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Solitary median maxillary central incisor syndrome like rare cause of congenital nasal obstruction

Tania Sofia Leano Martins, Gustavo Rocha and **Hercilia Guimaraes**
Centro Hospitalar Sao Joao, Portugal

Introduction: The solitary median maxillary central incisor syndrome is a rare disease with a prevalence of 1/50000 infants, predominantly female and of unknown etiology in most cases. Some cases are associated with chromosomal abnormalities (chromosome 7 and 18) or mutation of the *SHH* gene. It is characterized by the presence of changes in midline structures: cranial bones (single central incisor), airway (choanal atresia, intra-nasal stenosis and stenosis of the piriform aperture) and brain (holoprosencephaly).

Clinical Case: Preterm 30 weeks gestational age, female, twin pregnancy, bicoriónica/ biamniótica was diagnosed with ultrasound, prenatal: normal and amniocentesis: 46xx. Eutocic delivery at 37 week of gestation was induced with Index of apgar (1'/ 5') 6/7. By somatometry 1335 g/40 cm/27 cm needed invasive mechanical ventilation until day four life and exogenous surfactant for respiratory distress syndrome. In nCPAP until day 36. In 33 day by worsening signs of respiratory distress and suspected nasal obstruction was observed by Otorhinolaryngology. Nasofibroscopy revealed left choanal stenosis. CT nasal sinuses showed changes consistent with SICSS. Cerebral NMR showed no change. Echocardiogram showed persistence minimum ductus arteriosus and patent foramen ovale with left-right shunt. Abdominal ultrasound showed no change. Endocrinological study without changes and research of deletions of chromosomes 18 and 7 and research of mutation of the SHH gene is in progress. On day 43 life underwent septoplasty with placement of tubular stents with a multidisciplinary orientation.

Conclusion: In the presence of congenital nasal obstruction suspected SICSS should be placed as early diagnosis which allows the identification of other congenital anomalies and their correct orientation.

Biography

Tania Sofia Leano Martins has completed her Medical Schooling from Medical School of Porto with 15 values and now is pursuing Pediatrics Specialty. She is a Trainer of advanced support of pediatric life as part of the Portuguese pediatric resuscitation group and Volunteer Teacher of pre-graduated Pediatrics practical and theory classes of Master's degree in Medicine from Oporto Medical Schools. She has attended several courses with evaluation like 11th European Post-graduate course in Neonatal and Pediatric Intensive care. She has presented several clinical studies in national and international conferences and published in the form of book chapter themes: Tuberculosis in pediatrics age and Neutropenia in Pediatrics.

tania_sofia_martins@hotmail.com

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