

# Choanal Atresia - A Rare Case Report

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## Abstract

Choanal atresia is a rare congenital condition wherein there is unilateral or bilateral obstruction of posterior nasal passage. We report a case of bilateral choanal atresia in a two days old female presenting with chief complaints of respiratory distress since birth. Transnasal endoscopic surgery was used to obtain a patent airway.

**Keywords:** Bilateral Choanal Atresia, Endoscopic Diagnosis, Stenotic Choanae, Transnasal Endoscopic Canalization

## 1. Introduction

Choanal atresia is a rare congenital condition wherein there is unilateral or bilateral obstruction of posterior nasal passage. This abnormality was first described in 1755 by Roederer<sup>1</sup>. In 1854; Emmert reported the first successful surgical procedure for congenital choanal atresia in a 7 year old boy using a curved trocar transnasally<sup>2</sup>. 50-60 percent cases are unilateral. Incidence is 1 in 7000<sup>3</sup>. Female to male ratio is 2:1<sup>4</sup>. It could be purely bony (29%) or mixed (71%) i.e., bony and membranous variety<sup>3</sup>. The most acceptable theory regarding the pathophysiology is failure of breakdown of buccopharyngeal membrane on 45<sup>th</sup> day of gestation<sup>3</sup>. Bilateral choanal atresia presents as an acute respiratory emergency at birth since newborns are obligate nasal breathers<sup>4</sup>. The classical picture of cyclical cyanotic spells is observed as the patient cries. Unilateral nasal obstruction comes to light in relatively older children with profuse persistent unilateral nasal discharge with no history suggestive of foreign body insertion<sup>5</sup>. The diagnosis of choanal atresia is made clinically by the inability to pass a nasogastric tube through the nasal cavity and is confirmed with CT imaging. It may occur as an isolated anomaly or in association with other anomalies or syndromes; examples- CHARGE syndrome

(coloboma, heart-defects, atresia choanae, retardation of growth, genital anomalies and ear abnormalities), Achondroplasia, Crouzon syndrome, Treacher Collin syndrome and Fetal alcohol syndrome<sup>1,3-5</sup>.

## 2. Case Report

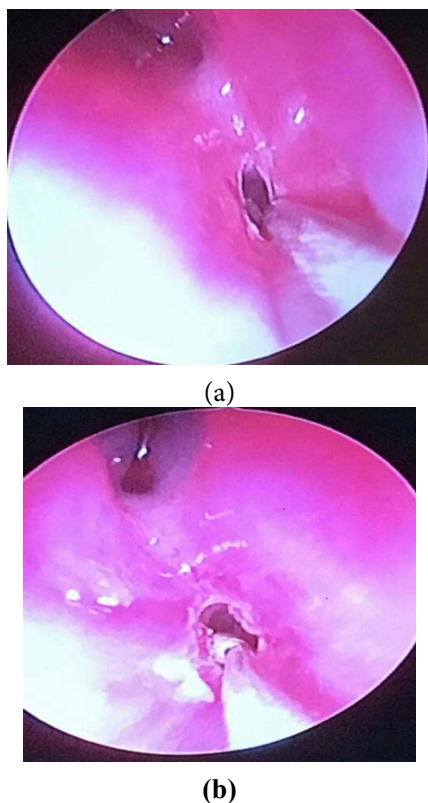
A 2 day old female child was brought by relatives to casualty with chief complaints of respiratory distress since birth. The patient was born on 15<sup>th</sup> August 2016 at 7 pm. It was a full term, normal delivery at hospital, birth weight being 3kg and baby had cried immediately at birth. Mother was G2P1A1L1 with history of oligo-hydramnios in antenatal period and no intrapartum or postpartum complications. She was immediately taken to a tertiary hospital for respiratory distress associated with noisy breathing.

On examination patient was conscious, crying and was maintaining 88 percent oxygen saturation without any supplemental support. Patient was tachypneic and was making grunting sounds however air entry was found to be equal on both sides without any respiratory adventitious sounds. Infant nasogastric tube could not be inserted in either nasal cavity and the patient was diagnosed as having bilateral choanal atresia. On examination

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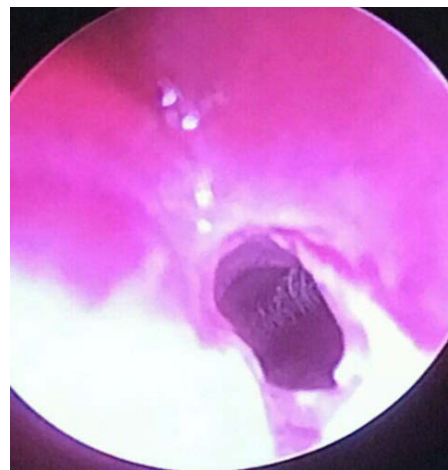
of other systems no abnormalities were detected. Nasal endoscopic examination confirmed bilateral membranous choanal atresia.

After due work up, patient was taken up for endoscopic canalization of stenotic choanae on 23<sup>rd</sup> August. Both the membranous choanae were incised with a sickle knife and serially dilated with Hegar's dilators with the help of a sinuscope. Patency was achieved bilaterally and 2.5 number endotracheal tubes were inserted bilaterally and fixed.



**Figure 1.** Intra-operative pictures showing the serial dilatation of the choanae.

After the surgery patient was stable and maintaining saturation. Patient was started on feeding by Ryles tube. The left nostril endotracheal tube came out spontaneously on 28<sup>th</sup> August 2016 and the right nostril endotracheal tube was taken out on 7<sup>th</sup> September 2016 and nasal endoscopy was done. On endoscopy bilateral posterior choana were open and patency was achieved bilaterally. Patient was thriving well and gained weight after the surgery. Patient was then sent home on 12<sup>th</sup> September 2016. The patient was followed up 3 months after surgery at which the choana was patent. No follow up was taken after this as the patient was fine.



**Figure 2.** Post-operative endoscopic picture.

### 3. Discussion

Complete nasal obstruction in a newborn as seen in bilateral choanal atresia may cause sudden death of the newborn due to asphyxia as during attempted inspiration the tongue is pulled to the palate and oral airway gets obstructed and since nasal airway is also obstructed. Vigorous respiratory efforts by the newborn produce respiratory distress and cyanosis. However, when the child cries and takes a deep breath through mouth the oral obstruction gets relieved momentarily and as soon as the crying stops both airways get obstructed. Various theories have been proposed to explain the pathophysiology of choanal atresia-

- Persistence of buccopharyngeal membrane.
- Failure of buconasal membrane of Hochstetter to rupture.
- Medial outgrowth of vertical and horizontal processes of the palatine bone.
- Abnormal mesodermal adhesions forming in the choanal area.
- Misdirection of mesodermal flow due to local factors.

In 2008, Barbero et al suggested that prenatal use of antithyroid (methimazole, carbimazole) medications was linked to choanal atresia<sup>6</sup>. Lee et al., evaluated the association between continuous and categorical infant T4 levels and non syndromic choanal atresia<sup>7</sup>. It was suggested that when there are low levels of T4 or low levels of thy-

roid hormones during the critical period of development there is a risk for choanal atresia. Using data published from the National Birth Defects Prevention Study; Kancherla et al., found evidence linking choanal atresia to maternal exposure to various nutrients, thyroid medications, and cigarettes. The investigators looked at pre pregnancy exposures (within 1 year prior to conception) and periconceptual exposure (between 1 month before and 3 months after conception) in 117 women who gave birth to infants with choanal atresia and 8350 control mothers. Positive associations were found between the following pre pregnancy exposures and choanal atresia<sup>8</sup>:

- Intake in the highest quartile: Vitamin B-12, zinc, niacin.
- Intake in the lowest quartile: Methionine, vitamin D.
- Coffee ( $\geq 3$  cups per day).

Positive associations were also found between the following periconceptual exposures and choanal atresia:

- Thyroid medications.
- Cigarette smoking.

A high index of suspicion is required to diagnose choanal atresia. The initial clinical evaluation includes a complete physical examination to assess for other congenital anomalies. A small feeding tube could be used to determine the patency of the choana, but a complete nasal and nasopharyngeal examination should be performed using a flexible fiberoptic endoscope to assess the deformity. CT scan is the diagnostic procedure of choice in evaluation of choanal atresia. Rhinography is another diagnostic procedure that involves the administration of radiopaque dye into the nasal cavity<sup>2</sup>. Other diagnostic procedures include<sup>2</sup>:

- Failure to pass an 8F catheter through the nasal cavity more than 5.5 cm from the alar rim.
- The lack of movement of a thin wisp of cotton under the nostrils while the mouth is closed.
- The absence of fog on a mirror when it is placed under the nostrils.

In the case discussed, we established the diagnosis of the patient by endoscopic examination. The treatment options for choanal atresia are emergency and elective.

Bilateral choanal atresia is an emergency that is initially best treated by inserting an oral airway to break the seal formed by the tongue with the palate and the oral airway pathway of the neonate can be kept patent by it for several days till the elective surgery is undertaken. Other alternative emergency treatments include use of a Mc Govern nipple and intubation. The elective treatment options include surgery to establish nasal patency which includes:

- Transnasal puncture, with or without a microscope - this approach has become unpopular due to high rate of complications, high rate of failure and need for revision surgeries.
- The transseptal technique consists of making a window in the septum anterior to the atretic plate.
- Transpalatal repair is a technique that provides excellent exposure and has a high success rate but requires more operative time and more care post-operatively with high incidence of complications.
- The endoscopic technique (nasal or retropalatal), with or without powered instrumentation, offers excellent visualization with great ease in removing the bony and membranous choanae<sup>9</sup>. This approach is employed during the case described and was done via the nasal endoscopic approach.
- Combined transoral-transnasal<sup>10</sup>.
- Use of microdebriders.
- Carbon dioxide and KTP lasers with adjuvant use of Mitomycin C to prevent restenosis<sup>11</sup>.

Postoperative stenting is usually done with either endotracheal tube or Foley's catheter and neonate is given care in a neonatal ICU setting.

## 4. Conclusion

Congenital choanal atresia has been recognized for more than 200 years and there are different methods used to diagnose and treat this condition. Diagnosis by checking patency using nasal catheter and then serial dilatations to maintain patency of the choanae are routinely done methods. Neonatal airway obstruction leading to respiratory distress is a rare problem in the new born, and it is important to be able to swiftly delineate the cause of the obstruction to give effective treatment in a timely manner. Bilateral choanal atresia should always be considered in these circumstances because, although it is a relatively rare cause, mortality is high even when surgery is done

with due risks of anaesthesia present as well as prolonged post-operative care required in NICU. The most important reason for increasing awareness about choanal atresia is in peripheral parts of our country where these patients often succumb due to misdiagnosis and inadequate treatment.

## 5. REFERENCES

1. Flake CG, Ferguson CF. Congenital choanal atresia in infants and children. *Ann Otol Rhinol Laryngol.* 1964; 73:458–73. <https://doi.org/10.1177/000348946407300216> PMID:14196687
2. Tewfik TL, Hagr A. Choanal atresia. *The e-Medicine Clinical Knowledge Base.* Institutional ed; 2007 Feb 23.
3. Ballenger's otorhinolaryngology head and neck surgery. 16th ed; 2004.
4. Sataloff's comprehensive textbook of otolaryngology, head and neck surgery.
5. Scott Brown's Otorhinolaryngology, Head and Neck Surgery. 7th ed. CRC Press; 2008.
6. Barbero P, Valdez R, Rodríguez H, et al. Choanal atresia associated with maternal hyperthyroidism treated with methimazole: A case-control study. *Am J Med Genet A.* 2008 Sep 15; 146A(18):2390–5. [Medline]. <https://doi.org/10.1002/ajmg.a.32497> PMID:18698631
7. Lee LJ, Canfield MA, Hashmi SS, Moffitt KB, Marengo L, Agopian AJ, et al. Association between thyroxine levels at birth and choanal atresia or stenosis among infants in Texas, 2004-2007. *Birth Defects Res A Clin Mol Teratol.* 2012 Nov. 94(11):951–4. [Medline]. <https://doi.org/10.1002/bdra.23091> PMID:23125093
8. Kancharla V, Romitti PA, Sun L, et al. Descriptive and risk factor analysis for choanal atresia: The national birth defects prevention study, 1997-2007. *Eur J Med Genet.* 2014 Apr; 57(5):220–9. [Medline]. <https://doi.org/10.1016/j.ejmg.2014.02.010> PMID:24576610 PMCid:PMC4520237
9. Assanasen P, Methetrairut C. Choanal atresia. *J Med Assoc Thai.* 2009 May; 92(5):699–706. [Medline]. PMID:19459535
10. Hassan M, AboEl-Ezz T, Youssef T. Combined transoral-transnasal approach in the repair of congenital posterior choanal atresia: clinical experience. *J Otolaryngol Head Neck Surg.* 2011 Jun 1; 40(3):271–6. [Medline]. PMID:21518653
11. Prasad M, Ward RF, April MM, Bent JP, Froehlich P. Topical mitomycin as an adjunct to choanal atresia repair. *Arch Otolaryngol Head Neck Surg.* 2002 Apr; 128(4):398–400. [Medline]. <https://doi.org/10.1001/archotol.128.4.398> PMID:11926914

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