



# Supposed otolaryngology emergency which presents late: Bilateral congenital choanal atresia

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## Abstract

Bilateral congenital choanal atresia is a life threatening emergency which always presents in new born. Cyclical respiratory distress, cyanosis and apnoea relieved by crying are common clinical features. We report a rare case of bilateral choanal atresia in a 5 year old girl with unremarkable neonatal history, whom presented to us with persistent bilateral nasal obstruction. The atresia was further confirmed by Computed Tomography (CT) scan and endoscopic examination. Patent airway was achieved successfully with transnasal endoscopic surgery.

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## Introduction

Choanal atresia is the congenital obstruction of posterior nasal cavity, and encountered at a rate of 1 in 5000 to 1 in 8000 live births, with slight female preponderance [1]. This anatomical anomaly is classified into bony (90 percent), or membranous (10 percent); unilateral or bilateral; complete or incomplete [2]. There is about 10 to 50 percent of cases show associated anomalies, CHARGE syndrome (coloboma, heart defects, atresia choanal, retarded growth, genital hypoplasia, ear deformities) the commonest [3]. Bilateral congenital choanal atresia always presents as upper airway emergency where the newborn

shows respiratory distress, cyanosis, apnoea which are relieved by crying. We report a rare care of bilateral congenital choanal atresia which presented late beyond her neonatal period.

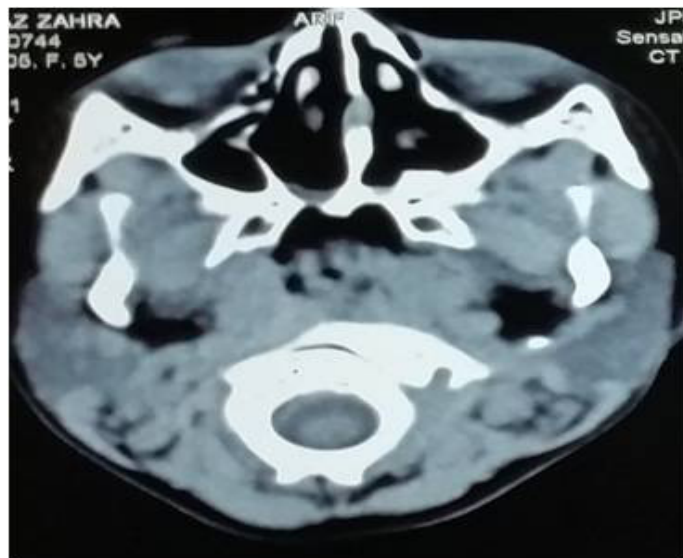
## Case report

A 5 year old girl presented to our otolaryngology clinic with bilateral persistent nasal blockage and rhinorrhea since birth. There was no frequent sneezing or nasal itchiness. Parents noticed that the child had persistent mouth breathing and did not seem to be interested in food stuffs that most children were.



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The child also was unable to differentiate smells from various sources. Her perinatal and neonatal periods were uneventful. She had never experienced any respiratory distress and cyanosis thus far. On examination, the child was not syndromic. Externally, the nasal framework was normal. Cold spatula test was negative as well as the cotton wool test. Internally, the nasal cavities were tight and mucoid secretion was copious. Passage of size 8 French Ryle's tube through nasopharynx failed. Careful endoscopic examination showed bilateral posterior choanal atresia. Both the atretic areas were soft on probing. Diagnosis was further confirmed by high resolution CT of paranasal sinus, which showed bilateral membranous atresia (Figure 1,2).



**Figure 1:** Axial cut CT scan showing bilateral post nasal choanal atresia



**Figure 2:** Axial cut CT scan showing bilateral post nasal choanal atresia

Transnasal endoscopic surgery was performed by using 0 degree Hodgkin endoscope 2.7mm. A cruciate incision was made on the atretic plate and the edges were curetted and cut with Stammberger 'mushroom' punch. Following that, mitomycin C (0.4mg/ml) soaked cotton pledget was applied onto edges for 5 minutes. We did not use stent in our case.

The posterior choanal patency was good until 3 months after follow up, it became Gradually stenosed at 6 months and remained static. Currently it is post-surgery 3 years, there is par-

tial re-stenosis of right choana but the left was patent (Figure 3,4). However, the child is asymptomatic except for anosmia that never resolved.



**Figure 3:** Post operative endoscopic view showing right posterior choanal partial stenosis



**Figure 4:** Post operative endoscopic view showing left patent posterior choana

To date, there are only few reports on late presentation of bilateral congenital choanal atresia. This is an life threatening emergency of the upper airway in newborn where a patent airway needs to be addressed as soon as possible, if not the child can die of asphyxia. They almost always present with cyclical respiratory distress, cyanosis and apnoea which are relieved by crying [4]. Feeding makes the situation worse. This condition is commonly detected even in labour room itself and the easiest way to come to diagnosis is failure to pass the feeding tube via nasopharynx with correlation with symptoms. Immediate way to establish patent airway is by inserting an oral airway. The reason being the newborns are obligate nasal breather till at least 4 to 6 months of life. It is extremely rare to see neonates to survive through this period with underlying choanal atresia without being noticed. There is a hypothesis by *Baker et al.* that newborn may compensate by rapidly learning mouth breathing [5]. In our report, the child responded well to surgery with relieved of nasal obstruction and rhinorrhea. However, anosmia

still persisted. *Gross Isseroff et al.* speculated that early olfactory stimulation is vital in establishing normal functioning olfactory nerves. Long term deprivation of stimuli could be the cause of persistent anosmia in our case [6].

Mainstay of treatment for bilateral congenital choanal atresia is surgery. Transnasal approach is preferred to transpalatal approach. The later carries higher risk of bleeding, disruption of facial growth and orthodontic problems. Transnasal approach is quick, safe and simple. The incidence of re-stenosis could be addressed with careful and more meticulous surgical technique to prevent granulation tissue formation such as use of prophylactic antibiotics anti-fibrotic agent such as mitomycin C [7].

### Conclusion

Choanal atresia is a rare entity especially if it escapes through the neonatal period without being detected. It maybe misdiagnosed as allergic rhinitis or chronic rhinosinusitis if not properly examined particularly in children whom are not cooperative with office procedure such as feeding tube passage or endoscopic examination.

Sensation of smell maynot revert to normal after successful surgery, but the relief of nasal blockage and rhinorrhea definitely helps in quality of life of patients.

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