

## Bilateral Congenital Choanal Atresia in a Female Neonate: Importance of Bedside Diagnosis

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### INTRODUCTION

Congenital bilateral choanal atresia is a neonatal emergency in which there is narrowing or imperforation of the posterior nares. The obstruction is usually at the junction of the posterior nasal cavity and the anterior aspect of nasopharynx. The aetiology has been attributed to a persistence of the embryological bucconasal membrane. This membrane separates the nasal cavity from the stomatodeum until it breaks down at about the seventh week of intrauterine development, allowing communication through the primitive posterior nares.<sup>1</sup> It has a prevalence rate of about 1 in 7000 – 10000 live birth<sup>2</sup> and affecting frequently the females about twice more than the males.<sup>3,4,5</sup> It can be bilateral or unilateral, complete or incomplete and purely membranous(10%), purely bony(30%) and mixed bony and membranous(70%).<sup>5</sup> Unilateral atresia is commoner than bilateral atresia in the proportion of 3:2<sup>1</sup> When it is unilateral, the right nostril is affected more than the left nostril at a ratio of 2: 1.<sup>2</sup> Unlike the unilateral atresia which may remain undiagnosed until persistent unilateral rhinorrhoea and nasal obstruction attracts attention, bilateral atresia presents immediately after birth with respiratory distress especially during feeding and requiring immediate intervention.<sup>6,7,8</sup> Congenital atresia is present in 10% to 30% of patients with “CHARGE” syndrome (coloboma of the eye, heart defects, atresia of the choanae, retardation of growth and/or development, genital and/or urinary defects, ear anomalies).<sup>5,8,9</sup> Other anomalies such as Crouzons, Di George, Amniotic band, Fetal alcohol and Treacher Collins Syndromes may be associated with choanal atresia.<sup>4</sup>

Although choanal atresia has been reported in literature from different parts of Nigeria, none to

our knowledge has been reported in Rivers State of Nigeria. We present a case of bilateral choanal atresia in a female neonate who was delivered after 35 weeks of gestation by caesarean section due to prolonged preterm rupture of the membrane and placenta previa. She was managed conservatively and referred to a tertiary centre for definitive care.

The objective of this case report is to describe the modalities of establishing the diagnosis of bilateral congenital choanal atresia in resource limited settings and confirmation using computed tomography.

### CASE REPORT

Baby EC was a 2.8 kg female delivered by a 28-year- old Gravida 2, Para 2 mother via caesarean section in a peripheral hospital in Port Harcourt. Pregnancy was supervised but complicated due to preterm premature rupture of the membrane (diagnosed with nitrazine test) and complete placenta previa (diagnosed by ultrasonography) at 24 weeks of gestation. She was on prolonged admission in the hospital and placed on steroids and antibiotics. She was planned for elective caesarean section at the 37<sup>th</sup> week of gestation but at the 35<sup>th</sup> week, she started bleeding per vaginam warranting an emergency caesarean section. The mother had no antenatal history of smoking, drugs intake, infections or exposure to teratogens.

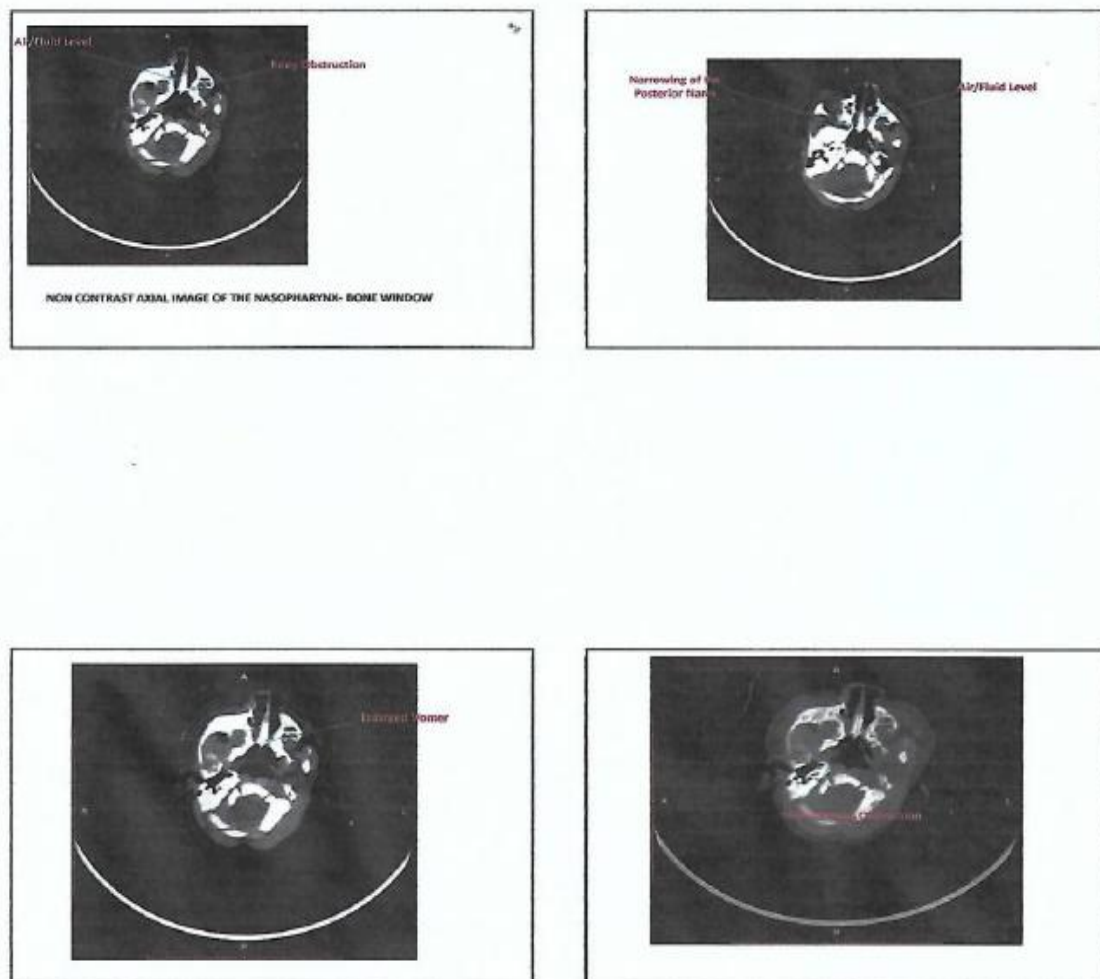
Her APGAR (Appearance, Pulse, Grimace, Activity and Respiration) score at delivery was 7 and 9 at 1st and 5th minute respectively. She developed respiratory distress immediately after birth. Attempts to suction mucous secretions from the nostrils met with resistance. Initial diagnosis of respiratory distress syndrome secondary to prematurity with a differential

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diagnosis of choanal atresia was made. Her PaO<sub>2</sub> was 87% at room air on pulse oxymetry. General physical examination revealed an active baby in respiratory distress evidenced by tachypnoea with a respiratory rate of 48 breaths per minute, substernal and intercostal recessions. The breathing difficulty was relieved by crying but aggravated by trying to suck breast but there was no cyanosis. The nasal examination showed a flat nasal bridge. Placing a wisp of cotton wool in front of the nostrils showed no movement with breathing suggesting nasal occlusion. A size 8 French nasogastric tube was passed into the nostrils. This revealed obstruction at 2.3 cm and 2.5 cm in the left and right nostrils respectively. This raised the possibility of a diagnosis of bilateral choanal atresia.

The cardiac and other systemic examinations were normal. She was placed on oxygen therapy by face mask and commenced on 4.3% dextrose in 0.18% normal saline intravenous infusion and prophylactic intravenous ceftriaxone. Her PaO<sub>2</sub> increased to 97% with the Oxygen therapy. The chest x-ray and echocardiogram were normal.

An orogastric tube was passed for feeding and an oropharyngeal tube passed to aid respiration and strapped with plaster to avoid being extruded. These kept the baby comfortable. She was referred to a tertiary centre for definitive diagnosis and surgery but was returned due to lack of bed space. A Computed Tomogram (CT) of the nose and paranasal sinuses was requested in the mean time to establish a diagnosis. This was done on the fourth day after delivery due to financial constraints. The existence of bilateral bony and membranous choanal atresia (the mixed variety) was confirmed (Figure 1). There were no other associated congenital anomalies found. She was finally moved to the University of Port Harcourt Teaching Hospital for surgical repair/recanalization on the seventh day. Four weeks after the surgery the child with stent in the nostrils (figure 2) was brought to the primary hospital for immunization. Her breathing through the stents was confirmed normal.



**Figure1.** Non contrast axial image of the nasopharynx- bone window



**Figure2.** *Baby EC with stent in the nose*

## **DISCUSSION**

Congenital bilateral choanal atresia is a neonatal emergency in which there is narrowing or imperforation of the posterior nares. Bilateral choanal atresia presents as an emergency immediately after birth with respiratory distress because infants are obligate nasal breathers in the first four months of life due to the nature of their larynx where it appears to have high cervical localization at this age.<sup>6</sup> The typical clinical picture is cyclic cyanosis and respiratory distress aggravated by feeding or sleeping with mouth closed and relieved by crying, as the mouth opens. Similar features were noted in this patient but there was no cyanosis, likely due to the prompt intervention of the attending physician. Due to its rarity, an initial diagnosis of respiratory distress syndrome secondary to prematurity was the first consideration but with a high index of suspicion and inability to pass size 8 French suction tube through the nasal cavity to the pharynx, bilateral congenital choanal atresia was considered as the diagnosis.

A comprehensive physical examination, chest x-ray and echocardiogram was done after the delivery of the baby since about 10% to 30% of patients with choanal atresia have other associated anomalies. Although none of these anomalies was found, further long term follow up will be required.

Prompt diagnosis of congenital bilateral choanal atresia is paramount in the survival of the patient. In the first place, a high index of suspicion is required in the diagnosis in neonates presenting with respiratory symptoms and signs.<sup>2</sup> In low resource settings, practical bedside tests such as: (a) passing a nasal catheter into each nostril and visualizing it at the oropharynx if the choana is patent, (b) placing a piece of cotton wool close to each nostril and observing flipping movement of such cotton wool, (c) introducing methylene blue dye into each nostril and observing its presence at the oropharynx, (d) auscultation over the nostrils to assess airflow and lastly (e) placing a mirror alternately close to one nostril after the other

while observing for the presence of fog on the mirror in presence of patent choanae.<sup>8,10,11,12</sup> Other methods of establishing the diagnosis of choanal atresia is by routine nasal endoscopy in suspected patients and by computerized tomography (CT) scan of the paranasal sinuses. These methods are not readily available in low resource settings and if available are expensive and beyond the reach of most patients. Although with the first two tests, the diagnosis was almost certain, CT scan was requested as a confirmatory investigation to determine the nature and extent of the lesion causing the nasal obstruction including other congenital causes.<sup>5,13,14</sup> This patient had to do a CT scan after a lot of persuasion on the fourth day post delivery because of the cost implications.

On the CT Paranasal Sinus, Axial View-bone window, findings include bony, membranous or mixed bony and membranous obstruction, Air/fluid level, and others include medial bowing of the posterior maxillary sinus, thickening/enlargement of the posterior vomer and airway less than 3mm-measurement at the reference level of the pterygoid plates in the axial plane. Bilateral mixed bony and membranous obstruction with air/fluid levels were clearly demonstrated in this patient(Figure 1). Some differential diagnoses of respiratory distress in a newborn on paranasal CT include - pyriform aperture stenosis, sinonasal mucoid impaction, nasoethmoid cephalocele.

### CONCLUSION AND RECOMMENDATION

It is possible that many of the cases diagnosed as neonatal respiratory distress syndrome could be attributed to choanal atresia. A high index of suspicion is therefore required by clinicians in the diagnosis of rare neonatal conditions such as congenital bilateral choanal atresia. Knowledge of the use of simple and practical bedside techniques when faced by neonatal patients with rhinological complaints is very important to avoid missing such rare but important diagnosis as bilateral choanal atresia. Clinicians should also bear in mind that other congenital anomalies may co-exist with this condition and examine such neonates thoroughly to avoid missing them. It is recommended that referral to appropriate specialists for definitive care should be done as soon as the patient is stabilized. Referred neonates with such life threatening conditions should be given priority attention in tertiary hospitals to reduce neonatal morbidity and mortality.

### ACKNOWLEDGEMENTS

The authors appreciate the management of Bethesda Family Hospital, Port Harcourt for allowing them access to this patient and her records.

### DECLARATION OF PATIENT CONSENT

The authors certify that they have obtained all appropriate patient consent forms. In the form the parents gave their consent for the images and other clinical information to be reported in the journal. The parents understand that their names will not be published and due efforts will be made to conceal their identity.

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**Citation:** Paul O. Diénye et.al, “Bilateral Congenital Choanal Atresia in a Female Neonate: Importance of Bedside Diagnosis”, *International Journal of Research Studies in Medical and Health Sciences*. 2020; 5(1): 03-07.

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