Pan Arab Rhinology Society
PARS Reader's Digest

Reader Digest

Digested by Dr. Tarek Kandil, MD. ENT Consultant, students Hospital, Cairo University

Introduction

This newsletter is intended to provide information that is useful to the student and specialist in the field of rhinology and allergic disorders.

The selected recent material represents important fundamental knowledge, current trends or recent developments in this field.

We hope that this newsletter will help the reader have a greater understanding of rhinology and allergic disorders .

1. Treacher Collins Syndrome

Sara Huston Katsanis 1, Ethylin Wang Jabs 2

Margaret P Adam, David B Everman, Ghayda M Mirzaa, Roberta A Pagon, Stephanie E Wallace, Lora JH Bean, Karen W Gripp, Anne Amemiya , editors.

Excerpt

Clinical characteristics: Treacher Collins syndrome (TCS) is characterized by bilateral and symmetric downslanting palpebral fissures, malar hypoplasia, micrognathia, and external ear abnormalities. Hypoplasia of the zygomatic bones and mandible can cause significant feeding and respiratory difficulties. About 40%-50% of individuals have conductive hearing loss attributed most commonly to malformation of the ossicles and hypoplasia of the middle ear cavities. Inner ear structures tend to be normal. Other, less common abnormalities include cleft palate and unilateral or bilateral choanal stenosis or atresia. Typically intellect is normal.

Diagnosis/testing: The diagnosis of TCS is established in about 97% of probands by detection of a heterozygous (autosomal dominant) pathogenic variant in TCOF1, POLR1D, or POLR1B or biallelic (autosomal recessive) pathogenic variants in POLR1C or POLR1D using molecular genetic testing and in about 3% of probands by clinical findings when molecular genetic testing has not been performed or does not identify pathogenic variants in one of the known genes.

Management: Treatment of manifestations: Treatment should be tailored to the specific needs of each individual, preferably by a multidisciplinary craniofacial management team. Neonates with airway issues may require special positioning or tracheostomy to facilitate ventilation.

Hearing loss is treated with bone conduction amplification, speech therapy, and educational intervention. Craniofacial reconstruction is often necessary. Cleft palate repair (if needed) occurs at about age one year; zygomatic and orbital reconstruction at about age five to seven years; and bilateral microtia and/or narrow ear canal reconstruction after age six years. The age of maxillomandibular reconstruction varies by severity; orthognathic therapies are typically before age 16 years.

Genetic counseling: Treacher Collins syndrome (TCS) can be inherited in an autosomal dominant or autosomal recessive manner.

Autosomal dominant TCS. About 55%-61% of probands have the disorder as the result of a de novo TCOF1, POLR1D, or POLR1B pathogenic variant. Each child of an individual with TCS has a 50% chance of inheriting the pathogenic variant.

Autosomal recessive TCS. The parents of a child with autosomal recessive TCS are obligate heterozygotes (i.e., carriers of one POLR1C or POLR1D pathogenic variant). At conception, each sib of an affected individual has a 25% chance of being affected, a 50% chance of being an asymptomatic carrier, and a 25% chance of being unaffected and not a carrier.

Once the TCS-related pathogenic variant(s) have been identified in an affected family member, prenatal and preimplantation genetic testing are possible

GeneReviews® [Internet]. Seattle (WA): University of Washington, Seattle; 1993.

2004 Jul 20 [updated 2020 Aug 20].

2. Efficacy of X-ray in the diagnosis of pediatric nasal fracture

Lizbeth Leapo 1, Mason Uemura 2, Maximilian C Stahl 3, Nirav Patil 4, Jay Shah 5, Todd Otteson 6

Abstract

Objective: Nasal fracture is one of the most common pediatric fractures, and diagnosis can be made with clinical findings or with radiographic imaging. The objective of this study is to determine the extent of x-ray utilization in decision-making regarding closed reduction of pediatric nasal fracture.

Methods: This a case-control study of 117 patients ages 0-18 with a diagnosis of nasal fracture seen at University Hospitals Cleveland Medical Center between January 2015 and January 2020. The exposure group had x-ray imaging of the nasal bones, and the control group had no x-ray imaging.

Results: A total of 59 (50.4%) patients had surgical intervention. The odds ratio to compare x-ray utilization to the control group for patients who underwent closed reduction surgery was .3951 (95% CI: 0.1848-0.8448, p-value = .0166).

Conclusion: The statistical analysis suggests that x-ray use is associated with decreased rates of closed reduction surgery. It is likely that while not necessary for the diagnosis of nasal fracture, x-ray serves as an additional data point for diagnosis confirmation, and may reduce the rate of unnecessary surgery for false positive cases of nasal fracture

Int J Pediatr Otorhinolaryngol. 2022 Nov;162:111305.

3. Effects of subcutaneous immunotherapy in allergic rhinitis children sensitive to dust mites

Yanfen Lin 1 2, Jinling Liu 1 2, Jing He 1 2, Lei Wu 1 2, Shuxian Li 1 2, BeiLei Cheng 1 2, Yanan Shao 1 2, Yuanyuan Zhang 1 2, Yingshuo Wang 1 2, Lanfang Tang 1 2, Zhimin Chen 1 3

Abstract

Background: Subcutaneous immunotherapy (SCIT) is now the only treatment that can modify the natural course of allergic rhinitis (AR). However, not all children with AR benefit from SCIT.

Objective: To evaluate the efficacy of SCIT in dust-mites-induced AR children and explore correlative factors predicting treatment response to SCIT.

Methods: 225 children aged 4-17 years old with AR were recruited from January 2016 to September 2019, and monitored at baseline, 4, 12, and 24 months after the start of SCIT treatment. The visual-analogue-score (VAS) was used to assess the clinical symptoms. Multivariate binary logistic regression analyses and receiver operating characteristic curves were used to explore correlative factors in predicting the efficacy of SCIT.

Results: The significant declines in VAS started after 4 months of SCIT and continued to improve throughout the study compared with baseline. An increase in children's age (OR=0.688, 95%CI: 0.479-0.988) and those with allergic history (OR=0.097, 95%CI: 0.009-1.095) were negatively associated with the risk of poor efficacy. Polysensitized children were more likely to suffer poor efficacy (OR=15.511 95%CI: 1.319-182.355). The clinical response at month 4 (r=0.707) and month 12 (r=0.925) was related to that at month 24. The area under the curve (AUC) for improvement at month 4 and month 12 was 0.746 and 0.860, respectively.

Conclusion: Our study confirmed the clinical efficacy of SCIT in AR children. Children with younger age, negative allergic history, and multiple allergens may predict a worse efficacy. The onset of action and the clinical response to SCIT in the second year can be predicted as early as by month 4

Allergol Immunopathol (Madr). 2023 Jan 1;51(1):84-91.

4. Unified Airway Disease: Future Directions

Jumah G Ahmad 1, Michael J Marino 2, Amber U Luong 3

Abstract

Unified airway disease describes the shared epidemiologic and pathophysiologic relationship among the chronic inflammatory diseases of the upper and lower airways including allergic rhinitis, chronic rhinosinusitis, asthma, and chronic otitis media. This concept proposes that these diseases are manifestations of a single inflammatory process and require an integrated diagnostic and therapeutic approach to achieve global disease control. Future directions to further establish this entity should focus on pathophysiology, diagnostic markers, flora microbes with particular emphasis on fungi, the role of type 3 inflammation, and targeted therapeutics including biologics, JAK inhibitors, and synthetic peptides.

Otolaryngol Clin North Am. 2023 Feb;56(1):181-195.

5. Laser nasal surgery (Review)

Mihai Ionuț Tănase 1, Mara Bulmaci 1, Constantin Stan 1 2, Rădeanu Gheorghe Doinel 1, Marcel Cosgarea 1, Septimiu Sever Pop 1, Alma Aurelia Maniu 1

Abstract

Laser nasal surgery has been an elusive subject in the last 10 to 15 years It was considered as a potential surgical staple for nasal surgery in the 1980s; however, it did not become one due to technical difficulties. Laser therapy has reemerged as an alternative to classical endoscopic surgery, and otorhinolaryngology surgeons are considering the benefits that it can offer. The advantages of this procedure are shorter hospitalization time, lack of nasal packing, high procedural precision with tissue sparing, and the unique capability of reducing both bacterial and fungal colonization at the level of the paranasal sinus. Therefore, laser therapy appears to be an invaluable tool for clinical practice. Due to the absence of a guaranteed cure for reoccurring nasal polyposis, laser therapy is worth investigating. For this therapy to evolve, an improved understanding of laser types and the effects that they produce is required. By investing in further developments of the equipment, the technique may become more widely used. With the current

accelerated rate of technological evolution and robotic capabilities, laser nasal surgery may become a gold standard in future years. The aim of the present review is to evaluate whether it is worth investing in nasal laser surgery as a future alternative to current treatment standards

Exp Ther Med. 2023 Jan 24;25(3):108.

6. Advances in Sinus Surgery for Nasal Polyps

T Graham Norwood 1, Jessica W Grayson 1, Bradford A Woodworth 1

Abstract

Background: Chronic rhinosinusitis with nasal polyps (CRSwNP) is comprised of a diverse group of endotypes that cause significant morbidity for afflicted patients. While endoscopic sinus surgery helps ameliorate the disease, polyps frequently recur. Newer strategies are intended to provide access for topical steroid irrigations in attempts to improve the disease process and quality of life, and decrease overall recurrence of polyps.

Objective: To review the current literature examining the latest surgical approaches for CRSwNP.

Methods: Review article.

Results: In dealing with the recalcitrant nature of CRSwNP, surgical techniques have simultaneously become more nuanced and aggressive. Bony resection in anatomically unfavorable areas such as the frontal, maxillary, and sphenoid outflow regions, replacing diseased or denuded mucosa with healthy grafts or flaps at the neo-ostia, and introducing drug-eluting biomaterials to newly opened sinus outflow tracts are highlights in the recent advancements in sinus surgery for CRSwNP. The Draf 3 or modified endoscopic Lothrop procedure has become a standard technique and demonstrated to improve quality of life and decrease polyp recurrence. A number of mucosal grafting or mucosal flap techniques have been described that cover exposed bone of the neo-ostium and evidence shows that this improves healing and diameter of the Draf 3. Partial middle turbinectomy, while controversial, appears to help decrease polyp recurrence in long-term follow-up studies. Modified endoscopic medial maxillectomy improves access to the maxillary sinus mucosa, facilitates debridement and, particularly, in the cystic fibrosis nasal polyp patient, improves overall management of the disease. Sphenoid drill-out procedure provides wider access for topical steroid irrigations and also may improve management of CRSwNP.

Conclusion: Surgical intervention remains a mainstay of therapy for CRSwNP. Newer techniques revolve around improving access for topical steroid therapy

Am J Rhinol Allergy. 2023 Mar;37(2):162-167.

7. Benign Tumors of the Anterior Cranial Base

Carl H Snyderman 1, Philippe Lavigne 2

Abstract

Benign tumors of the anterior cranial base may originate from intracranial, cranial, or extracranial sites. Intracranial tumors such as meningiomas may secondarily involve the cranial base and extend into the sinuses or nasal cavity. Bony tumors arising from the cranium include benign fibro-osseous lesions such as osteoma, fibrous dysplasia, and ossifying fibroma. The most common extracranial tumors that may extend to the skull base include angiofibroma and inverted papilloma. Symptoms are nonspecific and diagnosis is often delayed. In most cases, a diagnosis can be established based on the clinical presentation and radiographic features. Some small asymptomatic tumors may be observed for growth (meningioma, osteoma), whereas others should be treated due to continued destructive growth (angiofibromas) or potential for malignancy (inverted papilloma). Surgery remains the predominant treatment modality for benign tumors of the anterior cranial base. The major advance in recent decades has been the adoption of endoscopic techniques. Advances in endoscopic transnasal surgery have dramatically altered the surgical landscape, enabling the removal of tumors of the anterior cranial base with minimal morbidity. Due to decreased morbidity in comparison to transfacial or transcranial approaches, endoscopic transnasal surgery has lowered the threshold for surgery for benign tumors and can be applied to adult as well as pediatric populations. Anatomical limits include the anterior cranial base from the frontal sinus to the sella and optic canals and laterally to the mid-plane of the orbital roofs. Large dural defects can be reliably reconstructed using local (nasoseptal) and regional (extracranial pericranial) vascularized flaps

Adv Otorhinolaryngol. 2020;84:106-113.

8. Perioperative management of pediatric patients undergoing juvenile angiofibroma resection. A case series and educational review highlighting patient blood management

M M Longacre 1, S C Seshadri 1, E Adil 2, L C Baird 3, S M Goobie 1

Abstract

Background: Juvenile nasopharyngeal angiofibromas are one of the most enigmatic, bloody tumors encountered by otorhinolarygnologists, head and neck surgeons, neurosurgeons, and anesthesiologists. Juvenile nasopharyngeal angiofibromas are rare, benign, highly vascular tumors with a propensity towards aggressive local invasion. Surgery, open or endoscopic, to remove the growth is the primary treatment of choice for Juvenile nasopharyngeal

angiofibromas. Historically, surgical resection was associated with massive, rapid blood loss, traditionally managed by blood product transfusion and deliberate hypotension. Preventative management employing multimodal blood conservation strategies should be an essential standard of perioperative care for patients with Juvenile nasopharyngeal angiofibromas.

Methods: We describe a contemporary and comprehensive approach in the management of patients with high grade Juvenile nasopharyngeal angiofibromas. This includes surgical strategies such as preemptive external carotid artery embolization, endoscopic surgical approach, and staged operations, as well as anesthetic strategies including antifibrinolytic therapy and acute normovolemic hemodilution. These surgeries, once synonymous with massive transfusion, may potentially be performed without allogeneic blood transfusion, or deliberate hypotension.

Aims: Using a case series, the authors introduce a contemporary approach to multimodal, multidisciplinary blood conservation strategies for Juvenile nasopharyngeal angiofibromas surgery.

Results: Here in the authors report on an updated contemporary perioperative clinical approach to patients with Juvenile nasopharyngeal angiofibromas. From an anesthetic perspective, we describe the successful use of normal hemodynamic goals, restrictive transfusion strategy, antifibrinolytic therapy, autologous normovolemic hemodilution, and early extubation in the care of three adolescent males with highly invasive tumors. We demonstrate that new surgical and anesthetic strategies have yielded a significant decrease in intraoperative blood loss and eliminated the need for transfusion of autologous red blood cells, which enable improved outcomes.

Conclusions: The perioperative approach to elective surgery for Juvenile nasopharyngeal angiofibromas management is presented from a multidisciplinary patient blood management perspective.

Paediatr Anaesth. 2023 Mar 4.

9. Sinonasal Inverted Papilloma-Associated and De Novo Squamous Cell Carcinoma: A Tale of Two Cities or Not

Zekun Wang 1, Ye Zhang 1, Jianghu Zhang 1, Xuesong Chen 1, Jingbo Wang 1, Runye Wu 1, Kai Wang 1, Yuan Qu 1, Xiaodong Huang 1, Jingwei Luo 1, Li Gao 1, Guozhen Xu 1, Shaoyan Liu 2, Ye-Xiong Li 1, Junlin Yi 1 3

Abstract

Background: Sinonasal squamous cell carcinoma (SNSCC) can arise as either inverted papilloma-associated SCC (IP-SCC) or as de novo SCC (DN-SCC). It is controversial as to whether survival differences between IP-SCC and DN-SCC exist.

Methods: Between January 2000 and December 2016, 234 patients with SNSCC were analyzed retrospectively, including 68 with IP-SCC and 166 with DN-SCC. Propensity score matching (PSM) was performed to balance baseline characteristics. The Kaplan-Meier method and Cox proportional hazard model were used to determine risk factors on survival outcomes.

Results: The median follow-up time was 98.4 months. Before PSM, lymph node metastasis was noted to be lower in patients with IP-SCC. After PSM, the 5-year DFS, DSS and OS between IP-SCC and DN-SCC were 43.0% vs. 44.5% ($p = 0.701$), 49.2% vs. 56.2% ($p = 0.753$), and 48.2% vs. 52.9% ($p = 0.978$). The annual hazards of local failure, respectively, peaked at 28.4% and 27.8% for IP-SCC and DN-SCC within 12 months after treatment. Afterward, the hazards gradually decreased and the hazard for IP-SCC was always higher before approaching null.

Conclusions: This study provides novel evidence to support the clinical utility of improved distinction between IP-SCC and DN-SCC. Further studies are necessary to validate these findings before considering escalation of IP-SCC.

Cancers (Basel). 2022 Oct 24;14(21):5211.

10. Olfaction Now and in the Future in CRSwNP

Hannan A Qureshi 1, Andrew P Lane 1

Abstract

Background: Chronic rhinosinusitis (CRS) is the leading cause of olfactory dysfunction in the general population. Olfactory dysfunction is more common in patients with CRS with nasal polyposis (CRSwNP) compared to those without polyps.

Purpose: The present review aims to summarize the current literature on the mechanism behind olfactory dysfunction in CRSwNP and the impact of therapy on olfactory outcomes in this patient population.

Methods: A comprehensive review of the available literature on olfaction in CRSwNP was performed. We evaluated the most recent evidence from studies on the mechanisms behind smell loss in CRSwNP and the impact of medical and surgical therapy for CRS on olfactory outcomes.

Results: The mechanism behind olfactory dysfunction in CRSwNP is not completely understood, but evidence from clinical research and animal models suggests both an obstructive component causing conductive olfactory loss and an inflammatory response in the olfactory cleft

leading to sensorineural olfactory loss. Oral steroids and endoscopic sinus surgery have both shown efficacy in improving olfactory outcomes in CRSwNP in the short term; however, the long-term response of these treatments remains uncertain. Newer targeted biologic therapies, such as dupilumab, have also shown remarkable and durable improvement in smell loss for CRSwNP patients.

Conclusion: Olfactory dysfunction is highly prevalent in the CRSwNP population. Although significant advances have been made in our understanding of olfactory dysfunction in the setting of CRS, additional studies are needed to elucidate cellular and molecular changes mediated by type 2-mediated inflammation in the olfactory epithelium with potential downstream effects on the central olfactory system. Further identification of these underlying basic mechanisms will be vital for developing future therapies targeted to improve olfactory dysfunction in patients with CRSwNP

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