediate treatment is indicated. However, continued clinical follow-up is recommended since this abnormality may result in neurovascular compression and the development of thoracic outlet syndrome.

# PEDIATRIC HEAD AND NECK FIBROSARCOMAS: A DEMOGRAPHICAL, TREATMENT, AND SURVIVAL REVIEW

Tyler Janz

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Objective: To examine pediatric head and neck fibrosarcoma cases and review the demographics, management, and survival for these patients.

Methods: Pediatric patients in the Surveillance, Epidemiology, and End Results (SEER) database were included from 1973-2014 based on a diagnosis of a head and neck fibrosarcoma using ICD-O-3 head and neck primary sites and histology codes. Patients were included from ages 0-18 years.

Results: One hundred-eleven pediatric head and neck fibrosarcomas were identified within the SEER database over the study period. The mean age at diagnosis was 9.7 years (range: 0.0-18.0). Fifty (45.0%) patients were female. A majority (60.4%) of patients had dermatofibrosarcoma followed by 18 (16.2%) who had infantile fibrosarcomas. Nearly all patients (N=105, 94.6%) received surgical intervention. 27.5% of patients with an infantile fibrosarcoma received chemotherapy compared to 1.5% of patients with a dermatofibrosaroma (p=.004). The 5-year disease-specific survival was 97%.

Conclusions and Relevance: Pediatric patients with head and neck fibrosarcomas are most likely to present in Caucasian males or female during late childhood or early adolescence. Surgical management is common for pediatric head and neck fibrosarcomas. Additionally, chemotherapy may be used for infantile fibrosarcomas of the head and neck. Survival rates for pediatric patients with a head and neck fibrosarcoma are excellent.

# AUDIOLOGIC CREATIVITY DURING A MEDICAL MISSION: SOME LESSONS LEARNED

Alexa Murzyn

Alexa D. Murzyn (1)

1) Auditory Verbal Center, Inc.

Background: Medical mission work is often perceived as more giving than receiving when providing care and service not otherwise available to the people.

Purpose: Present insights from working a short-term medical mission to impoverished people with little access to healthcare. The poverty in Guatemala mirrors similar problems faced in both the United States, and other countries. Medical professionals face the dilemma of providing good care in an atmosphere of problematic politics while battling the challenges of episodic visits and limited resources in a foreign societal and medical culture. Audiologists need creative solutions to address equipment calibration, reconcile subjective versus test determined hearing problems, and practical acoustic amplification strategies.

Methods: Anecdotes include (1) trying to repair various types of hearing aids previously "fit" or purchased on the black market; (2) tethering hearing aids with paperclips and fishing line; (3) creation of domes from bulbous earpieces; (4) fabricating earmolds from irrigation syringes (5) making local language spondee lists; (6) cerumen management; and (7) counseling about noise toxicity precautions.

Summary of results: Beyond building trust, flexibility, creativity, and adaptability are crucial in mission trips into healthcare deserts.

Conclusion: The audiologist on a short-term medical mission receives more than gives. The needs for both individual patient and community hearing care are glaringly brought into focus.

# RECURRENT PERIORBITAL CELLULITIS IN A TODDLER

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1) Division of Otolaryngology, Dept of Surgery, Stony Brook Medicine

Periorbital cellulitis, is an acute infection of the soft tissue around the eye and anterior to the orbital septum. This is characterized by unilateral pain, swelling, and erythema of the upper and lower eyelid. The diagnosis of periorbital cellulitis is made primarily based on clinical findings, and can be supported by CT imaging. We present a healthy one year old child who experienced three episodes of periorbital cellulitis despite appropriate treatment with three weeks of antibiotics for each episode. The patient was noted to have complete recovery with no evidence of residual disease at completion of the antibiotic courses. Ultimately the patient underwent adenoidectomy and functional sinus surgery with no further infections during the two month follow up.

# REVIEW OF OUTPATIENT CLINICAL EMERGENCIES AT A TERTIARY CARE CENTER

Mary Frances Musso

Mary Frances Musso, DO(1) (2) Martin Worley, MBA(1) Susan Englemen(1) Mona Karimulah, MD(1) (2) Darlene Acorda RN(1) Gemma Elegores(1) Lauren Ivanhoe, MSN, RN-BC, CEN(1)

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## Background

The Otolaryngology clinic at a tertiary care center noted an increased incidence of outpatient clinical emergencies requiring activation of the incident management team (IMT). The IMT responds to emergent outpatient clinical situations to assess, triage, and provide needed support of care. Past activations included syncope, difficulty breathing, allergic reactions, and seizures. AIMS of this project included: reviewing past IMT events, evaluating staff confidence in activating and responding to emergent events and assessing current IMT process through staff survey.

## Methods

The project lead completed a retrospective review of outpatient IMT events at a tertiary care center from 1/2016-12/2016. A multidisciplinary project team created a needs assessment survey. The team piloted the survey with members of the IMT team and other staff. Post feedback analysis, the survey was sent out to all outpatient clinical staff.

## Results

Through retrospective review, the project team found 50 IMT events were voluntarily documented by the leader of IMT team, the house supervisor (HS) between

## Conclusion

1/2016-12/2016. HS recorded a larger percentage of IMT events in the following clinics: Pathology (16%), Allergy & Immunology (10%), Otolaryngology (10%), and Sleep lab (10%). Seventy percent of events involved a pediatric patient. Concerns from 157 survey respondents included consistent response time of the IMT team, availability of appropriate resources including medications, and improving treatment plans and procedures of adult patients.

Further investigation of the current IMT system is warranted. Better documentation of future events will allow a more thorough assessment of the current process.

# RIGHT-SIDED JUGULAR VENOUS ECTASIA IN A THREE-YEAR-OLD GIRL PRESENTING AS AN ENLARGING NECK MASS WITH CRYING

Steven P. Garbin

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Background: Jugular venous ectasia is an infrequently encountered but potentially underreported condition that generally presents as a soft swelling in the neck during straining (crying, screaming, etc.). Such ectasias are more common in males (1.4:1) and are more likely to be right-sided (5:1).

Clinical case: A 3-year-old girl presented to the pediatric otolaryngology department for evaluation of a right-sided neck mass of unknown duration noted by her parents to be worse with crying or screaming. The mass was described as soft and compressible. Swelling consistently subsided when the child was calmed. These findings were reproducible on physical exam with Valsalva maneuver. A contrast-enhanced CT scan revealed abnormal enlargement of the right internal jugular vein (IJV), nearly quadruple the size of the left IJV, extending from the level of the parotid gland to the region where it drains into the subclavian vein near the thoracic inlet. A baseline ultrasound was also obtained, revealing an ectatic right IJV which was substantially larger than the left IJV. The patient has no known underlying genetic or connective tissue disorders. At this time the parents and clinicians are in agreement to regularly monitor the lesion with observation and ultrasound.

Conclusion: Jugular venous ectasia is a potentially overlooked diagnosis in the setting of soft lower neck swelling in children. Diagnosis is made with neck ultrasound. Management is conservative, but serial ultrasounds should be performed to monitor the lesion. Surgery may be appropriate in the case of thrombus formation, phlebitis, cosmetic deformity, or rupture.

# SNHL RELATED TO ANATOMICAL VARIANT AND RARE GENETIC MUTATION

Mary Frances Musso

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Introduction:

The aplasia or hypoplasia of the vestibulocochlear nerve is a cause of Sensorineural Hearing Loss(SNHL). Genetic mutations including KCNQ1 are contributing factors to progressive SNHL.

Objective:

Present a case of SNHL related to anatomical variant and rare genetic mutation.

Case Report:

3 y/o male presented for evaluation of speech delay and failed hearing screen. Initial audiometry completed 2/1/17 was unable to detect responses to live voice below 30 dBHL and there were inconsistent responses to warbled tones and narrowband noise. Acoustic immittance measures yielded type A tympanograms and otoacoustic emissions were absent in the right ear and present in the left ear. Sedated ABR completed 2/7/17, notated normal sloping to moderate high-frequency

SNHL in the right and Auditory Neuropathy Spectrum Disorder in the left along with profound SNHL. MRI demonstrated no visible cochlear nerve on the left side, consistent with a hypoplastic or absent cochlear nerve. Patient was fitted with a hearing aid in the right ear on 06/16/2017. Subsequent, audiometry completed 2/13/18 revealed worsening of hearing in the right ear with mild sloping to profound SNHL. Genetic evaluation noted the patient was a carrier of a variant in KCNQ1 gene.

Discussion:

KCNQ1 is a voltage-gated potassium channel gene responsible for long QT syndrome. Heterozygous mutations in KCNQ1 typically cause Romano-Ward Syndrome, while homozygous mutations cause Jervell-Lange-Nielson syndrome. Herbert et al. found 100 families with mutations in this gene, most with novel private mutations.

Conclusion: It is important to consider anatomical and genetic factors when evaluating pediatric SNHL.

# INFANT WITH A RAPIDLY EXPANDING BUCCAL MASS: A RARE INITIAL PRESENTATION OF HEMOPHILIA A

Kimberly Luu (M.D.)

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PURPOSE:

To describe a rare etiology of an expanding buccal mass in an infant.

CLINICAL CASE:

Rapid growth of head and neck masses often requires emergent treatment but can sometimes present as a diagnostic dilemma. We report a case of an 8-month-old, previously healthy boy, who presented to the emergency department with left knee swelling and a rapidly expanding left buccal mass with overlying violaceous pigmentation. Initial investigations revealed extensive cystic and inflammatory changes on contrast-enhanced computed tomography scan concerning for infection and a possible congenital malformation, but in the context of a normal white blood count and C-reactive protein. The child was initially admitted to the pediatric intensive care unit and managed with intravenous antibiotics. Knee aspirate demonstrated a hemarthrosis and partial thromboplastin time was elevated, leading to the diagnosis of hemophilia A, with a severe factor VIII deficiency. Over the next day, the cheek mass evolved into an appearance consistent with a hematoma. The patient had no family history of coagulopathies and a normal circumcision.

CONCLUSION:

This case is the first known report of a rapidly expanding buccal hematoma as an initial presentation of hemophilia A. Hemophilia A is an X-linked, recessive disorder caused by deficiency of functional clotting factor VIII. It is an important entity for otolaryngologists to consider as it can first present as epistaxis, mucosal bleeding, or hemoptysis. This presentation emphasizes the need to maintain a broad differential diagnosis when encountering newly presenting head and neck lesions.

# BENIGN AND MALIGNANT SALIVARY MYOEPITHELIAL TUMORS IN CHILDREN: CASE REPORTS AND REVIEW OF THE LITERATURE

Steven M Andreoli (M.D.)

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Background: Pediatric myoepithelial tumors of the salivary glands are exceedingly rare with few reported cases. Benign myoepithelioma represents 1% of all salivary tumors and are treated with surgical resection. Salivary myoepithelial carcinoma is a rare locally aggressive tumor without a clearly defined treatment protocol. We present two cases: a benign myoepithelioma of the parotid and a myoepithelial carcinoma of the submandibular gland.

Case Histories:

Myoepithelioma: A 17 year old female presented with a six month history of a slowly enlarging left parotid mass. Outside fine needle aspirate pathology was consistent with either a basaloid or myoepithelial parotid neoplasm. MRI demonstrated a 2 cm diffusely enhancing parotid mass. She underwent a superficial parotidectomy via a microparotidectomy approach. Pathological evaluation demonstrated a completely excised benign myoepithelioma.

Myoepithelial carcinoma: A 9 year old female presented with a twice recurrent submandibular mass. She had undergone outside partial resections one and two years prior with pathology consistent with pleomorphic adenoma. MRI demonstrated a multifocal tumor involving the submandibular space invading the intrinsic tongue muscles. She underwent a transcervical resection, neck dissection, and pedicled sternocleidomastoid muscle flap for intrinsic tongue reconstruction. Pathology was consistent with stage IVB T3N2bM0 high grade myoepithelial carcinoma. She underwent adjuvant proton based radiation and seven cycles of chemotherapy with etoposide, cisplatin, and ifosfamide with no evidence of recurrence at two years.

Conclusion: Myoepithelial origin salivary tumors are very rare in children. We present a case series of benign and malignant variants with review of the literature.

# OTITIS MEDIA IN CHILDREN WITH UNILATERAL ISOLATED CHOANAL ATRESIA

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Background: The blocked nose is considered a contributor to the occurrence of otitis media. Choanal atresia puts patient in the worst group in Bellucci's classic classification of success of tympanoplasty.

Purpose: To describe the occurrence of otitis media in children with unilateral isolated non-syndromic choanal atresia.

Materials and Methods: For 25 children who underwent computed tomography for choanal atresia, 2000-2017, two indicators of otitis media were assessed: (1) water density in mesotympanum as seen at computed tomography; and, (2) tympanostomy-tube surgery.

Results: One patient had water density in each mesotympanums at CT. Only two patients underwent tympanostomy-tube placement during 124 patient-years, a rate lower than reported in the general population, $P<0.001$.

Conclusion: In contrast to children with syndromic or bilateral choanal atresia, children with unilateral isolated non-syndromic choanal atresia have less otitis media than the general population. The idea that blocked the nose contributes to otitis media, is questioned.

# SILENT STRIDOR: A DELAYED PRESENTATION OF TRACHEAL STENOSIS IN A CHILD BORN TO DEAF PARENTS

Madeline Pyle

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Introduction: Severe congenital tracheal stenosis traditionally presents within hours to days after birth with stridor, cyanotic spells, and apneic episodes. The disease can be life-threatening and usually requires prompt surgical repair.

Case details: We report the case of a 2-month-old premature male born to deaf parents who presented after noisy breathing was noted by an American Sign Language interpreter at an orthopaedic appointment. As his respiratory distress worsened, he aspirated during feeding resulting in acute respiratory failure with hypoxia. His symptoms prompted evaluation with bronchoscopy, which identified a severe distal tracheal stenosis. CT angiography better defined his airway anatomy, and he was found to have a 2.5mm short segment of severe narrowing of the distal trachea and carina without complete tracheal rings. Slide tracheoplasty and carinaplasty with interdigitated anastomosis was performed by cardiothoracic surgery. Post-operative bronchoscopy and CT imaging showed a normal tracheal lumen. There have been no new respiratory symptoms since surgery and follow-up rigid bronchoscopy at nine months of age demonstrated a patent airway at the site of surgical repair.

Conclusion: This case demonstrates a unique presentation of severe congenital tracheal stenosis with an excellent outcome in a 2-month-old patient born to deaf parents that were unable to hear symptoms of noisy breathing resulting in a delayed diagnosis. The excellent outcome exemplifies the necessity of a multidisciplinary approach to the care of these patients presenting with this rare anomaly.

# NASAL SEPTAL ANGIOFIBROMA: CASE REPORT AND LITERATURE REVIEW

Kaitlyn A. Brooks

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Background: Angiofibroma of the nasal septum is a rare nasal mass, with only 15 cases reported in the literature. While the histopathology is similar to juvenile nasopharyngeal angiofibroma, nasal septal angiofibroma occurs equally in both genders and has been reported in pediatric, adolescent, and middle-aged patients. We present a rare case of this unusual lesion.

Methods: Case report and review of literature.

Results: A 10-year-old male presented to the emergency department with an enlarging left anterior nasal mass and 1-month history of unilateral worsening epistaxis and nasal obstruction. On physical exam, the patient had a dark red, friable mass obstructing the left nasal vestibule. Facial MRI identified an enhancing, lobular soft tissue mass in the left nasal cavity attached to the nasal septum. Nasal endoscopy confirmed a 2 cm by 1.8 cm mass with its base attached at the anterior septum and a clinical appearance suggestive of papilloma. Radiological and gross examination increased suspicion for possible inverted papilloma or pyogenic granuloma. The mass was excised by detaching the base using an Aquamantis and removing the mass in its entirety. Endoscopic evaluation showed no further nasal cavity or nasopharyngeal extension of the mass. Histological examination of the specimen revealed a nasal septal angiofibroma.

Conclusions: Although extremely rare, nasal septal angiofibromas should be considered in the differential diagnosis of anterior nasal cavity masses among all patient demographics, even if the mass appears similar on gross examination to common nasal cavity tumors. Existing literature regarding these rare lesions will be reviewed.

# SUBCUTANEOUS EMPHYSEMA AND VOCAL FOLD PARESIS AS A COMPLICATION OF A DENTAL PROCEDURE

Nick Van Maele

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1) Central Michigan University 2) Medical College of Wisconsin

A 17 y.o. male without a significant past medical history presented with pain and swelling of his face and neck after third molar extraction with a pneumatic drill under local sedation. The procedure was halted after the patient was noted to have significant right facial swelling, and he was discharged on amoxicillin, hydrocodone, and decadron. He presented to the emergency room with progressive dysphagia and odynophagia. CT neck demonstrated extensive subcutaneous emphysema of right orbital, masticator, ptyregoid, parapharyngeal, retropharyngeal spaces and bilateral carotid and visceral spaces, in addition to pneumomediastinum. He was treated with transoral drainage and intravenous antibiotics, and he was discharged to home after resolution of subcutaneous emphysema. Due to persistent dysphonia, flexible laryngoscopy was performed, which demonstrated a left vocal fold paresis. Emphysema is a rare complication of restorative dentistry, periodontal surgery, endodontic treatment, repair of facial fractures, temporomandibular joint surgery, extraction of teeth and other procedures. Additional complications of third molar removal include pain, infection, bleeding, dry socket, nerve injuries and trismus. While several case reports document cervicofacial emphysema after dental procedure, vocal cord paresis has not been previously described. Here we describe a rare complication after inferior third molar extraction, demonstrating the importance of clinical monitoring in these cases.

# BILATERAL SILENT SINUS SYNDROME IN A CHILD WITH DOWN SYNDROME

Laurie Newton

Laurie Newton, APNP (1) Elizabeth Schueth, MS4 (2) Kayleigh Stabenau, MD (1) Cecille Sulman, MD (1)

1) Medical College of Wisconsin 2) Indiana University

A 9 year old female with a history of Down syndrome and chronic sinusitis with a history of prior bilateral maxillary antrostomies, presented with chronic sinus congestion (Insert additional history). Computed tomography of the sinuses demonstrated absent bilateral maxillary sinuses whereas review of prior imaging revealed hypoplastic maxillary sinuses that over time decreased in size. This presentation is consistent with silent sinus syndrome, a rare condition that usually presents in the third to fifth decade of life, and can pose a diagnostic challenge. Patients usually present with ocular symptoms, such as unilateral ptosis or retraction, a deep superior sulcus, or orbital asymmetry. This condition is characterized by unilateral spontaneous enophthalmos and hypoglobus due to increased orbital volume and retraction of the orbital floor. The pathophysiology is thought to be due to atelectasis of the ipsilateral maxillary sinus and, when the condition is left untreated, may result in complete obliteration of the sinus with worsening enophthalmos and hypoglobus. This is an unusual case of bilateral silent sinus syndrome in a pediatric patient. The patient did not manifest ocular symptoms; however, she will require monitoring of ocular changes moving forward.

# NEUROFIBROMATOSIS PRESENTING WITH PAROTID MASS: CASE REPORT AND LITERATURE REVIEW

Joshua B. Smith

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Background: Neurofibromas are an uncommon cause of masses in the head and neck which are often associated with Neurofibromatosis Type 1 (NF1). Furthermore, neurofibromas arising in the salivary glands are exceedingly rare, comprising only 0.4% of all salivary gland lesions. Diagnosis can be difficult due to non-specific presentation and radiologic findings. Pediatric parotid neurofibromas are rarely reported in the literature. We present an interesting case of this unusual lesion.

Methods: Case report and review of literature.

Results: A 5 year old male with autism presented to clinic with a left parotid mass initially thought to be a lymphatic malformation, however workup showed lack of vascular/lymphatic channels. Open biopsy of the lesion revealed a neurofibroma, patient was then diagnosed with NF1 by genetics. MRI showed a large left parotid mass with extension to skull base, parapharyngeal and carotid spaces. The patient was recommended for left parotidectomy and neck dissection. Intraoperatively, the mass was found to arise from the lower division of the facial nerve, coinciding with non-functional nerve branches on nerve stimulation. The temporal, zygomatic and buccal branches were identified and preserved. Neck dissection was performed, removing tumor from parapharyngeal, pre and post-styloid spaces. CN X-XII were identified and preserved. Facial function was intact postoperatively.

Conclusions: Neurofibromas presenting as a parotid mass are uncommon and often mimic other benign tumors or cysts. Neurofibromas should be considered in the differential diagnosis of patients demonstrating a parotid mass, especially in the presence of syndromic features. Existing literature regarding these lesions will be reviewed.

# COMBINED ENDOSCOPIC-MICROSCOPIC APPROACH TO ADVANCED CONGENITAL CHOLESTEATOMA RESECTION: CASE REPORT AND LITERATURE REVIEW

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Background: With an incidence of 0.12 per 100,000, congenital cholesteatoma is an uncommon otologic pathology thought to originate from residual embryological epithelial tissue. These lesions are benign, but bone expansion and erosion may cause compression of nearby structures. We describe a case of congenital cholesteatoma treated surgically at our institution with a combined microscopic and endoscopic approach.

Methods: Case report and literature review.

Results: A 14-year-old male was seen for right-sided conductive hearing loss. The patient was offered surgery in 2013, but declined at that time. Over the next 5 years, the hearing loss worsened, prompting the patient to return. On otoscopic examination, the right ear demonstrated subtle changes in ossicular chain landmarks. CT scan demonstrated a mass sitting on the head of the malleus and a small calcified lesion limiting ossicular chain mobility. The patient was referred for exploration tympanoplasty with possible ossicle chain reconstruction (OCR) and possible mastoidectomy. The resection was started endoscopically and the incus and malleus head were removed. The mass extended beyond the visual field afforded by endoscopy, so the decision was made to perform mastoidectomy and proceed microscopically. OCR was attempted but was too unstable to be successfully performed; the patient will undergo a second look procedure with OCR in the near future.

Conclusions: Congenital cholesteatomas are somewhat rare and often go unnoticed until they are very large. Surgical treatment should find an appropriate balance between sufficient resection and morbidity. A combined endoscopic-microscopic approach may prevent or reduce the incidence of complications or recurrences.

# IMPROVING AVS DOCUMENTATION IN A HIGH-VOLUME AMBULATORY CHILDREN'S HOSPITAL SURGICAL SUBSPECIALTY CLINIC SETTING

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An after visit summary (AVS) document contains important medical information and is provided for every patient as they are discharged from a clinic visit. The AVS is a required component of the Centers for Medicare and Medicaid Services (CMS) meaningful use criteria for physician/hospital reimbursement. After review of our AVS, we aimed to provide the patient/family with the information they would find most useful. In a fast-paced clinic that schedules over 32,000 patients annually, this was not an easy task.

The goal for improvement was to increase our compliance from 0% to 100% based on the EMR chart auditing. For an AVS to be considered compliant all 12 components had to be present:

1. Visit diagnosis
2. When to follow-up
3. Centralized scheduling's number
4. Nurse triage number
5. Department website link
6. New orders
7. New medications
8. Medication reconciliation
9. Medications given during visit
10. Three vital signs
11. Allergies
12. Special care instructions

Monthly independent audits were conducted for each provider. Formal nurse and surgeon education, creation of detailed patient education information and improved communication between surgeon and nurse significantly improved results.

The process was initiated in October 2015 and since that time several interventions have been implemented. Compliance with all quality parameters on audits went from 0% at baseline to 65% (p-value≤=0.001). The majority the AVS that were not perfect missed 1 or 2 of the 12 components. Audits of the data will continue with the goal of further improvement in the quality of AVS documentation.

# PATIENT COMPLIANCE WITH EMERGENCY TRACHEOSTOMY/AIRWAY QUICK REFERENCE CARDS

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Background

Patients with tracheostomies may have complex airway anatomy and communication with first responders and emergency care providers can be challenging. Tracheostomy/Airway quick reference cards including tracheostomy type/size/suction depth, airway status, and otolaryngologist contact information were developed and provided to patients prior to discharge.

Objective

The aim of this study is to evaluate patient/caregiver compliance with the tracheostomy/airway quick reference cards.

Design

Prospective survey-based study carried out in the Otolaryngology outpatient clinic at a tertiary hospital.

Methods

Tracheostomy/Airway cards were initiated in July 2017 to promote rapid and clear communication of trach and airway related information to caregivers and first responders. A total of 56 cards were provided and data was collected by oral survey at the time of clinic follow up. Caregivers were asked about airway card possession, input on content, location where card was kept, a situation where they found the card to be useful.

Results

From July 2017 to July 2018, a total of 56 airway/tracheostomy cards were provided to tracheostomized patients at the time of discharge. A total of 25 patients were interviewed during clinic follow up. 84% or 21 of 25 had the airway card in procession during the survey and found the card useful in communication with home health nurses and emergency room providers.

Conclusions

Our findings indicate that the tracheostomy/airway quick reference card promotes communication with caregivers, especially with home health nurses and emergency room providers. The program is ongoing, and the format of the card continues to evolve based on survey input.

# ABNORMALITIES OF THE THORACIC AIRWAY: CONTEMPORARY MULTI-DISCIPLINARY MANAGEMENT WITH FOCUS ON SURGICAL TREATMENT

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Anomalies of the thoracic airway, including tracheal abnormalities, foregut abnormalities, vascular rings and other vascular anomalies can cause tracheal or esophageal compression, most commonly manifesting in noisy breathing, other respiratory symptoms or dysphagia. They can result in urgent need for intervention

Typically diagnosed in infants, surgical treatment is the mainstay of therapy. For vascular anomalies, surgery is usually performed by Cardiothoracic Surgeons, but the Otolaryngologist often plays a vital role often by diagnosing, evaluating the airway before and after treatment with bronchoscopy.

At some institutions, Otolaryngologists and Cardiothoracic Surgeons collaborate for surgical repair of tracheal anomalies, however, this is not universal. These patients are often complex and require multidisciplinary care both preoperatively and postoperatively for optimal management.

Of particular interest to pediatric otolaryngologists, who are on the 'front line' for airway management, the purpose of this mini-seminar will be to provide a foundation for diagnostic techniques, imaging, surgical decision making , and medical and surgical treatment of complex thoracic airway abnormalities.

We'll present information in a case-based format with bronchoscopic and radiographic imaging, with input from surgeons from different specialties.

# UNPLANNED REVISITS FOLLOWING AMBULATORY PEDIATRIC SINUS SURGERY

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Objective: To determine the incidence, diagnoses, and risk factors associated with 30-day unplanned revisits following ambulatory pediatric sinus surgery.

Methods: We analyzed the State Ambulatory Surgery and Services, Emergency Department, and Inpatient Database from California for children less than 18 years of age who underwent sinus surgeries between the years 2008 and 2011. Rates, diagnoses, and patient-level risk factors for 30-day readmissions were examined.

Results: A total of 6,924 ambulatory sinus surgeries were performed in the 3 year period (mean: 3 years, 60.7% male). The 30-day unplanned revisit rate was 5.3% after surgery. Of all readmissions, 53.4% (173) were due to adenoidectomy. The most common readmission diagnoses were due to direct anesthesia complications (12.3%) [i.e. nausea, vomiting] and postoperative bleeding (10.2%). On univariate analysis, factors statistically significantly associated with hospital revisit were Black race (vs white: OR 1.92; 95% CI 1.12 - 3.29), Hispanic race (vs. white: OR 1.56; 95% CI 1.16 - 2.11), Adenoidectomy (OR 0.53; 95% CI 0.32- 0.87), and Private payer status (OR 0.47; 95% CI 0.37 to 0.60). On multivariate regression, statistically significant negative predictors for revisit were adenoidectomy (OR 0.37; 95% CI 0.19 - 0.71) and Private payer status (OR 0.44; 95% CI 0.31 - 0.61).

Conclusion: Our study demonstrated that 1 in 19 patients undergoing ambulatory sinus surgery revisit the hospital within 30 days. Revisits are most commonly due to anesthesia complications and post-operative bleeding. These specific complications should serve as targets for quality improvement after ambulatory sinus surgery.

# TEMPOROPARIETAL FASCIAL FLAP UTILIZATION IN STAGED REPAIR OF GRADE II MICROTIA: CASE REPORT AND LITERATURE REVIEW

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Background: Auricular reconstruction remains a significant challenge in the field of facial plastic surgery. The complex anatomy of the external ear, combined with the need to provide adequate projection and symmetry, make this type of reconstruction particularly difficult. Despite the technical challenges, recent advances in surgical techniques provide numerous options for reconstructive techniques. We describe a case of grade II microtia repaired using rib cartilage and a temporoparietal fascial (TPF) flap.

Methods: Case report and literature review.

Results: A 9-year-old boy with a complex past medical history including CHARGE syndrome presented to clinic seeking reconstruction options for right-sided microtia. On physical exam, he was found to have grade II right microtia with right external auditory canal atresia. Cartilage grafts from prior reconstruction were in place superiorly. We decided to use rib cartilage and a TPF flap to augment the existing right auricular framework. Intraoperatively, a 7x4cm TPF flap was elevated, a piece of rib cartilage was harvested, and a full-thickness skin graft was taken from the right chest. The harvested rib cartilage was used to augment the existing auricular framework and was subsequently covered by the TPF flap and skin graft, which was sutured in with 4-0 chromic sutures.

Conclusions: Various options for auricular reconstruction exist, such as autologous rib cartilage, alloplastic grafting, or prosthetic restoration. An effective approach to auricular reconstruction requires an adept surgeon experienced with multiple reconstructive techniques, affording each patient the best procedure available for them. Existing literature regarding reconstructive options and advances will be reviewed.

# A UNIQUE PRESENTATION OF LINGUAL THYROID IN AN INFANT

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Background: Failure of the thyroid to descend from the base of tongue to the neck during embryogenesis can lead to an ectopic lingual thyroid. This is a rare anomaly that most commonly presents with dysphagia or dysphonia, although lingual thyroid can be asymptomatic with a variable presentation.

Clinical Case: A 6-month-old female infant presented to the Otolaryngology clinic with hoarseness. She had a history significant for congenital hypothyroidism detected on newborn screening and treated with thyroid hormone replacement. Upon gross examination of the oral cavity, no abnormality or mass was observed. On laryngeal stroboscopy she was noted to have a left vocal cord cyst and a secondary reactive nodule on the right vocal cord. Additionally, a well-circumscribed mass at the base of the tongue was identified. Review of a prior neck ultrasound demonstrated absence of thyroid tissue in the neck, leading to a diagnosis of lingual thyroid. At 6 month follow-up, she had spontaneous resolution of her hoarseness and vocal cord cyst, and continued asymptomatic prominence of the lingual thyroid.

Conclusion: This case highlights the variable presentation of lingual thyroid in congenital hypothyroidism and the importance of physician awareness of this potential diagnosis. Clinical suspicion for this rare embryologic anomaly must be shared across various pediatric specialties in order to ensure the identification of these patients. In cases of thyroid absence on ultrasound, an evaluation by an Otolaryngologist should be considered.

# Z-PLASTY WITH GEOMETRIC BROKEN LINE CLOSURE OF A UNILATERAL TESSIER 7 CLEFT: CASE REPORT AND LITERATURE REVIEW

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Introduction: Tessier clefts are rare, with an incidence of only 1.43 to 4.85 per 100,000 live births, and Tessier 7 clefts comprise only 0.3-1.0% of the total facial cleft deformity spectrum. Facial clefts typically run parallel to relaxed skin tension lines (RSTLs), whereas Tessier 7 clefts are perpendicular to these lines. Z-plasty, w-plasty, and straight line closures have been reported in the literature for reparation. Geometric broken line closure (GBLC) creates a randomly irregular scar by interdigitating triangles and trapezoids in a random pattern to optimize the ultimate scar.

Case Presentation: We present a review of the literature and a case of a four-month-old female patient with Goldenhar, right-sided Tessier 7 cleft, macrostomia, preauricular appendages, a branchial cleft remnant on the right cheek, and a type 1A laryngeal cleft, for which a combined z-plasty and GBLC were chosen for surgical repair. The Tessier 7 cleft involved muscular diastasis at the commissure extending laterally toward the tragus. We describe a novel reconstruction technique wherein z-plasty reorients part of the scar parallel to RSTLs, and GBLC further camouflages the scar perpendicular to RSTLs.

Conclusion: Z-plasty repositions a portion of the scar to be parallel to the eventual nasolabial fold and RSTLs. GBLC further breaks up a scar that is otherwise both linear and perpendicular to RSTLs by creating small geometric shapes, which also makes the scar less noticeable. We present this case to expand the armamentarium of surgical options to address Tessier 7 clefts.

# TEACHING PRINCIPLES OF MEDICAL PHOTOGRAPHY TO PHYSICIANS: PRE AND POST - EVALUATION PILOT STUDY AND A SURVEY

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Introduction:

Current technology made digital photography broadly accessible, photographers along with physicians have been trying to establish gold standards when obtaining medical photos. Basic understanding of photographic principles, continuous practice of different techniques, and usage of accurately selected equipment contribute significantly to the standardization of photographs. Otolaryngologists do not have access to a validated and conclusive guide.

Objective:

To assess physicians' ability of crafting quality photographs before and after medical photography hands-on workshop, in order to validate a best-practice guide.

Study design:

A pilot study with pre-post-test quasi-experimental design, and a survey.

Methodology:

Twenty practicing physicians participated voluntarily in a hands-on training session. The latter presented a medical photography best-practice guide that was initially created by the authors. Structured assignment to evaluate understanding of ethical and technical principles of medical photography was given to all participants before and after the workshop. Assignments were evaluated and the results were compared. Additionally, a survey was sent to attendees, which collected their feedback and recommendations regarding the training session.

Results:

Nine out of twenty participants were able to reach a score of 100% after attending the workshop. Paired two tailed t-test for pre and post assignments revealed a statistically significant difference between participants' skills before and after the workshop. Nine attendees (45%) responded to the survey after the workshop and showed their satisfaction with the training session.

Conclusion:

Producing a high-quality medical photograph improved significantly and immediately in physicians using a medical photography guide acquired in a hands-on workshop.

# PEDIATRIC NASAL BURNS DURING OPERATIVE CAUTERY; ARE AURAL SPECULUMS MORE PROTECTIVE THAN NASAL SPECULUMS?

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INTRODUCTION: Operative intranasal electrocautery may be performed for turbinate reduction or epistaxis. As access occurs through the nares, the nasal alar skin is at risk for injury. To better protect the skin, we switched from the standard metal nasal speculum to a modified plastic aural speculum. Our aim with this study is to evaluate the number of nasal burns using each technique.

METHODS: Single center retrospective chart review of patients, aged 0-18, who underwent operative intranasal cautery between March 2011-March 2018 by a single attending surgeon. The incidence of external nasal burns was compared between groups.

RESULTS: 203 charts were reviewed. 81 cases involved the metal speculum, and 2 (.025%) were complicated by nasal skin burn. Both cases were small burns which went on to heal acceptably. 122 cases involved the aural speculum, and there were no burns. There were no other complications in either group. Fisher two tailed exact test shows p=0.158.

CONCLUSION: The standard metal nasal speculum provides surgical access, but the open design and conducting material does not provide skin protection. Burns, even small ones, should be prevented if possible. In this study, the use of a modified aural speculum resulted in fewer external burns, but statistical significance was not achieved. We believe this is a result of study underpower more than lack of effectiveness. We have embraced the aural speculum as a safer alternative; we contend that this is a worthwhile safety intervention based on both the protective material and the achievement of zero burns.

# DELAYED COMPLICATION OF TRACHEOCUTANEOUS FISTULA CLOSURE WITH SEVERE COMPROMISING SUBCUTANEOUS EMPHYSEMA

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Tracheocutaneous fistula (TCF) is commonly seen in pediatric patients with long term tracheostomy after decannulation. The fistula may be surgically closed by a variety of techniques. We report a significant complication of TCF excision in a 4 year old male with a history of restrictive lung disease due to a giant omphalocele who had been tracheostomy dependent since infancy. He was decannulated at age 3, and one year after decannulation had a persistent 3mm TCF. He underwent surgery to repair the TCF with excision of the fistula tract and intended closure by secondary intention. On post-operative day 2 the patient rapidly developed profound subcutaneous emphysema of the face, neck, arms and chest, as well as pneumomediastinum. Due to the rapid onset there was concern that swelling may compromise the airway leading to urgent intubation. He was extubated after 3 days as the swelling resolved and discharged from the hospital on post-operative day 7 in stable condition. At follow up he had complete resolution of subcutaneous emphysema as well as complete closure of the TCF. Subcutaneous emphysema is a rare complication of TCF repair. We discuss the main methods of TCF closure with the indications, benefits, and complications of each. Management of subcutaneous emphysema is also discussed along with the lessons learned from this case.

# DYSPHAGIA IN CHILDREN AFTER CONGENITAL CARDIAC SURGERY: A SYSTEMATIC REVIEW

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Purpose: As the morbidity and mortality of congenital cardiac surgery has improved, attention to postoperative feeding outcomes has become increasingly important. While centers have embarked on various strategies to achieve oral feeding postoperatively, there is still wide variability in common practice.

Methods: The literature was searched using the MEDLINE, EMBASE, and CINAHL databases for publications up to March 2018. All papers discussing pediatric patients with congenital heart defects, surgical intervention, and swallowing dysfunction were reviewed in a systematic fashion. Exclusion criteria were non-English publications, only >18-year old patients, and no outcome data regarding feeding or swallowing.

Results: In total, 19 studies were included. Of these, 6 studies focused on dysphagia specifically, but notably included heterogenous assessment methods and outcomes. 9 studies discussed vocal fold dysfunction (VFD). Among those, only 5 assessed swallowing function discreetly and none found dysphagia in more than half of the patients with VFD. There were 5 studies evaluating feeding disorders in general, and only 1 study evaluated long-term results greater than 2 years.

Conclusions: The available data on dysphagia after congenital cardiac surgery is heterogenous and sparse. In particular, there is a lack of common outcomes that are reported. Additional studies that focus on dysphagia in this population are needed to better isolate the risk factors and to develop comprehensive postoperative feeding strategies. A consensus of standardized, trackable outcomes will provide the basis for evidence driven feeding programs in this fragile population.

# ACQUIRED CHOLESTEATOMA IN SIBLINGS

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Objective: We report a rare familial grouping of acquired cholesteatoma.

Methods: Case reports and review of the literature concerning cholesteatoma and rare instances of hereditary grouping.

Results: Brothers separated in age by 2 years presented with history of of recurrent ear infections. Both were treated with placement of ear tubes. Subsequently, after tube extrusion, the brothers each developed extensive cholesteatoma disease of the right ear. Each child was 7 years of age when diagnosis of cholesteatoma was made and surgical intervention occurred.

Conclusion: Pathogenesis of cholesteatoma is the subject of continuing controversy however genetic causes are not usually emphasized. Reports of familial grouping of cholesteatoma are quite rare but reports in the literature may indicate a genetic predisposition is present for some individuals.

# SLEEP DISORDERED BREATHING IN TRISOMY 13 AND TRISOMY 18

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Purpose: Trisomy 18 (T18) and trisomy 13 (T13) are the most common autosomal trisomy syndromes after trisomy 21. While the prevalence of sleep disordered breathing (SDB) is well documented in trisomy 21, there has been little published on the incidence in T13 and T18. These syndromes have overlapping features that make patients prone to SDB. Due to the limited nature of the literature, we performed a retrospective chart review.

Methods: We reviewed the charts of children with T13 or T18 seen at our institution for SDB from 1/1/10 to 5/1/18. Candidates were selected based on ICD-9 diagnosis and procedural codes.

Findings: Eight patients met the inclusion criteria. All patients had clinical features that made them predisposed to SDB, including three patients with laryngomalacia or tracheomalacia, one with inferior turbinate hypertrophy, one with adenoid and lingual tonsil hypertrophy, five with a small mouth or micrognathia, six with hypotonia, and five with GERD. Seven patients were recommended a sleep study, six underwent endoscopy, one was fitted for a CPAP, and two were BiPAP dependent. One patient underwent adenoidectomy and lingual tonsillectomy. Two patients had tracheostomies. Two patients were intubated for respiratory insufficiency.

Discussion: The results suggest that T13 and T18 patients are at increased risk for SDB due to micrognathia, laryngomalacia, tracheomalacia, hypotonia, and GERD. These findings indicate a need for otolaryngologist intervention to increase both survival and quality of life.

Conclusion: As more T13 and T18 patients become surgical candidates, the present study provides an initial foundation for clinical decision making.