
Ear Anomalies in Apert and Crouzon Syndromes – The Description and Classification of Radiology Evaluation

ABSTRACT

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Introduction: Craniosynostosis (CS) is a premature fusion of cranial sutures associated with rare syndromes. Those syndromes have at least 150 genes identified, and the most common syndromes are associated with FGF. Although there are some abnormalities of external, middle and inner ear in those syndromes, there is a shortage in the literature about the main anomalies in the temporal bone (TB) on imaging examinations and their frequency in patients with Apert syndrome (AS) and Crouzon syndrome (CS).

Objectives: describe the main alterations in the temporal bone on Computed Tomography (CT) scans, classify them and their frequency in the AS and CS.

Methods: evaluation of the structures of the temporal bone using CT scans. Anomalies involving the external, middle and inner ear, large vessels, facial nerve, as well as other significant temporal bone anomalies were evaluated and classified by means of specific classifications and descriptive findings associated with each segment.

Results: Anomalies in the external ear were found 64,3% of AS ears and 81,9% CS ears, the middle ear anomalies were found 92% of AS ears and 81% of CS ears, the inner ear anomalies were found 69,6% of AS ears and 9% of CS ears, the facial nerve was abnormal 48,3% of AS ears and 47,8% of CS ears, the jugular was abnormal 37,5% of AS ears and 54,6 of CS ears the carotid artery was abnormal 14,3% of AS ears and 20,5% of CS ears.

Conclusion: The frequency of the main anomalies in the TB were demonstrated on these rare clinical conditions. The global management of those patients needs to embrace an evaluation of the TB with imaging exams. These findings can be considered phenotypic of the syndromes, and can compose protocols for their description. Furthermore, one can measure how challenging it can be to approach the TB of those patients.

Key words: craniosynostosis, Apert Syndrome, Crouzon Syndrome, ear/abnormalities, computed tomography
