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Choanal atresia in premature dizygotic twins - a transnasal approach with Holmium: YAG-laser*

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SUMMARY

Twins born prematurely presented with choanal atresia and were successfully treated using a transnasal, endoscopically and microscopically controlled Ho:YAG-Laser assisted technique. One twin, who had bilateral choanal atresia was operated immediately, the other one, with unilateral choanal atresia, received surgery 8 months after birth. The rare feature of twins both suffering from choanal atresia and a technique for definitive treatment of this disease in premature neonates are presented and discussed. This report of dizygotic twins with nonsyndromal choanal atresia suggests the possibility of an autosomal recessive inheritance with various penetration or an undefined teratogenic etiologic factor.

Key words: CHARGE association, choanal atresia, congenital anomalies, laser surgery, twins

INTRODUCTION

Bilateral choanal atresia is the most common cause of complete nasal obstruction in the neonate (Sprecher, 1997). The average prevalence of the anomaly is 0.8/10,000 births. Half of these cases are bilateral; unilateral cases occur equally often on the right and left side (Harris et al., 1997). Brown et al., (1996) reviewed 63 computed tomography scans of choanal atresia and found a combined incidence of 29% pure bony anomalies and 71% mixed bony-membranous anomalies and no pure membranous atresias. They suggest that the proper anatomic classification of choanal atresia should be bony or mixed bony-membranous, which should replace the former system of Fraser (1910). He estimated a 10% incidence for pure membranous atresias based upon a review of the literature. At that time, possibilities to assess the true nature of the anatomy accurately were limited. Now, CT-scan and nasal endoscopy are available. The newborn mandatorily breathes through the nose. Neonates with bilateral choanal atresia suffer from an acute respiratory distress syndrome and have feeding difficulties because of sucking impairment. Unilateral anomalies may remain unnoticed at birth. A catheter cannot be passed through the nose into the nasopharynx. The diagnosis is confirmed by CT-scan and endoscopy.

We report a case of premature twins with choanal atresia who were treated successfully using a Ho:YAG-Laser assisted technique.

CASE REPORT

History:

The fraternal, dizygotic twins were delivered December 26, 1997 by Caesarian section at 34 weeks gestation due to premature onset of labor. The mother, a 24-year-old primipara suffered from gestational diabetes and from preeclampsia after the 33rd week of pregnancy. She was treated with insulin and with nifedipine at the end of her pregnancy because of hypertension. The ultrasonographic examination showed a hydramnion.

Twin A:

S.S. weighed 1970 grams (P 10-50) and had APGAR scores of 3/5/8. The blood group was A, Rh-positive. The physical exam revealed a preterm newborn corresponding to its gestational age, with a transient respiratory distress that responded well to oxygen. The choana could be passed with a catheter on the right but not on the left side. The CT-scan revealed a unilateral mixed bony-membranous choanal atresia. No further malformations were found. Spontaneous breathing was undisturbed, and there were no nursing problems. Six months after birth, putrid rhinitis began on the left side, and it did not respond to conservative treatment with antibiotics and nasal lavage using saline water. Therefore, the choanal atresia was operated at the age of 8 months using the technique described.

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Twin B:
A.S. was a 1520-gram (P 10) white male with APGAR scores of 7/4/5. The blood group was O, Rh-positive. Immediately after birth, he developed an acute respiratory distress syndrome. Attempts to pass a nasal catheter were unsuccessful on both sides, hence bilateral choanal atresia was suspected. The infant required oral intubation and was referred to the neonatal intensive care unit. A CT-scan confirmed a bilateral mixed bony-membranous choanal atresia (Figure 1). Echocardiography revealed an asymptomatic pulmonary artery stenosis (gradient: 20 mmHg) and a patent foramen ovale; otherwise, no other congenital abnormalities were found. Five days after birth, the choanal atresias on both sides were opened according to the technique described.

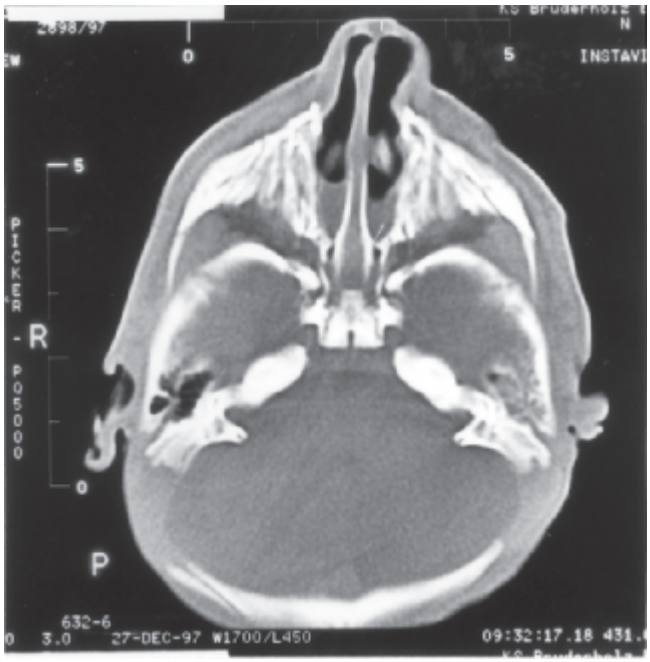

Figure 1. Axial CT-scan section of the bilateral mixed bony membranous choanal atresia.

Surgical technique:
The surgery is carried out under general anesthesia and oral, endotracheal intubation. The nose is decongested with xylometazoline on cotton pledgets. Endonasal inspection is performed using a rigid endoscope (Karl Storz®) with a 2.7 mm external diameter and a 30° deflection lens; epipharyngeal inspection is provided by a 4.0 mm rigid endoscope (Karl Storz®) with a 70° deflection lens. An aural speculum is placed in the nostril, and the atretic choana is viewed with the operating microscope (Wild Leitz®). The atresia is vaporized along the floor and the posterior nasal septum using the flexible, fiber-optic transmitted Ho:YAG laser (Coherent®; wave length: 2.1 μm, power: 6 Watt, pulse-mode: 6Hz, diameter of fiberoptic delivery system: 600 μm). The perforation is gradually enlarged with the laser and an otologic drill (Bien Air®, diamond drill diameter: 3.5 mm) until the neochoana has a diameter of about 4 mm. The operation at this stage is best viewed endoscopically from the postnasal space. The same procedure is performed on the other side, if required.

Figure 2. The stent is fashioned from a Portex® endotracheal tube.

A Portex® endotracheal tube (diameter: 3.0 mm) is used to fabricate the stent. An oval-shaped window is cut into the tube at a site equidistant from both ends and the tubing is folded upon itself (Figure 2). This window serves as the common posterior opening and is placed at the posterior edge of the septum. Anteriorly, a small piece of a Portex® endotracheal tube is sewed between both outer ends of the tube to prevent compression of the columella and to keep the stent in place. The same kind of stent is used in unilateral choanal atresia. Postoperatively, regular suction is carried out, following use of normal saline drops on a regular basis.

RESULTS
Twin A:
The stent was removed three days postoperatively under general anesthesia. It had to be replaced because of obstruction of the neochoana with mucous. This stent was removed 10 weeks later and patency has remained for a follow-up of 15 months.

Twin B:
The stent was removed six weeks after the operation under general anesthesia but had to be replaced because of obstruction of the otherwise patent neochoanae with thick mucous (Figure 3). This stent (Portex® endotracheal tube 3.5 mm) was

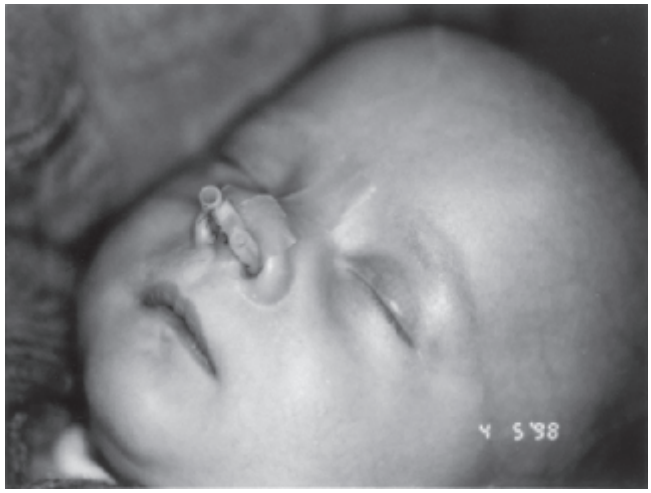

Figure 3. Twin B (18 weeks post partem) with the stent in both nostrils.

removed after twelve weeks. Both choanae have remained patent for a follow-up of 22 months.

DISCUSSION

Choanal atresia is a rare disease, whether identified as a single malformation in an otherwise normal individual or as a part of well-established syndromes such as Treacher Collins, Crouzon and Apert. Additional malformations are present in 47% of the infants without chromosomal anomalies. A weak non-random association can be demonstrated with the malformation entering the so-called CHARGE association (Coloboma, Heart Disease, Atresia of choanae, Retarded mental development and growth, Genital hypoplasia, Ear anomalies and deafness) (Harris et al., 1997).

There are only a few reports in the literature concerning twins presenting with choanal atresia not associated with the CHARGE complex. Umlauf described male, monozygotic twins with bilateral choanal atresia; Dickson reported female, monozygotic twins suffering from bilateral choanal atresia (in Dirlewanger, 1966; in Fendel, 1966). In their study, based on more than 5 million births, Harris et al. (1997) identified 444 infants with choanal atresia. There were 13 sets of twins and 2 sets of triplets. There was only one twin pair concordant for choanal atresia; both had associated malformations and were classified as CHARGE association. Two infants with choanal atresia belonged to a set of triplets and were of different sex. We believe that ours is the first published case report of dizygotic twins affected by nonsyndromal choanal atresia.

Nonsyndromal choanal atresia is usually sporadic and probably multifactorially determined. However, a few familial cases suggestive of both autosomal dominant and autosomal recessive modes of inheritance have been reported (Phelps, 1926; McGovern, 1950; Ransome, 1964; Fendel, 1966; Dirlewanger, 1966; Quazi et al., 1982; Gershoni-Baruch, 1992). Our report of dizygotic twins with phenotypically normal parents lends credence to the possibility of an autosomal recessive inheritance with various penetration or an undefined teratogenic etiologic factor. The autosomal recessive mode is often related to consanguinity (Fendel, 1966; Dirlewanger, 1966; Quazi et al., 1982; Gershoni-Baruch, 1992). However, the pedigree of the family presented here is inconspicuous. The relatives have not been accessible for examination, but none has a reported history of any nasal symptoms. Although there is no specific teratogenic agent identified, choanal anomalies have been reported in thalidomide embryopathy (Grahne and Kaltiokallio, 1966) and with maternal exposure to methimazole treatment for hyperthyroidism during pregnancy (Greenberg, 1987). There are CHARGE-like malformations described in infants of diabetic mothers (Brown, 1991).

There has existed controversy in the past concerning the preferred approach for treatment. In a survey of 94 articles about choanal atresia by Pirsig (1986), an equal failure rate for both approaches was reported. This result is supported by Black et al. (1985), who analyzed 48 cases of bony choanal atresias operated in the newborn period. Transpalatal (n=21) and transnasal (n=27) approaches yielded initial success rates (defined as the patency of a nasal airway for at least 6 months) of 57% and 59%, respectively. Thus, with regard to airway patency, both approaches seem to be equally effective. However, stunting of palatal growth and resultant orthodontic problems are major concerns of transpalatal surgery (Freng, 1978).

The microscopically and endoscopically controlled transnasal approach reported in this study allowed an excellent visualization of the operative field. The small diameter of the fiber-optic delivered Ho:YAG laser beam was a great advantage while working in the atretic area and was especially helpful during the opening of the choanal atresia. In contrast, the shaft of the otologic drill almost completely obstructed the view through the aural speculum. The Ho:YAG laser allowed precise vaporization of the membranous and bony parts of the atresia and caused minimal injury to the adjacent tissues. From this limited experience, it appears that this procedure has advantages over other conventional operations and deserves consideration as a modality of management for the difficult condition of choanal atresia of the premature neonate.

Regarding the duration of the stent placement, the consensus is that it should remain in place for 6 to 12 weeks (Pirsig, 1986). Illum (1986) and Panwar (1996) did not use stents after their laser-assisted corrections of choanal atresia, and they did not find an increased restenosis rate. However, they did not treat neonates; their patients had either unilateral choanal atresia or were undergoing revision surgery. We did not observe restenosis, but both twins presented with mucous-obstructed neochoanae after a few days without stenting. It is possible that the infants lungs were not mature enough to allow sufficient airflow to keep the neochoana free of secretion. We agree with Pirsig (1986) that maintenance of the stent seems necessary for at least 4 months if surgery is performed in premature and neonatal patients.

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