Transnasal Endoscopic Correction of Choanal Atresia

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Nasal Embryology

By the 3rd week of gestation, the nasal placodes appear. These placodes invaginate and extend posteriorly by the 5th week and by the 6th week the oronasal membrane ruptures to form the posterior choanae. Congenital Choanal Atresia (CCA) is due to the persistence of the bucconasal membrane. This persistence or failure to rupture may be due to factors acting on the misdirection of mucodermal flow resulting in the malrotation of burrowing nasal pits.

Epidemiology

CCA was first reported by Roederer in 1755 and the incidence appears to be 1 in 5000-8000 live births. The gender ratio is 2F: 1M. 50% of CCA's are associated with congenital anomalies. Whilst 65%-75% of cases are unilateral, 75% of the bilateral cases have anomalies. These anomalies include polydactly, Craniosynostosis and cleft palate. The best known one is the CHARGE association (**Table 1**) first described by Pagon in 1981. Since 1998, it has been renamed the CHARGE Syndrome.

Table 1 : CHARGE Association

Coloboma
Heart Abnormalities
Atresia of the Choanae
Retardation of Growth / Mental
Genitourinary defects
Ear Abnormalities

Diagnosis

A quick diagnosis is usually made in the nursery by the inability to pass suction catheters via the nostrils for unilateral cases. A flexible or rigid endoscopy usually confirm this. The 'gold' standard is the axial CT scans. It is important for pre-operative diagnosis and for discussing the surgical options with parents. The CT scan defines the thickness of the atretic plate, stenosis of the bony nasal cavity and its relationship to existing structures. 3D reconstruction is also possible. With the routine use of endoscopes and review of CT scans, we now know that unlike previously believed, the majority of CCA are mixed bony and membranous in nature (70%) and about 30% are truly bony alone.

Clinically, bilateral cases present with cyclic cyanosis that is relieved by crying. Unrecognised, these may lead to asphyxia and death. Unilateral cases may present later in life, is rarely emergent but parents may complain of tenacious secretions that never seen to clear up.

Management

Immediate management for bilateral CCA's is the use of an oral airway. 4 methods of surgical corrections have been generally described.

1. Transpalatal
2. Transeptal
3. Transantral
4. Transnasal (endoscopic, laser)

The author's method of choice is the transnasal technique, especially for very young patients. The advantages are:

a. Shorter operative time
b. Low blood loss
c. Preservation of the developing hard palate
d. With the use of powered instruments and videoendoscopic control, the results are comparable, if not superior, to the transpalatal technique.

The author first soaks the nasal cavity with neurosurgical patties impregnated with a combination of 1ml of 1:1000 adrenaline, 1ml of 10% cocaine, diluted with 3mls of normal saline. No more than 2mls is used in infants. The mucosa over the atretic plates are also injected with a 1:200,000 adrenaline / 1% xylocaine mixture. The infants mouth is kept open with a Dinghman's apparatus that is used by the plastic surgeons for cleft palate surgery. A suture is placed on the uvula and this is used to lift the soft palate up so that an assistant can insert a 120^0 Hopkin's Rod Telescope to give an excellent view of the choanae via the nasopharynx.

As the atretic plates are usually mixed, they are first perforated using Hegar dilators (used by gynaecologists to dilate the cervix)

A 2.9mm diamond burr is then used to break through the rest of the atretic plate, bone removed laterally and superiorly being done carefully with the view provided by the retropalatal view. The posterior end of the vomer can be removed using a backbiter. All soft tissue and mucosal tags are removed using a microdebrider. This will ensure that minimal granulation tissue forms when the cavity is reviewed in a months time under general anaesthesia and decrease the risk of restenosis. With adequate removal of bone and soft tissue, no postoperative stents are necessary. At review in 4 weeks, any granulation tissue present is removed with a microdebrider and Mitomycin C applied on the area of removal. Mitomycin C inhibits DNA and protein synthesis and decreases fibroblastic activity. The usual dose is 2 mg/ml applied for about 2 minutes and is prepared by the pharmacist half an hour before use.

With the use of power instrumentation and Mitomycin C and without indwelling stents, the author's last 7 cases did not restenose after a 2-5 years follow up.Unilateral cases present at birth are dealt with in a similar way when the child is about 1 year old as the nasal cavities are larger.

Recommended readings

1. G.D. Josephson, C.L. Vickery, W.C. Giles and C.W. Gross, Transnasal endoscopic repair of congenital choanal atresia: long term results, Arch. Otolaryngol. Head Neck Surg. 124 (1988), pp. 537-540.
2. T. Van Den Abbeele, M. Fracois and P. Narcy, Transnasal endoscopic treatment of choanal atresia without prolonged stenting, Arch. Otolaryngol. Head Neck Surg. 128 (2002), pp. 936-941.
3. W. H. Bradford and W.F. McGuirt Jr., Surgical management of choanal atresia: Improved outcome using mitomycin, Arch. Otolaryngol. Head Neck Surg. 127 (2001), pp. 1375-1381