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Case Report

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Choanal Atresia about Two Cases at the Hospital and University Center Gabriel Toure from Bamako (Mali)

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Abstract

Choanal atresia is a congenital defect in the permeabilization of the posterior end of the nasal cavities. Its bilateral form is responsible for respiratory distress that can be life-threatening. The unilateral form was suspected in view of the permanent unilateral right nasal obstruction. We report the observation of two clinical cases admitted to the Radiology and Medical Imaging department, CHU GT of Bamako (Mali). The aim of our study was to highlight the importance of screening for this condition at birth. Surgical treatment allowed us to obtain an overall success rate of 100%. Divulsion was used to pass the emergency stage after short-term palliative medical treatment in the case of bilateral choanal atresia. The operative techniques used were divulsion and nasal endoscopic route.

Keywords: Choanal Atresia, Nasal Obstruction, Divulsion, Nasal Endoscopic Approach.

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Introduction

Choanal atresia is a congenital malformation consisting of a total or subtotal obstruction of the posterior orifices of the nasal cavities. Its bilateral form is revealed by congenital respiratory distress resulting in a vital emergency. The unilateral form is compatible with life and can go unnoticed for many years. It is an overall rare condition, accounting for 1/5000 to 1/8000 births [1 - 2]. It is twice as common in girls as in boys.

In one out of two cases, choanal atresia is associated with other congenital anomalies, the complete form of which results in CHARGE syndrome (coloboma, heart disease, choanal atresia, mental and growth retardation, genital hypoplasia) [3].

Several pathogenic theories have been put forward, but they need to be confirmed; the most recent incriminates an anomaly in the migration of mesodermal tissue.

Diagnosis, primarily clinical, has been considerably facilitated by endoscopy. Currently, CT scans are the procedure of choice in the evaluation of

choanal atresia. Surgical treatment uses several techniques, but the most commonly used are: divulsion and endoscopic approach.

We propose to analyze the epidemio-clinical parameters of two clinical cases and, based on our experience and data from the literature, to conduct a comparative study of the results of different surgical procedures.

OBSERVATIONS

1st Clinical Case

We report the case of a newborn, female, 2 days old, hospitalized in the neonatology department of CHU Gabriel, for respiratory distress, admitted to the Radiology and Medical Imaging department of CHU Gabriel Touré for a scan of the nasal cavities.

Anamnesis. There was no family history of choanal atresia. No evidence from the interview suggested a hereditary condition. Furthermore, the child had no functional signs of cardiac or pulmonary involvement.

Clinical examination found an abolition of bilateral nostril flow.

A nasal-sinus CT scan revealed bilateral membranous choanal atresia on the left and mixed choanal atresia on the right.

Given this CT confirmation of bilateral membranous choanal atresia; additional examinations were carried out systematically to eliminate other associated malformations. The latter included:

- An ophthalmological examination with fundus examination and visual acuity.
- An examination of the external genitalia, a neurological, pulmonological and digestive examination.
- A brain scan, a cardiac ultrasound, a renal ultrasound and a chest X-ray. All these tests carried out did not reveal any abnormalities in the child.



Figure 1: Axial bone CT scan showing bilateral choanal obstruction secondary to linear membranes extending from the thickened vomer on the right to each posterior maxilla: Bilateral membranous choanal atresia on the left and mixed on the right

2nd Clinical Case

We report the case of a 3-month-old female infant, hospitalized for right nasal obstruction that had been developing since birth, referred to the Radiology and Medical Imaging department of CHU Gabriel Touré for a scan of the nasal passages.

Clinical examination found an abolition of right nostril

A nasal-sinus CT scan revealed right choanal atresia of bony origin.



Fig. 2: Axial bone CT scan, complete bony obstruction of the right choana with fusion of the enlarged vomer to the thickened posterior maxilla internally: Bony right choanal atresia

DISCUSSION

Choanal atresia is a rare condition with an estimated frequency of 1 case per 5,000 to 8,000 births [1-7]. Prescott in 1986 reported an estimated incidence of 1 case per 50,000 births [8]. A publication entitled "congenital choanal atresia in north africa infants" shows that the incidence of this malformation in the North African population is 1 case per 3,100 births [9]. The female predominance is reported by several authors [4-11] but does not find an etiopathogenic explanation and remains a simple epidemiological observation. These results are superimposable on our series, in which the two cases are female. The distribution of choanal atresia according to unilaterality and bilaterality varies according to the authors [4-13].

Clinically, bilateral choanal atresia occurs at birth in a picture of cyclical respiratory distress associated with major feeding difficulties. Unilateral or incomplete bilateral forms are pauci-symptomatic and may go unnoticed until adulthood. It occurs mainly during the first six weeks of life according to the work of Panda *et al.*, [14].

The diagnosis is primarily clinical. Bilateral and complete forms are evident in the newborn who presents with respiratory distress with cyanosis accentuated by crying [15]. This result is comparable to our first case of observation whose circumstance of discovery was also respiratory distress.

Unilateral or incomplete forms are insidious with non-specific symptoms. The diagnosis was most often made on the basis of the anamnesis and confirmed by CT scan, which is the procedure of choice in the evaluation of choanal atresia, to assess its characteristics, unilateral or bilateral involvement, the proportion of bone obstruction, its thickness, the bone anomalies that constitute it (inclination of the pterygoid processes, thickening of the Vomer) [16, 17]. With the contribution of CT scan, it would seem that mixed forms are in fact more frequent than is said in the literature, they represent 76% of cases in the Brow series [18]. Associated malformations (realizing CHARGE syndrome in their complete form) were sought during clinical examination.

The treatment of atresia aims to re-permeabilize the posterior part of the nasal cavity. The intervention will be proposed urgently in the case of bilateral form and deferred in the case of unilateral form.

Divulsion, which was considered the treatment of choice to get through the emergency phase [12-20], is currently being abandoned in favor of endoscopic approaches.

The endoscopic route has developed considerably since the first description by Stankiewitz. It has benefited from the appearance of modern

instrumentation, namely burs and the micro-debrider "powered instruments" [1].

It has the advantage of offering good visualization; of allowing extensive resection; of being a short intervention with low morbidity that can be proposed from birth for bilateral choanal atresia, whether mixed or bony, and in the treatment of recurrences.

CONCLUSION

We conclude through these two clinical cases that nasal endoscopy and CT scan have completely revolutionized the diagnostic and therapeutic approaches of choanal atresia. If the diagnosis has been facilitated, the frequent association with other anomalies requires the practice of a clinical and paraclinical assessment and prolonged follow-up by a pediatrician.

Advances in endonasal surgical instrumentation and practice have improved outcomes.

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