ORIGINAL ARTICLE

A 20-Year Experience in Microsurgical Treatment of Choanal Atresia

Hugo Rodríguez, Giselle Cuestas, Desiderio Passali

Servicio de Endoscopia Respiratoria, Hospital de Pediatría Prof. Dr. Juan P. Garrahan, Buenos Aires, Argentina

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KEYWORDS
Choanal atresia; Congenital; Microsurgical technique; Transnasal approach

Abstract
Introduction and objectives: Choanal atresia is the most common congenital nasal anomaly. Diagnosis is confirmed by endoscopic examination and computed tomography. The definitive treatment is surgical, and different surgical techniques and approaches are used. We describe our experience in transnasal microsurgical treatment of congenital choanal atresia.

Methods: We retrospectively evaluated 49 patients with congenital choanal atresia operated in the Department of Respiratory Endoscopy over a period of 20 years. The clinical variables analysed were type of atretic plate, age at diagnosis and surgery, associated malformations, maternal history of hyperthyroidism treated with methimazole during pregnancy, mode of airway stabilisation before surgery, surgical technique, complications, and outcome.

Results: Mixed bilateral choanal atresia was the most frequent (29 cases). Its incidence was higher in females (61.2%). Almost 51% of patients showed associated malformations, and 7 had a history of maternal hyperthyroidism treated with methimazole during pregnancy. The surgical procedure consisted of a transnasal microscopic approach and placement of a silicone endonasal stent for one to 12 weeks. Thirty-five patients required revision after surgery.

Nine patients had complications. Suitable nasal ventilation was achieved in 46 patients (93.9%). One patient died of causes unrelated to the surgery. Two patients with permeable choanae remain with tracheotomy.

Conclusion: The transnasal microsurgical repair with endonasal stent proved to be a safe and effective procedure.

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PALABRAS CLAVE
Atresia de coanas; Congénita

Experiencia de 20 años en el tratamiento microquirúrgico de la atresia de coanas

Resumen
Introducción y objetivos: La atresia de coanas es la anomalía congénita nasal más común. El diagnóstico se confirma mediante examen endoscópico nasal y tomografía computarizada de...
Introduction

Blockage of the nasal passages in newborns is a potentially fatal condition due to their necessarily nasal breathing. The most common congenital causes are choanal atresia (CA), dermoid cyst, glioma, encephalocele and congenital stenosis of the piniform aperture.¹

CA is the most common congenital nasal anomaly. It is estimated that its incidence is 1 case per every 8000–10 000 live births,² and it is more prevalent among females (2:1).³ It can appear in isolation or as part of multiple malformation syndromes such as CHARGE (acronym for coloboma, heart defect, atresia choanae, retarded growth, genital hypoplasia, ear abnormalities).⁴,⁵ Bilateral cases manifest as respiratory distress from the moment of birth. Unilateral atresia is manifested as respiratory failure and unilateral rhinorrea and may go unnoticed. The diagnosis is suspected by the absence of airflow in the nostrils and inability of a nasogastric tube to advance, and is confirmed by nasal endoscopic examination and computed tomography (CT) scan of the craniofacial complex. In most cases, the plates are mixed (bony and membranous components), and less often, bony.

The definitive treatment is surgical, through various possible techniques and surgical approaches. Surgical repair with transnasal endoscopic technique provides an excellent visualisation of the posterior nasal defect and has currently become the procedure of choice due to its safety and effectiveness, displacing the transpalatine approach.⁶,⁷

We describe our experience in the transnasal treatment of congenital CA using microscopy and placement of an intranasal stent.

Method

We retrospectively evaluated 49 patients undergoing surgery for congenital CA at the Respiratory Endoscopy Service over a period of 20 years (May 1992–May 2012).

The variables analysed were gender, location and type of atretic plate, age at diagnosis, associated malformations, maternal history of hyperthyroidism treated with methimazole during pregnancy, mode of airway stabilisation prior to surgery, age at surgery, surgical technique, duration of the nasal stent, need for other treatments, complications, follow-up time and evolution.

We performed a thorough prior assessment of patients by endoscopic examination of the nasal passages with a flexible fibroscope (2.2 mm or 3.5 mm) or a rigid endoscope (2.7 mm), and CT scan of the craniofacial complex to evaluate the characteristics of the lesion: laterality, thickness, bone and/or membranous component. All images were digitally documented.

A total of 8 patients had undergone surgery previously at other institutions (7 cases of transnasal approach with an endoscope and 1 case of palatal approach).

The surgical procedure consisted of a transnasal approach using microscopy. Under general anaesthesia, after placing a gauze soaked in vasoconstrictor (adrenaline 1:1000), both nostrils were visualised with a 0° 2.7 mm rigid endoscope. A protective gauze was placed in the nasopharynx. The affected nostril was approached under 300× microscopy. Following the bottom of the nasal fossa, the choana was opened medially and inferiorly. The incision of the mucosa was carried out with CO₂ laser (5 W continuous mode) or cold instrumentation. Once the nasal mucosa covering the atretic plate was removed, the plate and the posterior part of the vomer were extracted with microsurgical instrumentation (ear curette or drill). The edges of the neochoana were smoothed and the bone surface was covered with mucosa.

All patients underwent placement of a silicone stent (Foley probe number 12–18) as a support for the intranasal lumen for 1–12 weeks. The time of removal of the nasal tutor was based on laterality and type of atretic plate; it was shorter in unilateral CA and longer in bony plates. In unilateral cases the tutor was placed on the affected side, fixed to the columella by a transfixing point. In bilateral cases,
the stent was placed as a perforated “U” on the posterior part, without suture (Fig. 1).

From 2002 we began using topical mitomycin C (0.4 mg/ml for 2–4 min). This was used in 14 patients (28.6%) at the time of removing the tutor and after postoperative choanal dilatation.

In the immediate postoperative period, patients were admitted to the intermediate care ward, except for those who were intubated previously, who remained in the ICU until extubation at 24–72 h and were then transferred to intermediate care. Extubation time was dependent on concomitant diseases, previous medication and clinical condition.

These patients were treated with anti-reflux therapy (hygienic-dietary measures), antibiotics (ampicillin-sulbactam 50 mg/kg/day), analgesics as required (ibuprofen 10 mg/kg/dose), and feeding through an orogastric probe until their oral tolerance was tested. They also received local postoperative treatment with nasal humidification and suction as required.

During the time the nasal tutor was maintained, patients underwent nasal washes with saline solution through the stent, aspiration of secretions as required and nasal mupirocin to prevent lesions by decubitus position thereof. We performed weekly endoscopic controls during the first month after nasal stent extraction to evaluate the calibre of the neochoana and formation of granulation tissue.

Postoperative review was defined as any procedure under general anaesthesia conducted after the first intervention, including nasal stent removal and removal of granulation tissue with CO₂ laser or choanal dilatation with urethral catheters, with or without the use of topical mitomycin C, either to prevent restenosis or to improve choanal permeability.

**Results**

The study included 30 female patients (61.2%) and 19 male patients (38.8%) with congenital CA. There were 3 pairs of affected sisters (1 pair of twins). The atretic plate was bilateral in 33 patients (67.3%) and unilateral in 16 (32.7%), with 11 cases on the right side (68.8%) and 5 cases on the left side.

The most common plate was the mixed type (43 patients, 87.5%) (Table 1).

Among the bilateral cases, 90.9% (30 cases) were diagnosed in the neonatal period and among the unilateral, 81.2% (13 cases) after 2 months (range: 1 day–12 years).

A total of 25 patients (51%) presented associated malformations, with the most frequent being genetic syndromes (15 cases, 60%), heart disease (4 cases, 16%) and coloboma (3 cases, 12%) (Fig. 2). A total of 8 patients suffered a recognised syndrome, with CHARGE being the most common (4 cases), followed by Down (3 cases) and Turner (1 case).

A total of 7 patients (14.3%) had a history of maternal hyperthyroidism treated with methimazole during pregnancy. All were females (2 of them were twin sisters) and

<table>
<thead>
<tr>
<th>Table 1</th>
<th>General Characteristics of Patients With Choanal Atresia.</th>
</tr>
</thead>
<tbody>
<tr>
<td>Gender</td>
<td>Bilateral CA (n=33)</td>
</tr>
<tr>
<td>Female</td>
<td>20</td>
</tr>
<tr>
<td>Male</td>
<td>13</td>
</tr>
<tr>
<td>Type of CA</td>
<td></td>
</tr>
<tr>
<td>Mixed</td>
<td>29</td>
</tr>
<tr>
<td>Bony</td>
<td>4</td>
</tr>
<tr>
<td>Side</td>
<td></td>
</tr>
<tr>
<td>Right</td>
<td>33</td>
</tr>
<tr>
<td>Left</td>
<td>33</td>
</tr>
<tr>
<td>Associated malformations</td>
<td>19/33 (57.6%)</td>
</tr>
<tr>
<td>Exposure in utero to maternal hyperthyroidism treated with methimazole</td>
<td>4</td>
</tr>
<tr>
<td>Main symptoms</td>
<td>Breathing difficulty, cyanosis</td>
</tr>
</tbody>
</table>

CA: choanal atresia.
patients (93.9%). One case (2%) died 8 years after surgery for unrelated reasons. Two patients (4.1%) with patent chaenae are still tracheotomised due to aspiration syndrome, and subglottic stenosis and glossoptosis, respectively.

**Discussion**

CA is the most common congenital nasal anomaly characterised by the obliteration of the posterior nasal opening, due to the imperforation of the oronasal membrane and overgrowth of the horizontal and vertical process of the palatine bone during weeks 4–11 of the gestation process. Other theories about the origin of CA point to a persistence of the oro-pharyngeal membrane, incomplete reabsorption of the nasopharyngeal mesoderm and abnormal migration of neural crest cells.\(^6\)\(^7\)

While the worldwide incidence of CA documented in the literature is 1 for every 8000–10 000 live births,\(^2\) in Argentina it is lower (1 per 70 000 live births), given that there are approximately 700 000 live births per year and the annually reported cases of CA do not exceed 10.

It is more common among females and the most common type of atretic plate is the right unilateral.\(^10\)\(^11\) The mixed type (bony and membranous components) represents 70%, whilst the bony type accounts for 30%.\(^9\) In our series, 61.2% of patients were female, and bilateral CA was the most frequent (67.3%). The latter could be due to the fact that our service is a referral centre for infant airway disorders. The plates were mixed (87.8%) in most cases and bony (12.2%) to a lesser extent.

Depending on the type of atresia and age of the patient, the symptoms vary from mild respiratory distress with feeding to severe airway obstruction.

While bilateral atresia is present at birth and represents a medical emergency, unilateral cases tend to appear between 5 and 24 months after birth.\(^3\)\(^,\)\(^5\)\(^11\) Most bilateral cases were diagnosed in the neonatal period, and unilateral cases after 2 months.

Due to their nasal breathing, newborns with bilateral CA present dyspnoea and cyanosis cyclically, which can be relieved with crying. These cases require immediate

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**Table 3** Patients who Required Postoperative Revisions.

<table>
<thead>
<tr>
<th>Laterality</th>
<th>Total patients</th>
<th>Patients who required revisions</th>
</tr>
</thead>
<tbody>
<tr>
<td>Unilateral</td>
<td>16</td>
<td>9</td>
</tr>
<tr>
<td>Bilateral</td>
<td>33</td>
<td>26</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Type of atretic plate</th>
<th>Total patients</th>
<th>Patients who required revisions</th>
</tr>
</thead>
<tbody>
<tr>
<td>Bony</td>
<td>6</td>
<td>6</td>
</tr>
<tr>
<td>Mixed</td>
<td>43</td>
<td>29</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Age at the time of surgery</th>
<th>Total patients</th>
<th>Patients who required revisions</th>
</tr>
</thead>
<tbody>
<tr>
<td>&lt;10 days</td>
<td>6</td>
<td>6</td>
</tr>
<tr>
<td>10–20 days</td>
<td>11</td>
<td>8</td>
</tr>
<tr>
<td>&gt;20 days</td>
<td>32</td>
<td>21</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Associated malformations</th>
<th>Total patients</th>
<th>Patients who required revisions</th>
</tr>
</thead>
<tbody>
<tr>
<td>Yes</td>
<td>25</td>
<td>19</td>
</tr>
<tr>
<td>No</td>
<td>24</td>
<td>16</td>
</tr>
</tbody>
</table>

**Table 2** Postoperative Complications.

<table>
<thead>
<tr>
<th>Complications</th>
<th>Number of patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>Nasal synechiae Related to the nasal stent</td>
<td>2</td>
</tr>
<tr>
<td>Epistaxis</td>
<td>1</td>
</tr>
<tr>
<td>Accidental extraction</td>
<td>2</td>
</tr>
<tr>
<td>Lesion in nostril</td>
<td>1</td>
</tr>
<tr>
<td>Necrosis of nasal mucosa</td>
<td>1</td>
</tr>
<tr>
<td>Necrosis of posterior part of the septum</td>
<td>1</td>
</tr>
<tr>
<td>Respiratory arrest due to obstruction by stent</td>
<td>1</td>
</tr>
</tbody>
</table>
stabilisation of the airway and feeding through an orogastric tube. When CA is unilateral, sometimes the diagnosis takes place late during childhood or adolescence, manifesting as nasal obstruction and positional unilateral rhinorhea (increases by tilting the head forward), and occasionally as unilateral otitis media. During the physical examination, anterior rhinoscopy reveals pale mucosa and turbinates in the affected nostril, as well as the presence of accumulated secretions which do not drain towards the nasopharynx. CA is suspected upon an absence of motion of a wisp of cotton or absence of fogging in a mirror placed under the nostrils while the mouth is closed, and also by impossibility of introducing an 8 French probe (2.7 mm) beyond 3.5–5.5 cm from the alar edge. The definitive diagnosis is obtained by endoscopic examination of the nasal fossae and CT scan of the craniofacial complex, with axial and coronal sections, showing the type, thickness and location of the atretic plate, narrowing of the posterior nasal cavity and thickening of the vomer (Fig. 3). It also enables other possible sites of obstruction and anomalies of the nasopharynx and nasal fossae to be excluded.

The differential diagnosis should consider stenosis of the piriform aperture, encephalocele, nasal tumours, such as gliomas and dermoid cysts, septal deviation and foreign bodies (in unilateral cases), among other causes. Patients should be studied to identify and treat possible associated malformations (present in 20% to 50% of cases), like CHARGE. In total, 51% of our patients presented other associated anomalies, especially those with bilateral CA (19 cases, 76%). A total of 8 patients presented a known syndrome, with CHARGE being the most common (4 cases). Numerous malformations that are not part of the acronym have also been described in these children, including: retrognathia, glossoptosis, subglottic stenosis, vocal cord paralysis, laryngeal cleft, tracheoesophageal fistula and difficult intubation. CA would only play a role as an aggravating factor in respiratory instability. Regardless of choanal patency, tracheotomy is necessary in a significant number of these patients (10%–30%), as in 1 of our own cases, where the surgical nasal correction was successful, but decannulation could not be achieved.

In 7 patients (14.3%) CA was associated to prenatal exposure to maternal hyperthyroidism treated with methimazole. Some studies have shown that the use of certain antithyroid drugs (methimazole, carbimazole) during pregnancy may increase the risk of children being born with CA. Nevertheless, it is unknown whether the association is attributed to the medication, the maternal thyroid disease or abnormal levels of thyroid hormones in the newborn. It is thought that the maternal disease is the causal factor, rather than treatment with methimazole.

Patients with bilateral CA require expedited stabilisation of the airway, which can be achieved using a McGovern nipple, oral cannula or endotracheal intubation. Tracheostomy is reserved for situations in which other craniofacial anomalies are associated.

The definitive treatment is surgical and is recommended in the first days of life in bilateral cases, although it can be delayed if the CA is unilateral and no serious respiratory or feeding problems exist. Some authors prefer surgical correction of unilateral CA from 12 months of age. The rationale for this delayed repair is allowing the nasal cavity to become larger, and also so the surgically created choanal opening does not grow along the surrounding tissues, becoming narrower when the child matures.

Different techniques for the surgical correction of CA have been described, with the transnasal, transpalatal and transseptal approaches being the most common.

In recent years, the intranasal route has displaced the classical palatal pathway, as it is less invasive and provides excellent results.

Microsurgical techniques were used to treat CA in all 49 patients. The transnasal approach including the use of a microscope is a safe and effective procedure. It offers accuracy, with good visualisation of the atretic plate with amplification and binocular vision, rapid recovery, short hospital stay and low morbidity. It does not alter the growth of the hard palate or the nasal pyramid. Its main disadvantage appears when there are associated malformations and when the intranasal anatomy is unfavourable (septal deviation, turbinate hypertrophy, high palatal arch). In all our patients, we complemented the microscopic view with endoscopy.
The increasing range of instrumentation available allows surgeons to make increasingly accurate incisions in the mucosa and bone resections. The advantages of laser include less bleeding and limited damage to adjacent tissue.9

Due to the advancement of nasal endoscopy and the development of small and specialised instrumentation, several authors prefer endoscopic surgery for the treatment of CA.5,7,19-23 They argue that the use of the endoscope provides a better view of the posterior nasal cavity than microscopy, which would result in better outcomes. However, some studies have compared the endoscopic transnasal technique with the non-endoscopic option and have not found significant differences in the surgical results.7 Its main disadvantage appears when the nasal cavity is small, in narrow cases and when the nasal anatomy is abnormal.24 In these cases, endoscopic surgery can present a challenge even for the most experienced endoscopic surgeons. It is easier to apply this technique in older patients.5,17

Blind transnasal puncture with a trocar has ceased to be the method of choice. Its advantages are speed and availability, but it is associated to a need for several additional revisions and a high probability of complications (cerebrospinal fluid leak and meningitis caused by penetration of the cribiform plate).4,25

Transpalatal surgery offers excellent exposure, with its main disadvantage being its effect on the growing palate (severe deformities, crossbite). It is associated with increased operative time, more bleeding, increased risk of palatal fistula and dysfunction of the palatine muscle.4,18,25,26

Sublabial transseptal repair is reserved for newborns with unfavourable intranasal anatomy or craniofacial abnormalities.6

Some authors emphasise that there is no definitive evidence based on randomised controlled trials demonstrating the potential advantages and disadvantages of any particular surgical technique for patients with CA.27

The success of surgery depends on several factors:

- Wide resection of the vomer and/or the posterior septum to obtain a large choana.
- Respecting the anatomical reference points: opening of the choana should be carried out below the tail of the middle turbinate in order to avoid bleeding and damaging the basisphenoid. The floor of the nasal fossa should be followed and the opening should be done in a medial an inferior direction.
- Avoiding bone ridges (smoothing the edges, without leaving round-shaped orifices) and covering the bone surface with mucosa.

Mucosal flaps are technically complex, especially in neonates by a transnasal approach.24 Therefore, different authors employ them to avoid the use of nasal stents.25

The benefits and risks of placing a stent after CA repair are still debated in the otolaryngology literature.

We placed an intranasal, Silastic® prosthesis in all patients. Although the need for a tutor is not universally accepted, its use is justified by preventing restenosis during the reepithelialisation of the neochoana.18,29

The use of a tutor entails certain risks, including damage to the nasal mucosa by excessive pressure, resulting in granulation tissue and scar formation, bacterial overgrowth and blocking of mucus drainage.5,7,14,21 The placement of an endoprosthesis made with soft material for a short time reduces the possibility of formation of granulation tissue and the risk of postoperative infection, thus reducing the risk of choanal restenosis.5

In most cases, the nasal tutor was maintained in place for 1 month. This period was shorter in unilateral CA and longer in bony CA.

Frequent irrigation and suction by the caregiver are necessary to prevent obstruction by secretions and to maintain patency of the nasal tutor. Caregivers must carry out a good management and receive CPR training.

Mitomycin is an antineoplastic agent which inhibits fibroblast proliferation and the formation of granulation tissue.12,17 Its application to the choana reduces the risk of restenosis.7,13 Nevertheless, there is insufficient evidence of its long-term effects to support its routine use.7

In our series, mitomycin was used until the tutor was withdrawn and subsequently to choanal dilatation. It was not used routinely (only in cases of granulations or suspected restenosis), so no conclusions could be drawn about its effectiveness.

The most frequent complications were related to the intranasal tutor (7 cases). In 5 patients, the nasal tutor was extracted, and in the 2 patients in whom removal had been accidental we decided not to replace it. Two patients presented septoturbinal synchieae, which were resolved using CO₂ laser.

Despite progress in the different surgical techniques, postoperative choanal stenosis is still frequent.8,10,26 Predictive factors of restenosis include nasopharyngeal reflux, gastroesophageal reflux, age less than 10 days (due to the normally narrow noses of newborns, and limited resection of the vomer), and insufficient postoperative endoscopic control.3,7,10 Restenosis is more frequent in the treatment of bilateral CA, when the atretic plate is purely bony, and when there are associated malformations.11

The need for revision documented in the literature varies widely from 10.2% to 89%.9,14,17 This is because the term "revision" is defined differently by different authors. Some consider as revision the additional removal of tissue, whilst others define it as any procedure under general anaesthesia subsequent to surgery, including the removal of a nasal stent.14

In total, 71.4% of our patients required at least 1 additional procedure under general anaesthesia after the first surgery. This high percentage could be influenced by many years of monitoring, a high proportion of bilateral cases, and removal of the nasal stent under general anaesthesia (removal of the stent was only performed without general anaesthesia in the initial years).

We highlight the importance of frequent postoperative nasal washes with saline solution and periodic endoscopic revisions to remove crusts and granulation tissue in neochoanae, in order to prevent restenosis.

We achieved normal nasal ventilation in 46 patients (93.9%) (Fig. 4). These cases remain asymptomatic and without clinical evidence of restenosis at 3.5 years of follow-up. One patient died due to causes unrelated to the surgery
or the nasal condition. Two patients with patent choanae still have a tracheostomy, one of them due to aspiration syndrome, and the other due to subglottic stenosis and glossoptosis.

Conclusions

Patients with CA should be studied thoroughly to identify possible associated malformations. It is essential to assess other sites of airway obstruction, muscle tone and the need for tracheostomy.

We observed the association of this condition with pre-natal exposure to maternal hyperthyroidism treated with methimazole.

Transnasal microsurgical repair proved to be a safe and effective procedure, and may be considered as an option for intranasal treatment.

Conflict of Interests

The authors have no conflict of interests to declare.

References