

Six Years to Diagnose Bilateral Congenital Choanal Atresia: A Case Report

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Abstract

Introduction: Bilateral choanal atresia is a rare congenital malformation, which causes asphyxia neonatorum rarely compatible with life, making this pathology a diagnostic and therapeutic emergency. We report a very rare case of bilateral choanal atresia in a girl who survived to the age of 6 without neonatal care, and we discuss the clinical, endoscopic, radiological aspects and the treatment of this condition.

Presentation of case: this is a 6 years old girl who consulted at this age for bilateral nasal obstruction, night snoring, mouth breathing since birth. Endoscopic examination and CT of the face revealed bilateral choanal imperforation in its membranous form. Endoscopic surgical treatment was performed with good clinical improvement. Follow-up evaluation at postoperative 10th month showed that her symptoms improved significantly and, on endoscopic examination, both choanae remained patent.

Discussion and conclusion: the diagnosis of choanal imperforation is above all clinical, based on systematic screening in the delivery room for any newborn, and confirmed by endoscopy and CT. The revelation at an advanced age is exceptional. Endoscopic transnasal choanoplasty remains the gold standard in choanal atresia surgery. However, the value of additional treatments, such as mitomycin and laser-assisted techniques, is not proven.

Keywords: Bilateral Choanal Atresia; Congenital; 6 Years Old; Endoscopic Transnasal Choanoplasty; Case Report

List of abbreviations: CT: Computerized Tomography

Introduction

Bilateral choanal atresia is a rare congenital malformation that causes acute respiratory distress in the newborn, rarely compatible with life. According to the literature, bilateral choanal atresia accounts for 30% of patients [1]. It often necessitates early surgery because newborns are obligatory nasal breathers owing to the position of the epiglottis, tongue, and soft palate, especially during feeding [2]. Transnasal endoscopic repair has become the most widely accepted technique. In some cases, a transpalatal approach remains necessary, especially in neonates with severe craniofacial abnormalities or very small nasal fossae [3,4].

We present a case of a 6-year-old girl who survived with bilateral choanal atresia without any treatment.

Case Presentation

A 6-year-old girl presented at our department for chronic nasal obstruction with exclusive mouth breathing since birth, nocturnal snoring affecting sleep quality with tiredness on waking, episodes of recurrent infectious pneumonia. There was a history of episodes of cyanosis in childhood essentially while breastfeeding, with recovery on crying.

Clinical examination found an ogival palate without septal deviation, no obvious craniofacial abnormalities or other congenital malformation.

Endoscopic examination and CT of the face revealed bilateral choanal membranous obstruction (Figure 1).

Transnasal endoscopic repair was performed under general anesthesia, using 0° and 30° pediatric optics. The opening of the atretic plate on both sides was practiced by using the micro-resector with a 2 mm diameter suction cutting blade to create a first orifice of the same caliber that will be gradually expanded. The posterior edge of the vomer was then resected and the size of the neochoan is enlarged using the micro-resector with a protected strawberry. Finally, a calibration of the choans was carried out by intubation probes fixed by a tape on the nasal pyramid kept for a month (Figure 2 and 3).

The postoperative care included a daily nasal saline irrigation in the lying position, upside down and turned to the side by instilling saline serum into the nasal cavity. This allows the discharge of secretions and scabs by the opposite nasal cavity.

Regular evaluation at 10 months post-operative shows good clinical improvement with no restenosis.

Discussion

Choanal imperforation is a fairly rare congenital malformation, with an incidence estimated at 1/5000-9000 [5].

Bilateral choanal atresia accounts for 30% of patients, and unilateral choanal atresia accounts for approximately 70% [6]. Original reports suggested a 90% bony stenosis and 10% membranous obstruction [7], but more recent analysis suggests a mixed bony/membranous in 70% and pure bony in 30% [8].

Between 7 and 29% patients with choanal atresia will have CHARGE syndrome (Corrales and Koltai, 2009) Each patient with clinical symptoms should undergo a nasofiberscopy or a nasal rigid endoscopy in clinics to confirm the diagnosis of choanal atresia and a CT imaging in order to specify the type of obstruction and plan surgery [9].

The patient in our case suffered a delayed diagnosis due to the ignorance of her parents and those around her who trivialized the symptoms and attributed them to an enlarged tonsil with adenoids explaining the difficulty breathing and snoring at night considering the frequency of this affection in their family.

Treatment is surgical, however several techniques have been described: Transnasal endoscopic repair for choanal atresia is the preferred initial technique with transpalatal approach reserved for patients in whom transnasal repair would be impossible [9].

The patient survived until the age of 6 years without treatment probably due to the methods of unscrambling used by the parents including opening the mouth when it was difficult to breathe. Also, the possibility of incomplete choanal atresia at the birth which became complete at the time of diagnosis.

Bilateral choanal atresia often necessitates early surgery because newborns are obligatory nasal breathers owing to the position of the epiglottis, tongue, and soft palate, especially during feeding [2]. The timing of surgery for bilateral choanal atresia must be guided by clinical tolerance of the nasal obstruction and the presence of associated abnormalities. Globally, children with bilateral choanal atresia undergo surgery during the first week [6].

The use of choanal stenting and mitomycin C as an adjunct therapy to prevent restenosis are a controversial topic in the management of choanal atresia as there is no clear-cut evidence on the effectiveness of using stents or mitomycin after choanal atresia repair [7,8].

The Postoperative care is recommended using proton-pump inhibitors, nasal saline irrigation which should be applied after surgery with a duration less than or equal to four weeks. Intranasal corticosteroids are commonly used while systemic corticosteroids are not and the prescription of antibiotics is debated [9-11]. Endoscopic follow-up is necessary to detect a scar disorder or restenosis, the main complications of this management.

Conclusion

The diagnosis of a choanal imperforation is above all clinical, based on systematic screening in the delivery room for any newborn, and confirmed by endoscopy and CT allowing rapid and adequate care.



Figure 1: CT of bilateral choanal atresia demonstrating a membranous type obstruction



Figure 2: Transnasal endoscopic repair



Figure 3: An intraoperative Figure showing a patent choanae

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Declaration of competing interest

All authors have no conflict of interest or financial support with this article.

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