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An Italian multicentre experience in endoscopic endonasal treatment of congenital choanal atresia: Proposal for a novel classification system of surgical outcomes

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ABSTRACT

Objective: The purposes of this study were to report our experience with endoscopic treatment of choanal atresia (CA), to illustrate our surgical technique and analyse the different factors that may affect outcomes.
Material and methods: A retrospective review was performed of patients affected by congenital CA and treated between June 1996 and November 2013 at three referral centres which follow a uniform policy.
Results: Eighty-four patients with CA (55 unilateral and 29 bilateral), aged between one day and 76 years (mean, 13 years) were included. Associated malformations were present in 28 patients. The overall success rate was 93%, with 96.3% and 86.2% success rates for unilateral and bilateral CA respectively. Six patients (7%) required revision surgery for early symptomatic restenosis. Statistical analysis revealed that outcomes were not influenced by sex, previous surgeries, unilateral versus bilateral atresia or associated anomalies. However, age seemed to be a prognostic risk factor for patients under one year-old.
Conclusion: The endoscopic endonasal approach is safe and effective, with a very high success rate and low morbidity. The removal of the vomer and the use of mucoperiosteal flaps are the main keys to avoiding postoperative stenosis. The use of stents or Mitomycin C is therefore not mandatory.

1. Introduction

Choanal atresia (CA) is a rare malformation occurring in between 1:5000 and 1:8000 live births (Schwartz and Savetsky, 1986; Stankiewicz, 1990) and causing obstruction of the upper airways due to the obliteration of the posterior nasal aperture. The historical theories of the persistence of the buccopharyngeal membrane, the persistence of the nasobuccal membrane of Hochstetter, the incomplete resorption of the nasopharyngeal mesoderm and the

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locally misdirected mesodermal flow are nowadays associated with molecular and genetic mechanisms in the pathogenesis of CA (Corrales and Koltai, 2009). It is more frequent in females than in males, with unilateral presentation being more common (60%) than bilateral (40%). The atretic plate may be bony (~30%) or mixed bony-membranous (~70%), with no purely membranous anomalies found in recent CT scan studies (Brown et al., 1996). In 20–50% of cases, other congenital malformations are associated with CA. CHARGE syndrome is the most common of these and consists of coloboma, heart defect, CA, growth and mental retardation and genital and ear anomalies (Lazar and Younis, 1995).

Different surgical options have been proposed in the past, including transpalatal, transeptal, sublabial, transantral and transnasal approaches (Park et al., 2000). However, more recently the growing experience and technical advances in endoscopic sinus surgery have led many surgeons to make more frequent use of the endoscopic endonasal technique in the repair of CA which has provided better results and fewer surgical complications than in traditional procedures (Josephson et al., 1998). The use of choanal stenting, Mitomycin C and resurfacing of the surgical defect with mucosal flaps in order to prevent restenosis are some of the most controversial issues in the management of CA.

The present study reports the experience of three university centres with a uniform policy for the management of congenital CA. Particular emphasis is given to illustrating the surgical technique employed and analysis of the factors affecting the surgical outcomes. In addition, the safety and effectiveness of the endoscopic endonasal procedures are evaluated and the short and long-term success of repair are examined.

2. Material and methods

2.1. Patient population

A retrospective analysis of a database dedicated to patients with CA treated at the Departments of Otorhinolaryngology at the University of Bologna, Pavia and Varese (Italy) from June 1996 to November 2013 was performed. Acquired defects (e.g. post radiation therapy) and children with Crouzon syndrome (as they present only stenotic choanas and not real CA) were excluded from the present study. Records were analysed with respect to patients' age at surgery, sex, side of atresia, previous surgery, surgical technique, postoperative stenting, complications and follow up. All patients or their parents were fully informed about the method of treatment and gave their written consent to the therapy. The study met the approval of the local board of medical ethics.

2.2. Preoperative evaluation

All patients underwent preoperative nasopharyngoscopy with flexible or rigid endoscope and fine cut CT scan in axial and coronal projections in order to confirm the clinical diagnosis, determine the nature of the atresia and evaluate the characteristics of other possible congenital malformations (e.g. anatomical variations of the paranasal sinus, septal deviation and skull base defects).

2.3. Surgical technique

Surgery was performed with patients placed under general anaesthesia in a supine anti-Trendelenburg position. The nasal cavities were packed with neurosurgical pledges soaked, depending on the age of the patient, in 0.05–0.2% oxymetazoline, 0.5–1% oxybuprocaine and adrenaline (1:100,000–1:200,000) solution, for 5–10 min. Endoscopes with 0° lenses and diameters of 2.7 or 4 mm were employed. An irrigation sheath connected to an irrigation pump, when required, was used to keep the endoscope's lens clean during surgery. Mucosal incisions were performed with a beaver knife in almost all patients in order to avoid postoperative granulation tissue formation and subsequent restenosis. However, a diode laser was used in some adult cases to provide bloodless incisions. In selected cases, endonasal drills equipped with long handles were used with diamond burrs to remove the bone of the atretic plate without harming surrounding tissues. The surgical techniques varied mainly according to the type of CA, unilateral or bilateral, and on the available “surgical space”. Mitomycin C was never used in the present series.

a) Unilateral choanal atresia

In unilateral cases when the surgical space was generally adequate, the procedure started with a vertical incision of the nasal mucosa at the level of the posterior third of the vomer in order to raise two mucoperiosteal flaps. The first flap was developed from the anterior surface of the atretic plate and displaced laterally, whereas the second septal flap was harvested in a postero-anterior direction from the part of the vomer that had to be removed. The bone was then pierced in the middle and drilled away, up to complete ablation of the inferior portion of the sphenoidal rostrum. The infero-posterior part of the vomer was resected with the use of a back-bite forceps. The mucoperiosteal flap detached from the anterior surface of the atretic plate was used to cover the raw area of the lateral aspect of the neochoana, while the mucosa detached from the posterior aspect of the vomer was employed to cover the medial edge of the neochoana and septum (Fig. 1).

In more recent cases, we have adjusted the flap harvesting technique. The vertical incision was performed bilaterally at the level of the posterior third of the vomer. The first flap was harvested from the surface of the atretic plate and pedicled superiorly, while the second flap was harvested on the contralateral nasal fossa, at the level of the vomer that had to be removed, and pedicled inferiorly. At the end of the procedure the superiorly based flap was rotated to cover the raw area of the sphenoidal rostrum while the inferiorly based flap was adequately resized and rotated to cover

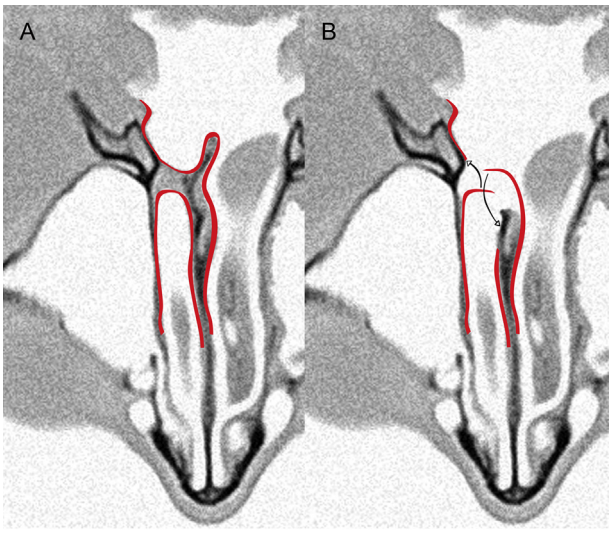

Fig. 1. (a) Axial view of a unilateral right choanal atresia. Enhanced in red are the anterior and posterior mucosal surfaces. (b) Axial view of the right neochoana after removal of the atretic plate, showing the placement of the anterior and posterior mucoperiosteal flaps in order to resurface respectively the lateral and medial aspects of the neochoana.