

## AURICULOCONDYLAR SYNDROME IN SIBLINGS: A CASE REPORT

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Auriculocondilar syndrome (ACS) is characterized by typical malformations of the ears called "question mark ears" due to submit constriction between the lower third and middle third, separating the lobe of the helix. Associated with this finding, there is a constellation of abnormalities of the temporomandibular joint. The phenotype is highly variable, which makes the differential diagnosis of syndromes with the 1st and 2nd pharyngeal arches and other craniofacial malformations. The mechanism of inheritance is autosomal dominant recognized. **METHOD / RESULTS:** We describe two brothers, male children of consanguineous couple. Case 1, 16 years with intellectual disabilities, facial asymmetry, palpebral fissures slanted upward, epicanthal folds, micrognathia, deviation from labial to the left, cupped ears, having the right, narrowing between helix and lobe. On the left, the lobe is separated and there is pre-auricular appendage. Patient 2, 23 years with intellectual disabilities, deficiency of coagulation factor XI and psychiatric disorder. Presents ptosis, palpebral fissures slanted upward, epicanthal folds, squint convergent to the right, crowding, micrognathia, cupped ears, with narrowing between the middle and lower third of the helices, lying at the site of lobules, structures similar to propellers and anti-helices. The karyotype of both was 46, XY normal. **CONCLUSION:** The patients presented in this work have a phenotype suggestive of ACS, but have intellectual disability and familial pattern compatible with autosomal recessive inheritance. We propose to manage the syndrome, an auditory initial and periodic and recommend a specialized approach by a multidisciplinary team.