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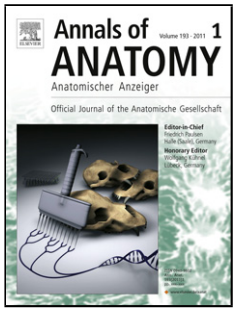

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Maxillary sinuses and midface in patients with cleidocranial dysostosis

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Abstract: The cleidocranial dysplasia is general skeletal disorder with an autosomal dominant inheritance. It is manifested by many craniofacial abnormalities, of which the maxillary hypoplasia is the most evident. The aim of the study was to use CBCT to evaluate the volume of the maxillary sinuses and the dimensions of maxillae in patients with CCD and compare them with healthy individuals. Files of four children with cleidocranial dysplasia were investigated.Volume of every maxillary sinus as well as two dimensional measurements of distances between particular points of interest located on surface of maxilla were calculated from reconstructed CBCT examination. Data were compared with a control group. Statistical analysis was performed. Linear and volumetric data obtained using CBCT was collected and compared with a control group.All affected children had both maxillary sinus volume and maxillary dimensions smaller than control values. The maxillary sinuses were underdeveloped up to half of normal values. The largest differences were recorded in vertical linear dimensions of the maxillae. Horizontal dimensions were also lower. There are morphological modifications of bone tissue which accompany CCD. It seems that these changes occur on the midfacial region and to a greater extent concern the maxillary sinus volume.

Keywords: CCD , CBCT , paranasal sinuses, maxillofacial growth

1. Introduction

The cleidocranial dysplasia (CCD) is general skeletal disorder with an autosomal dominant pattern of inheritance. It was originally thought to involve only bones of membranous origin (Mundlos 1999). Genetic studies report that CCD is caused by mutations involving the transcription factor CBFA1 on chromosome 6p21 (Mundlos et al.,, 1995, Mundlos et al.,, 1997, Quack et al.,, 1999). Abnormal ossification in the CCD results from altered RUNX2 regulation of hypertrophic chondrocyte specific genes during chondrocyte maturation (Zheng et al., 2005). Clinical findings in CCD include abnormalities within both the axial and appendicular skeleton, and are, for example, characterized