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Long-term outcomes of endonasal surgery for choanal atresia: 28 years experience in an academic medical centre

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Abstract This study aimed to evaluate the long-term outcomes of endonasal surgical treatment for choanal atresia (CA) and determine possible predictors of recurrence. A retrospective study of 50 cases of CA that were managed endonasally was conducted at an academic tertiary referral centre. Recurrence of disease was assessed with regard to the type and nature of atresia, presence of CHARGE association and stent usage. Bilateral CA cases ($n = 76$ sides) were operated at an early age and had an incidence of recurrence of 57%, while unilateral cases had a recurrence rate of 25% (3/12). Use of stents did not have a statistically significant effect on the rate of recurrence. However, a significantly higher recurrence rate was noted in patients with purely bony atretic plates ($p < 0.001$) or CHARGE association ($p = 0.049$). A tendency towards a higher recurrence rate in bilateral atresias was also found ($p = 0.085$). The transnasal approach is a safe and effective way to re-establish the nasal choanae. However, appropriate information on the likelihood of recurrence should be included in the preoperative counselling of patients and their parents, especially in cases with bilateral atresia or CHARGE association.

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Introduction

Choanal atresia (CA) is a rare congenital disorder of the posterior nasal cavity, first described by Johann Roederer in 1755 [1]. Congenital CA has an incidence of about 1 in 7,000–8,000 live births [2, 3], and affects females almost twice as often as males. It is the second most common congenital nasal disorder after dermoids [3, 4]. However, no large series have been reported in the English literature. The first report of surgical management of CA was from Emmert in 1853 [1]. Association with other anomalies and syndromes ranges from 20 to 50%, with CHARGE association being the most common (C-coloboma, H-heart disease, A-atresia choanae, R-retarded growth and development and/or CNS anomalies, G-genital hypoplasia, and E-ear anomalies and/or deafness) [2]. Unilateral CA may remain undetected for a long time and it commonly presents later in childhood or adolescence as recurrent rhinosinusitis or otitis media. By contrast, bilateral CA presents as a medical emergency immediately after birth [1]. Infants present with apnoea or paradoxical cyanosis, since they are obligate nasal breathers during the first weeks of life [5]. Bilateral atresia has also been reported in an adult with a normal neonatal history [3].

Until now, surgical treatment has been the only acceptable curative approach to CA [6]. The aim of CA surgery is to achieve complete and long lasting resolution of symptoms, combined with low morbidity, low risk of recurrence and short hospitalisation. Various surgical techniques have been proposed, including navigation-assisted endonasal, transpalatal, transeptal and transantral approaches [7]. Despite advances in the different surgical techniques, restenosis remains a significant issue for the surgeon [5].

The main aims of this study were to evaluate long-term outcomes of endoscopic–microscopic CA surgery and

determine the potential significance of various factors affecting restenosis. This could be of practical importance in providing patients and their parents with useful information on the probability of recurrence requiring revision surgery.

Materials and methods

We conducted a retrospective review of 50 patients who underwent primary endonasal surgical treatment for CA at an academic tertiary referral centre (Department of Otorhinolaryngology, Head and Neck Surgery, University of Erlangen-Nuremberg, Germany) between 1982 and 2010.

A total of 17 (34%) males and 33 (66%) female patients were included. Mean birth weight was 2,740 g (range 980–4,150 g). Diagnosis of atresia was typically based on nasal endoscopy and axial CT of the head, whereas the cold mirror test, the methylene blue dye test and nasal probing were used in isolated cases.

All surgical procedures were performed under general anaesthesia. For the endoscopic–microscopic approach, a 2.7 mm 0° endoscope was used (Storz, Tuttlingen, Germany). The nasopharynx was packed with ribbon gauze for protection. The mucosa over the atretic plate was then removed. While staying inferior and medial in the nasal cavity, the atretic plate was perforated using powered instrumentation (microdebriders) or perforators and dilators. For stenting, an endotracheal tube stent (no. 2.5–3.5 in neonates and larger in adults) was folded and fenestrated posteriorly. The posterior fenestrated end was placed in the neochoanae, bridging over the vomer, and was fixed in position with two atraumatic sutures.

Recurrence of CA was defined as a return of symptoms in combination with endoscopic presence of an atretic plate at any time after primary surgical intervention. Surgical revision was defined as secondary tissue removal or secondary dilatation of the choana after primary surgery [8–11].

Pertinent parameters, such as type and nature of CA, presence of CHARGE association and stent usage were assessed in relation to recurrence. Surgical outcomes were analysed using SPSS 18.0 for Windows (SPSS, Inc, Chicago, IL, USA). Comparisons were based on the Chi square or Fisher exact test for categorical variables. A p value of <0.05 was considered statistically significant.

Results

As shown in Table 1, there were 38 bilateral cases ($n = 76$ sides, 76%) and 12 unilateral cases (24%). The ratio of bilateral to unilateral cases was therefore 3.17:1. In unilateral cases, the right side was affected in 7 cases and the left side in 5 cases.

Dyspnoea and cyanosis were the most common clinical symptoms of bilateral choanal atresia, whereas nasal obstruction and chronic nasal discharge were the most common presenting symptoms of unilateral choanal atresia. CHARGE association was evident in 11 cases. The association of pertinent factors with recurrence can be seen in Table 2. In the unilateral atresia group, the average age at diagnosis was 10.6 years (SD = 8.3) and at the time of operation 12.1 years (SD = 8.7). In the group of bilateral cases, the mean age at the time of diagnosis was 1 day and at the time of operation 4 days.

Table 1 Clinical characteristics of choanal atresia

	Bilateral atresia ($n = 76$ sides)	Unilateral atresia ($n = 12$ sides)
Gender		
Male	12	5
Female	26	7
Type of atresia		
Bony	47	10
Membranous	23	2
Mixed	6	
Side		
Right	38	7
Left	38	5
Mean age		
Diagnosis	1 day	10.6 years (SD = 8.3)
Operation	4 days	12.1 years (SD = 8.7)
Major symptoms	Dyspnoea, cyanosis	Nasal obstruction, chronic nasal discharge

Table 2 Factors correlated with recurrence of atresia

	Recurrence (no of recurrences/no of sides) and percentages	p value
Type of atresia		
Bilateral	43/76	0.085
Unilateral	3/12	
Nature of atretic plate		
Bony	44/71	**<0.001**
Membranous	2/11	
Mixed	0/6	
CHARGE association		
With CHARGE	16/22	**0.049**
Without CHARGE	30/66	
Stent at primary surgery		
Yes	41/73	0.184
No	5/15	

Mean follow-up was 5.2 years (2.0–19.4 years). Revision surgery was required in 26/50 cases (52%). Additional procedures under general anaesthesia, including stent replacement or removal, were required in 32/50 cases (64%). Emergency intubation or, in rare cases, tracheotomy were necessary in 33 patients, 31 of whom suffered from bilateral CA. The remaining two cases with unilateral CA had an atretic plate on one side and a very narrow (but not atretic) choana on the other side.

Of the parameters examined, presence of a purely bony atretic plate and CHARGE syndrome were significantly associated with disease recurrence ($p < 0.001$ and $p = 0.049$, respectively). Bilateral CA ($p = 0.085$) and the use of stents ($p = 0.184$) showed slight correlations with the need for restenosis, but these correlations were not statistically significant.

Immediate postoperative complications were observed in 3/50 cases (6%). In two bilateral cases, operated primarily without stenting, a stent was later placed because of acute dyspnoea due to re-occlusion of the choanae during recovery from surgery. One patient presented postoperatively with epistaxis, which was managed with nasal packing that was removed 2 days later. With regard to long-term complications other than restenosis, one patient developed a malformed nose.

Discussion

Despite progress in surgical treatment of CA, restenosis remains a challenging problem for the head and neck surgeon. A careful review of the relevant literature reveals that the term ‘revision’ is defined differently by many authors. In many studies, revision is defined solely as removal of additional tissue [8–10, 12]. However, other authors define revision as any secondary intervention under general anaesthesia, including stent replacement or removal [5, 11]. Moreover, many reports include patients who were operated primarily in external institutions [10, 12]. Taking these aspects into consideration, the restenosis rate reported in the literature varies from 10.2% [10] up to 89% [5]. Consistent with published reports, revision surgery was necessary in half of our study patients. However, there was a relatively high proportion of bilateral cases in our series (76%).

Prevention of restenosis is an important challenge in surgery for CA [5]: intraoperative local application of mitomycin, steroid therapy and stenting have all been proposed [2], but indications, materials, and duration of stenting remain controversial [6, 13]. Moreover, adverse effects of stenting have often been reported. Friedman et al. [14] suggested that stents may stimulate formation of scar tissue due to injury of the surrounding tissues. Schoem [4] claimed that this prolonged stent-related mucosal trauma could lead to clinically relevant restenosis. Other possible complications include stent migration or damage of the septum, columella, and nasal ala [15]. Havel et al. [16] regarded stenting as indicated only for a limited period of time and only in cases where it is absolutely necessary (e.g. mechanical ventilation). Other authors support stenting in bilateral cases [17] or even in every case [18]. However, there is little evidence in the literature to show that stenting continues to prevent recurrence after the stent is removed [19]. Although there is a trend towards less frequent use of stents with the improvement of endoscopic techniques, they remain in use by many surgeons on a case-by-case basis [20]. In our study, we found no significant association between stenting and recurrence of atresia. It may be that stenting protects the choanae from occlusion due to secretions in the early postoperative phase, without affecting choanal patency on a long-term basis.

Another important issue is whether existence of a bilateral atretic plate is correlated with recurrence. Van Den Abbeele [11] found no correlation ($p = 0.41$). However, according to Teissier et al. [10] and Chia et al. [21], bilateral CA is associated with an increased risk of restenosis. This tendency was also found in our series ($p = 0.085$). The surgeon should take this relationship into consideration and counsel patients and their families accordingly.

As regards the nature of the atresia, Teissier et al. [10] found a tendency towards higher recurrence rates in patients with purely bony atretic plates ($p = 0.08$). We found a much stronger correlation in our patient sample ($p < 0.001$).

As mentioned, CHARGE association was present in 11 of our study cases (20.3%), and was by far the most common congenital anomaly. Interestingly, almost every third bilateral case was associated with this syndrome. This is consistent with reports in the literature, and suggests that further diagnostic investigation to detect CHARGE or other chromosomal anomalies is justifiable, especially in bilateral cases [22].

Few reports have investigated and analysed statistically the influence of several factors on recurrence of CA [10, 11]. The relatively high recurrence rate in our study could be attributed to the high proportion of bilateral cases. There was also a high incidence of bony atretic plates in our patient group (87%), contributing further to our high revision rate.

Conclusion

The transnasal endoscopic approach is currently considered to be the treatment of choice for CA, being safe, minimally invasive and reliable. Presence of a purely bony atretic plate was strongly correlated with restenosis of the

choanae. An obvious tendency towards restenosis was also found in cases with bilateral atresia and CHARGE association. Stenting seems to facilitate patient care by preventing occlusion of the choanae in the early postoperative phase. However, its role in preventing restenosis in the long term was not supported by our data.

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