Choanal atresia

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Authors

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Abstract: Choana is also known as posterior nasal aperture. Nasal airway continues with that of posterior nares. Air from nasal cavity finds its way into the lungs via the choanal apertures. In some children the choana may be congenitally closed. This causes either total (bilateral choanal atresia) or partial (unilateral choanal atreisa) nasal obstruction. Child being obligate nasal breathers, find it rather difficult to breathe when there is bilateral choanal atresia. This is more so during the first 6 weeks of life. Hence bilateral choanal atresia should be considered as an emergency in paediatric age group. This article attempts to discuss the etiopathology and management of this condition.

Introduction:

Choanal atresia is actually a developmental failure of the nasal cavity to communicate with nasopharynx.. This condition is rather rare occurring in about 1 in 10000 live births. This condition is more common in female children the ratio being 2:1 [1]. About 50% of of these patients have other associated congenital anamolies. The most commonly associated congenital anomaly is CHARGE Syndrome.

CHARGE Syndrome include [2]:

C- Coloboma

H- Heart disease

A- Choanal atresia

R- Mental and growth retardation

G- Genital hypoplasia

E- Ear deformities

Other anomalies associated with choanal atresia include [3]:

1. Polydactyly

2. Nasal / auricular deformities

3. Palatal deformities

4. Down’s syndrome

5. DiGeorge syndrome

6. Meningocele

7. Menigoencephalocele

8. Treacher Collin's syndrome

9.Mid face hypoplasia

Types of choanal atresia:

1. Unilateral / Bilateral : Unilateral choanal atresia is an incidental finding and is verycommon (about 70%) [4]. High degree of suspicion is necessary to identify this condition. An

infant with unilateral choanal atresia will have problems suckling milk from the breast opposite to the side of atresia.

2. Bony atresia – 90%, Membranous atresia – 6%, Combined atreisa 4% [5].

Anatomic deformitites associated with choanal atresia include [6]:

1. The bony atretic plate is situated infront of the posterior bony septum

2. Nasal cavity is narrow in these patients

3. Lateral pterygoid plates are found to be thickened compromising the nasal airway

4. Medially vomer is thickened

5. Inbetween lateral pterygoid and vomer is the membranous plate

6. High arched palate is common in these patients

History of choanal atresia:

This condition was first described by Johann George Roederer in 1755. This was later termed as an anatomical abnormality of palatine bone by Adolf Otto in 1854 [7].

Carl Emmert has been successfully credited with the first choanal atresia repair in 1854 [7].

Embryology of choanal atresia:

Development of face and cranial structures occur during the first 12 weeks of gestation. The development of choanae takes place between the 4th and 11th weeks of gestation [8]. Cranial

structures develop from neural crest cell migration. Development of nose begins during the 4th week of gestation. This is indicated by the formation of nasal pits. During the 5th week of gestation the nasal pits begin to fold inwards intothe mesenchyme forming nasal sacs. These primitive nasal sacs are separated from oral cavity by oronasal membranes. During the 8th week of gestation this oronasal membrane ruptures creating nasal cavity and a primitive choana located at the junction of nasal cavities and nasopharynx. During this phase of development there is gradual proliferation of neural crest cells. These cells contribute to the formation of skull base and nasal vaults. During the 10th week of gestation the nasal septum and developing palate fuse. The

primitive choanae gets pushed posteriorly. This choanae which forms during the 10th week of gestation is known as "Secondary choanae". In normal foetus these secondary choanae are patent

for a functioning airway between the anterior nasal cavity and nasopharynx [9].

Theories of development of choanal atresia [10]:

Four theories for the development of choanal atresia:

1. Persistence of a buccopharyngeal membrane from the foregut.

2. Persistence of the nasobuccal membrane of Hochstetter – most commonly accepted theory.

3. The abnormal persistence or location of mesodermal adhesions in the choanal region.

4. A misdirection of mesodermal flow secondary to local genetic factors better explains the popular theory of persistent nasobuccal membrane

Other theories that are not so widely accepted include:

1. Resorption of the floor of secondary nasal fossa

2. Incomplete dorsal extension of nasal cavity

3. Migration of dorsal part of fronto nasal process to fuse with the palatal shelves

Studies have revealed that cranio facial anamolies with mesenchymal damage and cell disruption were found in mothers who ingested high doses of vitamin A during their period of pregnancy.

This has been attributed to disturbances in migration pattern of neural crest cells, which is also followed by disturbances in mesoderm development in the cranio facial area [11].

In patients with choanal atresia the atretic plate has the following boundaries:

Superior – under surface of body of sphenoid

Lateral – medial pterygoid lamina

Medial – vomer

Inferior – horizontal plate of palatine bone

This anatomical knowledge of atrophic plate is highly valuable while performing surgery on these patients.

Clinical features:

Bilateral choanal atresia is considered to be a neonatal emergency. These infants present with asphyxia neonatorum. Bilateral choanal atresia is commonly associated with:

1. Nursing difficulties – Sucking difficulties

2. Respiratory distress – Cyclic. When the infant falls asleep it becomes breathless as the nose is blocked

3. Respiratory infections – can occur due to aspiration

4. Recurrent nasal allergies

5. Cyanosis which gets better when the child cries.

6. Cry is not normal (Rhinolalia clausa)

7. Bilateral choanal atresia is also commonly associated with other birth defects like orofacial defects, cardiac defects and limb defects [12].

Teratogenic syndromes causing bilateral choanal atresia include:

1. Methimazole embryopathy [13]

2. Carbimazole embryopathy [14]

Detailed drug intake history is a must in diagnosing embryopathies.

Unilateral choanal atresia is commonly missed. These infants find difficulty in sucking milk frombreast opposite to the side of choanal atresia. These children have unilateral nasal obstruction with nasal discharge. A strong degree of suspicion is a must to identify this condition.

Tests to identify choanal atresia:

1. Attempting to pass 6-8 sized French plastic catheter through the nose. If there is no atresia the catheter will effortlessly pass through the nasal cavity into the nasopharynx. If there is choanal atresia then a typical solid feeling would be encountered at about 3-3.5 cms from the alar rim. If obstruction is encountered within 1-2 cms from the anterior nares, then it could be caused due to traumatic deflection of nasal septum due to trauma.

2. When a wisp of cotton is placed closed to the nasal opening then it would move in the presence of air flow.

3. When methylene dye is instilled in to the anterior nasal cavity it can be seen passing through the nasopharynx. Obstruction due to choanal atresia will prevent flow of methylene dye into the nasopharynx.

CT scan imaging is virtually diagnostic:

It has the unique advantage of differentiating membranous choanal atresia from bony ones. In patients with combined atresia it will also reveal the contribution of these two elements to the atretic plate. Actual structures involved in the atretic plate would be clearly seen. In all these patients

vomer appears to be thickened, the lateral nasal wall bends medially to fuse with vomer thereby obstructing the nasal cavity.

Management:

In bilateral choanal atresia securing the air way takes the top priority. An oral airway can be introduced to tide over the immediate crisis.

Role of intraoral nipple:

A nipple with a large opening (McGovern) Nipple [15]can be introduced into the oral cavity of the infant to tide over the crisis. This provides adequate airway to the infant. A small infant feeding tube can be passed through another small opening present in the nipple or alongside the nipple for

gavage feeding. This helps to buy time till the child has gained adequate weight to withstand corrective surgery.

Tracheostomy:

This should be considered only on rarest of rare occasions when the patient is not able to adequately maintain oral airway.

Timing of repair in unilateral choanal atresia:

Choanal atresia repair in unilateral atresia is delayed till the child reaches its first birthday. This allows

the surgery site to enlarge thereby reducing the risk of post op stenosis. Bleeding is also reduced if surgery is delayed. Older infants tolerate stenting better than young ones.

Trans nasal approach:

Transnasal approach: (using endoscopes): The surgery is performed under general anesthesia. A self retaining nasal speculum is used to expose the nasal cavity and the atretic plate. If the atresia is membranous in nature a simple perforation of the same under endoscopic guidance would suffice.

The nasal cavity is decongested using 4% xylocaine with adrenaline in the concentration of 1 in 10,000 concentration. Under endoscopic guidance a mucosal incision is made and the mucosal flaps are elevated exposing the posterior vomer and lateral pterygoid lamina. A diamond burr on an angled hand piece is used to drill the atretic bony plate. It is perforated at the junction of the hard palate and the vomer. Incidentally this is the thinnest part of the atretic plate. This procedure was first described by Stankiewicz. To improve visualisation the inferior turbinate can be out fractured

or even be trimmed. After drilling care is taken to preserve the mucosal flaps. A silastic stent is placed into each nostril passing through the drilled neo choana. This helps in reducing the incidence of restenosis. Stent is kept in place for atleast 6 weeks 19.

Opening made should be large enough to allow smooth passage of suction catheter. 3-4 size Endotracheal tube can be used as stent to prevent restenosis. The size should be choosen carefully in such a way that it should be adequate to prevent restensosis and inadequate to cause nasal

regurgitation.

Caution:

While performing this procedure caution must be taken not to injure the sphenopalatine vessels behind the middle turbinate.

Advantages of this procedure:

1. This process is faster and easier

2. Blood loss is minimal

3. Can be performed in children of all ages who do not have associated external nasal deformities

4. Child can be immediately breast fed

5. Child can be discharged on the 3rd day itself

Disadvantages:

1. Vision is highly limited especially in the new born

2.Inability to adequately remove enough of the posterior vomerine septal bone and prevent restenosis

3. Longer stenting time

4. Endoscopes do not offer binocular vision

5. Cannot be done safely and with good results on patients with multiple nasal and nasopharyngeal

anomalies.

First transnasal repair of choanal atresia was performed by Dehaen[18] in 1985. He used microscope

for magnification and visulization of the atretic plate.

Endoscopic transnasal approach[16] is facilitated by advances in instrumentation, anesthesia, imaging etc. CT imaging reveals the amount of contribution to the stenotic segment by the lateral nasal wall. It is prudent to avoid drilling too much into the lateral nasal wall as it could damage the

sphenopalatine vessels[17]. Use of powered instrumentation like soft tissue shavers and drills have made this procedure a lot safer.

Use of Mitomycin C[19] to prevent restenosis:

In addition to routine stenting restenosis can be prevented by topical application of Mitomycin -C, which is an antimetabolite known to inhibit fibroblast formation.

Complications of use of stents following transnasal repair of choanal atresia:

1. Formation of granulation tissue

2. Crust formation

3. Septal perforation

4. Persistent nasal discharge

Transpalatal approach:

This approach is more suitable for bilateral choanal atresia. Under general anesthesia the palate is exposed with a mouth gag. Palatal mucosa is infiltrated with 2% xylocaine with 1 in 100000 adrenaline. This infiltration serves the dual purpose of helping in flap elevation and providing much needed hemostasis during the entire surgical procedure. A curved incision is given on the palate starting from just behind the maxillary tuberosity on one side and is carried medially along

the alveolar ridge up to the canine region. The same incision is carried out even on the opposite side. A “U” shaped palatal flap is elevated. This flap is elevated up to the edge of the hard palate.

The greater palatine neurovascular bundle is preserved at all costs. The soft palate is now retracted posteriorly and superiorly exposing the posterior edge of hard palate. This area is the area for dissection. The posterior edge of hard palate is taken down using kerrison’s punch or drill. The

nasal mucosa is exposed. This mucosal flap is lifted posteriorly till the choana is reached. The posterior portion of nasal septum and lateral superior nasal wall is also taken down. A stent is placed. The mucosal flap is then redraped in position.

Complications of transeptal approach:

1. Pressure necrosis of columella

2. Plugging of stent

3. Displacement of stent

4. Palatal dehiscence

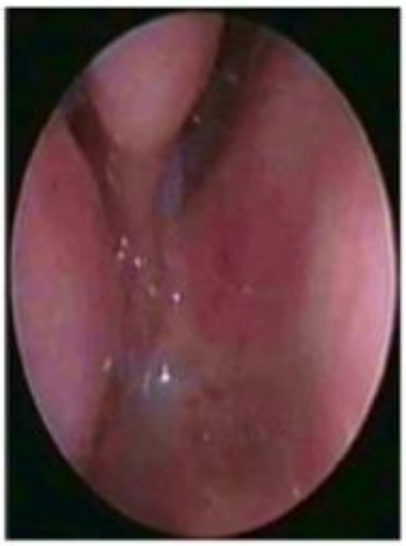
5. Maxillary hypoplasia causing malocclusion

6. Granulation tissue formation around the stents.

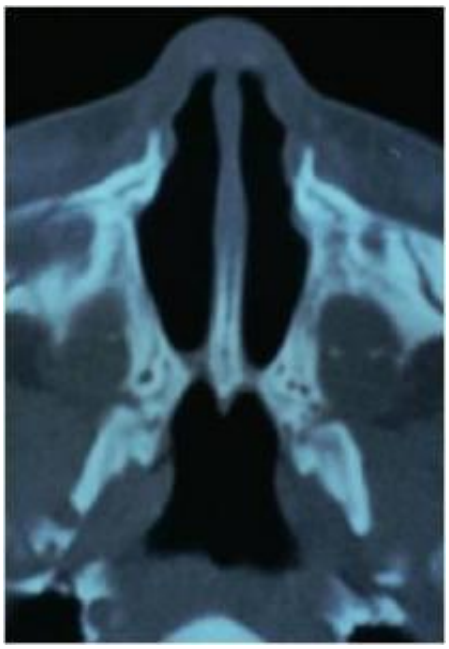
Conclusion:

The main aim of choanal atresia repair is maintenance of nasal airway at all costs.

Success of choanal atresia surgery is determined by the necessity of post op dilatation of the chonal orifice or revision surgery on the same patient due to dense re stenosis.



Endoscopic view of choanal atresia



CT scan showing bilateral choanal atresia

References

1. Theogaraj SD, Hoehn JG, Hagan KF. Practical management of congenital choanal atresia. Plast Reconstr Surg 1983; 72: 634-42.

2. Bergstrom L, Owens O. Posterior choanal atresia: a syndromal disorder. Laryngoscope 1984; 94: 1273-6.

3. Harris J, Robert E, Kallen B. Epidemiology of choanal atresia with special reference to the CHARGE association. Pediatrics 1997; 99: 363-7.

4. Dobrowski JM, Grundfast KM, Rosenbaum KN, Zajtchuk JT. Otorhinolaryngic manifestations of CHARGE association. Otolaryngol Head Neck Surg 1985; 93: 798-803.

5. Brown OE, Pownell P, Manning SC. Choanal atresia: a new anatomic classification and clinical management applications. Laryngoscope 1996; 106: 97-101.

6. Harner SG, McDonald TJ, Reese DF. The anatomy of congenital choanal atresia. Otolaryngol Head Neck Surg 1981; 89: 7-9.

7. Hengerer AS, Brickman TM, Jeyakumar A. Choanal atresia: embryologic analysis and evolution of treatment, a 30-year experience. Laryngoscope. May 2008;118(5):862-866.

8. Dunham ME, Miller RP. Bilateral choanal atresia associated with malformation of the anterior skull base: embryogenesis and clinical implications. Ann Otol Rhinol Laryngol. Nov 1992;101(11):916-919.

9. Johnston MC. The neural crest in abnormalities of the face and brain. Birth Defects Orig Artic Ser. 1975;11(7):1-18.

10. http://www.drtbalu.co.in/choa_atre.html

11. Mossey PA, Little J, Munger RG, Dixon MJ, Shaw WC. Cleft lip and palate. Lancet. Nov 21 2009;374(9703):1773-1785.

12. Burrow TA, Saal HM, de Alarcon A, Martin LJ, Cotton RT, Hopkin RJ. Characterization of congenital anomalies in individuals with choanal atresia. Arch Otolaryngol Head Neck Surg. Jun 2009;135(6):543-547

13. Clementi M, Di Gianantonio E, Pelo E, Mammi I, Basile RT, Tenconi R. Methimazole embryopathy: delineation of the phenotype. Am J Med Genet. Mar 5 1999;83(1):43-46.

14. Wolf D, Foulds N, Daya H. Antenatal carbimazole and choanal atresia: a new embryopathy. Arch Otolaryngol Head Neck Surg. Sep 2006;132(9):1009-1011.

15. McGovern, FH. Association of congenital choanal atresia and congenital heart disease. Reprot of two cases. Ann. Otol.Rhin and laryng 1953; 62:894-895

16. McGovern, FH. Association of congenital choanal atresia and congenital heart disease. Reprot of two cases. Ann. Otol.Rhin and laryng 1953; 62:894-895

17. Pasquini E, Sciarretta V, Saggese D, Cantaroni C, Macri G, Farneti G. Endoscopic treatment of congenital choanal atresia. Int J Pediatr Otorhinolaryngol 2003; 67: 271-6.

18. Dehaen F, Clement PA (1985) Endonasal surgical treatment of bilateral choanal atresia under optic control in the infant. J Otolaryngol 14: 95-98.

19. Selik YK, Biesman BS, Rebeiz EE (2000) Topical application of Mitomycin-C in endoscopic dacryocystorhinostomy. Am J Rhinol 14: 205-207.