AN EXPERIENCE WITH CHOANAL ATRESIA

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INTRODUCTION

Choana is derived from a Greek work meaning a funnel. The choanae are, by definition, the posterior apertures of the nose. Roederer first described atresia of the choanae in 1775 [1]. It is one of the more commonly observed congenital anomalies of the nose. The average rate of choanal atresia is .82 cases per 10,000 live births. Unilateral atresia occurs more frequently on the right side and rates of unilateral to bilateral is 2:1. A slightly increased risk exists in twins. It occurs in equal frequency in people of all races and females are affected more than males. A number of theories have been proposed to explain the occurrence of choanal atresia which are persistence of buccopharyngeal membrane, failure of bucconasal membrane of Hochstetter to rupture, medial outgrowth of vertical and horizontal processes of palatine bone, abnormal mesodermal adhesions forming in the choanal area and misdirection of mesodermal floor due to local factors [2]. In bilateral choanal atresia cases, complete nasal obstruction in newborn may cause death due to asphyxia [3].

Despite rarity of this condition, four cases of choanal atresia presented in CMH Rawalpindi in the last one and a half year. Two cases had bilateral disease while two had unilateral disease.

BRIEF ANATOMY OF CHOANAE

The choanae or posterior nasal apertures are divided medially by free posterior edge of vomer. Their roof is formed by the body of sphenoid with the overlapping, flared alae of the vomer and the vaginal process of medial pterygoid plate, and the floor by the free posterior edge of the horizontal plate of the palatine bone (fig. 1). During embryonic life choanae are closed by bucconasal membrane. This membrane lies slightly more anterior to the eventual position of choanae, which do not become established until the third month of intrauterine life [4].

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CASE REPORTS

Case No. 1

A 5 days old baby girl was referred as a case of bilateral choanal atresia. Soon after birth, the baby became blue and unable to breath. She was resuscitated at a hospital in Mirpur and then sent to CMH Rawalpindi with oral airway in mouth. At CMH Rawalpindi, she was admitted in the neonatal intensive care unit under supervision of pediatrician. Examination of child showed no airflow through the nares and even the smallest NG tube could not be passed through the nostrils. No other congenital abnormality could be found either by pediatrician or by otolaryngologist. The patient was given Oxygen inhalations and intravenous fluids. Antibiotics were started as the child was running fever. Relevant investigations like Blood Complete picture, urine routine examination, PT, PTTK, serum bilirubin were carried out and found normal. Arterial blood gases showed reduced PO2. CT scan showed bilateral choanal atresia with thin bony atresia plate on both sides (fig. 2).

Surgical intervention was planned as an emergency. She was built up and surgery was carried out at 7 days of age. Complete general anaesthesia with endotracheal intubation was given by an experienced anaesthetist. It was decided to approach and break the atresia plate by trans-nasal route, while keeping the trans-palatel approach in reserve. A mouth gag was applied to have a clear visualization of the throat. Sharp probe (fig. 3) was then introduced into the left nostril and force was applied to break the posterior choanae. When a give way was achieved, female uretheral dilators (fig. 3) were introduced through the opening to make them wider.

Plastic tubing was then introduced through the left nostril and pulled out through the mouth. The same procedure was then repeated for the right nostril (fig. 4).

The nasopharyngeal ends of the tubes were stitched together and tubes were pulled anteriorly

till the stitch holding them together engaged in the posterior edge of septum (fig. 5).

Anteriorly the tubes were then stitched to each other in front of columella (fig. 6).

Postoperatively the child was no longer cyanosed. Child was handed over to pediatrician for general care who started formula milk orally. As the child was very weak and emaciated, she was kept in incubator on intravenous nutrition. Child was discharged after a week. Regular suction and cleaning of the tubes were carried out in out-door, sometimes after instillation of saline. The tubes were removed after three months.

The child reported for follow up monthly during the first year of life (fig. 7). Regular suction and periodic dilatations were done at regular intervals. Patency of her nasal passages was reasonably adequate till one year of age, which was her last follow up visit. She was advised to have further follow-ups at local hospital in Mirpur.

Case No. 2

A 3 days old male child was admitted in CMH Rawalpindi by the pediatrician. He had a history of becoming cyanosed soon after birth. However when the child opened his mouth while crying, the cyanosis disappeared (Paradoxical Cyanosis). An oral airway was put which relieved the cyanosis. However the cyanosis reappeared when the airway was removed. On ENT examination, it was not possible to pass butterfly canula through either nostril. X-ray lateral view of the face taken after instillation of contrast in the nostrils confirmed bilateral Choanal atresia. Examination by pediatrician revealed no other congenital abnormality.

An emergency surgery under GA was planned as before. Patency of nasal airway was established by trans-nasal approach as in the previous case. Tubes were secured in either nostril as in previous case. Child made smooth post-operative recovery and was discharged on 7th postoperative day. Regular follow-ups were done and the tubes removed after 03 months. Periodic dilatation of nasal passages was done when required. At the last follow-up, child had grown above one year of age and was a nose breather.

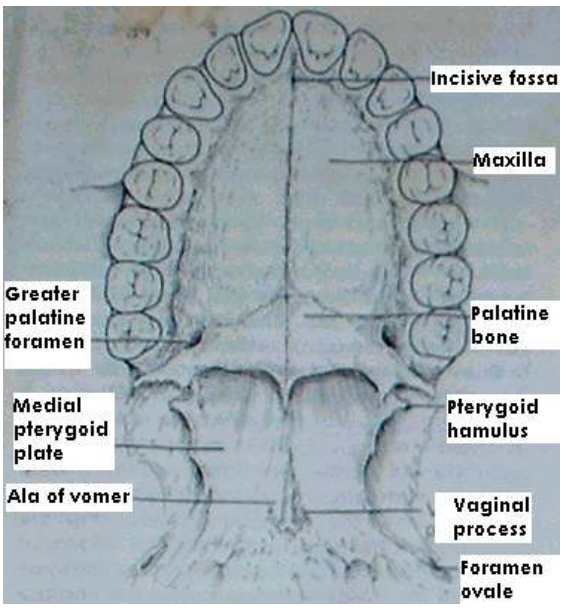

Fig. 1: The palate and posterior nasal apertures seen from below

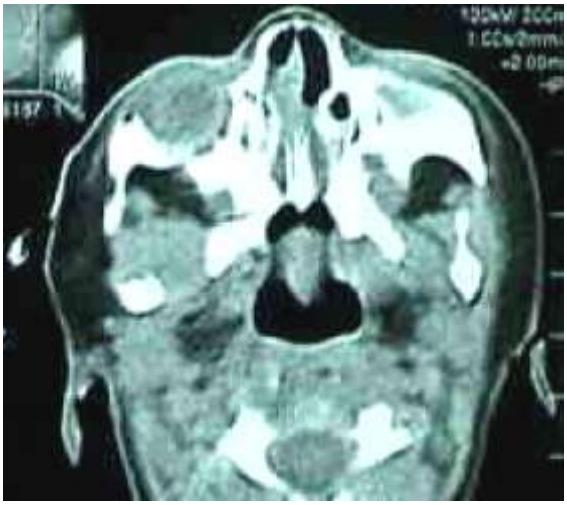

Fig. 2: Thin bony atresia plate seen on CT scan

Fig. 3: Artesia plate was broken by sharp probe (top). It was then dilated by female urethral dilators.

Case No. 3

A one year old child was brought to CMH Rawalpindi with a history of right sided nasal discharge since birth. On inspecting the nasal cavity after suction of nasal discharge, there was no evidence of foreign body in either nostril. Inability to pass small NG tube through the right nostril confirmed choanal atresia. CT Scan revealed a thin bony atresia plate on the right side. Examination by pediatrician revealed no other congenital abnormality. An elective surgery was planned under GA. Atresia plate on the right side was broken trans-nasally as in the previous cases. A tube was passed through the right nostril into the naso-pharynx and secured into place by applying a stitch at the columella. The child was discharged on the 3rd post-operative day. The tube was periodically inspected and cleaned. It was finally removed after 03 months. Following that regular follow-ups are continuing at monthly intervals. Child has grown to more than six months of age and air flow through right nostril is satisfactory.

Case No. 4

This case was different from the previous cases in the sense that the atresia was membranous. It was complete on one side and partial on the other. It was a part of generalized symptom complex in a three months old child with Apert Syndrome. Other features of the syndrome including lobster claw, syndactyly of feet, proptosis due to shallow orbit and high arched palate were present (fig. 8).

Partial nasal patency on one side had enabled the child to grow up to three months of age without any treatment. However occasional cyanotic episodes continued specially during breast-feeding. The diagnosis was confirmed as in previous cases. The child was also seen by pediatrician and investigated thoroughly to look for any other associated abnormality as may be present in Apert Syndrome. However not other abnormality except those written above could be found. Surgery was carried out as in previous cases (fig. 9) and the child was discharged on the second post-operative day. Child has visited ENT department twice since then. Tubes are still in place with plan to be removed at six months age. He is also under treatment of surgeon for skeletal abnormalities.

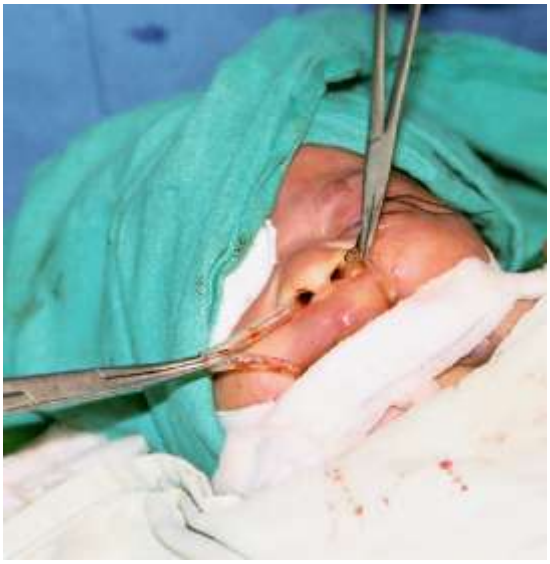

Fig. 4: Note the tubes passed through nose and brought out through mouth.

Fig. 5: The tubes pulled through mouth so that their posterior mutually stitched ends engage the septum.

Fig. 6: The tubes secured into place and anchored to each other in front of columella.

DISCUSSION

The essential aim of treatment of choanal atresia is creation of patent nasal passages. In the case of unilateral atresia, there is seldom any emergency at presentation and surgery can be undertaken as a planned cold procedure. Bilateral atresia however, always presents as a respiratory emergency and has to be dealt with as an emergency [5].

The treatment can be divided into emergent and definitive categories. Bilateral choanal atresia is best treated by inserting an oral airway to break the seal formed by the tongue pressing against the palate. This oral airway will be tolerated for several weeks. The method of repair is controversial with no technique having gained universal acceptance. Surgical procedures to correct choanal atresia can be broadly classified into transnasal and transpalatel approaches. However the decision rests on the surgeon's assessment of the choanal anatomy, composition and thickness of atretic plate, the depth and shape of the nasopharynx and presence of other anomalies [6]. The transnasal approach requires less time, causes slightly less morbidity and is best for thin membranous atresia in older children, however it does not allow sufficient exposure for extensive bone removal [7]. The thickened vomer and lateral pterygoid plates cannot be approached, as it is a blind procedure. A shallow nasopharynx is a relative contraindication to this approach. The transpalatal approach provides better exposure and more accurate bone removal. However there is increased operative time and blood loss but risk of major vascular injury, intracranial complications and chance of re-stenosis is reduced. The transpalatel approach is the best for thick bony atresia, bilateral atresia in neonates and in cases where there are anomalies affecting the anterior nasal cavities or nasopharynx.

Postoperative care includes regular suction of the tubes after instilling sterile saline drops. The nasal stents/tubes are usually removed after 6 – 12 weeks after the procedure and choanae inspected endoscopically to assess the healing. Periodic dilatation may be required if posterior choanae begins to restenose again.

Hamad A.M. [8] reviewed 30 cases of choanal atresia out of which three patients had other congenital abnormalities like Apert syndrome and Down syndrome. The atresia was unilateral in 16 and bilateral in 14 cases. 24

Fig. 7: The same child at six months of age.

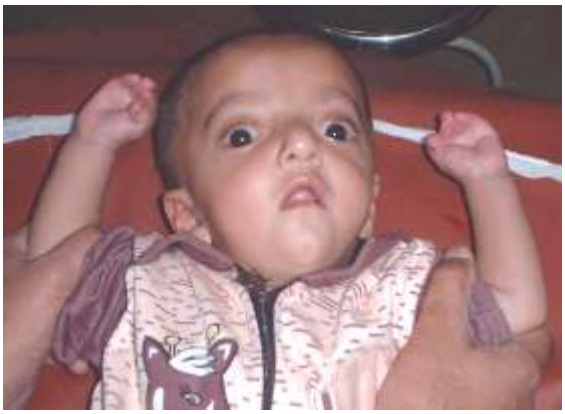

Fig. 8: Choanal atresia as a part of symptom complex in apert syndrome. Note the lobster hands, bilateral proptosis and mouth breathing due to choanal atresia,

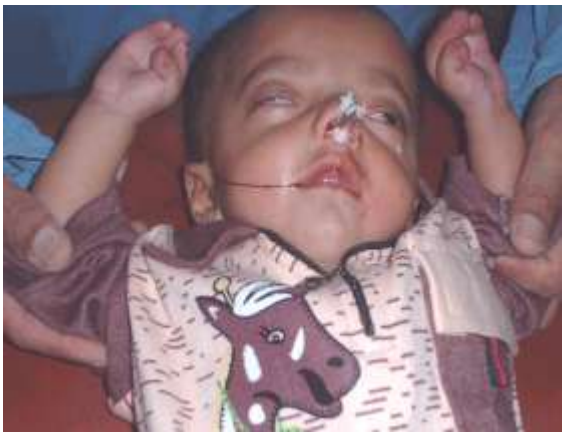

Fig. 9: Same child after surgical correction of choanal atresia. Note the tubes coming out of both anterior nares.

patients were operated by trans-nasal approach while 6 underwent transpalatel correction. Out of the first group that had underwent trans-nasal correction of defect, nasal patency remained adequate in 14 patients while 10 developed restenosis that required revision surgery.

These results are quiet similar to those observed in our four cases. Two cases out of the four (50 %) had unilateral choanal atresia. One case (25 %) had association with other congenital abnormality i.e. Apert syndrome. All of our four reported case had relatively thin atresia plate that could be broken by trans-nasal approach. In all the four cases periodic dilatation was carried out without anaesthesia, and so far restenosis has not occurred in any of the four cases.

CONCLUSION

The following conclusions can be drawn from our experience with four cases of choanal atresia-

- The frequency of unilateral and bilateral choanal atresia is almost the same.
- A child with unilateral choanal atresia does not have any acute symptoms at birth and usually first seen by otolaryngologist between few months to one year of age. On the contrary bilateral choanal atresia presents as an acute emergency immediately after birth.
- Once the diagnosis is confirmed, oral airway is passed to secure airway. Radiological investigations including CT scan must then be carried out immediately to assess the thickness of atresia plate.
- In most cases atresia plate is thin enough to be broken through trans-nasal approach using a sharp probe and then dilated by female urethral dilators under general anaesthesia. This procedure can easily be performed within the first few days of life.
- Once the nasal patency has sufficiently been established, it is maintained by keeping plastic tubing inside the nasal cavities. These tubes are removed after 3 months. Regular follow-ups and periodic dilatations may be done whenever restenosis is feared.

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