 Abstract: Congenital choanal atresia is the developmental failure of the communication between the posterior nasal cavity and the nasopharynx. Choanal atresia is an uncommon congenital anomaly of the nose with an incidence of approximately one in 7000 live births (1, 2). Choanal atresia is caused by failure of resorption of the bucco-nasal membrane during embryonic development (3). Choanal atresia has a significant incidence of associated defects (4). The most common associated congenital anomaly is CHARGE syndrome (5). Bilateral choanal atresia presents with severe respiratory distress and cyanosis at birth and is alleviated by crying whereas unilateral atresia often remains undetected until late in life (6). Bilateral choanal atresia is managed with an oropharyngeal airway (7). Surgical repair is recommended in the first weeks of life in bilateral atresia because this is a life-threatening situation in newborns with two main approaches, namely transnasal or transpalatal (8, 9). Here we discuss a case report of successful management of a neonate with bilateral choanal atresia who presented with intermittent attacks of cyanosis and respiratory distress soon after birth. **Keyword:** Bilateral Choanal Atresia, Neonate, Respiratory Distress, Transnasal Endoscopic surgery

Introduction: Congenital Choanal atresia is a rare malformation that causes airway obstruction in newborns. It affects girls twice as often as boys and is frequently unilateral and rightsided than bilateral (6). The atresia may be classified as bony, mixed bony and membranous or purely membranous although the latter is rare. About 30% are pure bony, whereas 70% are mixed bony-membranous (10). Most cases of choanal atresia are isolated malformations, but association with other congenital deformities is not an exception. Associated anomalies occur in 20% to 50% of infants with choanal atresia with CHARGE syndrome being the most common (4). The CHARGE syndrome includes
coloboma or other ophthalmic anomalies, heart defect, atresia choanae, restriction of growth and developmental, genitourinary defects, and ear anomalies with hearing loss. Bilateral choanal atresia is an emergency, because maintaining an airway and relieving the obstruction is a priority. Increased cyanosis and death may occur if appropriate interventions are not implemented. The immediate management of neonates presenting with intermittent cyanosis is the insertion of an oral airway and feeding via an oro-gastric tube. There are numerous methods for correcting this condition. Commonly used methods are the transnasal approach, and the trans-palatal approach. Diagnosing and managing this rare neonatal emergency in a symptomatic neonate with bilateral choanal atresia, requiring transnasal repair using serial dilators with postoperative stenting at a very early postnatal age of 8 days is presented.

Case Report:
A 2.3 Kgs, 38 weeks/SGA/Female neonate was born to Para 5 mother by labour naturalis at a hospital in Nellore (AP). She was born with APGAR scores of 8 and 9 at 1 and 5 minutes respectively. Soon after birth the baby developed respiratory distress with cyanosis. Septic workup and cardiac evaluation was planned. A chest x-ray was taken which was normal. Septic markers including complete blood count, CRP and blood culture were normal. The finding of cyclical or intermittent cyanosis (which resolved on crying) concomitant with an unremarkable respiratory and cardiovascular examination prompted them to exclude the possibility of choanal atresia. A size 6 French suction tube was passed into the nasal cavities but it failed, hence further workup was planned to confirm the diagnosis of bilateral choanal atresia. The baby’s airway was maintained with an oropharyngeal airway of size 0. There after CT scan of paranasal sinuses and nose was performed. CT paranasal sinuses confirmed the presence of bilateral bony-membranous type of congenital choanal atresia. The baby was referred on 6th day of life to our hospital in Chennai, for further management.
CT scan of the newborn showing bony-membranous type of congenital choanal atresia bilaterally in the sagittal view

Photograph 1 following successful bilateral surgical correction showing stents fashioned from endotracheal tubes in place and the baby breathing at room air

Photograph 2 following successful bilateral surgical correction showing stents fashioned from endotracheal tubes in place and the baby breathing at room air

Examination at our place did not reveal any dysmorphic features. An ENT consult was sought and diagnosis confirmed. She was started on feeding via oro-gastric tube and tolerated feeds well. A surgical correction of bilateral choanal atresia was carried out on the 8th day of life. Endoscopy showed complete choanal atresia of mixed type with bony and membranous parts on both the sides. Surgery was performed under general anesthesia using the transnasal approach. The bilateral choanal obstruction was relieved by serial dilatation using dilators of progressively larger diameter whereby the membranous and thin bony laminae were pierced through and neochoanae created. After surgery stents fashioned from size 3 endotracheal tubes were placed bilaterally and stitched anteriorly to the columella with silk to maintain the patency of the neochoanae. Baby remained stable postoperatively. Local antiseptic cream and nasal saline drops were instituted. Parents were taught regular suctioning and irrigation with a few drops of saline solution to maintain the patency of the endonasal tubes. Association with CHARGE syndrome was ruled out. The baby was discharged on 10th postoperative day while she was able to breast feed and the tubes were in situ. Patient is being followed up and was doing well at the last follow up at age of one and a half months with the endonasal tubes in situ and the neochoanae patent.

Discussion:
Congenital Choanal atresia is a disease of nasal airway where no connection exists between the nasal cavity and the aero digestive tract. It was first described by Johann Roderer in 1755. Female to male ratio is (2:1) and right unilateral atresia is more common than bilateral. It is a rare congenital disorder occurring approximately one in 7000 live births. Atresia itself may be classified as bony, mixed bony and membranous, and purely membranous. Current theories of Choanal atresia...
embryogenesis cite either the persistence of nasobuccal membrane of Hochstetter or the buccopharyngeal membrane from the foregut, impaired migration of neural crest or mesoderm cells, or overgrowth of the vertical and horizontal processes of the palatine bone as the cause of choanal atresia.

Unilateral choanal atresia may not be detected for years and patients may present with unilateral rhinorrhea or congestion. Bilateral choanal atresia causes complete nasal obstruction and leads to immediate respiratory distress and cyanosis at birth as newborns are obligate nose breathers until approximately 4-6 weeks of life at which time mouth breathing is learnt reinforcing the fact that ontogeny recapitulates phylogeny. The respiratory obstruction is cyclic — as the child falls asleep the mouth closes and a progressive obstruction ensues starting with stridor followed by increased respiratory effort and cyanosis. However, a high index of suspicion is required to diagnose bilateral choanal atresia. Symptoms of airway obstruction and cyclical cyanosis are the classic signs of neonatal bilateral choanal atresia. Bilateral congenital choanal atresia is diagnosed when the 6 F nasogastric tube fails to pass through both the nares down to oropharynx in a symptomatic neonate. Absence of misting on a mirror or the movement of a wisp of cotton wool in front of the nostrils are other bedside methods used for the diagnosis of choanal atresia. Most cases of choanal atresia are isolated malformations, but association with other congenital deformities is not exceptional and has been reported in literature in 20% to 50% of the case. Association with CHARGE syndrome is the most common which has an incidence of 0.1 to 1.2/10,000 births. The diagnosis can be confirmed by flexible fibreoptic endoscopes and nasopharyngeal choanography but the current investigation of choice is Computed Tomography (CT) scan.

For good results in CT scan careful suctioning is performed to clear excess mucus, and a topical decongestant is applied prior to the investigation. The purpose of CT scanning is outlined as follows:

Schematic presentation of normal choanal air space and vomer width in a newborn
Confirming the diagnosis of choanal atresia (unilateral or bilateral).
-Evaluating choanal atresia (vomer bone width and choanal airspace distance).
-Excluding other possible nasal sites of obstruction.
-Determining the degree of bony, membranous, or mixed atresia.
-Delineating other abnormalities in the nasal cavity and nasopharynx.

The surgical treatment of congenital atresia is one of the more challenging endeavors within the realm of pediatric otolaryngology. There are about five methods for correcting this condition – transnasal, transpalatal, sublabial transnasal, transseptal and transantral but the most commonly used ones currently are the transnasal and the transpalatal approach. Trans-nasal puncture and dilatation followed by stenting with modified endotracheal portex tubes for at least 4-6 weeks to allow complete healing of the neochoanae, is the initial surgical procedure of choice.
These nasal tubes need to be kept clear by regular suctioning and irrigation with a few drops of saline as required. The baby’s parents should be taught this process prior to discharge home. The transnasal approach works best with thin buccopharyngeal membranes and tends to have higher recurrence and reoperation rates\(^\text{14}\). The transpalatal approach entails direct surgical correction of the defect and is typically performed for thick bony membranes. However, despite the minimal rate of reoperation, this approach is associated with a higher rate of palatal growth deformities. Modern endoscopic devices have improved the transnasal technique and studies continue to demonstrate increasing support for the transnasal endoscopic repair of choanal atresia\(^\text{15}\). Improved outcomes have been reported using topical mitomycin C application at the time of surgery\(^\text{16}\).

**Conclusion:**
Bilateral congenital choanal atresia in the neonate is a combined medical and surgical emergency that should be treated promptly. It is to be suspected in any newborn with a history of *cyclical cyanosis* and diagnosed by failure to insert 6 F nasal catheters through the nares into the nasopharynx. CT scan of the nasal cavity and nasopharynx helps to firmly establish the diagnosis, to define the extent and type of atresia and to determine the surgical approach. These infants should have an adequate airway established and secured to prevent intermittent cyanosis and resultant hypoxia till further management with corrective surgery. Furthermore, the finding of choanal atresia, especially when it is bilateral, warrants careful search for other congenital anomalies in the newborn.


