NASAL OBSTRUCTION: NOT ONLY ADENOID AND TURBINATE HYPERTROPHY

R9

Breda MS, Menezes A, Pereira S, Silva DP, Moreira F, Ramalho S, Ribeiro D, Dias L
Department of Otorhinolaringology & Head and Neck Surgery – Hospital de Braga

INTRODUCTION

In pediatric population, nasal obstruction is mainly caused by adenoid and inferior turbinate hypertrophy. Choanal atresia (CA) is a less frequent cause characterized by a narrowing of the posterior opening of the nasal cavity into the nasopharynx. This condition occurs in 1:5000 to 1:8000 live births and affects females twice as often as males. Unilateral CA is much more common than bilateral CA (65- 75% of all cases). Bilateral CA is more related to syndromic cases like CHARGE or Crouzon. Historically, 90% has been classified as bony and 10% as membranous, but recent studies, using CT techniques, suggest only 30% as pure bony and 70% as mixed bony-membranous. Surgery for CA correction can be approached through transnasal, transseptal or transpalatal routes.

CLINICAL CASE

A female newborn (2010/11/20), with normal pregnancy, studied by a multidisciplinary team because presented at birth:

- **Pulmonary Valve Stenosis***
- **Heart atrial septal defect (*ostium secundum*)***
- **Low ears implantation***
- **Left eyelid ptosis ***

The baby was referred to our consultation by her 5th month of life, due to unilateral rhinorhea and nasal obstruction. ENT study revealed **right CA***. The remaining ENT study was normal (study of auditory pathway with otoacoustic emissions, brainstem auditory evoked potentials and magnetic resonance).

CHARGE syndrome (*) was our most likely diagnosis.

In ophtalmologic examination coloboma was excluded as well as vision loss.

Since CA was unilateral, she was proposed for elective corrective surgery only at 5 years old.

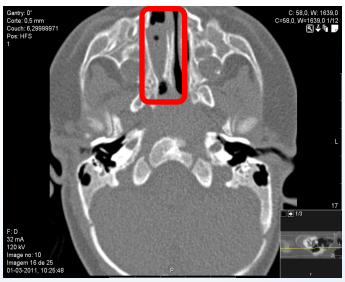


Fig.1 – Axial computed tomography showing **right mixed bony-membranous CA**.

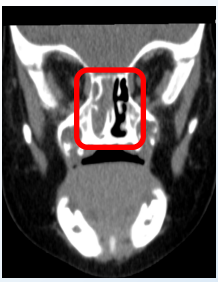


Fig.2 – Coronal computed tomography showing **right mixed bony-membranous CA.**

CA CORRECTION (2015/11/03)

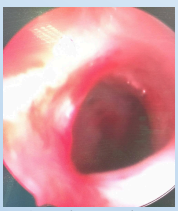


Fig.3 – Endoscopic nasal view (0º endoscope) during surgery.

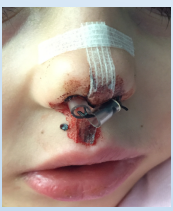


Fig.4 – Immediate post operative period with nasal stenting

6 weeks later

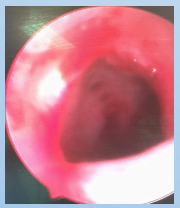
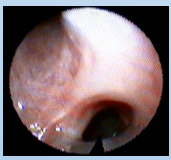


Fig.5 – Stent removal, in the OR under sedation, with choanal patency verified (0º endoscope)

5 Months later



QR code to watch with smartphone the last nasal endoscopy

Fig.6 – Follow-up – Right neochoana with partial stenosis

SURGICAL TECHNIQUE

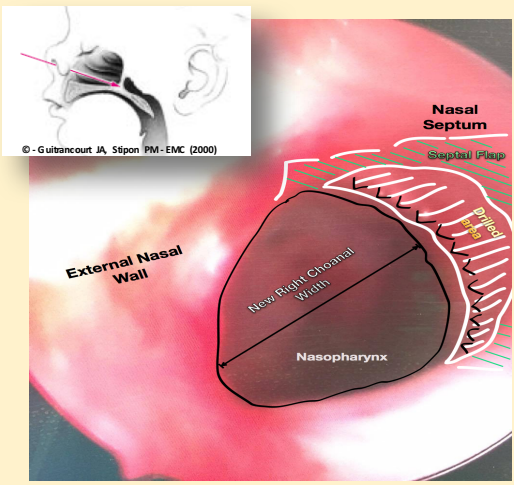


Fig.7 – **Schematic View of Surgery** - Transnasal correction with 0º endoscope. **1st step** – Performance of a mucoperiosteal flap immediately next to the anterior third of vomer. The flap was displaced in an antero-posterior direction (black arrow direction). The flap area created was wider than the expected drilled area. (Green lined) **2nd step** – The atretic plate/bone excess was drilled away. The infero-posterior part of the vomer was resected. (White lined) **3rd step** – The flap previously created was used to cover the drilled area. **4th step** – At the end of the procedure a Silicone Tube (4,5 mm diameter) was used to stent the neochoana. This tube was placed giving a complete turn posteriorly to the vomer (U-Shaped) and fixed anteriorly with Vycril. The silicone tube was kept in place 1 month.

DISCUSSION & CONCLUSION

CA, mainly the unilateral form, can resemble the most common causes of nasal obstruction, being only the bilateral form clinically obvious due to "cyclical cyanosis". CHARGE syndrome is an autossomal dominant genetic disorder typically caused by mutations in the chromodomain helicase DNA-binding protein-7 (CHD7) gene. No single feature is universally present or sufficient for the clinical diagnosis. Blake *et al* (1998) suggested that a diagnosis of CHARGE requires the presence of at least 4 major features or 3 major features plus at least 3 minor features. In our clinical case, CHARGE syndrome was hypothesized because the baby had 2 major features: unilateral CA and low ear implantation; and also had 2 minor features: left eyelid ptosis and congenital heart defects. The remaining workup study excluded other major features like coloboma/vision loss and hearing loss, present in CHARGE more than 90%. After 5 years of follow-up, CHARGE was definitively excluded, with the rest of her clinical findings well compensated or unnoticeable, with normal intellectual and physical development.

The use of endoscopic techniques for transnasal CA repair was first demonstrated by Stankiewicz (1990). Various studies have reported primary repair success rates ranging from 67 to 88%. In our clinical case, as we expected by literature, after 5 months of CA endoscopic correction, partial stenosis of the neochoana ocurred, but still with nasal patency and without any breathing complaint.

REFERENCES

- Leung MK, Krakovitz PR, Koltai PJ. Congenital Sinonasal Disorders. In: Kennedy DW, Hwang PH, editors. Rhinology: diseases of the nose, sinuses, and skull base. New York: Thieme; 2012. p.384-5.
- Andrieu-Guitrancourt J, Stipon PM. Cirurgia de la imperforación coanal. Encyclopédie Médico-Chirurgicale: Cirurgía ORL. 2000; E-46-230.
- Stankiewicz JA. The endoscopic repair of choanal atresia. Otolaryngol Head Neck Surg. 1990; 103: 931-7.
- Blake KD, Davenport SL, Hall BD, Hefner MA, Pagon RA, Williams MS. CHARGE association: an update and review for the primary pediatrician. *Clin Pediatr (Phila).* 1998 Mar. 37(3):159-73.

miguelbreda@gmail.com - June 2016