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The Role of Computed Tomography in the Evaluation of Bilateral Choanal Atresia: Two Case Reports

Congenital posterior choanal atresia is an uncommon anomaly which may have life-threatening consequences. This defect is frequently associated with other congenital abnormalities. CT scan confirms the diagnosis and accurately defines the anatomy of the atresia and differentiates osseous stenosis from membranous stenosis in the choanae. Therefore, CT may be the first-line imaging choice for the pre-operative diagnosis of congenital choanal atresia. We present two cases of bilateral bony choanal atresia, of which one is associated with bilateral coloboma.

Keywords: Choanal Atresia, Coloboma, Computed Tomography

Introduction

Choanal atresia (CA) is the developmental failure of the communication between the posterior nasal cavity and the nasopharynx. Congenital choanal atresia was first described by Roederer in 1755.¹ The reported incidence is 0.02-0.04 percent.² The anomaly is twice as frequent in girls as in boys.² About two-thirds of the cases are unilateral on record, and it involves the right side twice as often as the left.² This obstruction can be either bony or membranous, with bony atresia being more common.³ Concurrent congenital anomalies have been encountered in as many as 50 percent of the cases;⁴ of which coloboma is the most common.⁵ Data about the incidence and presentation of congenital choanal atresia among the Indian population is limited. In this paper, we present two cases of bilateral bony choanal atresia, in which one have bilateral colobomas, and we also discuss the role of CT in the appropriate diagnosis and management of these situations.

Case Presentation

Case 1

A full-term girl presented with severe respiratory difficulty, aspiration of feeds, and frequent attacks of cyanosis since birth. Attempts to pass a nasal catheter were unsuccessful, and the infant was examined with CT imaging for nasopharyngeal obstruction. Axial 1.5 mm CT scans were obtained through the nasopharynx without contrast administration. The CT imaging revealed osseous atresia of bilateral nasal choanae with medial bowing of lateral walls of the nasal cavity and an increased vomer width. Dependent fluid in both nasal airways is also evident (Fig. 1). Based on these findings, diagnosis of bilateral bony CA was considered. No associated congenital anomalies were present. Nasal surgery confirmed the diagnosis, which was then corrected surgically.

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Case 2:

A 10-day-old neonate presented with respiratory and feeding difficulties. Passing a nasal catheter was impossible. On clinical examination, the child had low birth weight and bilateral congenital convergent strabismus. Fundoscopic examination showed an increased physiological cupping/excavation of the optic disc bilaterally with a surrounding pigmented area suggestive of coloboma. Imaging workup for congenital nasopharyngeal obstruction was performed by CT scanning. CT scan showed medial fusion of the thickened bowed lateral walls of the nasal cavity, an enlarged vomer and dependent fluid in the nasal cavity bilaterally (Figs. 2A&B). In addition, there were conical protrusions at the posterior aspect of both globes and exaggerated cupping of both optic discs. Microphthalmia of the left globe was also evident (Figs. 2C&D).

Diagnosis of bilateral bony CA with associated bilateral colobomas was made based on imaging and fundoscopic findings. Although two major criteria (choanal atresia and coloboma) and one minor criteria (growth retardation) of CHARGE syndrome were seen in this case, confirmation of the diagnosis could not be made by molecular testing for CHD7 because

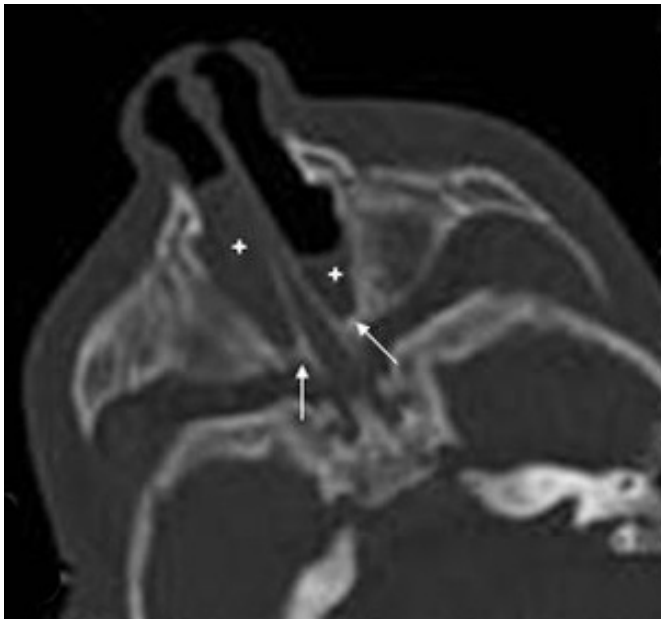


Fig. 1. A newborn girl with bilateral choanal atresia. Axial non-contrast computed tomography (NCCT) of the nasopharynx demonstrates medial bowing of bilateral lateral walls of the nasal cavity (arrows) with an increased vomer width and retained nasal secretions (asterisks).

of the patient's poor socio-economic condition. This patient subsequently underwent uncomplicated transnasal surgical repair of bilateral posterior choanal atresia.

Discussion

A high level of suspicion is required to diagnose bilateral choanal atresia. Symptoms of severe airway obstruction and cyclical cyanosis are the classic signs of neonatal bilateral atresia. Other ancillary signs are choking and gagging while nursing, pale or blue colored gums (as a result of oxygen deficiency), weakness and lack of weight gain. When crying alleviates respiratory distress in an obligate nasal breather, the neonatologist should bear in mind the probability of bilateral choanal atresia. Unilateral atresia may not be detected for years and patients may present with unilateral rhinorrhea or congestion.⁶

The clinical evaluation in a case of CA includes a complete physical examination to assess other congenital anomalies. A small feeding tube could be used to determine the patency of the choana, but a complete nasal and nasopharyngeal examination should be performed using a flexible fiberoptic endoscope to assess the deformity. Several abnormalities associated with choanal atresia have been reported.⁶ In a study of 130 cases of choanal malformation, 57 had other anomalies. Most of these were from the CHARGE association (C, coloboma; H, heart defects; A, atresia choanae; R, retarded growth; G, genitourinary defects; E, ear defects).⁷ In 1998, an expert group defined the major (the classical 4C's: Choanal atresia, Coloboma, Characteristic ears and Cranial nerve anomalies) and minor criteria of CHARGE syndrome. Individuals with all four major characteristics or three major and three minor characteristics are highly likely to have CHARGE syndrome. However, there have been individuals genetically identified with CHARGE syndrome without the classical choanal atresia and coloboma. Furthermore, mutations in the CHD7 gene (member of the chromodomain helicase DNA protein family) are detected in over 75% of patients with CHARGE syndrome. Molecular testing for CHD7 mutations may help to confirm the diagnosis.⁸ Coloboma (from the Greek koboboun, to mutilate), one of the major criteria associated with CHARGE syn-

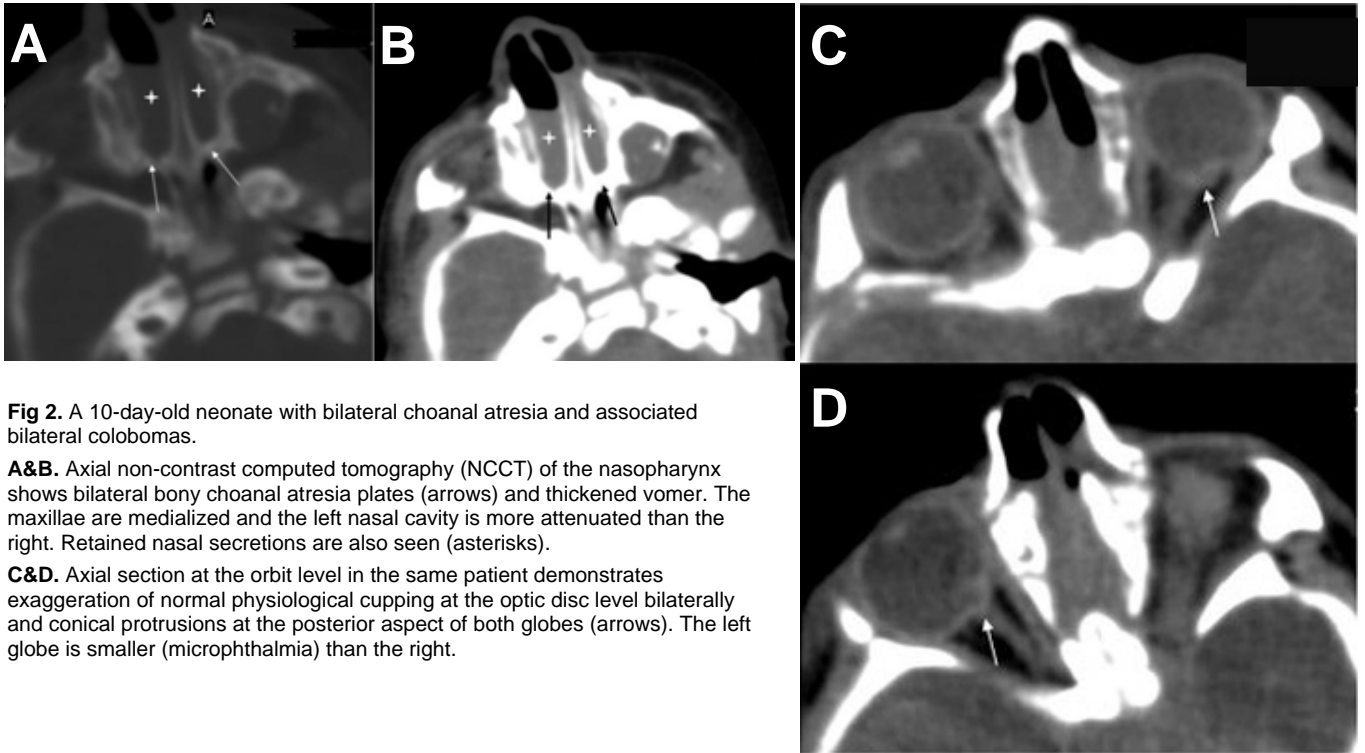


Fig 2. A 10-day-old neonate with bilateral choanal atresia and associated bilateral colobomas.

A&B. Axial non-contrast computed tomography (NCCT) of the nasopharynx shows bilateral bony choanal atresia plates (arrows) and thickened vomer. The maxillae are medialized and the left nasal cavity is more attenuated than the right. Retained nasal secretions are also seen (asterisks).

C&D. Axial section at the orbit level in the same patient demonstrates exaggeration of normal physiological cupping at the optic disc level bilaterally and conical protrusions at the posterior aspect of both globes (arrows). The left globe is smaller (microphthalmia) than the right.

drome is defined as an absence or defect of some ocular tissue, usually resulting from malclosure of the fetal intraocular fissure⁹ was first described by Walther in 1821. Cobobomas may involve the optic nerve, retina, choroid, iris, or lens and may be complete, when all of these structures are involved, or incomplete, when a variable part of the eye is spared.¹⁰

CT scanning is the radiographic procedure of choice in the evaluation of choanal atresia. For good results, careful suctioning is performed to clear excess mucus,

and a topical decongestant is applied. The purpose of CT scanning is outlined as follows:

- Confirming the diagnosis of choanal atresia (unilateral or bilateral).
- Evaluating choanal atresia (vomer bone width and choanal airspace distance).
- Excluding other possible nasal sites of obstruction.
- Determining the degree of bony, membranous, or mixed atresia.
- Delineating abnormalities in the nasal cavity and nasopharynx.

The choanal air space is defined as the distance from the lateral wall of the nasal cavity to the vomer bone (LWNC-V).¹¹ The boundaries of choanal air space are: the sphenoid superiorly, the medial pterygoid lamina laterally, the vomer medially and the horizontal portion of the palatal bone inferiorly. Both sides are measured and most often are symmetrical. In those instances of discrepancy between sides, the difference is always less than 0.1 cm. At birth, normal choanal airspace measures 0.67 cm (mean) [0.34-1.01 ($\pm 2SD$) cm], and normal vomer width is 0.23 cm (mean) \pm [0.11-0.34 ($\pm 2SD$) cm]¹¹ (Fig. 3).

In bony atresia, CT findings include narrowing of the posterior choanae (LWNC-V < 0.34 cm in new-

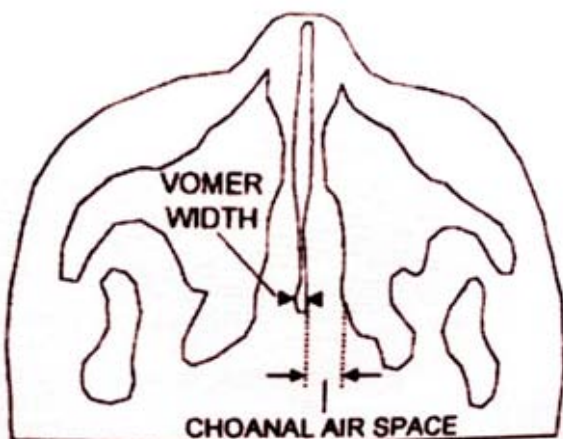


Fig. 3. Schematic presentation of normal choanal air space and vomer width in a newborn.

born), medial bowing and thickening of the posterior medial maxilla (which may be fused with the lateral margin of vomer), thickening of vomer (>0.34 cm) and retained fluid in the nasal cavity.¹¹ In membranous atresia, these pathologic anatomic findings are largely absent, but the air passage between the lateral wall of the nasal cavity and vomer is small. It should be noted, however, that while it is easy to define the membranous atretic segment, secretions in the nose may accumulate and give a falsely thick appearance of the atretic segment. Hence, it is important to have the nose properly suctioned and to have the mucosa pretreated with a topical vasoconstrictor.¹¹

Choanal atresia (CA) is due to failure of breakdown of the buccopharyngeal membrane in the fourth or fifth week of gestation.³ Newborn infants up to the third to sixth week of life cannot spontaneously breathe through the mouth.² Thus, when the obstruction is bilateral, the neonate exhibits severe respiratory distress, which can lead to a critical state and requires prompt relief of the obstruction. Here, CT plays a significant role in the diagnosis and therapeutic approach to congenital choanal atresia and should be the examination method of choice to evaluate neonates with nasal obstruction. CT accurately defines anatomy of the atresia and a more systematic surgical approach may be used which may prevent multiple surgical procedures.¹²

In conclusion, bilateral congenital CA in the neonate is a medical emergency that should be treated promptly. CT scan of the nasal cavity and nasopharynx helps to establish the diagnosis and to define the

extent and type of atresia. This precise anatomic evaluation helps in determining the surgical approach. Since computed tomography is a valuable and easily accessible diagnostic tool, it should be the choice examination method in evaluating neonates with nasal obstruction. Furthermore, the finding of choanal atresia, especially when it is bilateral, warrants careful search for other congenital anomalies in the newborn.

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