

Presentation of choanal atresia in Saudi children

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ABSTRACT

Objectives: To present data of cases with choanal atresia (CA) from Saudi patients, and to compare them to the data from the international literature.

Methods: A retrospective analysis of the data available from the files of 37 consecutive patients with the diagnosis of CA at King Abdul-Aziz University Hospital, Riyadh, Kingdom of Saudi Arabia between January 1999 and December 2005. This involved reviewing the age, gender, presenting symptoms, associated anomalies, surgical intervention, and outcomes.

Results: Twenty-three of our cases had unilateral and 14 had bilateral CA. Strikingly, 83% of unilateral CA involved

the right side. In our study, we found the female to male ratio to be 2:1. Approximately, 95% of the cases had mixed bony and membranous CA. Thirty-two percent of the cases had other associated congenital anomalies. Most of our cases had their surgical intervention by endoscopic technique.

Conclusion: Choanal atresia is a rare anomaly. In Saudi children, female is more commonly affected than male. There is a striking rate of involvement of the right side in the cases of unilateral CA.

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Nasal obstruction is an unpleasant dysfunction that can be very disturbing to the patient. This can result in loss of basic function of the nose, which include breathing, humidification, protection, adjusting temperature, and smell. However nasal obstruction in neonates may be life threatening situation. The reason for this is that newborn is an obligate nasal breather within the first 6-8 weeks of life.¹

Congenital choanal atresia (CA) is a rare anomaly, which can be responsible for nasal obstruction. It is reported in 1 in 5000 - 8000 births.²⁻⁴ It was first described by Roederer in 1755.^{2,5} Emmert reported the first successful operation for CA on a 7-year-old boy, which he had performed 3 years earlier using a curved trocar transnasally.⁶

Reviewing the English literature, many have been written regarding this anomaly. This raises the following question; do we have similarity between our findings of cases of CA in Saudi Arabia and the findings in the international literature?

Rejial et al⁷ in 1994, reported the association between CA and brain abnormalities in Saudi Arabia. In this study, we present cases of CA from Saudi patients, managed at a university hospital in Riyadh, Kingdom of Saudi Arabia (KSA), and compared the available data to international literature.

Methods. Thirty-seven consecutive patients diagnosed with choanal atresia (CA) were managed at King Abdul-Aziz University Hospital (KAUH), Riyadh, KSA between January 1999 and December

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2005. The diagnosis was based on clinical examination, utilizing fiberoptic scope, and radiological assessment by CT scan. The files of all of these cases were reviewed for; age, gender, presenting symptoms, associated anomalies, surgical interventions, as well as outcomes.

Mann-Whitney test was used to compare the association between 2 categorical variables. A *p*-value of less than 0.05 indicated statistical significance.

Results. Twenty-four of our patients were female and 13 were male with age ranging between 4 days and 25 years. There was only 2 adults, the rest of our cases were children. Twenty-three of our patients had unilateral CA, and 14 had bilateral atresia. Bilateral cases presented at relatively younger age (mean of 3 months) compared to the age in unilateral cases (mean of 4.5 years), this difference in age was statistically significant, with $p < 0.001$. Initial surgery was carried out at KAUH on 9 of the bilateral CA cases, presented before the age of 2 months. However, the rest of bilateral cases were managed surgically somewhere else but restenosed. For them revision surgery was carried out at KAUH. All of the unilateral cases were children except in 2 cases. All of them had their initial surgical management at KAUH. Nineteen (83%) of the unilateral CA involved the right side, and only 4 cases had left side CA. There was a need to secure the airway by oropharyngeal airway or endotracheal intubation for the neonates and young infants with bilateral CA in this study, until the time of the definitive surgery. On the other hand, the presentation of unilateral cases was mainly with unilateral nasal discharge, obstruction, and occasional feeding difficulties.

A total of 12 (32%) cases were associated with other congenital anomalies. Seven of them had unilateral CA (representing 30% of the unilateral cases), and 5 had bilateral CA (representing 36% of the bilateral cases). Isolated congenital anomalies were seen in 5 of the cases, 5 cases had variable syndromes, and 2 cases of bilateral CA had ocular colobomas, heart defects, choanal atresia, retarded growth and central nervous system issues genitourinary hypoplasia, and ear anomalies (CHARGE) association.

Thirty of our patients underwent endoscopic surgical repair of the posterior choana. Five cases had an alternative surgical technique; 3 had trans-palatal, one combined trans-palatal and endoscopic, one trans-nasal non-endoscopic surgical repair of the posterior choana. Two of the patients were not operated upon, because of medical reasons. Endoscopic repair involved the use of thin telescope (2.7 or 4 mm), use of microdebrider, drill and backbiting forceps. All of

the cases of CA showed mixed bony and membranous atresia, by CT scan or during surgical repair, except 2 cases with pure bony atresia. There was no case with a pure membranous atresia. Surgical intervention to open CA among all of our cases ranged between 1-3 times. We followed up our patients for a period ranging between 6 years and 4 months. All of our patients who underwent surgical intervention had successful outcome, with good size of posterior choana and normal function. Only 5 of our patients have less than 6 months follow up, and still showing patent posterior choana after repair.

Discussion. Congenital posterior choanal atresia is a relatively rare condition resulting from a failure of the breakdown of the wall between the nasal pits and the stomodeum in early embryogenesis. Commonly, it affects female more than male, with a ratio of 2:1.⁸ This is in agreement with our findings in this study. Clinical presentation of CA cases is based on the involvement of one side or both sides of the posterior choana. Bilateral CA almost always presents with respiratory distress and cyclical cyanosis at birth. The symptoms are classically relieved by crying and worsened by sucking. The diagnosis may be suspected by failure to pass size 6 or 8 French rubber catheter into the nasopharynx. This necessitates emergency management with either oropharyngeal airway, McGovern nipple, or intubation, until definite repair is performed.³ However, bilateral CA can still present for the first time in adult life, indicating that some neonates with bilateral CA can still breathe orally.^{1,9} Unilateral CA, on the other hand, usually presents with less acute features, such as unilateral nasal congestion and discharge, which commonly manifests later on life. From our results there was a statistically significant difference in age at presentation, with a mean age of 3 months for the bilateral cases and 4.5 years for the unilateral cases, $p < 0.001$. Unilateral was more common than bilateral CA in our study with a ratio of approximately 2:1. Ratio of unilateral to bilateral CA is reported to be approximately 3:2.⁸ Our finding, however, is not in agreement with the findings of Harris et al¹⁰ they found that the ratio of unilateral to bilateral CA was almost 1:1.

Choanal atresia involving the right side is more common than in the left side.¹¹ This finding is hard to explain. In our study, the right side was involved in 83% of unilateral cases. There is no association between maternal age and CA.⁷ Unfortunately, maternal age was not available in our records to evaluate its effect on the occurrence of this anomaly. The proportion of infants with CA and associated other malformations

may reach up to 50%.¹⁰ In our study, 12 cases (32%) were associated with other anomalies.

There was no major difference between unilateral and bilateral cases in association with other congenital anomalies. Unilateral CA had associated other anomalies in 30% of the cases. On the other hand, other anomalies were seen in 36% of bilateral cases.

The only 2 cases of CHARGE association were seen in cases with bilateral CA. The term CHARGE should be restricted to infants with multiple malformations, and choanal atresia or coloboma, combined with other cardinal malformations (heart, ear, and genital) and with a total of at least three cardinal malformations.^{7,12}

Almost all of our cases of CA (95%) had mixed bony and membranous atresia, except 2 (5%) of the cases, they showed bony atresia only. Reports in the literature are showing a grate variation.¹³ Brown et al¹⁴ found no pure membranous CA among their cases, 71% mixed, and 29% pure bony atresia. Surgical opening of the posterior choana is the way to restore normal airway in cases with CA. Many surgical approaches are used, which include; transpalatal, transnasal, transantral and trans-septal route.¹⁵ In the recent years repair of CA by endoscopic technique became very popular.^{16,17} Most of our cases are operated by endoscopic technique, which can visualize the bony margins very clearly. With the availability of thin telescopes (2.7 mm) and powered instruments (microdebrider and drill), this made telescopic technique very practical even for neonates.¹⁸

We followed up our patient for a period ranging between 6 years and 4 months. All of the patient that underwent surgical repair eventually had successful outcome. The frequency of surgical repair ranged between 1 - 3 times. This rate of surgical intervention is not far from the reported rate in the international literature.¹⁹

In conclusion, CA is a rare anomaly. In Saudi children, female is more commonly affected than male. There was a striking rate of involvement of the right side in the cases of unilateral CA in this study. This study is having the limitation of retrospective studies. However, it can be used as a baseline for further prospective studies and a larger study groups.

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