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Case Series

Ear, Nose and Throat Section

Neonatal Outcome of Choanal Atresia Surgical Corrections- Experience from a Tertiary Care Centre from Eastern India

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ABSTRACT

Choanal atresia is a congenital condition that occurs due to developmental failure of nasal cavity to communicate with the nasopharynx. There can be both unilateral and bilateral occlusion, bony, soft tissue and both. It can present at birth or early in the neonatal period or even later in life. This case series included three patients, who were admitted in the Neonatal Intensive Care Unit (NICU) and they underwent surgical correction in the hospital. All these cases had different presentations and clinical course. First case was admitted for respiratory failure and sepsis and, later was diagnosed as bilateral choanal atresia. Second case was admitted at 17 days of life with diagnosis of Hypoxic Ischaemic Encephalopathy (HIE) and multiple extubation failure and later was diagnosed as unilateral choanal atresia. Third case had respiratory distress soon after birth and was referred with a diagnosis of suspected choanal atresia. It was confirmed as bilateral choanal atresia and the child improved after surgical correction. All three cases were out born and referred in the hospital at different postnatal ages with varied symptoms. The lessons learnt in management of first case helped in subsequent cases resulting in better outcome.

Keywords: Choanal atresioplasty, Cyanosis, Neonates, Respiratory distress

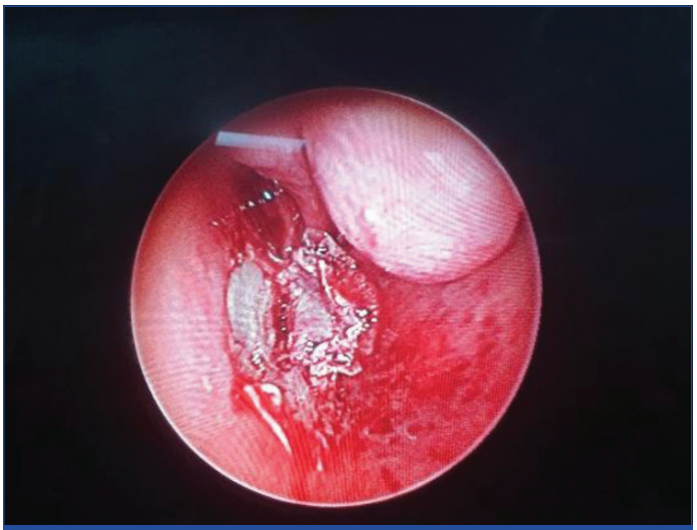
INTRODUCTION

Choanal atresia is a well recognised congenital craniofacial defect characterised by occlusion in the posterior nasal passage [1]. It may be unilateral, presenting with unilateral mucopurulent discharge or bilateral, in which the neonate presents with respiratory distress and cyanosis which is alleviated by crying. Since newborns are obligate nasal breathers, establishing an airway is an acute otolaryngologic emergency in such cases [2-4]. The series presented three patients of congenital choanal atresia, their clinical presentations, management and lessons learnt in management.

CASE 1

A two-day-old late preterm male neonate (36+5 weeks), born by Lower Segment Caesarean-Section (LSCS) {Premature Rupture Of Membranes (PROM)} subsequently developed respiratory distress, for which he had to be intubated and referred to our hospital. Clinical examination did not reveal any other congenital anomaly. Echocardiogram (Echo) and Ultrasound (USG) of abdomen was normal. Initially, the case was thought be of early onset neonatal sepsis presenting with congenital pneumonia, given the maternal history of PROM. However, the markers of sepsis were negative, and blood culture was sterile and X-ray chest (Anteroposterior view (AP view) showed no infiltrates. As the baby improved clinically, his ventilator settings were reduced to minimum and he was planned for extubation. However, the trial of extubation failed and the child started desaturating once he was shifted to Continuous Positive Airway Pressure (CPAP). He was reintubated and put on mechanical ventilation. As soon as he was mechanically ventilated, his distress went away, and saturation improved dramatically. As the attempt to pass a nasogastric tube for feeding purpose via both nostrils failed, it was then suspected that he may have bilateral choanal atresia. So, Computed Tomography (CT) scan for nose and paranasal sinuses was done which confirmed the bilateral choanal atresia of combined osseo-membranous type as diagnosis.

Baby underwent bilateral choanoplasty by a transnasal endoscopic approach. Surgical findings revealed osteomembranous type of bilateral choanal atresia. During choanoplasty, first the posterior septal flap was raised to expose the bony atretic plate [Table/Fig-1].

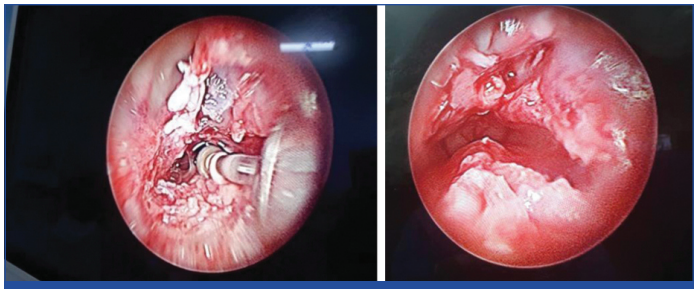


[Table/Fig-1]: Preoperative image showing posterior septal flap raised on left side.

The bony occlusion was broken with trochar and cannula with utmost care [Table/Fig-2] and choanal opening was widened with 1 mm size cutting or diamond burrs in order to expose the nasopharynx adequately [Table/Fig-3]. A cut portion of endotracheal tube with outer diameter 4 mm and length 5 cm was placed in both the nasal cavities and fixed with thread to keep the choanal opening patent. The portion of Endotracheal tube (ET) as a stent was planned to be kept for three weeks to make the operated nasal opening patent.

Baby was well in the immediate postoperative period, and was successfully extubated and feeding was established. The nasal tubes were removed after three weeks. The patient started desaturation after a few days and intubated again. Repeat endoscopy revealed mucosal tags growth which were occluding the choanae.

After the revision choanoplasty, in which bilateral silicon urinary catheter (12 gauze) size was placed, the child improved gradually, he



[Table/Fig-2]: Intraoperative image showing bony occlusion broken.
[Table/Fig-3]: Postoperative image showing nasopharynx. (Images from left to right)

was successfully weaned off mechanical ventilation and he began to take oral feeds [Table/Fig 4]. He was then discharged. The baby was followed-up monthly for three months than three monthly and in follow-up visits the baby was asymptomatic, growing well on last follow-up baby was one and a half year old and was well with normal voice.

[Table/Fig-4]: Postoperative image showing patient with bilateral silicone catheter adequately exposed after surgery.

CASE 2

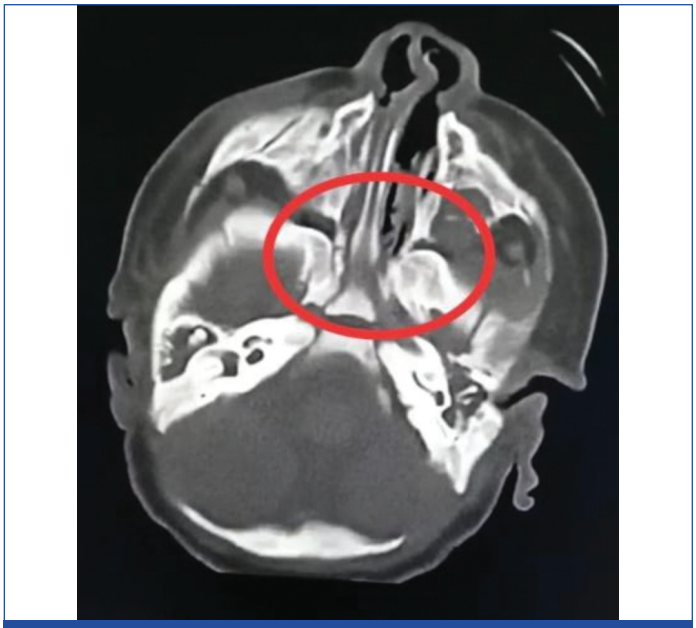
A 17-day-old male newborn presented in the NICU in a gasping condition. Baby was intubated and put on mechanical ventilation. He was born by normal vaginal delivery and had delayed cry at birth. He was on mechanical ventilation for 14 days and referred to our hospital as there was no improvement and successive attempts to wean the baby off the ventilator had failed, on nasal prongs.

There was history of seizures and the baby was on two anticonvulsants. He had hypertonia of bilateral lower limbs. The baby's respiratory condition improved in the following days and he was extubated on the 5th day of admission. However, he suffered extubation failure and intubated again and put on invasive mechanical ventilation.

He had profuse discharge from one nostril which was not amenable to suctioning. Inability to pass a 5 French tube through the right nostril into the nasopharynx led to the suspicion of unilateral choanal atresia. CT scan of nose and paranasal sinuses [Table/Fig-5] confirmed the diagnosis of unilateral (right sided) choanal atresia of combined type (bony and membranous) however the baby also had left sided soft tissue opacification (membranous stenosis) of posterior part on nasal cavity.

Surgical findings demonstrated right sided bony atresia, along with stenosis of the left choanae. Left choanal opening was widened, and atretic plate of right-side was removed using instruments as mentioned earlier. As the lesson was learnt from first case that chances of restenosis was high using ET tube stent so this time nasal cavities were packed with Merocel (non absorbable nasal packing material).

The baby was doing well postoperative period. He was on mechanical ventilation in the immediate postoperative period. Gradually, he was weaned off ventilation and feeding was established. Merocel was removed on the third postoperative day. His surgical wound healed with time and examination showed bilateral patent choanae. Magnetic Resonance Imaging (MRI) brain was done to look for any hypoxic injury and it showed features of HIE sequelae. Brainstem Evoked Response Audiometry (BERA) and Visual Evoked Potential (VEP) were within normal limits. When the baby was stable in room air and was taking feeds through bowl and spoon, he was discharged. On follow-up



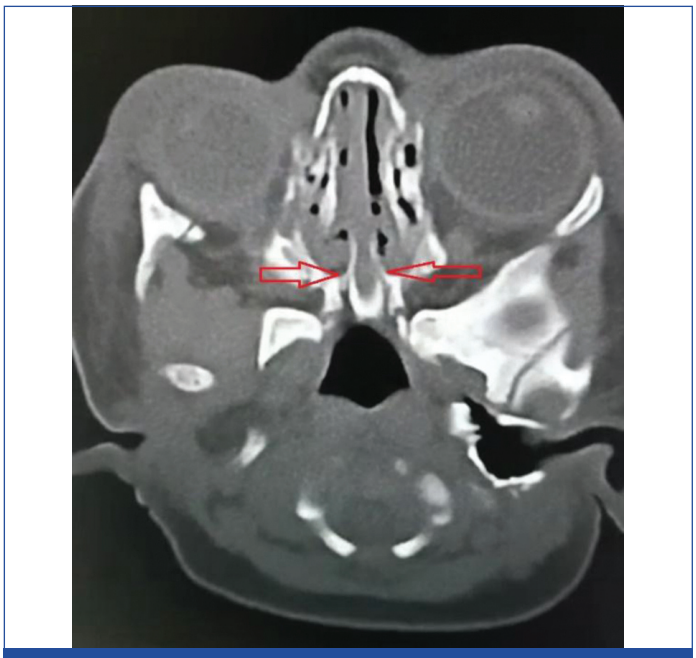
[Table/Fig-5]: CT Image of case 2 showing unilateral (right sided) combined choanal atresia with left sided membranous occlusion.

baby was haemodynamically stable on mixed feed and tone in lower extremities were high for which baby was referred to high risk clinic.

CASE 3

The third patient is a three-day-old single, term, female baby with a birth weight of 2.5 kg, who was born outside in a private hospital to a primigravida, by LSCS and developed respiratory distress soon after birth. The baby used to get cyanosed while feeding and got relieved on crying. She was suspected to have bilateral choanal atresia and was referred to the hospital. On examination, she had respiratory distress with intercostal and subcostal retractions, bilateral conducted sounds in lungs, oxygen saturation of 82% with O_2 at flow rate of 5L/min via oxygen hood. Examination findings of other systems were within normal limits. She did not have any apparent congenital anomaly.

She was immediately intubated with 3.5 mm tube fixed at 9 cm and put on invasive mechanical ventilation. Necessary investigations were sent. After failure of passing a nasogastric tube to both nostrils, a provisional diagnosis of choanal atresia was made. A CT scan of nose and paranasal sinuses was done to confirm the diagnosis. In this case, a complete bilateral bony variety of choanal atresia was seen [Table/Fig-6]. Echocardiography, USG abdomen,



[Table/Fig-6]: CT scan showing bilateral bony choanal atresia.

ophthalmologic examinations were done to rule out any associated congenital anomalies, but they were all normal.

Patient was posted for bilateral choanoplasty via a transnasal endoscopic approach. Surgical findings included bilateral persistent buccopharyngeal membrane. Bilateral posterior septectomy was done and single opening of choanae made. Merocel was placed in each nasal cavity.

Postoperative period was uneventful. Merocel was removed on the fourth postoperative day. Patient was successfully extubated, and postoperative nasal endoscopy was done which revealed patent choanae. Feeding was established and gradually increased. The child ultimately started taking breastfeeding and was then discharged. The baby was doing well in follow-up visits. The demographic and presentation characteristics of the cases are given in [Table/Fig-7,8] shows their laboratory findings.

DISCUSSION

Choanal atresia is defined as a malformation of the posterior nasal aperture that interferes with airflow from the nose to the rhinopharynx. It was first described in 1755 by Roederer [5]. A 55% of the cases are unilateral, while rest is bilateral. If unilateral, it occurs more commonly on the right-side [6]. It is more common in females [7].

Variables	CASE 1	CASE 2	CASE 3
Gestational age	36+5 weeks	39 weeks	38 weeks
Gender	Male	Male	Female
Laterality	Bilateral	Unilateral	Bilateral
Type	Osteo-membranous	Osteoid	Membranous
Presentation	Respiratory failure	Unilateral nasal discharge	Respiratory failure
Associated congenital anomalies	None	None	None
Other complications	None	HIE-II+Late onset neonatal sepsis	None
Type of surgery	Endoscopic transnasal bilteral atresioplasty	Endoscopic transnasal Unilateral atresioplasty	Endoscopic transnasal bilateral atresioplasty
Postoperative complications	Restenosis due to mucosal tag formation	None	None

[Table/Fig-7]: Comparison of characteristics of the three cases.
HIE-II: Hypoxic ischaemic encephalopathy

Investigations	CASE 1			CASE 2			CASE 3		
	Day 3 (Day 1 of admission)	Day 7	Day 14	Day 17 (Day 1 of admission)	Day 29	Day 39	Day 3 (Day 1 of admission)	Day 9	Day 16
Total Leucocyte Count (TLC) (/µL)	7200	12300	12300	8800	21860	11670	15220	11440	6020
Differential Leucocyte Count (DLC) (%)	$N_{77.4}L_{15.7}$	$N_{74.6}L_{18}$	$N_{74.6}L_{18}$	$N_{73.6}L_{21.3}$	$N_{68.2}L_{26.7}$	$N_{43}L_{44}$	$N_{75.6}L_{17}$	$N_{55.1}L_{25.9}$	$N_{77}L_{17.4}$
Haemoglobin (g/dL)	11.6	11.8	11.8	15.2	14.2	8.5	16.3	12.3	12
Platelet count (/µL)	305000	416000	416000	215000	292000	449000	330000	187000	453000
C-reactive protein test (mg/L)	2.13	-	-	-	59.6	29.82	<2.8	-	-
Blood C/S	Sterile	-	-	Contaminant growth	-	-	Sterile	-	-
Blood urea/Serum creatinine (mg/dL)	38.1/0.74	-	--	22.8/0.5	-	-	39.4	13.9/0.55	-
Serum ($Na^+/K^+/Cl^-$) (meq/L)	134.08/5.05/99.29	-	-	(136/6.3/103)	-	-	139/6.5	139.3/4.59	-
Serum bilirubin (Total/ Direct/Indirect) (mg/dL)	-	-	-	(1.1/0.53/0.57)	-	-	11.48/0.74/10.74	2.79/0.72/2.07	1.09/0.43/0.66
Serum protein (Albumin)	-	-	-	5.1/3.07	-	-	6.45/4.44	3.63/2.44	5.15/3.03
SGPT/SGOT	-	-	-	39.7/59.1	-	-	26/92	8.6/33.6	6.7/17.0
PT/APTT/INR	-	-	18.3/33.1/1.55	14.6/45.2/1.16	-	-	19.5/18.2/1.67	15.8/35.6/1.28	-
CSF examination (Cell/ Sugar/Protein)	-	-	-	5 (L_{100})/93/170	-	-	-	-	-
CSF culture	-	-	-	Sterile	-	-	-	-	-
MRI brain	Abnormal hyperintense T1/FLAIR signal intensities seen involving bilteral thalamus, bilteral basal ganglia, midbrain, pons, medulla, and bilteral perirolandic area. Marked T1 hyperintensities seen along cortical grey matter of bilteral frontal region. Features suggestive of HIE sequelae.								
Echocardiography	Situs solitus. Valves and chamber size normal. Small secundum type of ASD of size 4 mm. Biventricular systolic function normal.			Small sized PDA, severe PPHN			Within normal limits		
Brain Evoked Response Auditory (BERA)	Bilteral hearing sensitivities within normal limits.			Bilteral hearing sensitivities within normal limits.			Bilteral hearing sensitivities within normal limits.		
Visual Evoked Potentials (VEP)	Normal			Normal			Normal		
USG whole abdomen	No gross abnormality seen.			No gross abnormality seen.			No gross abnormality seen.		

[Table/Fig-8]: Comparison of investigations done for the three cases arranged by day of life (MRI Brain done in case 2 only).
SGPT/SGOT: Serum glutamic oxaloacetic transaminase/Serum glutamic pyruvic transaminase; PT: Prothrombin time; INR: Normalised ratio; aPTT: activated partial thromboplastin time; CSF: Cerebrospinal fluid; MRI: Magnetic resonance imaging; USG: Ultrasound; HIE: Hypoxic Ischaemic encephalopathy

The nasal pit deepens and cavities extend posteriorly during development with the posteriorly directed fusion of the palatal processes. The nasobuccal membrane, which separates the nasal cavities from the oral cavity thins out gradually and by 6th week of development, it ruptures and forms the choanae. Failure of this rupture results in choanal atresia [1]. Choanal atresia is a relatively uncommon disease entity with an estimated incidence of 1:5000-7000 birth [5]. The atresia can be bony, membranous, or a combination of both. Purely membranous atresia is rare [6]. Most cases of choanal atresia are isolated malformations, but it may be associated with CHARGE syndrome (includes coloboma, heart defect, atresia choanae, retardation of growth, genital anomalies), and ear anomalies like hypoplasia of the external ear and hearing loss [8]. Presentation of unilateral and bilateral atresia is different. While bilateral atresia usually presents in the early neonatal period with respiratory distress and/or cyanosis as was seen in case 1 and 3 of the series, the unilateral cases although being commoner, diagnosis of unilateral atresia is much delayed. In this series, patient 2 unilateral atresia, but still needed respiratory support as his neonatal period was complicated by HIE II and late onset neonatal sepsis. Choanal atresia was an incidental finding in this case and it was suspected when there was profuse discharge from one nostril along with inability to pass feeding tube through it. However, there are rare cases of bilateral atresia presenting in childhood [9], adolescente [10], and even adults [11].

With regards to aetiology of the defect, a commonly proposed molecular theory is disruption of neural crest cell migration between the 4th and 11th weeks of gestation [12]. Exposure to agents like alcohol, retinoic acid, and anti-thyroid medications may contribute to choanal atresia. Off late molecular mechanisms related to retinoic acid receptor development has been implicated in the pathogenesis of choanal atresia [12].

Most of the cases of choanal atresia are isolated sporadic defects, but association with certain syndromes like Apert's syndrome, DiGeorge syndrome, trisomy 18, Treacher Collins syndrome, camptomelic dysplasia, and CHARGE association (coloboma, heart defects, atresia choanae, retardation of growth and development, genitourinary problems, and ear anomalies) has been seen. So, it is imperative to do a comprehensive general physical examination in order to exclude any associated congenital anomalies. In these three cases, no other significant congenital anomaly was identified. In all three of our cases, detailed physical and radiological assessment did not reveal any syndromic association.

Bilateral choanal atresia is a respiratory emergency, requiring surgical correction in the first few days of life. Temporary measures including oral airway, McGovern nipple and intubation may be used prior to surgical correction. Gujrathi CS et al., reported a series of patients who were all intubated prior to surgical management [13]. In this case series also, all three babies required mechanical ventilation before surgery.

The approaches to surgical management can be transpalatal, or transnasal. Increasingly, the transpalatal approach has been reserved for revision cases in older patients. Endoscopic transnasal repair of choanal atresia allows for excellent visualisation [14]. This approach was followed for the management of the index cases. Two cases had uneventful postoperative period, while one had to undergo revision surgery as there was growth of mucosal tags obstructing the choanae.

Having learnt from first case, in which there was restenosis while using cut ET tube as nasal stent, Merocoel (non absorbable suture material) was used for packing of nasal cavity during subsequent operations, which prevented restenosis.

CONCLUSION(S)

Choanal atresia is a rare anomaly which can be difficult to diagnose, particularly the unilateral ones. It is prudent to suspect choanal atresia in a neonate who presents with intermittent cyanosis, respiratory failure not attributable to any other cause, or unilateral nasal discharge (in case of unilateral atresia). The lessons learnt in management of first case helped in subsequent cases, resulting in a better outcome.

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