Clinical retrospective analysis of 15 cases of choanal atresia — Our experience

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Objective: Choanal atresia is a rare congenital disorder due to failed recanalization of the nasal fossae during fetal development. This article focuses on our experience in dealing with choanal atresia and its management. Here we discuss the varied clinical symptoms that the patients presented with, the clinical tests and investigations that were specific in diagnosing this condition and surgical management of these cases with endoscopic transnasal choanaloplasty with stenting and follow up topical Mitomycin C application.

Material and methods: This is a retrospective study based on computerized medical record review of the patients born in Department of ENT of Sur Hospital between 2002 and 2017. The patients were assessed with detailed history, presentation of clinical symptoms and all underwent nasal endoscopy and CT scans for assessing the atretic type. These patients underwent transnasal endoscopic choanaloplasty under general anesthesia using microdebrider and stented using endotracheal tube. The patients were regularly followed up for review with nasal endoscopy after discharge between 4th and 7th postoperative period. The parents were educated on nursing care and the stent was removed in 4 weeks, all patients had Mitomycin C applied to the neochoana and were on regular follow up for a year with no recurrence.

Results: Fifteen patient records were analyzed, 10 females and 5 males, ages varying from newborns up to 14 years old. Unilateral to bilateral choanal atresia was 4:1 ratio and female-male showed 2:1 ratio. Right malformation was predominant in both sexes in unilateral atresia. Mixed imperforation (bone-membranous) was the most frequently observed type, followed by bone malformation. The commonest symptom during diagnosis was rhinorrhea and the least one was respiratory failure. Majority of cases were diagnosed by CT scans of sinuses. Around 10% patients presented with cardiac problems. None had restenosis in one year follow up.

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Introduction

Choanal atresia is an uncommon congenital disorder, initially reported by Roederer in 1755, where the back of the nasal passage (choana) is blocked, usually by abnormal bony or soft tissue (membranous) due to failed recanalization of the nasal fossae during fetal development. The incidence of newborns born is about 1:5000–1:8000 live births, with a female preponderance in the ratio of 2:1. It could be unilateral or bilateral. When only one nostril is affected (unilateral atresia), it will typically be the right nostril. Twice as many babies are born with only one nostril affected (unilateral) as with both nostrils obstructed (bilateral choanal atresia). There is a slightly increased risk in twins. It occurs in all races. Approximately two thirds of cases are unilateral, 65%–75% happens unilateral and 75% of bilateral cases have other anomalies. Thirty percent of atretic plate is bony and 70% is mixed. Bilateral choanal atresia is a very serious life-threatening condition because the patient will then be unable to breathe directly after birth as babies are obligate nasal breathers. In some cases, this may present as cyanosis while the baby is feeding, because the oral air passages are blocked by the tongue, further restricting the airway. The cyanosis may improve when the baby cries, as the oral airway is used at this time. These babies may require airway resuscitation soon after birth.

A number of theories have been proposed to explain the occurrence of choanal atresia such as persistence of the bucchoaryngeal membrane, failure of the bucconasal membrane of Hochstetter to rupture, abnormal mesodermal adhesions forming in the choanal area, and misdirection of mesodermal flow due to local factors.

The clinical evaluation includes a complete physical examination to assess for other congenital anomalies. A small feeding tube could be used to determine the patency of the choana, but a complete nasal and nasopharyngeal examination should be performed using a flexible fiberoptic endoscope to assess the deformity. Symptoms of severe airway obstruction and cyclical cyanosis are the classic signs of neonatal bilateral atresia. When crying alleviates respiratory distress in an obligate nasal breather, the neonatologist should be alerted to the probability of bilateral choanal atresia. Unilateral atresia may not be detected for years, and patients may present with unilateral rhinorrea or congestion. Many patients have an associated narrowed nasopharynx, widened vomer, medialized lateral nasal wall, and/or arched hard palate. Nonrandom association of malformations can be demonstrated using the CHARGE association, which appears to be overused in clinical practice. Various treatment options are described by different authors, surgical option being the preferred treatment of choice, most commonly endoscopic transnasal choanaplasty with stenting. Here we discuss the ways we dealt with fifteen patients in our hospital between 2002 and 2017.

Material and methods

This is a retrospective study based on computerized medical record review of the patients born in our secondary referral hospital between 2002 and 2017. The patients included in the study were those submitted to corrective surgery of choanal atresia, assessed according to time of diagnosis and treatment, sex, associated defects/anomalies, duration of stent, time of follow-up, unilateral or bilateral involvement, and surgical technique adopted. A detailed prenatal, perinatal and postnasal history was taken in all newborns and neonates taking into account the associated anomalies and they all had nasoendoscopy and CT scans to rule out the type of atretic plates (Figs. 1–2). All the new born and pediatric patients were taken up for surgical repair via nasal route, with endoscopes and debriber, under general anesthesia. After decongestion and suction removal of thick mucus, suction cautery was used to remove the mucosa at choana and opening made with controlled pressure of metallic nasal suction cannula, under vision from 2.7 mm, 0° nasal endoscope. Further, opening was widened using microdebrider and posterior septectomy was done using back biting forces (Fig. 3). Removal of posterior septum/vomer helped in making a single large rectangular common opening, rather than 2 small circular ones, which are more likely to stenose by circumferential

Conclusions: Neonates with acute respiratory insufficiency due to choanal atresia can be diagnosed with simple bedside tests like cold spatula test, less invasive tests like failure to pass intranasal catheter, CT scan. Surgical correction with endoscopic intranasal choanaplasty is the way to address this problem and could avoid radical palatal approach, less morbidity and high success rate.

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fibrosis. Hemostasis achieved intraoperatively, with no need of nasal packs, after surgery endotracheal tube lumen size 2 to 3 or 3.5 mm, cut to appropriate length (posterior end of this stent crosses the posterior choana and doesn’t touch posterior wall of nasopharynx), bent and partial wall removed from middle (to achieve patency of airway and allow easy passage of suction catheter), was placed bilaterally and stay suture applied anteriorly with silk. Routine intravenous antibiotic coverage besides normal saline instillation and regular suction was given. Patients remained stable post-operatively on continuous positive airway pressure for one day and on room air subsequently. With no intraoperative or postoperative complications documented, the patient was discharged between 4th post-operative and 7th post-operative day. All the patients were on breast feeding at the time of discharge with no respiratory difficulty. The parents were taught to do repeated suctioning of the tubes with a portable machine and disposable catheters and prophylactic antibiotics were advised. The nasal stent was removed 4 weeks post-operatively in all patients. Nasal endoscopy was performed in all the patients postoperatively and Mitomycin C (1 ml of 0.4 mg/ml) was applied topically to the neochoana for 4 min and repeat endoscopy was performed 2 months and 4 months later which showed no signs of stenosis and satisfactory functional patency of the choanae, without respiratory discomfort or secretions in the follow-up, and definite choanal patency was confirmed with nasal endoscopy (Fig. 5). The patients were under follow up for a year with no complaints of recurrence of symptoms or any complications. Our overall revision rate after initial endoscopic surgery in this study was very less and those two patients who had revision surgery were disease free at the last documented clinic visit (Fig. 4).

Results

Fifteen computerized medical records were selected and analyzed, including 10 females and 5 males, ages varying from newborns up to 14 years old. Unilateral choanal atresia was observed in 80% (12) of the patients and bilateral choanal atresia in the other 20% (3). In patients with unilateral atresia, right malformation was predominant in both sexes; bilateral imperforation was frequent in females. Mixed imperforation (bone-membranous) was the most frequently observed type, followed by bone malformations. The most common symptoms at diagnosis were respiratory failure (1 case, 6.7%), nasal obstruction (5 cases, 33.3%) and rhinorrhea (7 cases, 46.7%). Diagnosis was confirmed by CT of sinuses in 12 cases (80%) and by nasoendoscopy in 3 cases (20%).

Associated diseases were found in 10 patients presenting cardiac defects/valve heart problems, craniofacial anomalies and facial dysmorphisms.

Surgery was performed via transnasal endoscopy access with stent placement in all patients, except for one patient who did very well without any stent placement. None had restenosis. Outpatient follow-up lasted 1 year on average. The demographics of the observed patients are listed in Table 1.

Discussion

Choanal atresia is a rare condition that is present from birth, in which the nasal passages are blocked by bone or tissue. This condition can affect one or both nasal passages. Five percent of patients have monogenic syndromes or conditions. The other associations could be polydactyly, nasal-auricular and palatal deformities, crouzon’s, craniosynostosis, microencephaly, meningocele, meningoencephalocoele, facial asymmetry, hypoplasia of orbit and midface, hypertelorism and cleft palate. CHARGE association (coloboma, cardiovascular malformations, growth and mental retardation, urogenital and ear anomalies) is seen in around 20%–50% of new born. Chromosomal anomalies are found in 6% of infants with choanal atresia.

The cause of choanal atresia is unknown. It is thought to occur when the thin tissue separating the nose and mouth area during fetal development remains after birth. There are several theories explaining choanal atresia: persistence
of oral-pharyngeal membrane, failure of the physiological oral-nasal membrane rupture, mesodermic tissue adherence and growth of palatal processes.\(^6\) Another epidemiological report in 2010 found even higher associations between increased incidents of choanal atresia and exposure to second-hand-smoke, coffee consumption, high maternal zinc and B-12 intake and exposure to anti-infective urinary tract medications. In 2008, Barbero et al suggested that prenatal use of antithyroid (methimazole, carbimazole) medications was linked to choanal atresia.\(^7\) Teratogenic effects were caused by early pregnancy use of antithyroid drugs.

Choanal atresia can be suspected if it is impossible to insert a nasal catheter. Choanal atresia is diagnosed at birth when babies with choanal atresia have difficulty in breathing unless they are crying. Newborns generally prefer to breathe through their nose. Typically, infants only mouth breathe when they cry. Choanal atresia may affect one or both sides of the nasal airway. Choanal atresia blocking both sides (bilateral) of the nose causes acute breathing problems with cyanosis and breathing failure. Infants with bilateral choanal atresia may need resuscitation at delivery. More than half of infants have a blockage on only one side, which causes less severe problems. Chest retracts

Table 1  Demographics of the observed patients.

| Patients order | Age/Sex | Side and type | Presenting symptoms | Associations | Procedure done | Stent and duration | Revision surgery |
|---------------|---------|--------------|---------------------|--------------|----------------|--------------------|------------------|
| Case 1        | 2 days, male | right, mixed | nasal block         | cardiac valve defects none | transnasal endoscopy | yes, 4 days | no |
| Case 2        | 3 months, female | right, mixed | rhinorrhea          | none         | transnasal endoscopy | yes, 5 days | no |
| Case 3        | 2 months, female | right, membranous | nasal block       | cleft palate | transnasal endoscopy | yes, 5 days | no |
| Case 4        | new born day 1, female | bilateral, membranous | respiratory failure | microcephaly | transnasal endoscopy | yes, 7 days | yes |
| Case 5        | 1 month, male | left, bony | rhinorrhea/sinusitis nasal block | craniofacial dysmorphim pierre robin sequence imperforate anus | transnasal endoscopy | yes, 7 days | no |
| Case 6        | 3 months, male | left, mixed | nasal block         | none         | transnasal endoscopy | yes, 5 days | no |
| Case 7        | newborn day 1, female | bilateral, membranous | nasal block       | craniofacial anomaly facial dysmorphim | transnasal endoscopy | yes, 5 days | no |
| Case 8        | 15 days, female | right, mixed | nasal block         | craniofacial dysmorphim cardiac anomaly | transnasal endoscopy | yes, 6 days | no |
| Case 9        | 2 years, male | left, mixed | rhinorrhea/sinusitis | none         | transnasal endoscopy | yes, 4 days | no |
| Case 10       | 8 years, female | left, mixed | rhinorrhea          | craniofacial defect | transnasal endoscopy | yes, 10 days | yes |
| Case 11       | 12 years, female | right, membranous | nasal block       | transnasal endoscopy | yes, 6 days | no |
| Case 12       | 9 years, female | right, membranous | rhinorrhea/sinusitis rhinorrhea | none         | transnasal endoscopy | yes, 4 d | no |
| Case 13       | 5 years, female | right, mixed | rhinorrhea/sinusitis rhinorrhea | none         | transnasal endoscopy | yes, 5 days | no |
| Case 14       | 14 years, male | left, mixed | nasal block         | none         | transnasal endoscopy | no | no |

Figure 5  Postoperative follow up of patients following repair of choanal atresia with nasal endoscopy (A–C).
unless the child is breathing through mouth or crying. Difficulty breathing following birth, which may result in cyanosis (bluish discoloration), unless infant is crying, inability to nurse and breathe at same time, inability to pass a catheter through each side of the nose into the throat, persistent one-sided nasal blockage or discharge can all be suspicious of choanal atresia.

A physical examination may show an obstruction of the nose. Also, if one notices a continuous stream of mucus draining from one or both nostrils, it could be a sign of an atresia. Another common sign is cyanosis in an infant while breast feeding, as breathing is dependent on nasal patency in this situation. Diagnosis is confirmed by radiological imaging, usually CT scan. CT scan will be done to evaluate the extent of the choanal atresia once a feeding tube fails to pass through the nasal passage. A small plastic tube will be surgically inserted in the nostril to create an airway for the baby until more extensive surgery can be done. The baby will need very close observation and careful suctioning of mucus to be sure that breathing is maintained. Some babies do learn to mouth-breathe so surgery can be delayed. However, feeding is very difficult, so an airway tube through the mouth or a tracheostomy (breathing tube in the neck) may be necessary.

Surgery will be necessary, perhaps involving both an otolaryngologist as well as a plastic surgeon to create a functioning airway and to obtain a good cosmetic appearance of the nose. Surgery may be delayed a few months so that the baby’s facial structures can grow and repair will be more successful if the child’s airway and feeding can be maintained. Sometimes further surgical correction is necessary. The ultimate outcome for the child should be very good if there is no problem with resuscitating the baby at birth or with tolerating the surgical repairs. Babies who have complex syndromes and other conditions will have variable outcomes depending on those other problems. Their choanal atresia, once detected and repaired, should not pose long term problems for those babies with complex conditions. Today, several types are performed: microscopic and endoscopic transnasal, transpalatal, transseptal, transeptal, transtral techniques and intranasal dilatations with stenting. If the baby is resuscitated successfully at birth and the condition is detected before significant problems occur (i.e. brain damage from lack of oxygen during the cyanotic spells), there should not be any long-term implications for the baby’s development. However, nearly half of the children will have some other defect of the nose or palate, so there may be a number of surgeries that may cause discomfort and affect the child’s appearance. Choanal atresia ranges from very mild to severe. It may be impossible to detect once the nasal passage has been surgically repaired, or in other cases, it may significantly involve the appearance of the face, causing the child to be self-conscious.

Building the child’s self-confidence in light of the problem will be a very important task for parents and others who interact with the child at home, at school and in medical settings. Dental abnormalities might also be associated with the condition, making it important that the child be cared for in a pediatric multi-disciplinary craniofacial clinic that brings together all the professionals who care for the child, including genetics, speech therapy, nursing, nutrition, plastic surgery, and ear-nose-throat specialties.

Monitoring the child’s growth and nutritional status will be important, and social workers might also be helpful to the family coping with raising a child with complex, long-term health care needs.

Conclusion

In neonates with acute respiratory insufficiency, it’s worthwhile to rule out bilateral atretic plates. The diagnosis is easily achieved by simple bedside tests like cold spatula misting, passing nasal catheter, failure to pass a 6 FG nasal catheter would confirm the diagnosis and it is supplemented with radiological scans. It is mandatory to obtain second opinion if other congenital anomalies are suspected. Surgical correction is needed to address the problem. Endoscopic transnasal choanalplasty followed by stenting with endotracheal portex tube for 4–6 weeks is the standard operation of choice due to ease in accessibility, economical timing, minimal chance of bleeding, does not involve radical approach to the palate and alveolar arch development is not affected, less morbidity and high success rate.

Conflicts of interest

There is no conflict of interest.

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