Masking of the Symptoms of Bilateral Congenital Choanal Atresia by a Coexisting Anomaly of the Upper Lip

Congenital choanal atresia (CCA) is the developmental failure of the nasal cavity to communicate with nasopharynx. Bilateral CCA is an acute emergency in an obligatory nasal breathing neonate. The bilateral involvement in this anomaly presents immediately after birth as respiratory distress and cyanosis, except while crying. We herein describe a newborn with bilateral CCA presenting with no distress either at birth or subsequently during rest, due to a co-existing anomaly.

A full-term male neonate was born by normal vaginal delivery in an island hospital at Andamans, India. Mother had uneventful antenatal and intrapartum course. At birth, he weighed 3100 grams and cried well. Baby was noted to have an incomplete cleft lip along with coloboma of the left eye lid flanked by a tissue tag. Neonate had a broad and prominent nasal bridge. Baby was breathing normally with an oxygen saturation of 100% in room air. Neonate was roomed-in with mother after clinically ruling out other serious anomalies like cardiac defects. After one hour of delivery, the baby was noted to have developed restlessness, hypoxia and cyanosis on attempt to breast feed. A possibility of upper airway obstruction, particularly bilateral CCA, was entertained. An attempt to pass the catheter through both the nostrils was unsuccessful, confirming the clinical suspicion. A large button of tissue of lip adjacent to the narrow cleft on upper lip was keeping the mouth open during rest resulting in good breathing through the oropharynx with distress appearing only while feeding (Fig. 1). As facilities to investigate further and manage were not available at the remotely located island hospital, the baby and mother were transferred by air to a tertiary care centre at Kolkata within 24 hours of diagnosis. The imaging studies confirmed bilateral CCA with membranous atresia on the left side and bony atresia on the right. Cardiac echocardiogram was normal. There were no renal, ear, genital or other ocular abnormalities. Obstruction due to membranous atresia was relieved surgically within 24 hours and intervention for the bony obstruction was scheduled for a later date. Baby made an uneventful recovery and was thriving well during follow up review at 6 weeks of life.

Fig. 1 The anomalous button of tissue adjacent to upper lip cleft, keeping the mouth wide.

This infant had persistent opening of the mouth due to a large button of tissue adjacent to a narrow cleft at upper lip, preventing approximation of lips and leaving the mouth sufficiently open to breathe. This association masked the clinical manifestation at rest, even with the mouth ‘closed’. Belegere, et al. [1] described a case in which bilateral CCA associated with craniofacial anomalies did not present with respiratory distress in the neonatal period as the baby had a complete unilateral cleft lip which facilitated oropharyngeal respiration. The case described in this report had a very narrow and incomplete cleft; however, the anomalous large tissue of upper lip prevented closure of mouth and ensured oral airway patency.

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*K M ADHIKARI and #KANNAN VENKATNARAYAN
*Department of Pediatrics, INHS Dhanvantari-Tri Services Naval Hospital Port Blair, A & N Island; and #Department of Pediatrics and Neonatology, Command Hospital, Kolkata; India.
*kmadhikari@gmail.com

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