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Review Article

Choanal atresia

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Introduction:

Choanal atresia is not an uncommon congenital anomaly. This causes complete airway obstruction in neonates and infants in bilateral cases. Incidence of choanal atresia is 1:7000 - 8000 births.[1,2] Female to male ratio is 2:1. This is more frequently unilateral then bilateral and right sided twice often then left sided. 90% of choanal atresia is bony and 10% of which is membranous. 30% of such malformations consist of purely bony obstructions and 70% of a mixed bony - membranous obstruction.[3,4] Incomplete atresia is termed as choanal stenosis. Associated congenital anomalies occur in association with choanal atresia, about 50% of the patients with CHARGE.[5-7]

Embryology: There are four theories for the development of choanal atresia. The theories are (i) Persistence of the nasobuccal membrane, (ii) Persistence of the foregut buccopharyngeal membrane,(iii) Abnormal mesodermal adhesion in the nasal choanae and (iv) Misdirection of mesodermal flow due to local factors.[8]

Anatomy: The boundaries of the atresia plate are: superiorly - the under surface of the body of the sphenoid bones, laterally - the medial pterygoid lamina, medially - the vomer and inferiorly - the horizontal portion of the palatal bone.[4, 9]

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Clinical presentation:

a. Bilateral choanal atresia:

i) Complete nasal obstruction: there will be immediate respiratory distress and even potential death due to asphyxia as newborns are obligate nose breathers until approximately 4-6 weeks at which time mouth breathing is learnt.

ii) Cyclical respiratory obstruction: As the child falls asleep the mouth closes and a progressive obstruction starts followed by stridor, increased respiratory effort and cyanosis. Either the observer opens the child's mouth or the child walkes up and the obstruction is cleared.

iii) Presentation as tracheo- esophageal fistula: Bilateral choanal atresia may present like tracheo- esophageal fistula due to aspiration of milk. Progressive airway obstruction, cyanosis & choking develops in both cases.[10]

b. Unilateral choanal atresia:

Unilateral mucoid discharge is usual phenomenon. Rarely respiratory distress may occur.

c. Associated congenital Anomalies:

There are many congenital anomalies which are associated with choanal atresia.

i) CHARGE ASSOCIATION: Component of this association are **C** : Coloboma **H**: Congenital heart disease **A** : Choanal Atresia **R** : Retarded growth and development **G** : Genital Anomalies **E** : Ear anomalies & deafness.

ii) 59% of patient with CHARGE association have bilateral choanal atresia. The rest have unilateral choanal atresia.

iii) 2.75% of all patients with multiple congenital anomalies & with bilateral choanal atresia.[11]

Differential diagnoses of choanal atresia are deviated nasal septum, dislocated nasal septum, septal haematoma, mucosal swelling, turbinate hypertrophy, encephalocele, nasal dermoid, hamartoma, chordoma, teratoma, glioma, rhabdomyosarcoma etc.

Assessment and planning:

A. Physical Examination:

1. It depends on level of obstruction of soft fine catheter. Failure to pass # 6 to 8 french plastic catheter through the nares into the pharynx. Obstruction approximately 3-3.5 cm from the alar rim is suspected for choanal atresia. Obstruction approximately 1-2 cm from alar rim is usually traumatic defection of the nasal septum during delivery. Failure to pass more than 5.5 cm from alar rim is diagnostic.

   No definite obstruction is found in case of mucosal swelling, hypertrophied inferior turbinate etc. Other methods to detect choanal atresia are failure to pass soft metal probes, movement of wisps of cotton due to nasal airflow, Methylene blue dye in the nares not passing into pharynx, Acoustic rhinometry etc.

B. Imaging studies:

1. An axial non-contrast high resolution CT scan with thin section (2-5 mm) is the image study of choice. According to a study, the mean choanal air space distance (lateral wall of the nasal cavity to vomer) is 0.67 cm and it increases 0.027 cm/year until age 20 year of age.

2. Rhinography is a procedure that involves the administration of radiopaque dye into the nasal cavity.[11]

Treatment:

Treatment can be divided in to two types

a) Initial emergency management[5, 7, 12]

b) Elective definitive management

Initial emergency management:

A. Establishment of airway is essential in new born with bilateral choanal artesia. This can be achieved by- An oral airway, MC Govern nipple, Intubation, orogastric tube. Tracheostomy may be required when CHARGE association is present.

B. Feeding: Usually Gavage feeding is required until the child has learnt to mouth breathe. When once the child learns mouth breathing, then he will swallow air and need frequent "breathing/burping breaks."

Elective definitive treatment: Surgical management can be done in three ways:

1. Transnasal blind puncture, 2.Transnasal approach with endoscopy, 3. Trans palatine approach

Transnasal endoscopic choanal atresia repair showed a primary success rate of 70% and after second look endoscopic debridement the success rate was 100%. In a series it was reported that using a cruciate incision for exposure of the mucoperiosteum covering the atretic plate, then a diamond burr to create a hole at the level of the inferomedial portion of the plate (considered the thinnest and softest area.)[6]

Stenting with a nasal tube for a suitable period is favourable.

Outcome of endoscopic repair of congenital choanal atresia has been improved with new interventions like-

- Use of mitomycin-C, potassium-titanyl-phosphate laser and diode laser, optical, CT data based navigation system etc.

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