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Forty-one cases of congenital choanal atresia over 26 years - retrospective analysis of outcome and technique*

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SUMMARY

This retrospective analysis reflects the outcome of various techniques used in a series of 41 cases of choanal atresia treated at the Department of Otoloaryngology, Head- and Neck Surgery at the University of Mainz between 1980 and 2006. Thirteen bilateral and 28 unilateral cases are included. After endonasal management in 38 and a transpalatine approach in 3 cases a total of 15 patients needed revision surgery between 1 and 5 times to establish a stable result. Postoperative stenting was used in 23 patients with a failure rate of 35%, whereas only 11% of the 18 patients without stenting had to be revised. None of those 5 cases where Mitomycin C had been applied intraoperatively in combination with postoperative transnasal dilations needed surgical revision. We conclude that the endonasal micro-endoscopic surgical approach is successful if combined with postoperative dilations for up to one year. Stenting should be abandoned as it stimulates granulation formation that frequently leads to restenosis. The intraoperative application of Mitomycin C offers a promising adjunct in achieving a stable result.

Key words: choanal atresia,surgical technique, follow-up, stenting, mitomycin c

INTRODUCTION

In choanal atresia a widened vomer fuses with the narrow posterior nasal airway to an atretic plate, which can be either bony or membranous, resulting in an hourglass configuration to the nasopharynx and the choanal region. Among the reasons for failure after attempts of surgical correction in the past was sometimes inadequate analysis of the exact dimensions of anatomical components of the atretic plate but more frequently early restenosis of the choanal opening due to granulation and scarring. It is one of the most frequently observed congenital abnormalities of the nose with an incidence of 1 in 7000-8000 live births. Since its first description in 1755 its demographic characteristics have been repeatedly confirmed with a female predominance of 2:1 and a ratio of bilateral to unilateral of 40% to 60% [1,2]. The atretic plate is either purely bony (30%) or mixed of bony and membranous components (70%) [3]. Associations to other congenital abnormalities are present in about 50% and up to 75% in bilateral manifestations. The by far most frequent syndromal association in about 20% is the CHARGE – malformation. As suggested by Davenport et al. [4], two of the following defects must be present to establish this diagnosis: **C**oloboma, **H**eart, **C**hoanal **A**tresia, **R**etardation, **G**enito-urinary and **E**ar. Other isolated malformations frequently observed are meningoceles, hypertelorism and clefts. Different embryologic origins have been proposed, including a failure of the buccopharyngeal membrane to break and more recently another theory is the misdirection of mesoderm in the nasal cavities leading to a medial outgrowth of the horizontal and vertical process of the bony palate [5-7].

Bilateral atresia presents as a neonatal emergency as newborns are known to be obligate nasal breathers during the first 3-4 weeks of life. Immediately after birth the tongue is in contact with both soft and hard palate with the epiglottis being positioned above the soft palate. These infants therefore present with periodic respiratory distress and cyanosis, released by crying and pallor as well as severe feeding problems and aspiration. Immediate airway management and oropharyngeal airway intubation within the first hours of life are needed before planning surgical correction, which is performed within the first days of life. The diagnosis in most cases can be established by the inability to pass a catheter through either nostril into the pharynx and or endoscopic examination. However, today, state of the art workup includes a thorough endoscopic examination and a multi slice high resolution CT scan to analyze the individual anatomical topography as well as the bony or mixed bony / membranous nature of the atretic plate [3,8].

Over the last century four approaches have been discussed for surgical correction: Transseptal, transantral, transpalatine and trans- or endonasal, with the latter two still being frequently used. Since the advent of endonasal endoscopy and the

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endonasal microscopic technique, the endonasal approach has evolved to be the most common and is preferred today because of its excellent visualization and magnification that results in increased safety and reduced surgical time [9,10]. The use of various instruments has been described, including the use of bougies, dissectors, biting and cutting instruments, drills and lasers. To increase patency rates, some authors suggest the use of mucosal flaps to minimize the amount of corresponding raw surfaces while others have reported on the intraoperative application of Mitomycin C [8,11-13]. The most controversial aspect appears to be the use of stents for anywhere between 4 and 12 weeks [3,12]. However, stents have been associated with an overall failure rate of 30% regardless of the chosen approach or the respective surgical technique.

It is the objective of this retrospective analysis to discuss controversial aspects in the treatment of uni- or bilateral congenital choanal atresia and present a protocol based on the extensive experience gained from a large series of 41 consecutive cases at the Department of Otolaryngology, Head and Neck Surgery at the University of Mainz, Medical School in Germany.

MATERIAL AND METHODS

This is a retrospective, longitudinal and descriptive review of the surgical outcomes of a series of 41 patients with congenital choanal atresia. Of all 53 patients, who underwent surgery for uni- or bilateral choanal atresia between 1980 and 2006 at the Department of Otolaryngology, Head and Neck Surgery at the University of Mainz, Medical School in Germany, complete files, meeting the criteria for this retrospective analysis, were available in 41 cases. These inclusion criteria were: Cases of congenital choanal atresia with a complete chart available including medical notes on preoperative clinical course and symptomatology, documentation on diagnostic methods applied and clinical findings, operative reports on every surgical procedure, medical notes and / or reports on the postoperative course. Absence of symptoms or of endoscopic findings indicating restenosis had to be documented for at least one year postoperatively before the surgical procedure was assumed successful. However, in most cases more than one-year follow up was available, so long-term success could be evaluated. As documented in the patients' charts the individual follow up period varied between 14 months and 19 years. The mean follow up in our 41 patients was 41 months. All cases of acquired choanal obstruction or stenosis secondary to inflammatory or neoplastic disease, to trauma or to collagen disorders were excluded. Any respective files of our pediatric department or from other institutions involved with our patients were also included in the data acquisition. Such additional information was available in 31 of our cases. Of the 41 patients investigated 27 were female and 14 male. At the time of first surgical intervention the youngest girl was three days old and the oldest patient at the age of 67 years. Variables analyzed were: specifics of the individual malformation (bilateral, right – left), concomitant malformations, sex, age at first surgery, number of interventions necessary, initial symptoms, diagnostic procedures, surgical approach and surgical technique used, use of stents, use of Mitomycin C, surgical complications, postoperative choanal patency.

Statistics

Using SPSS 12.0 for windows, the complete database with all values for each variable was analyzed. Due to the small size of the studied patient group, the Fischer exact test was applied for statistical evaluation of the respective parameters and the relation between outcomes and surgical techniques

RESULTS

Thirteen patients had bilateral manifestations, 14 had unilateral disease on the right and 14 on the left side. There was no influence on the affected side by the respective gender of the patients. The most common syndromal manifestation was in our patient population was CHARGE association in 5 infants (12%), followed by Francescetti syndrome in 2 (5%), Fraley syndrome in 1, trisomia 4q in 1 and trisomia 22 in 1. In 5 of these infants with identified syndromal disorders and in 7 others, further isolated abnormalities were observed (Figure 1).

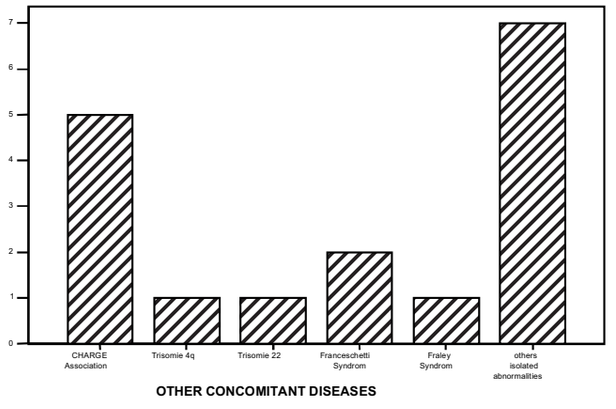

Figure 1. Incidence of concomitant diseases and deformities in 41 cases of uni- or bilateral congenital choanal atresia.

The most common diagnostic sign to establish the diagnosis was the inability to pass a small rubber catheter through the nose. This was the case in 28 patients, which included the 19 neonates, diagnosed in the delivery room. Nasal endoscopy was used diagnostically in 15 cases, a mirror test to ensure nasal breathing was diagnostically applied in 2 cases and one in 1984 in whom a plain radiograph with radiopaque medium was performed. Only 6 patients in this long-term review had a nasal CT scan for further evaluation. The classification of the nature of the atretic plate into bony or mixed bony / membranous was made based on the intraoperative findings. A purely bony plate was present in 15 cases (37%) while it revealed to have bony and membranous components were present in 26 cases (63%). The preferred approach for surgical correction of con-

genital atresia at present in the Department of Otolaryngology, Head and Neck Surgery in Mainz is the micro-endoscopic endonasal one. All, except 3, were operated endonasaly. The other three were operated on via a transpalatine approach due to an insufficient nasal passage. In a 40-year-old lady with unilateral atresia this was due to a severe endonasal deformity, which was corrected by an open septorhinoplasty at a later stage. In two secondary cases nasal passage challenged subsequent to complete restenosis by scarring and synechia formation. Between 1980 and 2006 this surgery was performed by 8 different senior otolaryngologists.

Looking at the number of surgical interventions necessary, 26 patients (63%) underwent only one procedure, 10 (24%) underwent two, 3 (7%) had three interventions while one needed four and another one five operations to achieve a satisfactory long-term result. The surgical instruments chosen depended on the nature and thickness of the atretic plate; these included

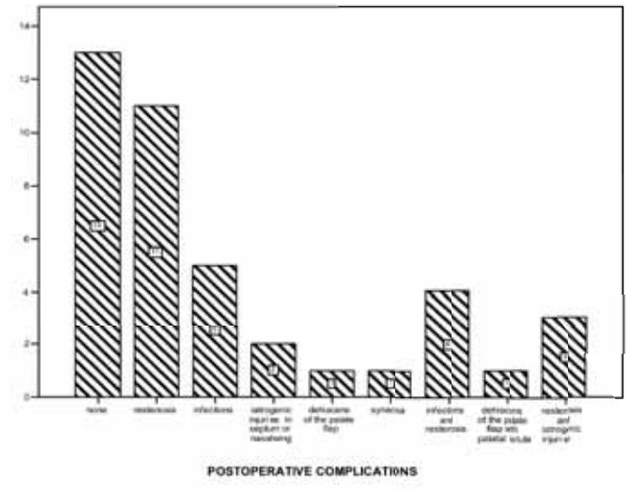

Figure 2. Frequency of various postoperative complications observed both short and long term.

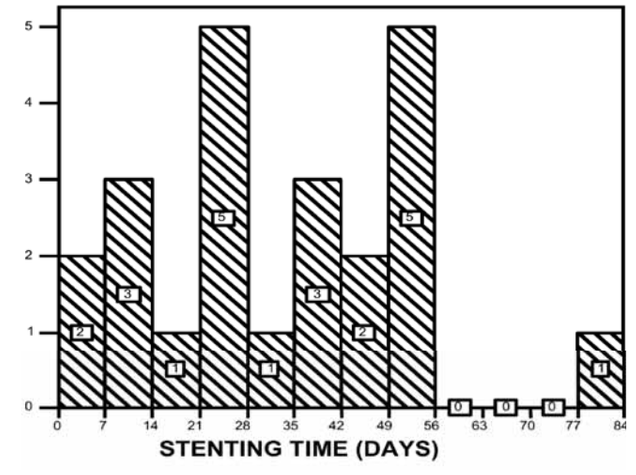

Figure 3. Analysis of the respective duration of postoperative stenting time in all individuals, in which stents were used.

Table 1. Comparison of surgical outcomes in patients treated with postoperative stenting as opposed to those treated without.

			surgical outcomes		
			success	**failure**	**Total**
Stents_vs._	Stents	Frequency	15	8	23
no stents		% of patients with stents	65%	35%	100%
	No Stents	Frequency	16	2	18
		% of patients without stens	89%	11%	100%
Total		subtotal	31	10	41
		Percentages	76%	24%	100%

bougies, dilators, cutting forceps and punches as well as powered drills, mostly used in combination. The drill was the most frequently used, serving as the most suitable tool to widen the medial aspect of the aperture by taking down the thickened vomer and posterior septum.

There were no life threatening intraoperative events such as spinal or skull base lacerations or severe bleedings. The postoperative complications observed are shown in figure two (Figure 2). To minimize the risk of the most common postoperative problem, which is restenosis of the created airway, stents were placed transnasally in 23 of our patients. The stenting time varied between 1 and 12 weeks (Figure 3). Comparing the rates of restenosis our data clearly reveal, that the group of the 23 stented patients had a failure rate of 35% compared to only 11% in the 18 patients where no stents were used (Table 1). The latter were followed by regular postoperative endoscopic examinations and repeated dilations with a soft rubber bougie. Dilations were performed in increasing intervals, performed by the surgeon initially on a daily basis. Starting after three weeks the number of dilations was reduced step by step to once a week and then performed by parents or patients themselves in a further outpatient follow up for up to one year. In five of the most recent cases we applied Mitomycin C intraoperatively (0.4 mg / ml for 10 minutes) as an additional effort to prevent restenosis. None of these individuals received stents. All were followed for more than 12 months so far and none had to be revised surgically.

DISCUSSION

The present retrospective analysis of the treatment of congenital choanal atresia over a period of 26 years, including 41 patients, constitutes one of the few reports on such a large number of cases in the literature [7,14-17]. The female/male ratio of 2 : 1 in this malformation [2,6,7,18] is in accordance with the literature. The right side predominance within the group of unilateral atresia, which was seen in other series [1,5,19] was not present in our patients, as we found both sides were equally affected with 14 manifestations. Brown et al. [3] reported a distribution of bony versus mixed bony and membranous atretic plates of 29% versus 71%, which is similar to our series. The timing of surgical intervention in bilateral cases is dictated by

the life threatening nature of the situation [18]. However, there is some debate on the most suitable timing for correction of unilateral choanal atresia. As there are few symptoms with the most common complaint being mucoid nasal discharge, sometimes accompanied by upper airway infections, headache and rhinolalia, some authors have suggested surgical repair when children are between 5 and 8 years old [6,20]. Stamm in contrast has performed unilateral correction in infants at the age of 6 months [8]. The benefit of an open airway and restored ventilation and drainage of the nasal and paranasal cavities have to be weighed against the potential need for revision due to relative narrowing as the created opening will not increase in size with growth of the child. In our series, over time there was a tendency towards earlier correction before school age, preferably around the age of two years. The embryologic etiology of congential choanal atresia has been attributed to a failure of resorption of the buccopharyngeal membrane and / or persistence of the nasobuccal membrane respectively in the past. After an evaluation of 37 cases by Hengerer et al. in 1982 [7] these theories appeared to have only little validity in a majority of cases. Hengerer instead hypothesized a misdirection of mesodermal flow secondary to genetic or environmental factors. The well described association of congenital choanal atresia with other malformations is indeed confirmed in our series with both syndromal and isolated deformities as has been detailed above. Besides CHARGE, Francescetti and Fraley syndromes we saw cases of trisomia 4q and a partial trisomia 22. However, the most common trisomia associated with choanal atresia in the literature is trisomia 21, known as Down´s syndrome, which none of our patients was diagnosed with. The partial trisomia 22 was attributed to a balanced translocation 11/22 in the mother. Complete trisomia 22 is the second most common reason for miscarriages after trisomia 16 and is rarely present in living neonates. However, the partial form is known to go along with a variety of pathologic physical findings with a cleft palate being among the most frequent. The male infant of our series with trisomia 4q presented with a right-sided choanal atresia. This genetic disorder is also rarely diagnosed and goes along with craniofacial disorders, malformation of the external ears and a prominent nasal bridge. Myers et al. [21] pointed out the potential role of maternal drug use, specifically of carbimazole. In our series however, we did not identify any clear pharmacological or environmental pathogenetic cofactors.

The majority in our series had the typical “delta deformity” of the widened vomer [16], which along with the medialized lateral walls of the posterior nasal cavity results in an hourglass shaped configuration of the choanal region and the nasopharynx. However, there was a broad variety of findings that varied with different craniofacial deformities, potentially associated with skull base defects. We would therefore, as a consequence of this retrospective analysis, strongly encourage any surgeon to whenever possible and available, achieve multiplanar CT scans with three-dimensional reconstructions to ensure adequate preoperative orientation and tailor the intervention to the individual pathology.

Our series reflects a clear preference for the endonasal approach, which has been widely described in the literature [19,22-24]. The excellent visualization allows for atraumatic and time effective repair using powered, miniaturized and endoscopic instrumentation. The combined micro- endoscopic endonasal approach has evolved as the best approach in our hands. The stereoscopic vision and magnification along with bimanual surgical manipulation offer a valuable adjunct to the use of the endoscopes, which combine excellent visualization with angled vision [10,25]. Figure 4 displays a case of right-sided bony atresia before (a) and immediately after (b) micro-endoscopic correction (Figure 4). Another option, especially in combination with this approach, is the use of CO2 lasers, which has also been applied in choanal atresia surgery [2,26]. The transpalatine approach was used in three of our patients

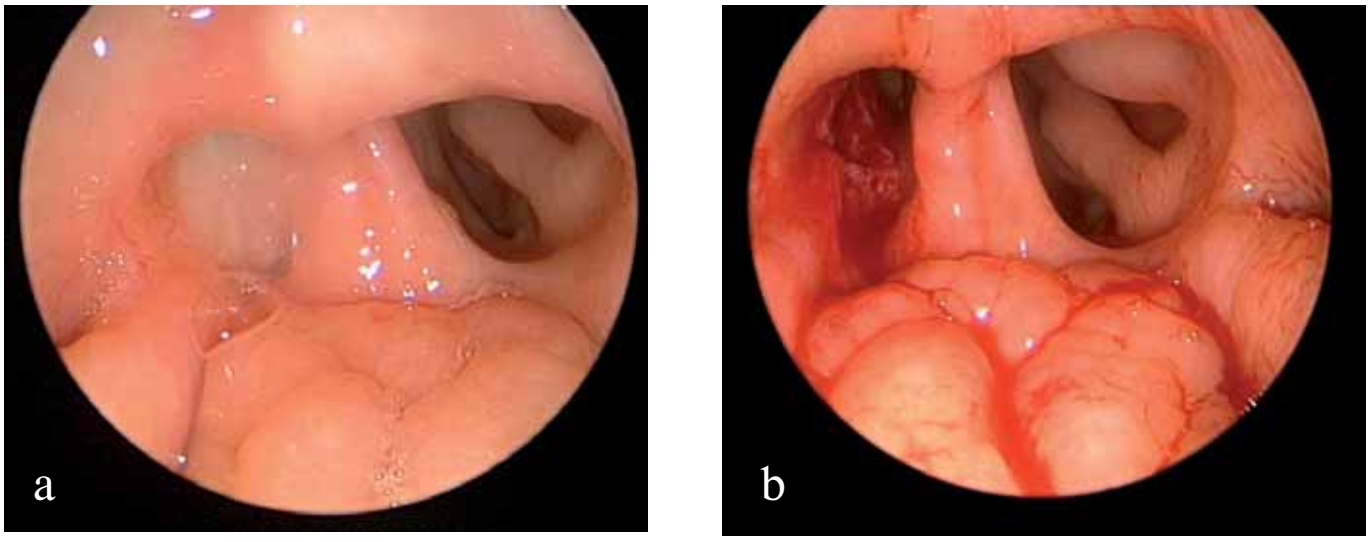

Figure 4. Endoscopic view of a combined bony-membranous atretic plate preoperative (a) and immediately postoperative (b).

and offers a good visualization in cases where the nasal passage is stenosed or obliterated to a degree that does not allow for endonasal instrumentation. However, a marked drawback of this procedure is the known long-term complication rate, resulting from palatal muscle dysfunction and consecutive mandibular joint pathology as well as orthodontic problems in 50% of the cases [1,18,19]. As a consequence, we would reserve this technique for cases of insufficient endonasal visualization in revisions or children older than 5 years.

Any outcome analysis of congenital choanal atresia centers on intra- and postoperative measures to ensure a stable result over time. Some authors propose mucoperiosteal flaps to be created around the created opening by asymmetric incisions or in a cross over technique in an effort to minimize or eliminate corresponding raw surfaces [8,11,12]. From the data of our series we cannot evaluate the effectiveness of these techniques. They have not become part of our routine due to rather inhomogeneous anatomical situations and the difficulty of respective adequate postoperative control in neonates and small children. A clear difference was seen between our patients with and without postoperative stents with an incidence of restenosis of 35% and 11% respectively. Difficulties related to stenting in repair of choanal atresia have been discussed in the literature mostly as a consequence of the circumferential pressure, leading to ischemia, osteoblastic and fibroblastic reactions and a lack of reepithelization in the choanal aperture as well as pressure related lesions of the collumella, alar cartilages and more posterior parts of the septal cartilage [6,12,22]. Granulation tissue has been associated with a 30% failure rate with the use of stents regardless of the surgical technique or approach [27]. As a consequence of our own experience and in accordance with the cited publications of other authors we have abandoned the use of stents.

As a more recent change in treatment modalities used in our series, we have applied topical Mitomycin C intraoperatively in the last 5 cases, combined with repeated postoperative dilations. After sufficient long term follow up of over at least 1 year, we did not have to revise any of these 5 patients. There is plenty of evidence from the literature on the efficacy of topical Mitomycin C in reducing scar formation and restenosis, especially in the aerodigesitve tract [28]. The cytostatic effects of this antibiotic substance and its inhibitory effects on fibroblast proliferation have, in accordance with our data, already been described as helpful in choanal atresia repair [13,29].

Taking all information gained from this series of 41 patients into account and relating it to the recent literature we are now using the following protocol for the treatment of congenital choanal atresia:

- Subsequent to initial airway management, bilateral choanal atresia is corrected within the first week of life. In cases of unilateral atresia surgical correction should be electively performed before school age, preferably around the age of 2 years.
- If the nasal airway is not severely compromised by concomitant malformations and deformities, the micro- endoscopic, endonasal approach is preferable and offers numerous technical options and variations.
- The atretic plate is to be removed with all compounds, based on preoperative analysis of the individual deformity and knowledge of potential concomitant malformations with the help of multiplanar high resolution CT scans and 3D reconstructions.
- Topical Mitomycin C is applied intraoperatively for 10 minutes in a dilution of 0.4 mg / ml to reduce excessive granulation and scar formation.
- In the postoperative management, repeated endoscopic controls are combined with transnasal dilations with a soft rubber bougie in increasing intervals, performed by the surgeon initially and by parents or patients in a further outpatient follow up for up to one year.
- No stents are applied into the nasal airways.

CONCLUSION

In conclusion, this thourough analysis of a series of patients presents a protocol for the treatment of congenital choanal atresia. The endonasal micro-endoscopic surgical approach is successful if combined with postoperative dilations for up to one year. Stenting should be abandoned due to immanent granulation formation frequently leading to restenosis. The intraoperative application of Mitomycin C offers a promising adjunct and may play a role in achieving stable long-term results.

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