Choanal atresia – the last years of experience at the Hospital Dona Estefânia

Original Article

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Abstract

Objectives: Report and characterize the group of children diagnosed with coanal atresia at a terciary referral pediatric hospital during the last 13 years.
Study design: Retrospective observational study of sixteen children with choanal atresia who underwent surgical intervention.
Results: thirteen patients were female and three male. We identified nine bilateral cases and seven unilateral. Regarding the type of obstruction, twelve cases were mixed and four were bone obstructions. We found an association to CHARGE syndrome in six cases. We found six recurrences requiring surgical reintervention.
Conclusions: Our epidemiological results have similarities with the literature. Our surgical success rates were superior in bilateral cases.

Keywords: choanal atresia, surgical repair, transnasal

Introduction

Choanal atresia (CA) is a rare congenital malformation characterized by the obstruction of the posterior nasal apertures caused by a failure in their permeability during embryonic development.

The incidence of CA among newborns (NBs) is 1 in every 5,000–8,000 live births,1 and the anomaly predominates among females (at a ratio of 2:1). Currently, CA is characterized as either osteo-membranous/mixed, accounting for 70% of cases, or completely osseous. Unilateral CA occurs in two-thirds of cases and is more common on the right.[1]

Several theories explain the occurrence of CA, namely persistence of the buccopharyngeal membrane, failure in the rupture of the Hochstetter bucconasal membrane, or the presence of mesodermal adhesions.[2] The higher incidence observed in genetic syndromes such as CHARGE syndrome or

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Treacher Collins syndrome can be explained by abnormal migration of the neural crest cells, which causes simultaneous craniofacial anomalies.[1,2]

Association with other congenital malformations occurs in 75% of cases, such as CHARGE syndrome,[3,4] where the letters comprising the acronym describe the clinical characteristics of the syndrome: coloboma, heart disorders, choanal atresia, growth retardation, and malformations of the genital apparatus and of the ear. Treatment success in these cases is lower than in cases of isolated CA.[3,4]

Clinical presentation of CA differs between unilateral and bilateral cases. In the former, patients can remain without a definite diagnosis for many years and may present with unilateral chronic nasal obstruction and mucoid rhinorrhea.[5]

Bilateral CA is a medical emergency because NBs are obligate nose breathers.[2,3,4] It manifests with signs of acute breathing difficulty, bilateral rhinorrhea, feeding difficulty and, specifically, cyclical and paradoxical cyanosis, which improves with crying and reappears during rest. In these cases, crying should be encouraged, or a finger, a Guedel tube, or a McGovern nipple should be inserted into the mouth of the NB to ensure patency of the airway.[1,2]

Failure to pass a nasogastric tube (NGT) through the nasal fossa is often the first sign that arouses clinical suspicion.[5,6] After the patient is clinically stabilized, the diagnosis should be confirmed via nasal endoscopy. Computed tomography (CT) is the gold standard imaging exam1 because it can facilitate determination of the nature, position, thickness, and size of the obstruction, as well as exclude the presence of other congenital anomalies.

Treatment is essentially surgical, and several approaches may be taken: transnasal, transpalatinal, transantral, and transeptal.[7,8] The objective of each approach is to conservatively restore the patency of the choanae, avoiding lesions in the adjacent structures. The timing for performing surgical correction varies depending on several factors including the type of obstruction and the patient's age.[9,10] In cases of unilateral CA without significant breathing or feeding difficulty, surgical correction should be delayed to promote growth of the facial structures and reduce the surgical risk.

The use of endoscopic techniques has drastically changed CA surgery, and the transnasal endoscopic technique is currently the most used. It is minimally invasive, less traumatic, and allows for better visualization, amplification, and lighting of the atresia plate.[10,11,12]

This study is a retrospective analysis of patients diagnosed with CA at Dona Estefânia Hospital (HDE) over the last 13 years, with group characterization and evaluation of the results through a comparison with the literature.

Material and Methods

This was a retrospective study based on the analysis of 16 medical records of children diagnosed with CA and submitted to surgical intervention in the period between March 2008 and March 2021 at the HDE. The data analyzed include sex, age at diagnosis, type of CA, association with other diseases, and use of adjuvant treatments. All patients underwent pre-, peri-, and postnatal evaluation, nasal endoscopy, and CT of the paranasal sinuses to determine the type of CA.

The analysis was performed through a review of the medical records using Sclinico®software.

Results

The medical records of 16 children, comprising 13 girls (81.3%) and three boys (18.7%), were analyzed. Age at diagnosis varied between 4 years and 13 years, specifically between 4 days and 20 days for bilateral CA and between 3 days and 13 years for unilateral CA. With regard to the type of obstruction, 12 cases (75%) were mixed CA, and four cases (25%) were osseous CA. Nine patients (56.3%) had bilateral CA, and seven patients (43.7%) had unilateral CA, four of which were on the right and three on the left. In six cases (37.5%), there was a clinical

association with CHARGE syndrome, and in five cases (31.3%), CA was the only congenital malformation. The remaining patients had several comorbidities: congenital heart disease, hypoxic-ischemic encephalopathy, epilepsy, cleft palate, and cataracts.

All children's diagnoses were confirmed by CT. With regard to adjuvant therapies, transnasal tubes were inserted in nine patients with bilateral CA and in one patient with unilateral CA (62.5%) for a period between four and weeks. Only three patients (18.8%), those with CHARGE syndrome, received intraoperative topical application of mitomycin C (2 mg/mL). Ten patients (62.5%) underwent only one intervention, of which seven (43.8%) had bilateral CA and three (18.7%) had unilateral CA. Six children (37.5%) were submitted to at least two surgical corrections, of which five had unilateral CA and one had unilateral type. With regard to peri- and postoperative complications, only the patients with bilateral CA with stent placement had scabs and granulations. In these cases, nasal irrigation with saline through the stents was sufficient to eliminate them.

Discussion

Although rare, CA is the main anatomic cause of congenital nasal obstruction. It presents as a surgical challenge because it involves multiple structures, namely the nasal septum, the atresia plate, and the base of the skull.

The female/male ratio in the analyzed sample was 2.4:1, and 75% of cases were mixed CA, which is in line with the literature.[1,2,3,6]

In the present study, the first surgery had a success rate of 62.5%, and most patients (43.8%) had bilateral CA, while 37.5% required at least one reintervention. Of these, 83.3% were cases of unilateral CA. A similar surgical procedure was used in the reinterventions, with the only change being the use of adjuvant therapies. Although several studies found no statistically significant difference between the two types of obstruction after the first surgery, there appears to be a higher risk of restenosis in cases of bilateral CA after the second intervention.[10,13] However, assessing this as an independent factor is difficult because there are several risk factors for restenosis (type of obstruction, age at the time of intervention, comorbidities, technique used) that often occur simultaneously.

The post-surgery risk of restenosis is high due to local inflammation that promotes the formation of granulation tissue.[10] For this reason, the use of adjuvant therapies may be very important to maintain the permeability of the choanal region. Adjuvant therapies include the use of transnasal tubes or stents, CO2 or Nd-YAG laser, and topical administration of mitomycin C.[13-15] At HDE, 62.5% of patients received a transnasal silastic stent with a diameter between 3.5 mm and 5.5 mm. Initially, its use was suggested to avoid the risk of restenosis and allow the patency of the choana until completion of the healing process. However, it carries additional risks. The local pressure exerted at the level of the alar cartilage or the septum may cause damage or infection and septal perforation, respectively. These patients need more frequent reevaluations and saline irrigations to avoid obstruction by secretions. Additionally, it may contribute to perpetuating local inflammation and to the consequent formation of granulation tissue, synechiae, and worsening pain.[1,5,10] Some authors report that the use of stents in patients with bilateral CA is a major factor to reduce the risk of restenosis, especially in younger children. In contrast, some authors recommend refraining from using stents to avoid additional postoperative complications. They assert that transnasal endoscopic surgery is a safe and effective technique, regardless of its use.[11,12]

In sum, the surgical procedure is performed transnasally using sinonasal endoscopic surgical material. After nasal congestion relief with phenylephrine chloride 2.5 mg/mL for 10–15 mins, a 0° endoscope is used to confirm the absence of choana permeability. A flap is usually created with the mucosa of the nasal septum, and permeabilization is performed in the most inferior internal area, using cold

instruments. In the case of mixed CA, the osseous component may be removed with a diamond bur.

Mitomycin C is an antibiotic that inhibits migration and proliferation of fibroblasts and therefore has several therapeutic indications, including CA.[13] At HDE, mitomycin C was used in three patients with CHARGE syndrome presented for surgical revision. The drug was used at a concentration of 2 mg/mL and applied topically for 4 mins. Some authors have reported the benefits of its use, but no statistically significant differences have been observed;[10,12,16] thus, at HDE, mitomycin C is only used in reinterventions.[10,12] Evaluation of the drug's independent efficacy is complex because this drug is frequently used with other adjuvant agents. Although it is widely used, two recent studies have shown a potential carcinogenic effect of this drug, but further studies are required to confirm this finding.[16,17]

Neither CO2 nor Nd-YAG laser were used in the analyzed population. The advantages of their use have been increasingly shown, as both represent a safe and precise method to remove granulation or scar tissue.[1,14,17]

The fact that CA is clinically rare makes drawing statistically significant conclusions difficult because the study samples are small and present different characteristics. Conducting multicenter studies may yield more robust conclusions and a more uniform approach to CA.

Conclusion

The demographic characteristics of the present study's sample were in line with those in the literature. Endoscopic sinonasal surgery accommodates a more precise and detailed approach, which leads to better surgical outcomes. However, new adjuvant treatments appear to play an important role in treating CA relapse. In the present study, therapeutic success in bilateral CA cases was superior, which may be explained by the advantage of using transnasal stents.

Conflicts of Interest

The authors declare that there is no conflict of interests regarding the publication of this paper.

Data Confidentiality

The authors declare having followed the protocols in use at their working center regarding patients' data publication.

Protection of humans and animals

The authors declare that the procedures were followed according to the regulations established by the Clinical Research and Ethics Committee and to the 2013 Helsinki Declaration of the World Medical Association.

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Availability of scientific data

There are no datasets available, publicly related to this work.

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