Endoscopic Treatment of Choanal Atresia

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Abstract

Introduction: Choanal atresia (CA) is an infrequent congenital obliteration of the airway at the level of the posterior nasal aperture resulting in the absence of connection between the nasal cavity and the aerodigestive tract. We present our experience with an endoscopic technique for congenital CA without the use of intranasal stents.

Material and methods: We analysed a series of 10 patients with CA treated in our department from 2006 to 2012 through endoscopic surgery. We present a description of the sample and the surgical technique used.

Results: The sample consisted of 5 men and 5 women. Mean patient age was 8 years (range: 5 days–32 years). Fifty percent of patients were cases with re-stenosis requiring revision surgery. Bilateral presentation was 7 and unilateral was 3. All CA were mixed (bony-membranous). Fifty per cent of patients had an associated malformation. All patients underwent nasal endoscopic surgery without stenting. After a mean follow up of 27 months (range: 11–78 months), the success rate was 100%. No complications were observed.

Conclusion: Transnasal endoscopic repair for both unilateral and bilateral CA without intranasal stenting was found to be a safe, expedient procedure that afforded minimal complications with a high success rate. Endoscopic endonasal surgery may be considered as the mainstay of treatment.

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Tratamiento endoscópico de la atresia de coanas

Resumen

Introducción: La atresia de coanas (AC) es una obliteración congénita y poco frecuente de la vía aérea, resultante de la ausencia de conexión entre la cavidad nasal y el tracto aerodigestivo. Presentamos nuestra experiencia en el manejo de la AC mediante endoscopia nasal sin utilización de stents.
Introduction

Choanal atresia (CA) consists of the narrowing or imperforation of the posterior nasal opening or choana, preventing proper communication between the nasal fossa and the nasopharynx. It was first described by Johan Roederer in 1755, although the first attempt to correct it surgically was performed by Emmert in 1851. It is an unusual congenital malformation that affects 1 out of every 5000–8000 live births and may be unilateral (more common, 60% of all cases) or bilateral (more serious). CA is more common in females (2:1) and there is a certain predominance on the right side. CA is associated with other, more or less serious congenital craniofacial anomalies in up to 50% of the cases, with CHARGE syndrome being the most commonly associated malformation.

Several different theories have been put forward to explain its aetiopathogenesis: (1) persistence of the nasobuccal membrane of Hochstetter (which is normally reabsorbed during the sixth week of gestation) (to date, this is the most widely accepted); (2) incomplete reabsorption of the mesoderm in the choanal region; (3) persistence of the buccopharyngeal membrane, and (4) abnormal migration of neural crest cells. It manifests anatomically as a narrow nasal cavity, altered growth of the vertical and horizontal palatine processes and thickening of the vomer and/or of the medial alae of the pterygoid processes. Histological studies reveal that the lateral pterygoid plate and the posterior vomer are expanded by the formation of endochondral bone and lined by a delicate fibroepithelial membrane that obstructs the choana.

Due to the fact that newborns are obligate nasal breathers (given that the larynx is located behind the velum), bilateral CA may present as potentially fatal neonatal asphyxia, particularly while swallowing, which requires emergency intervention. Unilateral CA generally presents with unilateral nasal obstruction and persistent rhinorrhea in late childhood, with some cases diagnosed in adulthood. Surgery in cases of unilateral CA may be delayed until the nasal fossae are larger, making surgery easier.

The diagnosis of CA, which is routinely screened for by the paediatrician immediately following delivery, is based on clinical examination. Computerised tomography (CT) confirms diagnosis in suspected cases and is fundamental for surgical treatment. Most CA are mixed (70%), while bony CA are less common (30%). Membranous CA are considered to be non-existent.

Treatment is always surgical. Various different surgical techniques have been described over time for the treatment of CA. Moreover, there is a fair degree of controversy regarding the usefulness of placing stents, adjuvant mitomycin and timing of surgery in unilateral cases.

The purpose of this study is to present our experience in treating CA, focusing largely on the type of surgical approach used, and on revising the management of this pathology based on the existing bibliography and on our own experience.

Material and methods

The ENT department’s surgical registry at our hospital from 2006 to 2012 was reviewed, collecting data from the case histories of patients with a diagnosis of CA. The study population was comprised of 10 patients who underwent surgery consecutively.

Data collection was based on reviewing case histories, recording data as to age, gender, prior surgeries, type of CA, associated malformations, personal and/or family history, clinical presentation, surgical technique, complications and follow-up. We have defined restenosis as a 50% decrease in the diameter of the neochoana.

A CT was taken in all cases prior to surgery. In some of the cases of bilateral CA, a choanography was performed (Fig. 1).

Preoperative preparation

We did not use a navigation system in any of the cases. The procedure is performed with general anaesthesia and orotracheal intubation. In all cases, disks impregnated in local anaesthetic (tetracaine) and a vasoconstrictor (adrenaline 1%) are applied in both nasal fossae. Patients are placed in supine position, with their head slightly elevated in anti-Trendelenburg. All patients are given preoperative antibiotic prophylaxis with intravenous cefazolin (in the event of allergy to β-lactams, clindamycin and gentamycin are used) at the appropriate doses based on age and weight.
Surgical technique

Endoscopic naso-sinus surgery (ENSS) is carried out in all cases. The surgery is performed simultaneously by 2 ENT surgeons. A 0°, 4 mm nasal endoscope (Karl Storz) is used. In those cases in which the CA is bilateral, surgery begins with the larger nostril that allows for better exposure. In these cases, initial septovomerian resection makes it possible to later work through both nostrils.

The surgical technique used is similar to that used by Stam et al.4 Prior to beginning the surgery, it is useful to place one or more disks orally in the cavum (perfectly attached and counted) to fill the nasopharynx and make orientation easier once the atretic plate is perforated.

Initially, on the more favourable side, a vertical, hemitransfixion incision is made in the septum, approximately 1 cm anterior to the atretic plate, using an electric knife. This incision continues along the floor of the fossa obliquely in the direction of the tail of the inferior concha. A mucosal flap (that will be L-shaped in the left nasal fossa and J-shaped in the right nasal fossa) is then raised to expose the osteocartilaginous septum and the atretic plate. This flap must be preserved during the entire surgery. On the contralateral side, a vertical incision is made in the septum at the level of the contralateral incision. This incision is extended superiorly, at a right angle, at the level of the free edge of the middle concha to the choanal border. Analogous to the previous side, a mucosal flap is raised to expose the septum.

Once both flaps have been raised, a posterior septectomy is performed and the atretic plate is eliminated. To do so, different instruments are used, depending on the type of atresia: punch, curettes, electric knife, Kerrison forceps, microdebrider or, preferably in our case, a burr.

Initially, after retracting the mucosa covering it, the atretic plate is punctured at the weakest point and inferomedially in order to avoid inadvertently penetrating the intracranial area or the eye socket. The puncture is generally made with the tip of the aspirator, even in purely osseous cases (in those cases in which this is not possible, a burr or chisel might be useful). After opening the communication with the nasopharynx (the disks previously placed there are easily identified), it is extended circumferentially with forceps or microdebrider for the soft tissues and with the burr for the bone in the pterygoid and sphenopalatine region. It is important to separate the pharyngeal mucosa from the nasal mucosa properly on the lateral edge of the neochoana in order to expose the medial plate of the pterygoid processes. The septectomy, which must include part of the perpendicular plate of the ethmoid bone and the posterior part of the vomer and the rostrum sphenoidale (that are generally hypertrophic), must go superiorly to include the inferior edge of the middle concha. This step is fundamental to avoid restenosis.

Once a broad communication between the nasal fossae and nasopharynx is established, the flaps must be carefully replaced to cover the bone exposed on the floor and roof of the neochoana. The flaps are placed in the most convenient way possible; however, we usually rotate the ipsilateral flap superiorly to cover the choanal arch and the contralateral flap is rotated inferiorly to cover the floor of the nasal cavity (Fig. 2A). At times, depending on the amount of mucosa preserved, it may be advisable to resect the excess mucosa in order to adapt it properly, albeit at all times avoiding any areas being left denuded of mucosa that might heal at the second intent with the formation of granulation, thus favouring restenosis. Prior to placing the packing, the adenosids are generally curettaged, with the surgeon’s preferred technique, since it will prevent the discomfort of the eventual bleeding that will take place during the surgery if performed first.

Packing

In all cases, after performing meticulous haemostasis, Merocel® or Netcell® (MedTronic Xomed, Jacksonville, FL, USA) or packing with a neurosurgical disc (in infants) is put in place and then removed one or two days after surgery. Stents were not placed in any case. No topical mitomycin or corticosteroids were applied.
Postoperative care

After removing the nasal packing, we recommend nasal lavage with saline, as well as ointment containing hydrocortisone and oxytetracyclin, and Vaseline, later on, in each nasal fossa.

If evolution is satisfactory, after being discharged from hospital, the first check-up is one month post-op. For most patients, we recommend that they continue lavages with water and applying topical ointment or Vaseline, and in those cases in which the mucosa has no scabbing, a spray with topical corticosteroid is recommended instead. Subsequent check-ups are scheduled annually. Follow-up and verification of choanal patency are verified by means of targeted history and nasal endoscopy (Fig. 2B).

Results

Table 1 presents patient characteristics. The sample consisted of 10 patients (5 males and 5 females) having a mean age at the time of surgery of 8 years (range, 5 days–32 years). Half of the cases (5 cases) were re-interventions in patients who had previously undergone surgery at other departments using an open transpalatine approach. Three of the patients had had stents placed in a prior surgery. Seven cases (70%) presented a bilateral CA and, in 3 cases (30%), the CA was unilateral. In total, 17 sides were treated. Of the cases with bilateral CA, 4 were interventions due to restenosis; hence, the patients were over the age of 8 years. In the 3 cases of bilateral CA undergoing surgery for the first time, the procedure was carried out at 5, 8 and 23 days of age.

Fifty per cent of patients (5 cases) had associated malformations, the most common finding being Treacher–Collins syndrome, since it appeared in 2 of the cases. Of the patients who had already undergone surgery (5 cases), 3 (60%) presented an associated malformation. Two patients had a premature birth and in one of them, the mother had hyperthyroidism, as well as ulcerative colitis under treatment with thyroid hormone and an immunosuppressant (azathioprine), respectively.

In all the cases, the CT study revealed the presence of a mixed CA.

In both the cases of bilateral CA, as well as those of unilateral CA, the surgery was scheduled immediately after the visit to the clinic.

<table>
<thead>
<tr>
<th>Cases</th>
<th>Age</th>
<th>Gender</th>
<th>Personal history</th>
<th>Family history</th>
<th>Associated malformations</th>
<th>Laterality</th>
<th>Type</th>
<th>Re-intervention</th>
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<tr>
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<tr>
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<tr>
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<td>M</td>
<td>Premature</td>
<td>Maternal H.</td>
<td>Treacher–Collins</td>
<td>Bilateral</td>
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</tbody>
</table>

EAC, external auditory canal; H, hypothyroidism; F, female; M, male.
The mean surgical time was 45 min (range 25–85 min). No intra-operative or postoperative complications were seen in any of the cases, with a very good postoperative evolution. After surgery, nose breathing was observed in all patients both while awake and while sleeping. In the infants, oral nursing was initiated the same day the packing was removed without any type of respiratory difficulty or aspirations. All patients were discharged on the second day following surgery.

After a mean follow-up time of 27 months (range 11–78 months), there have been no restenosis detected in any of the cases and no further surgery or dilatations were required.

Discussion

The analysis of the outcomes achieved in this 10-patient series (17 sides) who underwent surgery via ENSS in our service is satisfactory, given that no further surgery was required and we have not observed a single case of restenosis. Although only a few patients underwent the surgery, given that this is not a highly prevalent pathology and that most of the studies in English present a heterogeneous case series consisting of fewer than 30 patients undergoing surgery, our optimal outcomes provide a result worthy of being taken into consideration.

The mean follow-up time for our patients was 27 months. All were examined on the basis of their history, in search of symptoms and signs that would be grounds for suspecting possible restenosis (difficulty eating or mouth breathing while sleeping in the cases of bilateral CA and rhinorrhoea and nasal respiratory insufficiency in the cases of unilateral CA), as well as by endoscopy, until such time as it was deemed unnecessary as it was unlikely for any reduction of the choanal framework to occur. As has been previously shown, if restenosis is to take place, it will do so within the first year after surgery; hence, we can be fairly confident in accepting the outcomes achieved in our study as definitive.

Although we have not been able to confirm this in our series, given the 100% success rate, it appears that CA having a large bony component display a higher rate of restenosis as they exhibit neo-osteogenesis phenomena. It is therefore important for the choanal framework to be sufficiently expanded and lined with mucosa, as per our description. In some series, the need for revision surgery is greater in CA associated with other malformations. However, Teissier et al. in their series of 80 patients did not detect this relationship. In our series, 60% of the revision surgeries were performed in patients with CA associated with another malformation.

In some cases of bilateral CA, a certain autosomal dominant genetic component has been observed (largely in cases associated with other malformations caused by a defect in the development of the neural crest) with incomplete penetrance and variable expressivity, although it is generally considered to be a sporadic condition, as in the patients included in our series. CA has been associated with the use of several different medications, such as thyroid drugs or immunosuppressants, which is consistent with our observations in one case of bilateral CA.

The aim of treatment for CA is to restore the flow of nasal air without injuring structures that produce craniofacial development by means of safe and efficacious techniques. Bilateral CA diagnosed in the newborn are usually a neonatal paediatric emergency. Oral feeding in these children can lead to pulmonary aspiration. Symptoms can vary from mild respiratory distress with feeding to severe obstruction of the airway. Surgery should be performed early and, in the meantime, special devices should be used to control the airway (McGovern-type pacifiers, Guedel cannulae) and even, orotracheal intubation if there are signs of respiratory distress. In our series, the primary cases of bilateral CA underwent surgery in the first few days of life. In contrast, the revision surgery of bilateral CA was performed in patients over 8 years of age, who were mouth breathers, making their treatment less urgent. It is possible for unilateral CA to be detected later on and, although emergency treatment is not required, it is wise to intervene as soon as possible in order to improve the children’s quality of life. Achieving proper bilateral nasal patency is extremely important for craniofacial development in the first years of life and, later, for optimal quality of life. As seen in neonates, the endoscopic approach has no technical or space limitations.

High resolution CT is the key radiological study to confirming diagnosis. Performing a diagnostic CT scan, particularly in early infancy, has the disadvantage that it may be necessary to do so under sedation or general anaesthesia. Nonetheless, we believe that it is an indispensable study to identify possible anatomical variations and, particularly, bearing in mind that these patients may have more than one malformation.

One essential aspect to be assessed on the CT is the status of the development of the rhinopharynx. Its proper development will make the surgery much easier as regards orientation and safety, as well as providing a more favourable prognosis. Slovis et al. have established certain parameters for appraising choanal atresia. In neonates, the vomer has a width of 0.23 cm, increasing to 0.28 cm by the age of 8 years. The distance from the lateral nasal wall to the vomer in the neonate is 0.67 cm, and later increases by 0.027 cm per year until the age of 20 years. Neonates with unoperated CA have a broadened vomer (0.6 cm on average) and the choanal air space is missing. The use of intra-operative navigation may be useful, but not essential as demonstrated in our series, in which it was never used.

Different surgical approaches have been used to treat CA: transmaxillary, transseptal, transpalatine, transnasal, sublabial, paralateronasal and endoscopic nasal. Depending on the time and school of thought, one or another have been used, although with experiences such as ours, we have tried to show that the endoscopic nasal approach brings together the greatest advantages and should be considered the standard. The ideal method should be whichever one achieves the highest rate of permanent success in restoring communication between the nasal fossae and the nasopharynx, while causing as little morbidity as possible.

The transpalatine approach has been the most widely used throughout history. Advocates of this approach report that it provides them with a large surgical field and, therefore, greater probability of resolving the problem. Nevertheless, this approach is associated with a higher rate of postoperative complications (palatine necrosis, palatine
fissures, haemorrhaging, ...) and greater morbidity (up to 52% of dental malocclusions and alterations in craniofacial development). In our series, we must highlight the fact that all the patients who had surgery due to restenosis had had a previous surgery using a transpalatine approach and lasting nasal patency was achieved after the ENSs surgery.

Ever since Stankiewicz performed the first endoscopic correction of a CA in the 1990s, ENSs is the most widely used technique thanks to its high success rate, as exhibited by our series, its safety, the lack of external scars and its scant postoperative morbidity. ENSs reduces surgical time, allows direct vision, excellent lighting and avoids palatomedical complications. In the children and, especially in newborns, insertion of the endoscope in the nasal fossae at the beginning of the surgery scarcely allows any other instrument to be inserted. However, the dilatation of the nostrils, the lateral luxation of the inferior turbinates and working via both fossae favours the surgery in all the cases without requiring the use of paediatric endoscopes.

It is important to point out two fundamental steps in this kind of surgery: increasing the surface of the choanal frame by eliminating the posterior part of the vomer until a "single" neochoana is formed and the creation of the mucosal flaps that will cover the exposed bony surfaces on the edge of the choana, thus decreasing the formation of granulation tissue and scarring phenomena. Both these surgical gestures lower the incidence of restenosis. It has been shown that the all but total elimination of the vomer does not affect facial growth. There are authors who do not believe that the creation of mucosal flaps is of value, unlike our group and other authors. The results obtained with the ENSs reported in the literature report variable rates of restenosis, due to the heterogeneity of the cases (uni- or bi-lateral CA and the use or not of adjuvant therapies), around 10%–20%. These figures are comparable to those that correspond to the transpalatine approach, albeit with hardly any complications (<1%). One important aspect is the learning curve needed for CA repair via ENSs, since the success rate is linked to greater experience.

After obtaining adequate opening of the choanae, some authors apply topical mitomycin C or place a temporary stent to prevent choanal closure. In our series, we have not used either and the outcomes achieved have been optimal. The use of both adjuvant methods is subject to controversy.

The use of the antimetotic mitomycin C is supported by several authors to prevent the growth of scar tissue and thereby prolong nasal patency. However, other studies have failed to find significant differences. Recently, Newman et al. did not find significant differences in the rate of restenosis in their series of 31 patients undergoing an ENSs procedure, although they observed a higher rate with its use (32% versus 10%). The use of mitomycin C is not routine and is often used in more difficult cases, which would explain the increased incidence of restenosis among patients treated with this drug.

Classically, stents have been used to avoid restenosis. However, fewer and fewer authors promote their use, as is our case. The premise for the placement of a stent is that it would hold the mucosal flaps in place and would prevent the narrowing of the choanal lumen as healing takes place. Nevertheless, authors who are detractors of its use argue that the stents damage the nasal mucosa, which would lead to an increase in granulation tissue, scarring (particularly in the columella), bacterial overgrowth and biofilms, and alteration of mucous drainage. All of these factors would work against the ultimate aim of achieving lasting patency.

Several publications and a review of the Cochrane database conducted by Cedén et al. has concluded that there are no differences in the use of stents with respect to the rate of restenosis. In the event that stents are placed, it appears that they must remain in place for longer than 15 days to be useful. As in the case of the use of mitomycin C, although there does not appear to be any difference regarding the rate of restenosis, this might be due to a selection bias, since in most of the series the decision as to whether to use stents or not is made based on a greater severity of the disease. Furthermore, most of the studies conducted have been retrospective in nature. For this reason, it cannot be concluded that stents should be universally used or abandoned; however, our impression is that they are unnecessary when an appropriate surgical technique is used.

We have observed that postoperative irrigation with saline and suction of the nasal cavities is beneficial, as is the use of topical corticosteroids starting on postoperative day 15. These manoeuvres seek to maintain the nasal fossae free of waste or scabbing and to minimise inflammation during the first 2–3 weeks following surgery. This would decrease the possibility of infection and the formation of granulation tissue and, hence, restenosis.

Conclusions

Both the surgical sequence proposed, which achieves outstanding visualisation of the choana and a broad neochoana, and the postoperative care administered in our series make ENSS without stent placement a simple, fast and safe procedure with excellent patient recovery. ENSS without placing stents and without the use of adjuvant therapies makes it possible to achieve stable, long-term choanal patency with no associated morbidity, in both unilateral and bilateral settings, regardless of the type of atresia, the existence of associated malformations and of the age at the time of intervention. Therefore, we believe that ENSS should currently be considered the surgical standard of care in the treatment of unilateral or bilateral CA.

Conflict of interest

The authors declare no conflict of interest.

References