Paranasal Sinus Development and Choanal Atresia

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Background: Although the determinates of paranasal sinus development and sinusitis are not well defined, a candidate factor is blockage of the choana.

Hypothesis: Maxillary sinuses ipsilateral to unilateral choanal atresia are comparatively small and have more evidence of sinusitis than do the contralateral sinuses.

Design: Retrospective.

Setting: Children's hospitals.

Patients: Sixteen nonsyndromic children with isolated unilateral congenital choanal atresia.

Main Outcome Measures: Determination of maxillary sinus volumes and mucoperiosteal thickening on preoperative computed tomograms.

Results: Maxillary sinuses ipsilateral to unilateral choanal atresia have slightly larger volumes than, and mucoperiosteal thickening that is similar to, the contralateral sinuses.

Conclusion: These data suggest that maxillary sinus development and sinusitis are independent of posterior nasal ventilation and drainage.


Obstruction of sinonasal drainage and ventilation is a well-accepted contributor to paranasal sinusitis.1-11 Such a situation exists in children with choanal atresia. In 1927, Grove12 reported a case of unilateral choanal atresia, about the 180th at the time, and noted that:

the free ventilation of the nose is one of the most potent factors in the prevention of sinus disease and in the cure of an early existing sinus infection. As no air current is possible [through the nose with unilateral choanal atresia], it must become apparent that the sinuses on the side of a choanal atresia are particularly susceptible to infection.

Proetz,6 emphasized, in 1941, the importance of sinus ventilation and drainage, noting that “it is generally agreed that stagnation of mucus in the closed side is apt to result ultimately in sinus disease.” Later, Friday et al13 and then Wald10 put choanal atresia at the top of the list of mechanical obstructions predisposing to sinusitis.

Because ventilation of the middle ear contributes to temporal bone pneumatization, nasal ventilation may be important in the development of the paranasal sinuses.5,14-16 Extensive paranasal sinus and mastoid development indicate sinus and middle ear health during a person’s growth period.6 Conversely, growth of the maxillary sinuses is adversely affected by severe sinus infection. This compromised growth is exhibited by the small paranasal sinus volumes of patients with cystic fibrosis.17 Interestingly, Kim et al17 reported that children who meet the criteria for chronic sinusitis, but who do not have cystic fibrosis, have maxillary sinus development that is comparable to that of normal subjects.

In children with unilateral choanal atresia, ipsilateral anterior nasal discharge is a presenting symptom.18 These children are frequently treated for months for presumptive sinusitis before the choanal atresia is recognized. Because nasal breathing is absent on the side of the atresia, poor gas exchange, leading to a lower partial pressure of oxygen and improved environment for bacterial growth, would be expected.2,5 The combination of abnormal mucociliary flow, pooling of secretions, and poor nasal ventilation would seem to predispose these children, in particular, to sinusitis on the atretic side.

With this background understanding, we hypothesize that children with unilateral choanal atresia have abnormal mucociliary flow, relatively poor ventilation, and smaller sinuses and sinusitis on the atretic side. However, we were surprised and intrigued to encounter a child with unilateral choanal atresia and well-developed, aerated maxillary sinuses. Was this one case a fluke? Unilateral choanal...
atresia may be considered an experiment of nature, providing an opportunity to explore the relationship of nasal obstruction and sinusitis in children.

RESULTS

Sixteen children, aged 1 month to 17 years, with isolated unilateral choanal atresia were identified. Twelve children were female and 4 were male, similar to the 2:1 ratio reported by Brown.19 Nine patients had right-sided atresia and 7 had left-sided atresia. A wide range of maxillary sinus volumes was noted (Figure). The mean maxillary sinus volume ipsilateral to the choanal atresia was 4.84 mL, compared with 4.48 mL for the contralateral side. The sinuses ipsilateral to the atresia were, on average, larger than the contralateral sinuses (P<.06). Symmetry of maxillary sinus size was apparent (r = 0.97; P<.001).

The volumes of the soft tissue (presumably mucoperiosteal thickening) in the maxillary sinuses appeared symmetrical: 6 patients had more soft tissue in the sinus ipsilateral to the atresia; 5 patients had more soft tissue in the sinus contralateral to the atresia; and 5 patients had complete opacification of both maxillary sinuses; ie, sinusitis as assessed by the preoperative CT scans was unrelated to the unilateral choanal atresia.

COMMENT

Classically, chronic sinusitis in children is associated with both local and systemic contributing factors. Systemic factors include upper respiratory tract infections, both allergic and nonallergic rhinitis, immunodeficiency states, primary ciliary dyskinesia, cystic fibrosis, Down syndrome, inhalant pollutants, and aspirin sensitivity. The list of local factors includes dental infections, cleft palate, and swimming, but mostly items related to nasal obstruction: adenoid hypertrophy or infection, foreign bodies, tumors, polyps, septal deviation, and choanal atresia.10,11,12 All of these conditions presumably interfere with normal nasal ventilation and sinonasal mucociliary flow and lead to obstruction of the osteomeatal unit.

Lifelong, persistent, severe paranasal sinusitis is thought to be represented radiographically by nonaerated sinuses that are smaller than usual. The nonaerated (ie, opaque) sinus is attributed to stagnated mucus or pus or to thickened mucoperiosteum. Proetz6 suggested that “the failure of some sinuses to develop may be on a kindred basis [to Wittmaack’s contention that inflammatory middle ear disease limits mastoid pneumatization].” However, Ritter2 stated, “For unknown reasons, during growth the maxillary sinuses may cease development and produce a sinus lumen of less than normal size.”

Notwithstanding the consensus of conventional wisdom, sinusitis is not universal in children with choanal atresia. Proetz6 observed that newborns with bilateral choanal atresia do not have clinical infection of the maxillary sinuses. Stewart’s22 1931 report included the radiographs of 3 patients with unilateral choanal atresia: “The air sinuses on both sides of the skull were found to be well developed.” Mehta4 in describing an adult with unilateral choanal atresia, reported near-symmetrical, aer-
lated, pneumatized maxillary sinuses. Similarly, Mittermaier et al 23 illustrated a case of unilateral choanal atresia and stated in the study’s figure legend, “The sinuses are normally developed.” Klossek et al 24 and Diner et al 25 recently described 2 small series of patients (numbering 6 and 11, respectively) with unilateral choanal atresia: no sinusitis!

As Proetz 6 suggested:

A difference apparently exists between the behavior of a nasal mucosa deprived of air after breathing has been established, and one in which, through congenital posterior closure, no breathing has ever taken place. In the latter condition normal, healthy tissues are often found in the nasal fossae.

There are several potential explanations for our findings of maxillary sinus symmetry in children with unilateral choanal atresia, including (1) stagnation of sinonasal mucus per se does not limit development of the maxillary sinuses; (2) absence of nasal ventilation alone does not result in chronic sinusitis or limit paranasal sinus pneumatization; (3) the blocked choana decreases the risk of viral and other pathogens getting into the nose; and (4) our findings, and those of other investigators, are flukes (statistically, this seems unlikely).

That children with congenital unilateral choanal atresia manifest healthy, well-developed paranasal sinuses bilateraly is surprising and incongruous with the time-honored teachings that nasal ventilation and drainage are necessary for sinus health. The explanation for this unexpected finding is unknown, and may be a fertile investigative tract for improved understanding of paranasal sinuses.

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REFERENCES