J Maxillofac Oral Surg 8(1):52–54

CLINICAL STUDY

Management of choanal atresia in cases of craniofacial malformation

Martin Stieve[1] ✉ · **Kempf HG**[2] · **Lenarz Th**[1]

[1] Department of Otolaryngology, Medical University of Hannover, Germany
[2] Helios Klinik, Wuppertal, Germany

Received: 10 October 2007 / Accepted: 08 February 2009

Abstract
Objective To report the method and results of endonasal endoscopic approach in congenital choanal atresia in cases of craniofacial malformation.
Patients The pathology of unilateral and bilateral choanal atresia and the treatment results in seven children treated between 1999 and 2006 are presented. 5 infants suffering from bilateral atresia also had severe malformations (Charge syndrome [2 patients], trisomy 18, microcephalus, central cranioschisis, cleft lip and anophthalmia 9). The surgical intervention was carried out immediately after birth. 2 patients with unilateral atresia were treated in their second and sixth year of life respectively. CT scan was the diagnostic procedure of choice. Intraoperative endoscopy showed both membraneous and osseous atresia.
Surgical procedure Trocars of different sizes were used to open the atresia plate, while the osseous parts were removed with diamond drills. Silicone tubes were inserted transnasally and remained in place for several months to improve the infants'ability to breath and drink.
Results In all cases of bilateral atresia the tubes either had to be changed repeatedly or replaced with larger tubes due to dislocation and head growth. The septum was perforated in one case. No further stenoses were detected following the removal of the tubes (after 3–6 months).
Conclusion The transnasal access is particularly suited to newborns and infants because it induces a minor surgical trauma and carries a low risk of bleeding if endoscopes are used. The results show that the risk of restenoses can be minimised with sufficient fixating and in-patient care.

Keywords Choanal atresia · Craniofacial malformation · Surgical procedure

Address for correspondence:

Martin Stieve
Department of Otolaryngology
Medical University of Hannover
Carl-Neuberg-Str. 1
D-30625 Hannover
Germany
Ph: +49-511-532-6565
Fax: +49-511-532-5558
E-mail: sti@hno.mh-hannover.de

Introduction

Unilateral and bilateral choanal atresia are one of the most frequent congenital malformations of the skull base [1]. Unilateral atresia occurs more frequently than bilateral atresia. 20–50 % of the patients have severe congenital malformations such as Charge association or trisomy 18 [2–4].

The development of congenital choanal atresia has not been completely investigated. On the one hand it may be rooted in embryonal malformation in the area of the primitive posterior nasal meatus at the opening of the nasopharynx with persistence of the bucconasal membrane. On the other hand excessive growth of the vertical and horizontal process of the maxilla with septum fusion at the time of development of the obturator may be involved [1,5,6].

A combination of osseous and membraneous atresia is very common, whereas purely membraneous or purely osseous atresia occurs only very rarely [7,8]. Bilateral choanal atresia immediately after birth leads to a life-threatening condition due to dyspnea, aspiration and inability to drink. Most infants suffering from several malformations require extensive care, oxygen supply and gastric tubing [8,9].

Unilateral atresia may initially remain undetected and be manifested in rhinitis and sinusitis. Bilateral atresia, however, is diagnosed immediately postnatally on the basis of the clinical symptoms and by means of flexible endoscopy [10]. A coronary and axial computed tomography of the paranasal sinuses and the skull base is a prerequisite for surgery [11].

The surgical methods available allow a choice between three different approaches, i.e. the transnasal, transpalatinal and transmaxillary approach. This paper focuses on the results with the endoscopic approach. It has advantages of very little surgical trauma compared to other approaches. The long term results are satisfying and no further surgery is required.

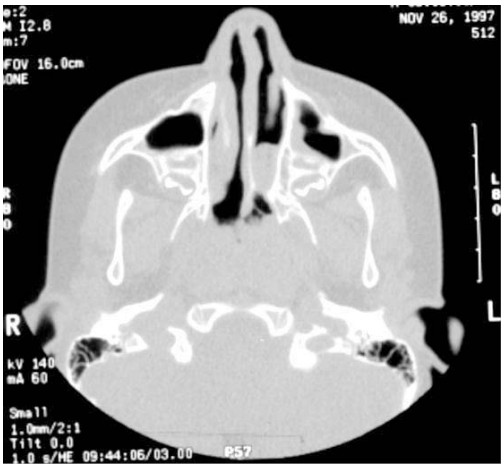

Fig. 1 Axial CT scan showing left unilateral choanal atresia

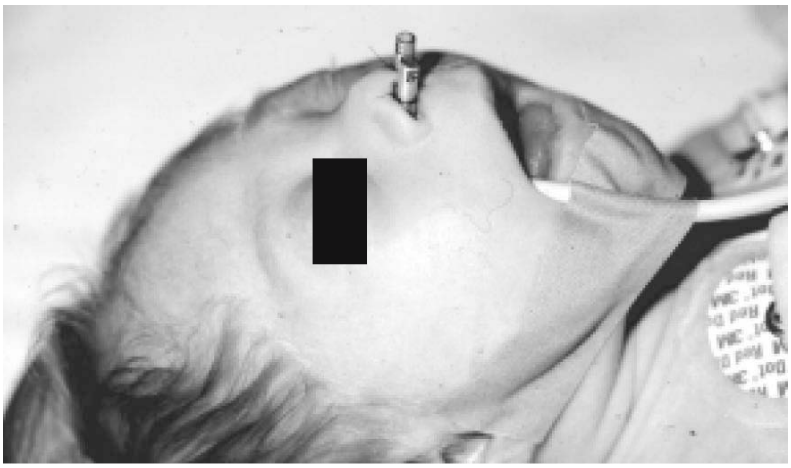

Fig. 2 Postoperative site

Patients

The group of patients consists of 7 children suffering from unilateral and bilateral choanal atresia, who were treated between 1999 and 2003. 5 infants had severe malformations (Charge syndrome [2 patients], trisomy 18, microencephalus, central cranioschisis, cleft lip and anophthalmia) in addition to bilateral atresia. Two patients suffering from unilateral atresia were treated in their fourth and eleventh year of life respectively (Table 1).

Results

The neonates (4 females, 1 male) suffering from bilateral stenosis were operated 1–2 days after birth. In one case surgery was not possible until 20 days after birth, due to pulmonary problems. The surgery in children with unilateral stenoses (2 males) was performed in their fourth and eleventh years due to later diagnosis and no life threatening symptoms.

The surgery was carried out after a coronary and axial CT scan of the paranasal sinuses and the skull base had been taken (Figs. 1 and 2). The intraoperative site showed predominantly mixed stenoses with membraneous and osseous parts. Initially, an endonasal and enoral endoscopy (30°) was performed. The membranous parts were incised with trocars of increasing sizes. The covering osseous parts were completely removed by means of diamond drills and bone punches. Silicone tubes with a diameter of 3–6 mm, which could be inserted and fixated at the anterior septum without complications, served as stents.

Table 1 Clinical data of infants with choanal atresia

Pat	Atresia	Other anomalies	Age of surgery (days)	Revision after first surgery	Outcome
1.	Bilateral	Charge Association: heart anomaly Facial paralysis coloboma	2	4 months	6 months
2.	Bilateral	Trisomie 18	1	4 months	6 months
3.	Bilateral	Central Cranioschisis Cleft Lip Anophthalmia	20	3 months	5 months
4.	Bilateral	Charge Association: heart anomaly Facial paralysis coloboma	1	6 months	12 months
5.	Bilateral	Microcephalus	2	4 weeks	3 months
6.	Unilateral	None	4 year of life		24 months
7.	Unilateral	None	11 year of life transpalatina 13 year of life transnasal		24 months

Postoperative breathing was considerably improved. As a result, food intake no longer posed a problem. The older children with unilateral stenoses had a good nasal air passage with the tubes in place. No dislocations occurred, and no revision surgeries were necessary. The postoperative observation period was 6 to 12 months. No restenosis occurred.

All bilateral stenoses required revision surgeries due to dislocation of the tubes. The tubes could be changed without complications under general anesthesia. A septum perforation occurred in one case. At time of replacement the choanal perforation were enlarged to adapt to head growth.

The postoperative observation period was 6–24 months. No restenosis occurred after removal of the tubes.

Discussion

As neonates must rely on intake of food and beverage on their nasal respiration due to the location of their larynx, which is elevated as compared to the adult larynx, bilateral choanal atresia is an immediate postoperative emergency [10,12,13]. Intubation or even tracheostomy may become necessary to ensure sufficient oxygen supply. The diagnosis of bilateral atresia is confirmed following pharyngeal suction through the nose immediately after birth. Flexible nasal endoscopy or transoral inspection of the nasopharynx confirms the diagnosis and excludes space occupying lesions of the nasopharynx [10].

Computed tomography in the axial and coronal plane of the paranasal sinuses and the skull base is a prerequisite for surgery. Immediate surgery is indicated with bilateral atresia. Transnasal, transmaxillary and transpalatinal access may be used.

The transmaxillary access should be avoided in infants as it may lead to damage of the tooth buds. Transnasal and transpalatinal access are commonly used. In the transpalatinal approach (first described by Brunk in 1909 [2]) the atresial plate is totally removed with a drill or bone punches via the hard palate while protecting the palate vessels. The perforation is subsequently closed with a nasopharyngeal mucous membrane flap [9].

In cases of unilateral choanal atresia Rudert [13] describes a method without using stents. After dissection he covers the wound with special flaps. The flaps are fixed with fibrin glue and with nasal tamponade for two days. In his series no restenosis was found.

The advantage of the transnasal access consists in the good endoscopical view of the surgical site. The risk of haemorrhage and the length of the surgery can be limited in this way. Electrosurgical and lasersurgical methods have been developed in the last few years to remove the atresial plates and have generally produced good results without restenosis [7,13]. Similar results were achieved with the procedure presented. Trokars, diamond drills and bone punches ensures a careful procedure. It is necessary to control the trocar position endoscopically during the surgical procedure to avoid damage to the skull base.

The dislocation of the silicone tube, however, is critical; revisions had to be carried out in cases of bilateral atresia. The reasons for dislocation are problems in fixation. In addition tubes have to be inspected to adapt to the growing head size of the newborns. No further stenoses occurred following removal of the tubes (after 3–12 months).

Conclusion

The transnasal access is particularly suited to newborns and infants because it induces a minor surgical trauma and carries a low risk of bleeding if endoscopes are used. The results show that the risk of restenoses can be minimised with sufficient fixating and in-patient care.

References

1. Belenky WM (1990) Nasal obstruktion and rhinorrhea. In: Bluestone CDE Stool (eds) Pediatric Otolaryngology, 2nd edition. Philadelphia. London: W.B. Saunders Company
2. Brunk A (1909) Ein neuer Fall von einseitigem knöchernem Choanalverschluß. Operationsversuch vom Gaumen. Z. Ohrenheilkunde 59: 221
3. Deitmer T (1997) Ätiologisch unklare Nasenstenosen bei Neugeborenen. Laryng Rhinol Otol 73: 591–594
4. Grevers G, Vogl T (1988) Computertomographische Darstellung der Choanalatresie. Laryng Rhinol Otol 67: 23–24
5. Hengerer AS, Strome M (1982) Choanal atresia: A new embryologic theory and its influence on surgical management. Laryngoscope 92 (8 Pt 1): 913–921
6. Josephson GD, Vickery CL, Giles WC, Gross CW (1998) Transnasal endoscopic repair of congenital choanal atresia: long term results. Arch. Otlaryngol Head and Neck Surg 124(5): 537–540
7. Jung H (1994) Die angeborene Choanalatresie und deren operative Korrektur. Laryng Rhinol Otol 73: 586–590
8. Legler U (1974) Mißbildungen der Nase (mit Ausnahme der Gaumenspalten), Fremdkörper, Nasenbluten. In Berendes, Link JR, Zöllner F (eds): Hals-Nasen-Ohrenheilkunde in Praxis und Klinik. Stuttgart: Thieme
9. Panwar SS, Martin FW (1996) Transnasal endoscopic holmium: YAG laser correction of choanal atresia J Laryngol Otol 110(5): 429–431
10. Peters H, Pontz BF (1988) Klinisches Bild und Verlauf von Kindern mit Charge-Association. Monatsschr. Leinderheilkd. 136: 690–693
11. Lazar RH, Younis RT (1995) Transnasal repair of choanal atresia using telescopes. Arch Otolaryngol Head Neck Surg 121(5): 517–520
12. Robert J (1977) Shprintzen Genetics, syndromes, and communication disorders, London: Singular Publishing
13. Rudert H (1999) Kombiniert transseptale-transnasale Chirurgie einseitiger Choanalatresien ohne Verwendung von Platzhaltern. Laryngo-Rhino-Otol 78: 697–702

Source of Support: Nil, **Conflict of interest:** None declared.