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Pediatric anosmia: A case series

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ABSTRACT

Introduction: Little is known about the etiology of olfactory dysfunction in the pediatric population. The aim of this study is to characterize the etiology and clinical features of anosmia and to explore evaluation options in a pediatric population.

Methods: Olfactory dysfunction was identified at a tertiary pediatric hospital between January 2003 and October 2014 using a text-based and ICD-9 search of the electronic health record system. Clinical information gathered included history, physical examination and imaging study. A phone questionnaire was completed to determine persistence and development of other rhinologic, endocrine, or neurologic symptoms.

Results: 37 children (male/female = 17/20) with mean/median ages of 13.28/14. 19 years were identified. The distribution of etiology was: rhinologic disease (N = 16), congenital (N = 4), trauma (N = 1), neoplasm (N = 1) and unknown (N = 15). Rhinologic disease included chronic rhinosinusitis (N = 3) and other nasal anatomic lesions. None of the four subjects with congenital anosmia had classic Kallmann syndrome. The utility of imaging in confirming an etiology of anosmia was noted in 1 of 8 CT and 5 of 22 MRI. The most significant finding of the questionnaire was confirmation of normal puberty in the congenital group.

Conclusion: Similar to the adult population, rhinologic disease is the most common cause. Absence or hypoplasia of the olfactory bulbs without associated delayed puberty is the presentation of congenital anosmia in our cohort. MRI had a higher utility than CT in evaluating anosmia in general and congenital anosmia in specific. MRI to evaluate children with a history of congenital olfactory dysfunction is recommended.

1. Introduction

Olfactory disorders have been well studied in the adult population, but little is known about these disorders in the pediatric population. The incidence of decreased olfactory function increases with age, with up to 50% of patients over the age of 65 affected [1]. Mucosal inflammation caused by chronic rhinosinusitis is the most common cause of decreased sense of smell in the otolaryngology patient population [2,3]. Head trauma, congenital causes, aging, viral etiologies, autoimmune diseases, neurodegenerative diseases, and toxic exposures have been linked to olfactory disorders in adults [1,4,5]. Specific pediatric populations have been reported to suffer from anosmia, including cystic fibrosis [6], CHARGE syndrome [7] and 22q11 deletion syndrome [8].

Congenital anosmia, defined as absence of smell function since birth, is rare accounting for only about 1% of the anosmic population. There are syndromic and nonsyndromic causes [9,10]. Kallmann syndrome is the best described genetic syndrome associated with anosmia and consists of hypogonadotropic hypogonadism and anosmia, occasionally in combination with other congenital abnormalities [9]. Case reports have documented an isolated congenital anosmia in the absence of other congenital abnormalities [9–11]; however, little is known about these disorders. No studies have specifically looked at the incidence and etiology of olfactory disorders in the pediatric population likely due to the difficulty in data collection.

One study evaluating 1,000 patients with smell disorders included a small fraction of pediatric patients [2]. The most common etiologies in this group included head trauma and congenital anosmia. Given this was a small subset of their population, no further detailed discussion was included, but it was noted that these patients had significantly lower odor thresholds compared to those with smell disorders from other causes, indicating that anosmia in the pediatric population can have a significant clinical impact [2]. The goal of this study was to evaluate children presenting with olfactory dysfunction to the pediatric otolaryngology clinic at a tertiary children's hospital and to further understand the natural history of this cohort with a follow up telephone questionnaire.

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Table 1
Demographics.

Total	37
Gender, N (%)	
Male	17 (45.9)
Female	20 (54.1)
Age at Diagnosis – mean/median (range) in years	13.28/14.19 (4.13–21.04)
Symptom Duration – mean/median (range) in years	5.80/2.78 (0.13–18.20)
Ethnicity, N (%)	
Caucasian	21 (56.8)
Hispanic/Latino	10 (27.0)
African-American	1 (2.7)
Asian	1 (2.7)
Other	3 (8.1)
Unknown/Not Reported	7 (18.9)

2. Materials and methods

This retrospective study was approved by the Colorado Multiple Institutional Review Board. Olfactory dysfunction was identified at a tertiary pediatric hospital by searching the electronic medical record for Otolaryngology encounters related to olfactory disturbances between January 2003 and October 2014. A text-based and ICD-9 search was utilized. ICD-9 codes included V41.5 “problems with smell and taste,” 781.1 “disturbances of sensation of smell and taste: anosmia, parageusia, parosmia,” 352.0 “disorders of olfactory nerve,” and 951.8 “injury to other cranial nerves: olfactory, traumatic anosmia NOS.” Text-based search included “anosmia, hyposmia, olfactory disorder, olfactory dysfunction, smell disorder, smell dysfunction, decreased sense of smell, absent sense of smell, loss of sense of smell, problem with sense of smell, and smell and taste disorder.” Specifically, only patients whose presenting symptoms matching these diagnoses were included. All subjects that carried the diagnosis of cystic fibrosis were excluded from the cohort. The electronic health records of the cohort were reviewed with special emphasis given to nasal examination (nasal endoscopy if performed), visits to cardiology, neurology and endocrinology clinics, results of the University of Pennsylvania Smell Identification Test (UPSIT) and imaging studies.

A phone questionnaire was completed to determine persistence of olfactory dysfunction, the development of other rhinologic or neurologic symptoms, or the presence of delayed or absent pubertal development.

3. Results

A total of 37 children (17 males and 20 females) with mean/median age of 13.28/14.19 years were included the study cohort. Caucasians and Hispanics/Latinos made up 84% of the cohort. The remaining demographic characteristics are outlined in Table 1.

The age of onset for subjective olfactory dysfunction ranged from 0 years to 19 years of age (mean 8 years). Nine children reported anosmia since birth and seven had an unknown age of onset. One child had an associated head trauma. There were no reports of upper respiratory tract infections coinciding with the onset of anosmia. Seven children had associated subjective taste disturbances.

Six subjects underwent olfactory testing. One subject completed the University of Pennsylvania Smell Identification Test, with a score of 25% confirming complete anosmia. Five subjects had partial testing, showing two with correct responses to the odors presented, two with incorrect responses, and one with unknown results. Ten subjects were referred to the Rocky Mountain Smell and Taste Center, however, this clinic ceased to exist at the time of this project and the results of any testing performed during this evaluation were not available. The remaining 31 subjects did not undergo any documented olfactory testing.

Computed tomography (CT) scan was performed on eight subjects (21.6%), with all but one yielding negative findings. The one positive scan was from a girl who suffered facial trauma during a fall with reconstruction of her LeFort and naso-orbito-ethmoid type and fractures. CT revealed involvement of her anterior ethmoid skull base and

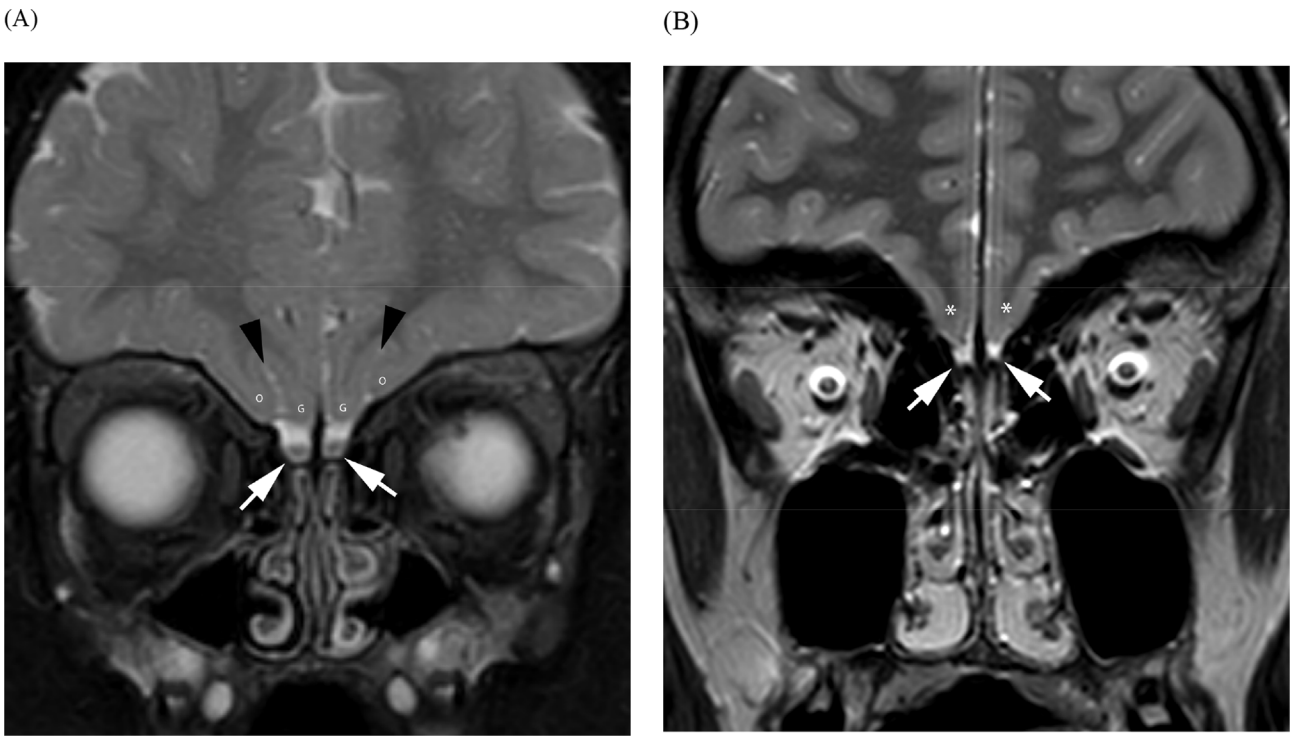

Fig. 1. (A) Coronal T2-weighted fat-saturated MR image illustrates a child with normal olfactory bulbs (white arrows), olfactory sulci (black arrowheads), orbital gyri (O), and gyri recti (G). (B) Coronal T2-weighted MR image in a child with congenital anosmia demonstrates absent olfactory bulbs (white arrows) and absent olfactory sulci (white asterisk).