Congenital bilateral choanal atresia- Endonasal endoscopic surgery- Report of two cases

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Abstract

Congenital choanal atresia (CCA) is the development failure of the nasal cavity to communicate with nasopharynx. It is an uncommon congenital anomaly of nose with an incidence of approximately 1 in 5000-7000 live births. Choanal atresia is caused by failure of resorption of the nasobuccal membrane during embryonic development. Choanal atresia has a significant association with CHARGE syndrome. Surgical intervention is recommended in the first weeks of life in bilateral cases because this is a life threatening condition. Two cases of congenital bilateral choanla atresia (CCA) was admitted in the department of Otolaryngology & Head-Neck Surgery at Bangabandhu Sheikh Mujib Medical University (BSMMU) with the complaints of intermittent attacks of cyanosis, dyspnea, respiratory distress and history of inability to take feeds. On examination both babies had respiratory distress, mucoid discharge from both nasal cavities. A soft rubber catheter was passed into both nares revealed the diagnosis of bilateral CCA which was confirmed by flexible nasoendoscopy. Bilateral CCA was managed with a nasopharyngeal airway. Surgery is the definitive treatment with two main approaches namely transnasal and transpalatal. We discuss successful management of two neonates with bilateral CCA by endonasal endoscopic approach.

Keywords: choanal atresia, congenital;

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Introduction

Congenital choanal atresia (CCA) is a rare malformation that causes airway obstruction in newborns and infants, with an incidence of 1 in 5000-7000 births. It seems to occur more commonly in females than males and ratio is 2:1 and is frequently unilateral and right–sided than bilateral.[1] The atresia maybe classified as bony, mixed bony and membranous or purely membranous although the latter is rare. Nature of obstructing atretic plate has often been described as 90% bony and 10% membranous. Most of the cases of CCA are isolated malformations, but association with other congenital deformities as in CHARGE association which includes coloboma of the eyelid, heart disease retarded growth, genital hypoplasia, and ear anomalies.[2] Bilateral CCA is a medical emergency, because maintain an airway and relieving the obstruction is a priority.[3] Increased cyanosis and death may occur if appropriate treatments are not available. The immediate management of neonates presenting with intermittent cyanosis is the insertion of an oral airway and feeding via an oro-gastric tube.[2,3] There are numerous methods for correcting this condition, commonly used methods are the transnasal, transpalatal, transeptal approach[4] and the endoscopic transnasal approach.[5]

Case report

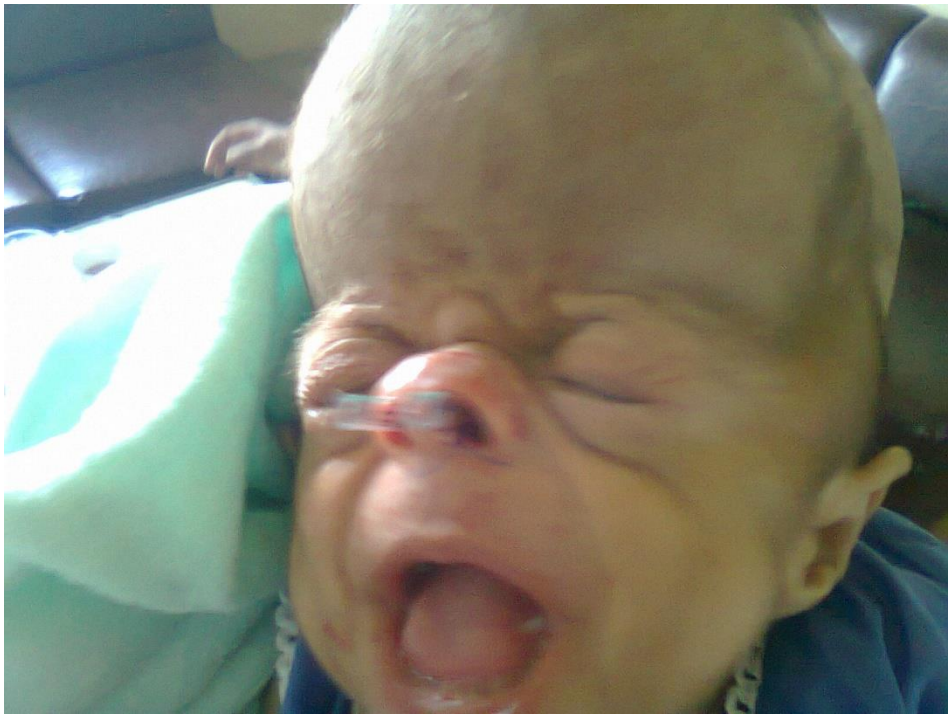
Two male babies of bilateral CCA, one was 3.1 kg and other was 2.8 kg were admitted in the Department of Otolaryngology & Head-Neck Surgery at Bangabandhu Sheikh Mujib Medical University with the complaints of intermittent attacks of cyanosis, dyspnoea and inability to take feeds. On examination both babies had respiratory distress, cyanosis and bilateral mucoid nasal discharge. One of them had hydrocephalous and another one had syndectile. Soft rubber catheter was passed into the both anterior nares which could not cross the posterior nares. Flexible nasoendoscope showed atresia of posterior nares.

An oropharyneal airway was put and fixed with tape in their mouth to relieve respiratory distress and intravenous fluid and antibiotic was started. Auscultation revealed bilateral crepitations and spasm in the chest in one baby. Heart sound was normal in both babies. Chest X-ray of one baby showed bilateral consolidation.

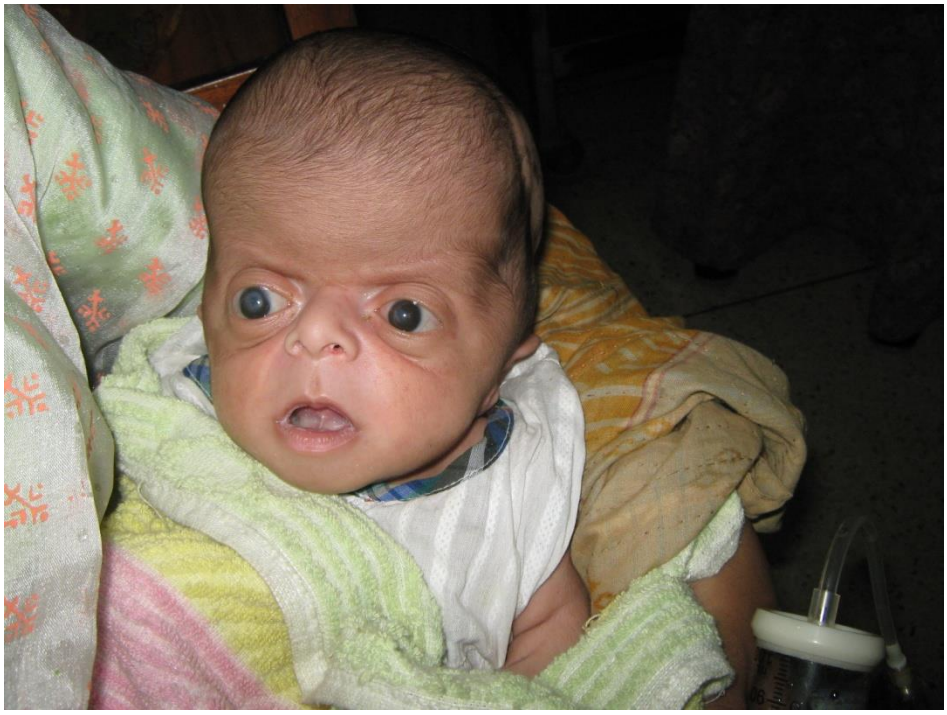
Iintermittent cyanosis was present which was disappearing on crying. A size 5 suction tube was passed into the both nasal cavity but failed to puss through the nares into the pharynx; flexible nesoendoscopy reveals atresia of posterior chonea of both babies.

Both patient was operated by endoscopic transnasal approach under general anaesthesia. Bone plate was perforated with gradual cervical dilators and suction tube. Stenting was done by 4 mm portex endotracheal tube to maintain the patency of newly formed lumen. A endotracheal tube fixed by stitching anteriorly. Both babies remained stable postoperatively and were discharged from hospital on 6th and 7th postoperative days. Both of them where on breast feeding at the time of discharge and advised weekly follow up. Stent was removed after 8 weeks. Both patients were alright with normal feeding and satisfactory weight gain until last follow up at 7 months and 6 months of age respectively.

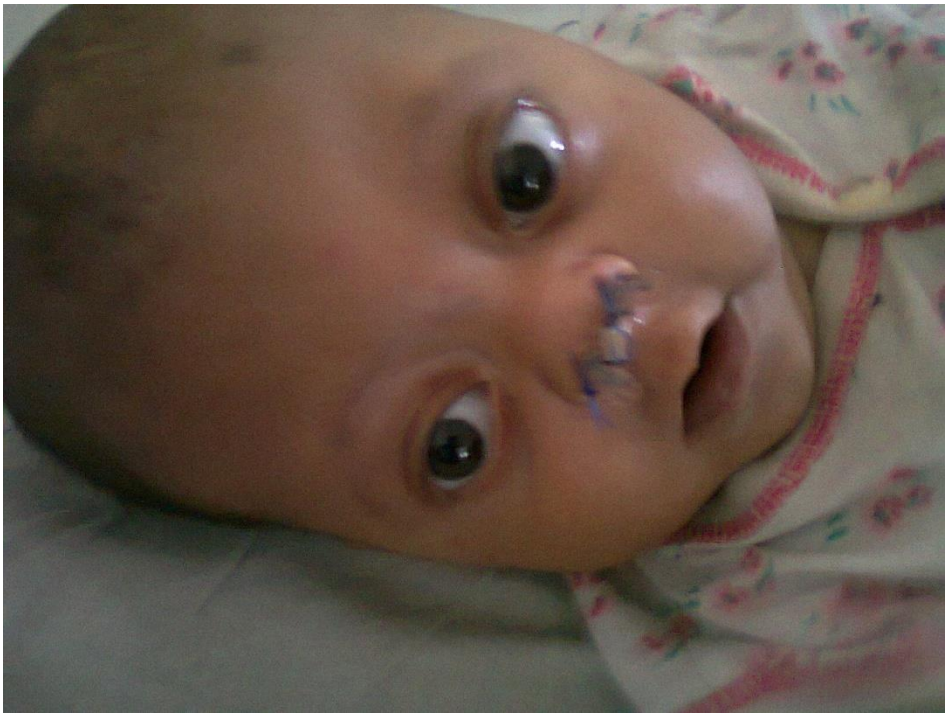
Photographs



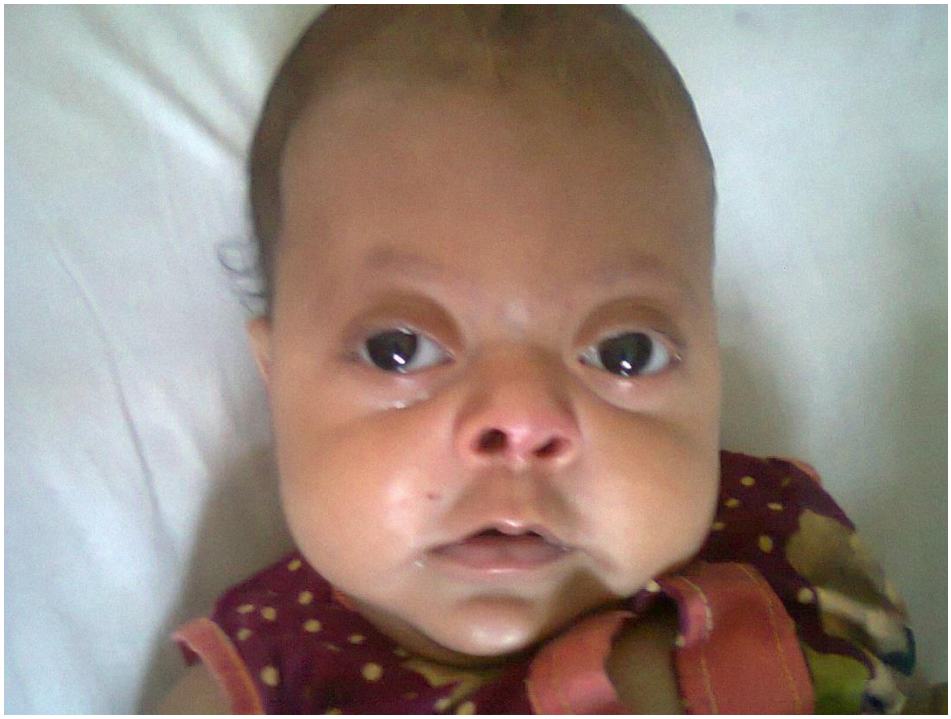
Stents in situ



After removal of stents



Stents in situ



After removal of stents

Discussion

Congenital choanal atresia is a disease of nasal airway where no connection exists between the nasal cavity and the aerodigestic tract. It was first described by Johann Roderer in 1755.[3] Current theories of choanal atresia is failed oronasal membrane rupture or abnormal migration of neural crest cells into the nasal vault.[5,6]

Bilateral chonal atresia will as an acute respiratory emergency at birth as newborns are obligate nasal breathers.[7] Symptoms of airway obstructive and cyclical cyanosis are the classical signs of newnatal bilateral atresia. Choanal atresia can be unilateral or bilateral. Bilateral choanal atresia presents very early in the life. Most patients with bilateral CCA are detected within the first month of life.

There are numerous ways to diagnosis choanal atresia. The simplest method is to pass soft rubber catheter into the nares. There is no air entry by cold spatula test. Radio opaque oil can be instilled into the nose and lateral radiograph showed the site of atresia. Flexible endoscopy, choanography and CT scan can help in diagnosis.

The surgical treatment of congenital atresia is challenging within the realm of paediatric otolaryngology, clinical evaluation should include a complete physical examination to look for others congenital anomalies. Numerous aproaches for operations are Trans nasal, transpalatal, endoscopic transnasal Co_2 laser resection.

Approach depend upon the age of the patient, size of the nasopharynx, thickness of atresia, bilateral vs unilateral use of postoperative stenting.[8] We performed endoscopic transnasal approaches. Advantages of endoscopic approach are faster and easier, minimum blood loss, can performed in all ages, child can be

immediately breast feed, less restenosis and discharge may be 3rd or 4th postoperative day, successful in upto 80% of the cases.[9] Postoperative close follow revealed that both of them remained in stable condition and breast fed without difficulty and gaining satisfactory weight.

Conclusion

In bilateral congenital choanal atresia (CCA) can be managed perfectly and precisely by endonasal endoscopic approach.

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