

Case Report

A Rare Case Report of Bilateral Choanal Atresia in an Adult at BMC Tanzania

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Abstract: Bilateral choanal atresia is uncommon in our environment. CA is caused by the failure of resorption of the bucco-nasal membrane during embryonic development. This occurs in the posterior nasal cavity whereby they will be complete obturation of the choanal. BCA presents with breathing problems immediately after birth this calls for emergency care. For our case we have 18 years old female presented in our department with complains of difficulty in breathing, persistence nasal discharge, nasal obstruction, mouth breathing, and history of on and off visiting to the hospital with the same complains. Also reports to be taken to the hospital twice and got some improvement for few days but can not recall what was done. Surgical intervention is needed immediately to help such patient as soon as the diagnosis is laid down.

Keywords: Bilateral Choanal Atresia bucco-nasal.

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INTRODUCTION

Choanal atresia in an adult is a rare congenital abnormality of the posterior nasal cavity. Congenital choanal atresia is due to the embryological failure of the primitive bucconasal membrane to rupture before birth. This results in the persistence of a bony plate (90%), membrane, or both, obstructing the posterior nares [1]. The condition may be unilateral (most commonly) or bilateral. It occurs in about 1 in 7000 births, and there is a family tendency [2]. Its frequent unilateral and right sided than bilateral [3].

Bilateral choanal atresia presents as an emergency at birth. The newborn is a near-obligate nasal breather and the nasal obstruction will therefore produce difficulty in breathing. The alae nasi dilate and the accessory muscles of respiration are used to no avail. There is pallor and cyanosis until the mouth is opened and after a few quick breaths are taken the infant cries. This sequence of events continues. The diagnosis should be suspected immediately an oral airway inserted to assist respiration. Feeding via an orogastric tube. Surgery is the definitive mode of treatment

CASE REPORT

- A 18 yrs old female presented at our ENT department with the following complains of difficulty in breathing, nasal obstruction, nasal discharge,

persistent mouth breathing, soiling and inability to recognise smell. All complains were present since birth. When the patient was a baby had difficulties in breast feeding whereby was fed using a spoon. Mode of delivery was SVD and birth weight was 3kg. Since then had severe DIB, persistent with episodes of apnea, and blue discoloration of lips, palms, and tongue: worsened by breast feeding relieved by cry. This was also associated with episodes of convulsion and loss of consciousness. History of been rushed to neonatal ICU and admitted for 6 months.

- On examination had good nutrition status, adenoid face, breathing through mouth with dry oral mucosa. Not pale, not jaundice and had no finger clubbing; also no lymphadenopathy. Vitals were within the normal range and physical examination was normal. On ENT examination anterior rhinoscopy revealed nasal discharge were from both nasal cavities interchanging between thick purulent, serous and mucoid. Ears and oral cavity plus the throat were normal. Basic and special investigations were both done. FBP, LFT, RFT were normal. Endoscopic examination showed both sides were obstructed. As shown in fig 2. Investigation done radiologically was CT-scan and it was as shown below.



Fig-1: CT-scan (axial view) illustrating bilateral choanal atresia which is bony and membranous occlusion

Procedure

We perforated the bone using a perforator. Below is the endoscopy view before the procedure and after the procedure.

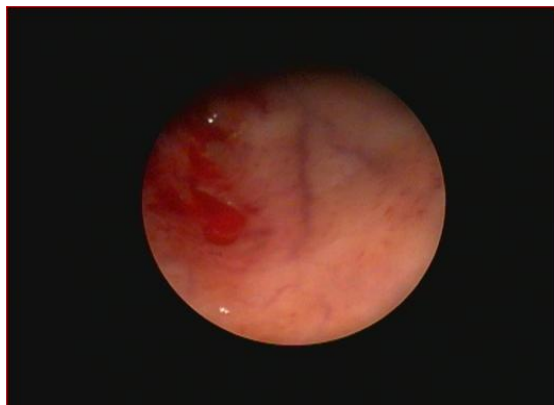


Fig-2: Endoscopic view of Right posterior choanal before release

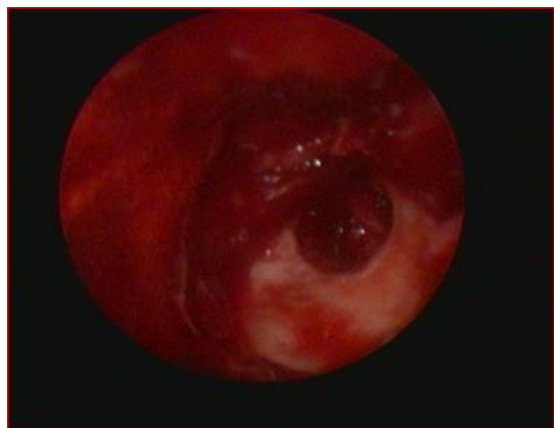


Fig-3: Endoscopic view of Right posterior choanal after release

DISCUSSION

Congenital choanal atresia is due to the embryological failure of the primitive buconasal membrane to rupture before birth. This results in the persistence of a bony plate (90%), membrane, or both,

obstructing the posterior nares. The condition may be unilateral (most commonly) or bilateral. It occurs in about 1 in 7000 births, and there is a family tendency. Male to female ratio of occurrence is 1:2 [1].

The mouth, palate, nose and paranasal sinuses all develop from the cranial portion of the primitive foregut. The nose begins as two epithelial thickenings known as the nasal placodes, which appear above the stomatodeum about the fourth week in utero. The placodes deepen to form olfactory pits which lie between the medial and lateral nasal processes. The medial processes fuse to form the frontonasal process. This is compressed to form the nasal septum as the lateral nasal processes approach each other. The nasal septum will then grow posteriorly to divide the two nasal cavities. Each nasal cavity is closed posteriorly by the thinned out posterior wall of the nasal sac, called the buconasal membrane. This usually breaks down around the sixth week in utero. Its persistence is thought to be the cause of choanal atresia. Although some cases are familial suggesting monogenic inheritance, the majority are sporadic [6].

Bilateral choanal atresia presents as an emergency at birth. The newborn is a near-obligate nasal breather and the nasal obstruction will therefore produce difficulty in breathing [10]. The alae nasi dilate and the accessory muscles of respiration are used to no avail. There is pallor and cyanosis until the mouth is opened and after a few quick breaths are taken the infant cries [11]. This sequence of events continues. The diagnosis should be suspected immediately and an oral airway inserted to assist respiration. About half of infants with choanal atresia display other abnormalities, including the CHARGE association. This refers to a problem of multiple congenital anomalies: C, colobama; H, heart disease; A, atresia choanae; R, retarded growth and development; G, genital anomalies; E, ear abnormalities and deafness (4 & 5). In addition, half exhibit facial nerve palsies and one third of cases have laryngotracheal anomalies [12].

The diagnosis can be confirmed by the mirror test or by attempting to pass a catheter through the nose into the nasopharynx. This will not be possible if there is an obstruction. Fibreoptic endoscopy will confirm the diagnosis. If there is still doubt the lesion can be demonstrated radiologically. A CT scan is the method of choice to delineate the nature and thickness of the obstruction [6].

Cases of unilateral atresia can initially be observed without treatment. In the case of the newborn with bilateral atresia the first priority is to insert and maintain an oral airway. The treatment of choanal atresia is surgical [7]. The challenge is to provide a nasal airway which has an adequate mucosal lining, and to prevent granulation tissue formation and subsequent stenosis.

Two approaches are in common use.

1. Transnasal. This is the usual approach in infants. A membranous occlusion may require no more than perforation with a probe. This can also be accomplished with electrocautery or laser. In the more common bony occlusions it will be necessary to perform a trephine and remove the obstruction. A stent should be inserted and a series of dilations of the choana will then be required to maintain an adequate lumen.
2. Transpalatal. This is preferred by some surgeons, particularly when the atresia is unilateral or if a previous transnasal opening has later closed. The palate is incised just in front of the posterior edge of the hard palate. The soft palate is retracted, and the occlusion removed together with part of the vomer and border of the hard palate.

Follow-up and aftercare

Maintenance of the opening following corrective surgery with regular bouginage is now mostly preferred to the use of indwelling tubes. Dilatation will probably be necessary every 2 months initially, but this period can be extended as the child grows.

Repair of choanal atresia has evolved significantly over the ages. Open trans-palatal techniques has led to minimal invasive endoscopic repair [8]. Novel used application of steroid in Eluting sinus stent in minimizing scarring following choanal atresia repair by endoscopy which results with high success [9].

CONCLUSION

Bilateral congenital choanal atresia is not a common disorder once occur requires a medical, surgical emergency. It should be suspected to neonates with cyclic cyanosis CT-scan of the nasal cavity and nasopharynx confirms the diagnosis and surgical approach to be used as it gives the location, type and thickness of atresia also it helps the surgeon to establish a plan for the associated abnormalities like high arched hard palate, thickened vomer, septum and lateral nasal structures, which further reduce their diameter of nasal fossa and the nasopharynx.

RECOMMENDATIONS

Midwives should be sensitized so as to have High Index of Suspicion.

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