SJIF Impact Factor: 5.464

WORLD JOURNAL OF ADVANCE HEALTHCARE RESEARCH

ISSN: 2457-0400
Volume: 4.
Issue: 3.
Page N. 01-05
Year: 2020

Case Report www.wjahr.com

NEONATAL BILATERAL CHOANAL ATRESIA: A CASE REPORT AND REVIEW OF LITERATURE

Mohammad H[1*], Otene AT[2], Adekwu A[3], Ochoga M[4], Eneojo A[1], Ausha JT[2], Azih NK[4], Ijiko BE[4] and Ojo B[5], Amodu A.[6]

[1]Department of Radiology, Benue State University Teaching Hospital, Makurdi.
[2]Department of Surgery, Federal Medical Centre, Makurdi.
[3]Department of Surgery, Benue State University Teaching Hospital, Makurdi.
[4]Department of Pediatrics, Benue State University Teaching Hospital, Makurdi.
[5]Department of Morbid Anatomy, Benue State University Teaching Hospital, Makurdi.
[6]Department of Family Medicine, Federal Medical Centre, Makurdi.

Received date: 02 February 2020 **Revised date:** 13 March 2020 **Accepted date:** 03 April 2020

***Corresponding Author: Mohammad H.**

Department of Radiology, Benue State University Teaching Hospital, Makurdi.

ABSTRACT

Bilateral choanal atresia (BCA) is a rare variant of choanal atresia. It presents in neonates with severe respiratory distress. The index case is a full term neonate which presented with acute respiratory distress. Emergency resuscitation was done. Thereafter, the diagnosis was made on choanogram and confirmed on CT scan. The patient had transnasal repair and stenting. This case highlights that BCA is not unusual. It should be considered as a differential diagnosis of severe respiratory distress in the newborn.

KEYWORDS: Neonate, Diagnosis, Bilateral Choanal atresia.

INTRODUCTION

Choanal Atresia is a congenital anomaly in which there is blockage of the posterior choanae in the nasal cavity by bone, soft tissue, or both. The condition may be unilateral or bilateral. The true incidence of choanal atresia in the general population is not known. However, figures of 1:5,000 and 1:10,000 live births have been reported with the bilateral lesion less common than unilateral (40%:60%) and females more commonly affected than males(2:1).[1,2] It is also documented that more than half of the affected infants also have other congenital anomalies.[2]

Though the etiology of choanal atresia is unknown, thioamides such as methiomazole, carbimazole and propylthiouracil which are antithyroid drugs when given in first trimester of pregnancy have been implicated. Other workers however, are of the opinion that the elevated thyroid stimulating hormone (TSH) which is associated with thyroid disease is the offending culprit rather than the antithyroid drugs.[3,4]

It has also been documented that there is higher association between increased incidence of this congenital malformation and exposure to second-hand-smoke, high maternal zinc and coffee consumption.[5] Some workers have also found positive associations between the following pre-pregnancy exposures and choanal atresia: Intake in the highest quartile: Vitamin B-12, zinc, niacin; Intake in the lowest quartile: Methionine, vitamin D; Coffee (≥ 3 cups per day).[6]

The clinical presentation of a unilateral choanal atresia maybe delayed until adult life and the only pointer to the anomaly maybe a unilateral mucopurulent discharge. However, the bilateral type presents at birth with severe asphyxia which may lead to death. Since newborns are obligate nasal breathers, establishing an airway is an acute Otolaryngologic emergency.

We present a case of BCA in a two weeks old termed baby with review of the clinical/radiological features and management challenges in a resource-poor setting.

CASE REPORT

A two weeks old term female neonate admitted at the 12th hour of life with complaint of difficulty in breathing immediately after birth following delivery via emergency lower caesarean section done on account of severe oligohydramnios at estimated gestational age(EGA) of 41 weeks + 5days. Pregnancy was booked at 13 weeks. Mother is a known asthmatic who had several acute exacerbations which were managed on outpatient basis using salbutamol nebulizer and hydrocortisone. There

was no history suggestive of thyroid disease. There was however a history of premature rupture of membrane(PROM) noticed in the last trimester and continued till delivery. Mother is a 31 year old now P3+1 (3 alive) food vendor with tertiary level of education. Examination revealed a term female neonate in obvious respiratory distress, marked snotting and salivation, acyanosed, not dehydrated and no pedal oedema. Her weight was 2.89kg, length – 50cm, occipitofrontal circumference (OFC) – 33cm, chest circumference – 31.5cm. She was dyspnoeic (grunting with intercostal and subcostal recessions) with a respiratory rate of 46cycles per minute; breath sounds were vesicular with transmitted sounds, and $Sp0_2$ – 78-89% in ambient air. Heart rate was 146 beats per minutes, S_1& S_2 only were heard. She was conscious and active, anterior fontanelle was patent and normotensive, tone and reflexes were normal. Attempts to pass nasogastric (NG) tube met resistance at 6cm into the nasal cavity; however other findings in the digestive system were normal.

A working diagnosis of Esophageal atresia was made. Baby was placed on nil per oral(NPO), intravenous (IV) fluid, IV antibiotics, oropharyngeal airway introduced and investigations were requested for.

Full Blood Count, Electrolyte/Urea/Creatinine, Calcium, Phosphate were essentially normal. Chest x ray revealed normal sized heart with no active focal lung lesion. However, choanogram outlined only the nasal cavities (Fig, 1 & 2). Computed Tomography (CT) examination was done and it showed bilateral choanal atresia. The vomer is thickened and on the left, there is a fusion of the bony elements posteriorly. The contralateral side shows membrane occupying the posterior segment of the cavity (Fig 3).

Surgical procedure: Patient was laid in supine position, pre-oxygenated and intubated. The throat was then packed with wet gauze. Shoulder support was placed and head stabilized on a head-ring. Blair's head dressing applied and the rest of the body covered with sterile drape.

The nasal cavities were prepared using drops of 0.5% Xylometazoline to decongest the nose which was later sucked with fine nasal catheter. The atresia was approached via the transnasal route. Lister's urethral dilator was introduced through the nasal cavity while the surgeon's left index finger was placed in the choana at the nasopharynx as a guide to the urethral dilator. The right atresia was gently punctured with size 3 -6 which was later dilated to size 6-9. The same procedure was done for the left side. Thereafter size 14 FG NG tube was fashioned and passed as stent and secured in both nasal cavities. The patient did well postoperatively and was discharged after one week. She was brought back after 6 weeks for re-dilation which was very successful and a bigger size 16 FG NG tube was left in-situ for another 8 weeks. Patient is doing well but has occasional partial nasal obstruction which is being managed conservatively at the Ear, Nose and Throat clinic.

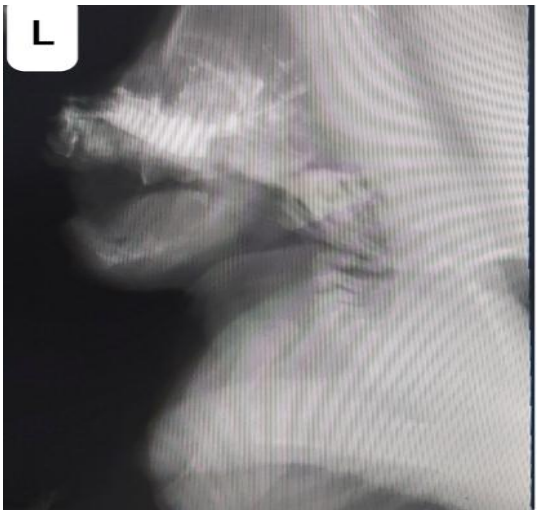

Figure 1: Choanogram showing contrast restricted to the nasal cavity.

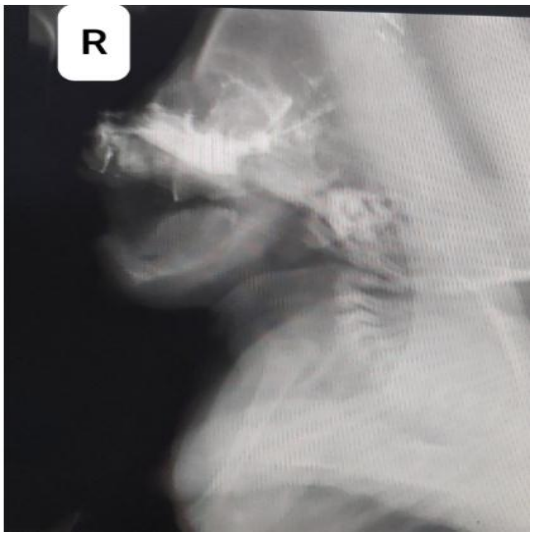

Figure 2: Choanogram showing contrast restricted to the nasal cavity.

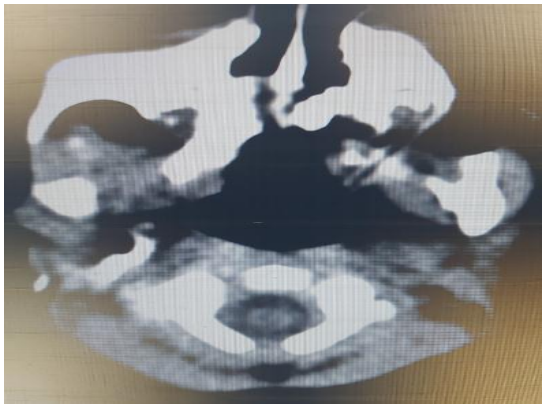

Figure 3: Axial CT image at the level of the nasopharynx showing bilateral choanal atresia with membranous type on the right and bony type on the left side.

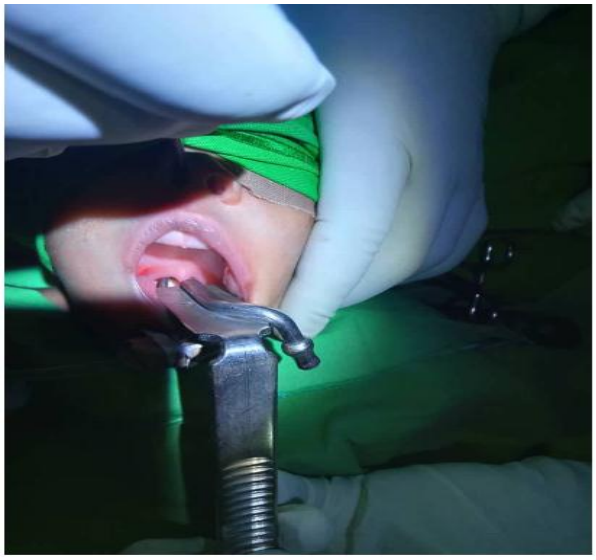

Figure 4: Tongue blade used to expose the oral cavity and oropharynx.

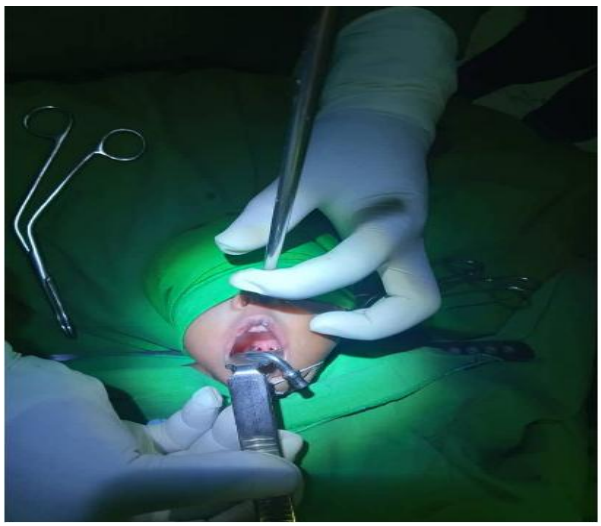

Figure 5: Lister's urethral dilator in the left nasal cavity with tongue blade exposing the oropharynx.

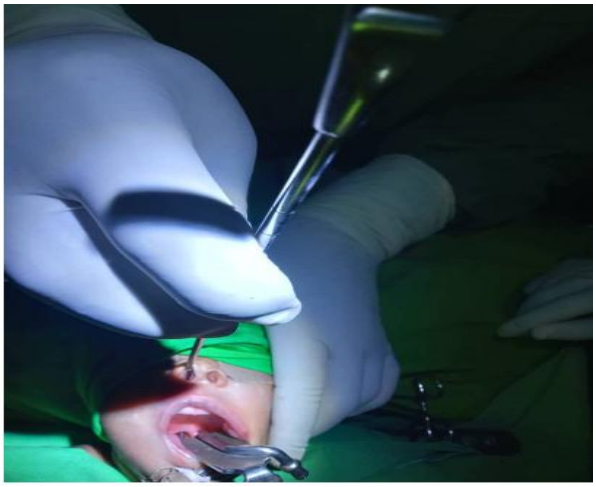

Figure 6: Lister's urethral dilator in the right nasal cavity with tongue blade exposing the oropharynx.

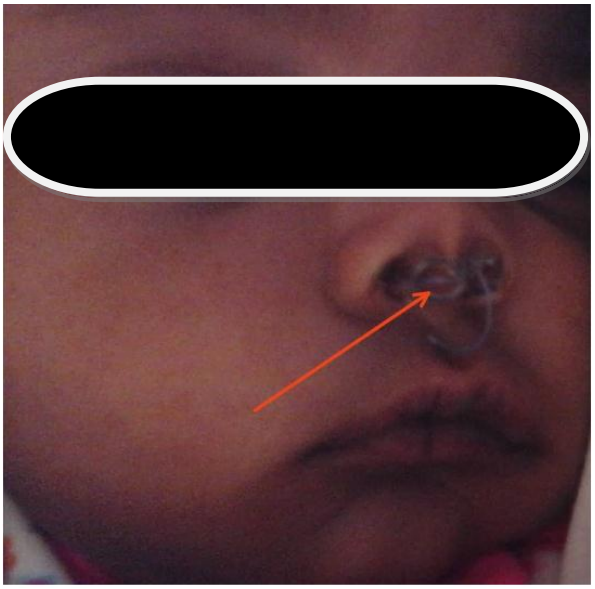

Figure 7: Post-operative Stent in-situ anchored to the columella.

DISCUSSION

Clinical features associated with Choanal atresia are many and varies in onset and severity depending on whether the lesion is unilateral or bilateral. These features may also depend on other co-existing congenital anomalies.[7] Classically, bilateral choanal atresia presents with acute respiratory difficulty which may result in cyanosis. This respiratory difficulty with the cyanosis maybe relieved when the child is crying only to return back when the child is at rest; this is termed paradoxical cyanosis.[7,8] Also, because of the inability to breath, the patient may present with choking episodes during feeding. Our patient presented to us with respiratory distress, though acyanotic.

Embryological studies showed that the nasal choanae develop between the 3rd and 7th week of intrauterine life. During this gestational period, there is a rupture or resorption of the vertical epithelial fold between the roof of the choanae, primary oral cavity and the olfactory groove. To explain the anomaly, many theories have been proposed. These include: the persistence of nasobuccal membrane, the persistence of buccopharyngeal membrane, the incomplete resorption of the nasopharygeal mesoderm, and the local misdirection of neural crest.[9]

Choanal Atresia maybe classified on the basis of the cause of the atresia into bony, mixed bony/membranous and purely membranous. Recent findings suggest that mixed bony/membranous are more common, occurring about 70% of the time.[10] The index case is purely membranous on the right side and bony on the left.

The anomaly has no racial predilection and maternal age or parity does not increase the frequency of occurrence. Bilateral cases occur less frequently than unilateral ones and females are more commonly affected than males.[2,11]

Our patient is a termed female and presented with bilateral atresia.

Patients with Choanal atresia may present with other defects including syndromes. These defects/syndromes include CHARGE (coloboma, heart disease, choanal atresia, retardation, genital hypoplasia, and ear anomalies) syndrome, Teacher-Collins syndrome, Crouzon syndrome and Marshall-Smith syndromes.[6,7] None of this defect/syndrome was seen in the index case.

In the clinical evaluation of choanal atresia, it is important to do a thorough physical examination in order not only to establish the diagnosis but also to rule out other associated congenital anomalies. As in the case under review, the lesion maybe suspected when there is failure to pass an 8F catheter through the nasal cavity more than 5.5 cm from the rim of the alar nasi. Other tests that may also point to the diagnosis include negative nasal patency tests demonstrated by lack of movement of a thin wisp of cotton wool held close to the alar nasi while the mouth is closed, the absence of fog on a metallic tongue blade when it is placed under the nose, or administering drops of a colored solution into the nose that is visible in the oropharynx.[11,12]

Radiological evaluation of a patient with choanal atresia involves the use of choanography and Computerize Tomographic Scan (CT scan). These two modalities were employed in the diagnostic workup for this patient. Choanography may only outline the nasal cavity but CT scan which is regarded as the modality of choice will clearly delineate the full anatomic abnormalities of choanal atresia. Indeed, CT scan plays a significant role not only in the diagnosis but also in the therapeutic approach in the management of this lesion.

Patient with bilateral choanal atresia as in this case under review, usually presents with respiratory difficulty and the management involves immediate passage of oropharyngeal airway, orotracheal intubation or tracheostomy. However, the definitive treatment is nasal recanalization through transnasal, transpalatal, transeptal, external rhinoplasty or endoscopic technique route.[13,14]

The choice of the procedure depends on the age of the patient, whether the atresia is unilateral or bilateral, bony or membranous, size of nasopharynx, thickness of the atresia and the need of post-operative stenting.[14]

In our index case, the transnasal approach was adopted not only because the procedure is simple and easy to carry out but also due to lack of endoscopic instruments in our facility. Though transnasal approach is simple and easy to do, it may become difficult when there is the presence of septal deviation, turbinate hypertrophy and nasal discharge. None of these problems was encountered in our patient.

The transpalatal approach is preferred in unilateral choanal atresia. The transpalatal approach is more popular because it provides good access to the operation field. However, this approach is associated with longer operation time, greater blood loss and longer period of recovery from the procedure. Apart from this, the procedure can be complicated by abnormal palatal growth, palatal fistula and further orthodontic problems, like cross bite. The index case has bilateral atresia, so this approach was not considered.

The transseptal approach is impractical in neonates like our index case while rhinoplasty and endoscopic techniques were not considered because of lack of such instruments in our Centre.

REFERENCES

1. Andaloro C, La Mantia I. Choanal Atresia. *StatPearls*. 2019 Jan. https://www.ncbi.nlm.nih.gov/books/NBK507724/
2. Ted L T, Yaser AA, Abdulrahman A H. Choanal Atresia. https://emedicine.medscape.com/article/872409-overview
3. John HL. Antithyroid Drug Treatment in Pregnancy. *The Journal of Clinical Endocrinology & Metabolism*, 2012; 97(7): 2289-2291.
4. Barbero P, Valdez R, Rodríguez H, Tiscornia C, Mansilla E, Allons A, et al. Choanal atresia associated with maternal hyperthyroidism treated with methimazole: a case-control study. *Am J MedGenet A*., 2008; 146A(18): 2390-5.
5. Miller EA, Manning SE, Rasmussen SA, Reefhuis J, Honein MA. National Birth Defects Prevention Study. Paediatr Perinat Epidemiol, 2009; 23(1): 9-17.
6. Vijaya K, Paul A. R, Lixian S, John C. C, Trudy L. B, Anna M S, et al. Descriptive and risk factor Analysis for choanal atresia: The National Birth Defects Prevention Study 7–2007. Eur J Med Genet, 2014; 57(5): 220–22.
7. Andrew T B, Howard S, Alessandro A, Lisa J M, Robin TC, Robert J. H. Characterization of Congenital Anomalies in Individuals With Choanal Atresia. Arch Otolaryngol Head Neck Surg, 2009; 135(6): 543-547.
8. Paraya A, Choakchai M. Choanal Atresia. J Med Assoc Thai., 2009; 92(5): 699-706.
9. NishimuraY. Embryological study of nasal cavity development in human embryos with reference to congenital nostril atresia. Acta Anat (Basel), 1993; 147: 140–144.
10. Khaled A, Alsaid L. Role of Multislice Computed Tomography and Local Contrast in the Diagnosis and Characterization of Choanal Atresia. https://www.hindawi.com/journals/ijpedi/2011/280763/
11. Gupta M, Kour C .Congenital Bilateral Choanal Atresia: A Rare Case. J Rare Disord Diagn Ther., 2017; 3: 9. doi: 10.21767/2380-7245.100162

12. Kwong KM. Current Updates on Choanal Atresia. *Front Pediatr*, 2015; 3: 52.
13. Newman JR, Harmon P, Shirley WP, Hill JS, Woolley AL, Wiatrak BJ. Operative management of choanal atresia: a 15-year experience. *JAMA Otolaryngol Head Neck Surg*, 2013; 139(1): 71-5.
14. Moreddu E, Rizzi M, Adil E, et al. International Pediatric Otolaryngology Group (IPOG) consensus recommendations: Diagnosis, pre-operative, operative and post-operative pediatric choanal atresia care. *Int J Pediatr Otorhinolaryngol*, 2019; 123: 151-5.