

## **Congenital Nasal Pyriform Aperture Stenosis: An Infrequent Cause Upper Airway Obstruction Leading to Respiratory Distress in Neonates**

**Vinod K Hasija<sup>1</sup>, Maryam Iqbal<sup>2</sup>, Adnan Mirza<sup>3\*</sup>, Sadia Sheikh<sup>3</sup> and Waqar Khowaja<sup>4</sup>**

<sup>1</sup>*Fellow of Neonatal medicine, The Aga Khan University Hospital, Karachi, Pakistan*

<sup>2</sup>*Resident of Paediatrics, The Aga Khan University Hospital, Karachi, Pakistan*

<sup>3</sup>*Faculty of Paediatrics, The Aga Khan University Hospital, Karachi, Pakistan*

<sup>4</sup>*Staff Neonatologist, The Aga Khan University Hospital, Karachi, Pakistan*

**\*Corresponding Author:** Adnan Mirza, Faculty of Paediatrics, The Aga Khan University Hospital, Karachi, Pakistan.

**Received:** December 09, 2019; **Published:** January 18, 2020

### **Abstract**

Congenital nasal pyriform aperture stenosis is a rare but serious cause of respiratory distress in neonates. It is caused by narrowing in cross-sectional area of pyriform aperture present in the anterior part of nasal airway leading to severe respiratory problems in obligate nasal breather infants. Prompt recognition and intervention is necessary to avoid lethal outcome. A detailed history and clinical examination help in making the clinical diagnosis, which can be confirmed by computed tomography scan. Treatment options depends on the severity of obstruction, it is managed either conservatively or surgically.

**Keywords:** Congenital Nasal Pyriform Aperture Stenosis; Respiratory Distress; Solitary Maxillary Central Incisor; Hegar Dilator; Neonate

### **Introduction**

Congenital nasal pyriform aperture stenosis (CNPAS) is an uncommon, potentially fatal form of neonatal nasal obstruction [1]. Anatomically, the pyriform aperture is the anterior most narrow opening of the nasal bony airways. Laterally, It is limited by the nasal process of the maxilla, inferiorly by the junction of the horizontal process of the maxilla and the anterior nasal spine, and superiorly by the nasal bones [2]. Congenital stenosis of pyriform region is an infrequent condition that may present as an isolated malformation or as a part of association with other craniofacial anomalies. Clinically, CNPS presents with signs of respiratory distress, Poor nursing, Apnea and Intermittent cyanosis, pointing towards nasal obstruction. Because infants are obligatory nasal breathers until 6 - 8 weeks of age, obstruction at any level of nasal airway can lead to severe respiratory distress; thus, early diagnosis and timely treatment is mandatory [3,4].

The purpose of this case report is to sensitize the general pediatrician and neonatologist to think about these uncommon causes of neonatal respiratory distress especially when the common causes has been ruled out. Moreover, this is the first case in our resource limited setup which has been treated successfully through a minimally invasive surgical intervention using hegar dilator.

### **Case Summary**

A full-term female baby, the 1<sup>st</sup> child of nonconsanguineous parents, was delivered via normal Vaginal delivery at 38 weeks of gestation. The antenatal period was uneventful. Baby cried immediately at birth, weighed 3.15 kg, with an APGAR of 8 and 9 at 1 and 5 minutes re-

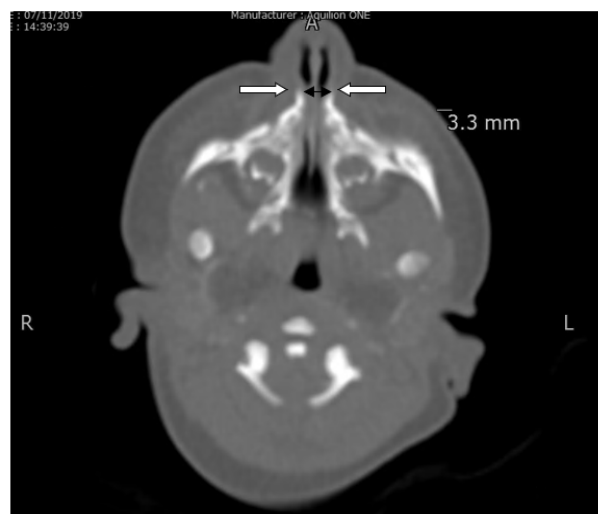
spectively. After few hours of delivery baby developed respiratory distress, necessitating admission in neonatal intensive care unit (NICU) for further evaluation and management.

On admission, her pulse oximetry at room air showed 95% oxygen saturation without any difference between upper and lower extremities. Her physical examination revealed a heart rate of 140 per minute, and respiratory rate of 70 per minute with obvious signs of respiratory distress. As a part of our institutional protocol nasal patency was checked via 5 French NG tube, to screen for upper airway obstructions, which was passed only in the right side of the nares with some local trauma. The heart sounds were normal with no significant murmur. Capillary refill time was < 3 seconds with regular and normal volume femoral pulses. Chest x-ray and ABG was normal. Sepsis screening also done and results were shown in normal limits.

Because of progressive worsening of respiratory distress and desaturation on high flow nasal therapy, baby was electively intubated and put on ventilator with Low requiring settings. After critical thinking and ruling out primary cardiopulmonary, infective and metabolic causes of respiratory distress in a neonate, upper airway obstruction was considered to be the most likely cause. The differential diagnosis of choanal atresia, congenital pyriform aperture stenosis, nasopharyngeal encephalocele and congenital nasal tumors were thought of and computed tomography (CT) scan was planned to evaluate upper airway.

CT scan demonstrated a narrow pyriform aperture in combination with a small nasal cavity more at the left side, due to abnormal medialization of the anterior part of the maxilla bilaterally resulting in narrowing of nasal airway likely representing congenital pyriform aperture stenosis (Figure 1), associated with abnormal dentition along with solitary median maxillary central incisor resulting in malalignment of the normal central and lateral maxillary incisors (Figure 2 and 3). Workup for possible associated anomalies including holoprosencephaly and pan hypopituitarism was unremarkable.

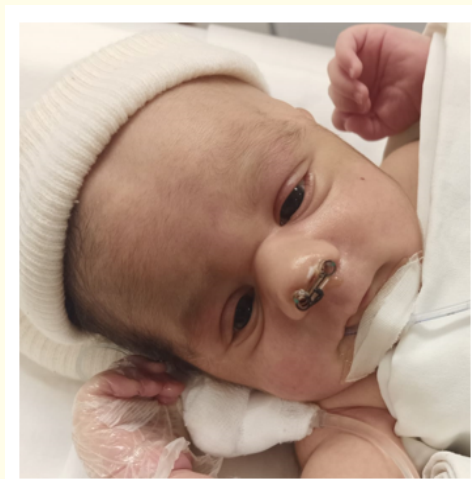
For the symptomatic Congenital Pyriform aperture stenosis, ENT surgeon was involved and the baby underwent hegar dilatation of the anterior nasal cavity, followed by stent placement using a size 3 mm ET tube. Baby was then successfully extubated on 2<sup>nd</sup> post-operative



**Figure 1:** CT image: axial view of the narrowed pyriform aperture measuring 3.3 mm. Note the abnormal medialisation of anterior part of maxillary bilaterally resulting in narrowing of nasal cavity (white arrows).



**Figure 2:** CT Image axial view solitary median maxillary central incisor (Circle).



**Figure 3:** Picture of the baby after hegar dilation showing stent placement.

day and was discharged in a stable condition with a plan of close follow up visit in ENT clinic and review for possible need of redilatation in 6 weeks.

### **Discussion**

The recent literature reports the incidence of CNPAS being 1 in 5000 - 8000 live births and the occurrence of solitary median maxillary central incisor (SMMCI) is 1 in 50,000 live births [5]. Neonates, being obligate nasal breather, require structurally patent nasal airway to

adopt in the extrauterine life. Any form of nasal obstruction will lead to respiratory distress at this age and needs immediate intervention [5]. CNPAS is an unusual cause of congenital upper airway obstruction in a neonate. Anatomically pyriform aperture is the most narrowest part of the nasal cavity, that if become congenitally stenosed, will cause respiratory distress, cyclical cyanosis, and difficulty in feeding. It may presented as isolated anomaly or associated with other anomalies like holoprosencephaly, midline facial defects, clinodactyly, pituitary dysfunction and most commonly mega incisor (75% of cases) that is our case as well [6].

To confirm the diagnosis CT scan with axial sections in a plane parallel to the hard palate are necessary, demonstrating narrowing of the pyriform aperture and free posterior choanae [7].

Classical radiological findings are narrowed bony inlet, triangular shaped palate, bony overgrowth in the nasal process of maxilla and abnormal dentition like mega incisor [8].

The normal width of pyriform sinus in the age group 0 - 6 months is 8.8 - 17.2 mm. Each pyriform aperture width < 3 mm or whole of the pyriform aperture < 8 mm in term neonate at the level of the inferior meatus between the medial aspects of the nasal process of the maxilla is diagnostic of CNPAS [8].

Embryologically SMMCI arises from 4<sup>th</sup> month of fetal development, by itself is not problematic for the neonate, and it is usually left untreated. Deletion of chromosome 7 and 8 are associated with SMMCI; these are the chromosomes, which carry the genes for holoprosencephaly and hence SMMCI is considered as a marker or microform of holoprosencephaly. Thus, co-occurrence of SMMCI with CNPAS mandates magnetic resonance imaging brain to rule out the associated features of holoprosencephaly sequence and pituitary disorders [9].

Once the diagnosis of CNPAS has been made, initial conservative treatment with topical nasal decongestants, intranasal steroids 0.1% dexamethasone drops, 3 - 3.5 mm endotracheal tube stent, humidification of air, insertion of oral airway and gavage feeding are tried [10].

In case of failed conservative pharmacologic management, surgical treatment with widening of bony inlet via sublabbial approach is performed with possible complications; including septal perforation, columellar necrosis and synechiae [10,11].

### Conclusion

CNPAS is an infrequent, potentially fetal but treatable cause of airway obstruction in neonates. It may present as an isolated congenital defect or in association with other anomalies. The neonates with respiratory distress should be screened for possible nasal obstruction by passing a 5 mm NG tube through the anterior nasal valve, especially if the symptoms are not explained. The goals of management are to establish the nasal patency early in the course of evaluation by doing an screening bed side test through NG tube and to manage the other associated anomalies with multidisciplinary teams.

### Bibliography

1. Brown OE., et al. "Congenital nasal pyriform aperture stenosis". *The Laryngoscope* 99.1 (1989): 86-91.
2. Pereira KD and Wang BS. "Congenital nasal pyriform aperture stenosis". *Operative Techniques in Otolaryngology-Head and Neck Surgery* 16.3 (2005): 194-197.
3. Burstein FD and Cohen SR. "Piriform aperture stenosis: a rare cause of neonatal airway obstruction". *Annals of Plastic Surgery* 34.1 (1995): 56.
4. Arlis H and Ward RF. "Congenital nasal pyriform aperture stenosis: isolated abnormality vs developmental field defect". *Archives of Otolaryngology-Head and Neck Surgery* 118.9 (1992): 989-991.

5. Günther L., *et al.* "Clinical course and implications of congenital nasal pyriform stenosis and solitary median maxillary central incisor in a newborn: a case report". *Journal of Medical Case Reports* 8.1 (2014): 215.
6. Utreja A., *et al.* "Solitary median maxillary central incisor in association with hemifacial microsomia: A rare case report and review of literature". *Contemporary Clinical Dentistry* 2.4 (2011): 385.
7. Ramsden JD., *et al.* "Choanal atresia and choanal stenosis". *Otolaryngologic Clinics of North America* 42.2 (2009): 339-352.
8. Sesenna E., *et al.* "Congenital nasal pyriform aperture stenosis: diagnosis and management". *Italian Journal of Pediatrics* 38.1 (2012): 28.
9. Hall RK. "Solitary median maxillary central incisor (SMMCI) syndrome". *Orphanet Journal of Rare Diseases* 1.1 (2006): 12.
10. Collins B., *et al.* "Congenital nasal pyriform aperture stenosis: conservative management". *Annals of Otology, Rhinology and Laryngology* 122.10 (2013): 601-604.
11. Devambez M., *et al.* "Congenital nasal pyriform aperture stenosis: diagnosis and management". *The Cleft Palate-Craniofacial Journal* 46.3 (2009): 262-267.

**Volume 9 Issue 2 February 2020**

**©All rights reserved by Adnan Mirza., *et al.***