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Study of a Series of Choanal Atresia Patients: Presentation, Surgical Management and Outcome

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Abstract
Congenital choanal atresia is a rare but well recognized entity. It is characterized by the developmental failure of nasal cavity to communicate with nasopharynx. While Roederer first described this condition in 1755 it was Oto who further described it in relation to the deformity involving palatine bones. Congenital atresia can be unilateral or bilateral. The reported incidence is 1 out of 6000-7000 live births. Females are more commonly affected than males. It may be associated with other congenital anomalies like coloboma, heart defect, retarded growth, genitourinary abnormalities, and ear anomalies (CHARGE anomalies). In 2/3 of the cases choanal atresia is bony-membranous type while approximately 1/3 cases are that of pure bony type. While unilateral choanal atresia may be subtle in presentation and the diagnosis may be missed in neonates and infants bilateral choanal atresia is life threatening and may present in immediate postnatal life with signs of severe airway obstruction and cyanosis.

Aims and Objectives: *(1) To study the cases of choanal atresia through clinical evaluation (2) To study the surgical management of choanal atresia (3) Post-operative follow-up of patients with choanal atresia.*

Materials and Methods:*This was a multicentric prospective cohort study conducted in Department of Oto-Rhino-laryngeology at a government medical college and a ENT hospital. Detailed history was noted and clinical examination was done. A pediatric reference was done in each patient to rule out associated congenital anomalies. CT-PNS was done in all cases to confirm the diagnosis and type of choanal atresia. Surgery was done using transnasal endoscopic approach. Nasal stents were kept for 7-10 days after which they were removed under general anaesthesia. Follow up diagnostic nasal endoscopy was done at 6 weeks and 6 months to assess the size of posterior nasal aperture. The data was analyzed with a special emphasis on demographic characteristics, clinical presentation, surgical intervention required and postoperative outcome.*

Results: *Total 7 cases with either unilateral or bilateral choanal atresia were included in this study. Out of these cases 4 were males and 3 females with a M: F ratio being 1:0.75. 4 patients had bilateral choanal atresia while 3 patients had unilateral choanal atresia. Out of the 3 patients who had unilateral choanal atresia 2 had right sided and 1 had left sided choanal atresia. Common signs present in bilateral choanal atresia were tachypnoea and central cyanosis while in children with unilateral choanal atresia the most*

common complaint was refusal to take proper feeding. The median age of surgery in children with unilateral choanal atresia was 6 years. Since bilateral choanal atresia is an emergency immediate intervention was done and median age of these patients at surgery was 12 days. Follow up of the patients was done every 6 monthly.

Conclusion: *Choanal atresia is a rare but well recognised entity. While the clinical features of unilateral choanal atresia may be subtle bilateral choanal atresia usually present with respiratory distress and cyanosis in immediate postnatal period. Trans-nasal endoscopic surgical repair of choanal atresia with postoperative stenting achieves good results in patients.*

Keywords: *Choanal Atresia, CHARGE, Trans-nasal endoscopic surgery, Respiratory distress.*

Introduction

Choanal atresia was first described by Roederer in 1755 [1]. It is defined as malformation of the posterior nasal aperture interfering with airflow from the nose to the rhino-pharynx. The reported incidence is 1-2 per 10,000 live births with a female preponderance[2]. The exact etiology of choanal atresia is not known but it is thought to be multifactorial. Some cases are reported to be associated with single gene defects that include both autosomal dominant and autosomal recessive mode of inheritance [3]. It may be isolated or can be part of a syndrome. The most frequently associated syndrome is known by the acronym CHARGE (Coloboma, Heart Defect, Atresia of Choanae, Retarded Growth and Development, Genital Anomaly, Ear Defect) [4]. Other well-known syndromes having choanal atresia as a component are Crouzon syndrome, pfeiffars syndrome and treacher-colin syndrome. Alcohol, retinoic acid, methimazole, propranolol and anti-thyroid medication use, is thought to contribute to occurrence of choanal atresia by influencing neural crest cell migration [5]. The malformation may either be a bony plate or soft membranous tissue due to failed re-canalization of nasal fossae during fetal development. In some cases the malformation is mixed type having bony as well as membranous components. Choanal atresia either may be unilateral or bilateral. While unilateral choanal atresia sometimes go unnoticed, bilateral choanal atresia is a life-threatening condition as newborns are obligate nasal breathers (they mainly use their noses to breath) they may present as cyanosis while baby is feeding, because oral air passage is also blocked by the tongue, further restricting the airway. The cyanosis may improve when the baby cries as oral breathing restarts. Many of these newborns require airway resuscitation in immediate postnatal period [6].

Several studies have been published on this subject, mainly case series without adequate standardization. There are many doubts about timing of surgery, access to surgical field, procedure technique, pharmacological treatment, stent application and maintenance. Moreover differentiation between patients having unilateral and bilateral choanal atresia remains a neglected aspect of many studies. We undertook this study to describe the case characteristics of patients presenting with unilateral and bilateral choanal atresia.

Materials and Methods

This was a multi-centric prospective cohort study comprising of 7 patients diagnosed with either unilateral or bilateral choanal atresia conducted at a government medical college and a ENT hospital.

Inclusion Criteria

- All patients presenting either with unilateral or bilateral choanal atresia.

Exclusion Criteria

- Patients whose parents refused consent to be part of the study.
- Patients who already have undergone surgical intervention.
- Patients in whom other congenital anomalies were present or in whom choanal atresia was part of a syndrome.

Detailed history was taken and ENT examination was done along with examination by pediatrician to rule out any associated medical condition.

Diagnosis

For confirmation of diagnosis following investigations were done

a) Passing infant feeding tube through both nostrils.
b) X-ray skull lateral view with contrast.
c) Diagnostic nasal endoscopy.
d) CT-PNS.

Surgical procedure

Surgical procedure was performed with transnasal endoscopy. Obstructing septae was perforated first with Hegar's dilators. Progressive increasing in size curved Hegar's dilators were used to perforate the atretic portion. Maximum size of Hegar's dilator was approximately equal to external nares of the infant. Care was taken to perforate the obstructing membrane in inferio-medial part. Surrounding obstructing soft tissue was removed with micro-debrider in membranous and mixed types of atresia. Osseous atresias were widened with drilling the bony obstructive plate. In all cases few millimeters of posterior septum was removed. Haemostatis was achieved with bipolar cautery and mitomycin was applied followed by stent using portex endotrachial tube. Patients were discharged on 3rd day .parents training was given regarding maintenance of nasal stents. Stent was kept for 7 to 10 days and removed under general anesthesia. Diagnostic nasal endoscopy was done at the time of stent removal. Nasal secretions and slough was removed and patients were kept on normal saline nose drops for 6 weeks, diagnostic nasal endoscopy was done at 6 weeks and 6 months to access the size of posterior nasal aperture.

Discharge and follow up:

Patients were discharged on 3rd day. Parents were trained regarding maintenance of nasal stents. Stent was kept for 7 to 10 days and after that removed under general anaesthesia. Diagnostic nasal endoscopy was done at the time of stent removal. Nasal secretions and slough was removed and patients were kept on normal saline nasal drops for 6 weeks. Follow up diagnostic nasal endoscopy was done at 6 weeks and 6 months to access the size of posterior nasal aperture.

Results

We studied 7 cases of choanal atresia. The cases either had unilateral or bilateral choanal atresia. Out of the studied cases 4 were males and 3 were females with a M: F ratio being 1:0.75.

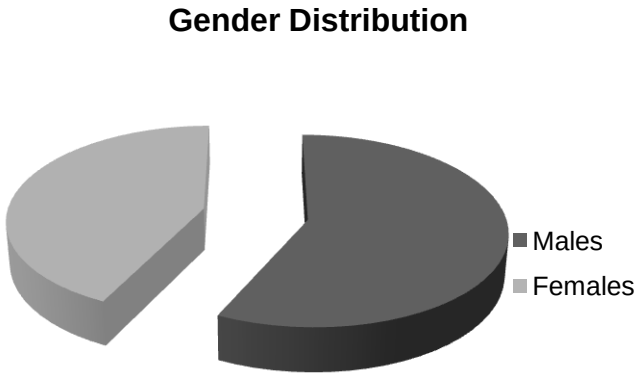

Figure 1 Gender Distribution of the studied cases.

4 (57.14%) patients had bilateral choanal atresia while 3 (42.86%) patients had unilateral choanal atresia. This is important to differentiate between unilateral and bilateral choanal atresia because later may present as life threatening emergency in postnatal period and may need an immediate intervention.

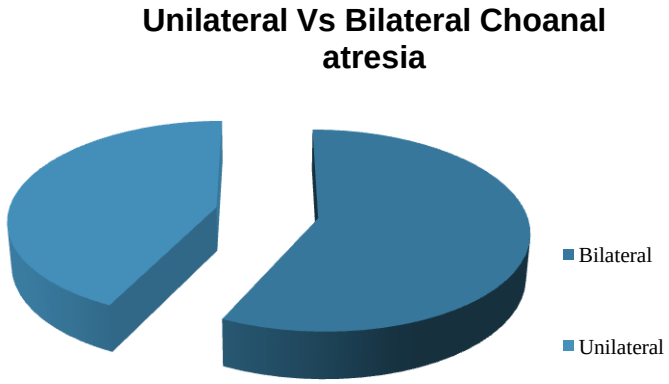

Figure 2: Unilateral and bilateral choanal atresia in studied cases.

The choanal atresia in 2 patients was osseous type (28.57%) and membranous type in1 (14.29%)

patient. While mixed (osseous as well as membranous type) was seen in 4 patients (57.14%). So the most common type of choanal atresia found in our study was that of mixed type comprising of osseous as well as membranous type of choanal atresia.

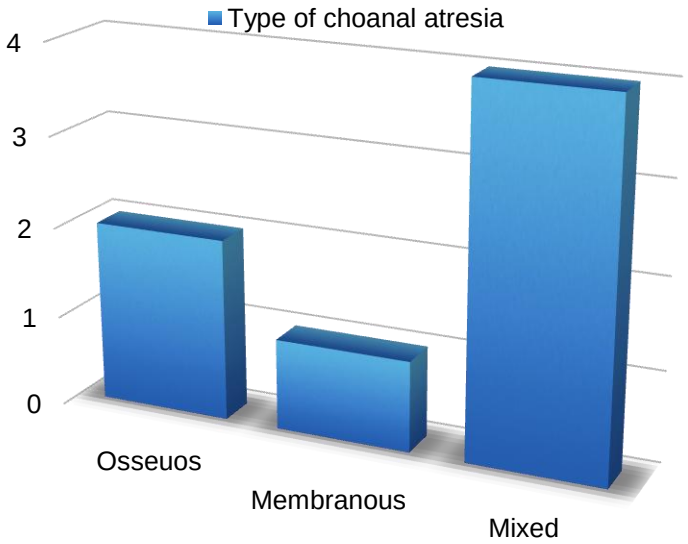

Figure 3: Type of choanal atresia in studied cases.

The group characteristics of the studied cases are given in the table below (Table 1).

Table 1: Group characteristics f the studied cases

	Sex		Side		Composition		
	Male	female	right	Left	Osseous	Mixed	membranous
Bilateral n=4	2	2	--	--	2	2	--
Unilateral n=3	1	2	2	1	--	2	1
Total n=7	3	4	2	1	2	4	1

The common symptoms in unilateral choanal atresia were blocked nose, mouth breathing and persistent rhinorrhea. While in bilateral choanal atresia newborns presented with difficulty in breathing, cyanosis and choking while feeding.

Table 2: Presenting signs and symptoms in patients with choanal atresia

Signs and Symptoms	No of patients	Unilateral/ Bilateral	percentage
Difficulty in Breathing	4	Bilateral	57.14%
Cyanosis	4	Bilateral	57.14%
Choking while feeding	4	Bilateral	57.14%
Blocked nose	3	Unilateral	42.86%
Mouth Breathing	3	Unilateral	42.86%
Persistent rhinorrhea	3	Unilateral	42.86%

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The study of surgical intervention of the studied cases revealed that all patients with unilateral choanal atresia demanded only one surgical procedure. One patient of bilateral choanal atresia of osseous type required additional procedures at 6 weeks to attain adequate airway patency. Stenting in all patients were done using portex non- cuffed endotrachel tubes and stent was maintained for 7-10 days.

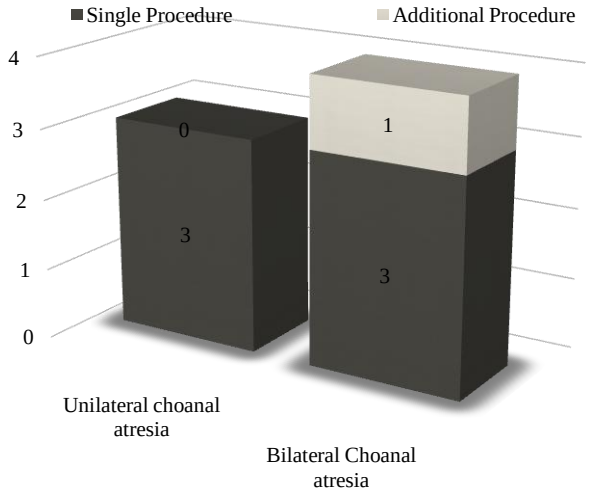

Figure 4: Single vs additional procedures in cases with choanal atresia.

Median age at surgery for patients with unilateral atresia was 6 yrs ranging from 3 yrs to 10 yrs. For patients with bilateral choanal atresia median age was 12 days. Median time from diagnosis to surgery was 3 months in unilateral group and 1 day in bilateral group. Follow up of patients range from 6 months to 7yrs.

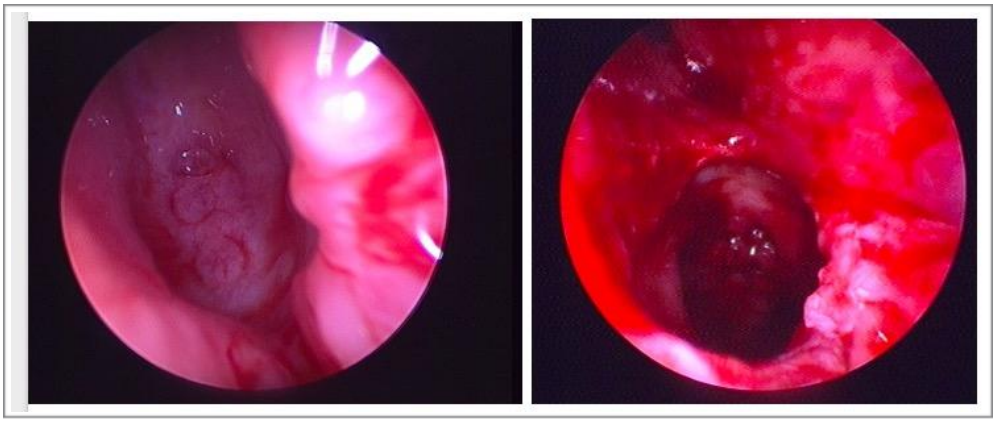

Figure 5: Nasal endoscopy showing right sided choanal atresia before (Left) and after endoscopic surgery (Right).

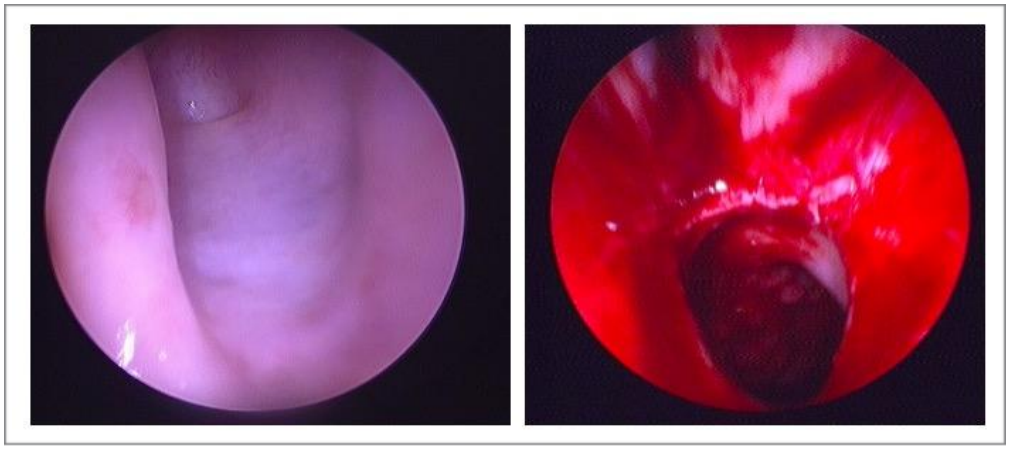

Figure 6: Nasal endoscopy showing left sided choanal atresia before (Left) and after endoscopic surgery (Right).

Discussion

Choanal atresia is one of the important causes of respiratory distress in immediate postnatal period. The neonates are by their nature obligate nasal breathers and if the choanal atresia is bilateral then the baby may present as severe respiratory distress with central cyanosis. The reflex to take breathing through mouth in the event of obstruction to nasal passage only develops after some months. The cyanosis is relieved if baby cries because during crying there is co-occurrence of breathing [7]. In contrast to bilateral choanal atresia unilateral choanal atresia present in relatively subtle ways and many cases of unilateral choanal atresia go unnoticed because of rarity of the condition and unfamiliarity with the symptoms [8,9].

Repair in the neonatal period is difficult and require expert surgeons. Four approaches described for surgical correction of choanal atresia include transnasal, transpalatal, transantral and trans-septal routes[10]. The most common approaches used for surgical corrections include transnasal and transpalatal route and last 2 approaches namely transantral and transseptal routes are rarely used.

We preferred transnasal route because it is a safe and simple procedure with less bleeding and minimal tissue handling. It is important to understand that being a congenital anomaly choanal atresia, particularly bilateral type, is usually present in infants and neonates who are the risk of severe hemodynamic imbalance, sepsis and multiorgan dysfunction[11,12]. The drawback of this approach is that it can only be used in membranous type of choanal atresia or when bony plate is thin [13]. Fortunately since all our studied cases were either having membranous or thin bony atresias we could successfully treat them transnasal endoscopic procedures. Many studies have been done on the subject of choanal atresia but most of the authors have clubbed the cases of unilateral and bilateral choanal atresia and very few studies have separated them [14,15].

In this study we have studied them as separate entity. While the features like respiratory distress, cyanosis and choking while feeding was more common n bilateral choanal atresia, rhinorrhea, nasal blockade and mouth breathing was more common in unilateral choanal atresia. Similarly there was a significant difference in the median age of patients at surgery for patients with unilateral (6 years) and bilateral choanal atresia (12 days). The difference is expected as bilateral choanal atresia will present as respiratory difficulty and cyanosis in early neonatal period and will demand an immediate intervention while unilateral choanal atresia presents with subtle signs and symptoms and may go unnoticed for years.

Conclusion

Transnasal endoscopic surgical repair of choanal atresia with postoperative stenting achieves good results in patients with choanal atresia. However literature should describe bilateral and unilateral types separately (as we did) as they seem to concern different clinical presentation & have different patient characteristic. Some more clinical trials should be undertaken to have comparative and standardized results.

Conflict of interest: None

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