

AN UNUSUAL ASSOCIATION BETWEEN CRANIOSYNOSTOSIS AND OAVS. REPORT ON TWO CASES

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INTRODUCTION: The first and second pharyngeal arches are embryonic primordium that contributes to craniofacial development. Interferences in normal development of these structures result in variable maxillary, mandibular, and ear abnormalities. These anomalies can be isolated or as part of some unknown and known conditions such as the oculoauriculovertebral spectrum (OAVS). **PURPOSE:** To describe the clinical aspects of two unrelated Brazilian cases with clinical diagnostic of OAVS with craniosynostosis. **METHODS:** Minimal diagnostic criteria were the presence of asymmetric external ear anomaly and craniosynostosis. **CLINICAL REPORT:** Both individuals were examined presenting typical clinical signs of OAVS and an unusual association with craniosynostosis. These individuals were categorized as having in common trigonocephaly, plagiocephaly, facial asymmetry, facial palsy, lagophthalmia, mandibular hypoplastic, anotia/microtia, hearing loss, and developmental and speech delay. Extracraniofacial anomalies such as branchial pit, thumbs hypoplastic, thenar hypoplasia, vertebral abnormality, and heart defect were observed only one of the cases. Cytogenetic study was normal in both cases. **CONCLUSION:** Therefore is possible that the association between craniosynostosis and OAVS represent a subset of OAVS characterizing a new condition within OAVS.