Respiratory Distress in Term Newborn Baby - Think Out of The Box: A Case Report

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1. Abstract
Respiratory distress is a common symptom in newborn. It may be a manifestation of benign self-limiting condition to life threatening conditions involving different organs. Upper airway obstruction is a rare cause of respiratory distress. We report a case of respiratory distress in term newborn secondary to congenital pyramid aperture stenosis. Our case highlights the need to think out of the box when faced with difficult situation.

2. Introduction
Respiratory distress in a term baby, presenting soon after birth, may vary in severity. Commonly, it is caused by delay in the resorption of fluid from the lungs. The other common causes are infection, air leak syndromes, meconium aspiration syndrome. Upper airway obstruction is a rare cause of respiratory distress in newborn babies. We present a case of respiratory distress in term newborn secondary to congenital pyramid aperture stenosis.

3. Patient and Observation
Baby girl was born at term with a birth weight of 3.256 Kg. Mother had uneventful current pregnancy with normal antenatal scans. Baby was delivered by Caesarean Section under spinal anaesthesia. There were no risk factors for sepsis. Baby needed bag & mask ventilation to initiate spontaneous breathing. APGARs were 3 and 9 at 1 and 5 minutes of age respectively.

At 7 hours of age, she had sub costal recession and was grunting. Her temperature was 36.4-degree C. Capillary gas showed pH: 7.19, PCO2: 9.53 kpa and Base Excess: - 0.8. She did not require oxygen to maintain normal saturation at that time. Nasogastric tube could be passed through both nostrils. She received intravenous fluids and antibiotics. Chest ray was normal.

At 9 hours of age, she developed oxygen requirement of 40% to maintain oxygen saturation above 93%. She continued to have sub costal recession and grunting. Arterial blood gas showed pH: 7.27, PCO2: 6.48 kpa and Base Excess: -4.1. Over the next 8 hours, oxygen requirement reduced to 25% with improvement in respiratory distress and respiratory acidosis. However, by 24 hours of age, there was worsening of respiratory distress. She was started on CPAP at 6 cms of H2O. Sub costal recessions improved with saturations of 100% in air. Her initial WBC count was 31.27.

It decreased to 22.6 after 2 days. CRP was normal and her blood culture did not show any growth. She received antibiotics for 48 hours. Repeat blood tests to rule out infection were normal. On further review, baby was noticed to have biphasic squeal on auscultation suggesting upper airway noise. Her soft palate was intact. In the presence of normal septic markers and cyclical nature of the respiratory distress that disappeared on insertion of Guedel’s oropharyngeal airway, anatomical airway (nasopharyngeal) obstruction was suspected. CT scan was done to obtain the images from the orbits through the nasal cavity. It showed anterior pyramid aperture stenosis, cleft mandible and an epidermoid in the midline. There was a central mega incisor. She also had an MRI scan of the head which was normal. She was reviewed by ENT surgeons and her larynx was normal. Any other upper airway obstruction was ruled out by ENT surgeons (Figure 1).

She underwent surgery which involved drilling of nasal floor and lateral nasal wall at around 2 weeks of life. She had a stent inserted during surgery. However, postoperatively, she developed worsening of respiratory distress. Stent was noted to be kinked and hence was removed. She had a nasopharyngeal airway inserted at the same time. Cleft mandible did not need any intervention. Following this, she progressed well, had normal weight gain on full feeds and discharged from NICU with adequate training of the parents at around 1.5 months of age.
On discharge from the hospital, parents had learnt to change the nasopharyngeal tube daily. She was doing very well, when reviewed in outpatient clinic at 9 weeks and 5 months of age.

*Figure 1: CT image axial view showing Pyriform aperture stenosis*

4. Discussion

Upper airway obstructions are important cause of respiratory distress in newborn babies. The obstructions range from mild to complete obstructions such as bilateral choanal atresia. One diagnostic clue to nasal obstruction is inability to pass nasogastric tube through the nostrils [1]. However, in our case, nasogastric tube was passed without any difficulty. This can easily mislead the clinician to believe the patency of nasopharynx. Congenital anterior pyriform aperture stenosis is a rare cause of nasal obstruction. First described by Brown et al. in 1989, it is caused by excessive growth of medial nasal process of maxilla that leads to narrowing of the bony part of nasal cavity [2]. This deformity is identified as apart of developmental field defect consisting of midfacial dysostosis associated with endocrine and central nervous system abnormalities [3]. In our case, investigations did not reveal any serious midline abnormality except for cleft in the mandible and central mega incisor. Neonates are preferably nasal breathers, and any cause of nasal obstruction may take them to severe consequences. Pyriform aperture is the narrowest part of the nasal airway, and even minor changes in its cross-sectional area can lead to markedly increase in nasal airway resistance [4]. Timely recognition and appropriate treatment are necessary to prevent asphyxia. A respiratory pattern of cyclic cyanosis relieved by cry and feeding difficulties are reported [5]. These symptoms may also occur in patients with bilateral choanal atresia.

CT scan of head with axial sections obtained from palate to roof of the orbit is the investigation of choice to make the diagnosis and rule out other causes of nasal obstruction. It is equally important to rule out other central nervous system association with appropriate investigations. Milder obstructions can be conservatively managed with McGovern nipple for feeding as well as maintaining oral airway. Surgery is considered for severe cases (4). In a case study of 13 patients by Marrugo, the mean pyriform aperture width was 5.5mm and 31% patients had congenital midnasal stenosis (6). A pyriform aperture width of less than 5.7 mm in a neonate is 88% sensitive and specific in predicting the need for surgical intervention (7).

5. Conclusion

Though rare congenital anterior pyriform aperture stenosis is an important cause of upper airway obstruction in newborns. Bilateral choanal atresia is a differential diagnosis. Inability to pass nasogastric tube through nostril is an important finding but may not be always present. Timely diagnosis and proper treatment can prevent asphyxia.

6. Competing Interests

The authors declare no competing interest.

7. Authors’ Contributions

All authors have read and approved the manuscript. SS: Drafted and critically reviewed the manuscript and approved the final manuscript as submitted. DJ: Drafted the manuscript and approved the final manuscript as submitted.

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