

OP29 CRANIOFACIAL MORPHOLOGY AND DENTAL OCCLUSION IN ADULTS WITH OSTEOGENESIS IMPERFECTA – A COMPARISON ACCORDING TO SEVERITY OF DISEASE

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AIM: To compare craniofacial characteristics and variation in dental occlusion according to severity of osteogenesis imperfecta (OI). OI is a rare inherited disease with fragility of bone and teeth because of abnormalities in the formation of collagen.

SUBJECTS AND METHOD: A total of 73 patients with a genetically confirmed diagnosis of OI were recruited. Standardized profile cephalograms were obtained in 68 patients. Cephalometric analyses were performed and cephalometric variables were compared according to OI severity [54 OI type 1 (mild) versus 14 OI type 3 or 4 (severe)]. The comparison was performed by multiple linear regression analyses with the cephalometric variables as outcome variable adjusted for the effect of age, gender, and family clustering. Patients had clinical photographs taken: overjet, overbite, crossbite and posterior open bite were assessed. Statistical analyses were by Fisher's exact test.

RESULTS: In comparison with mildly affected patients, the severely affected patients had significantly ($P < 0.05$) reduced mean (sd) lengths sella-basion [mild: 49 (3.6); severe: 45 (3.1)], basion-bregma [mild: 156 (7.7); severe: 149 (10.3)], basion-lambda [mild: 132 (7.0); severe: 124 (8.6)], and sella-lambda [mild: 136 (6.3); severe: 130 (8.5)], reduced mean (sd) thickness of the parietal theca [mild: 8 (1.8); severe: 6 (1.0)], a reduced maxillary mean (sd) length [mild: 95 mm (6.2); severe: 89 (8.3)] and maxillary posterior mean (sd) height [mild: 50 (3.7); severe: 46 (4.1)], a reduced maxillary prognathism [mild: 83 (3.4); severe: 80 (3.4)], increased mean (sd) maxillary inclination [mild: 6 (4.1); severe: 12 (4.1)], a reduced mandibular mean (sd) length [mild: 129 (7.2); severe: 120 (9.5)] and a reduced mean distance nasion-gnathion [mild: 129 (7.6); severe: 121 (10)]. More severely than mildly affected patients had mandibular overjet (≤ 0 mm) (mild OI: 4%, severe OI 64%) and posterior crossbite (mild OI: 15%, severe OI: 88%).

CONCLUSIONS: The degree of deviation in neurocranial morphology is associated with the severity of OI. The morphology and position of the maxilla is more affected in severe OI than in mild OI, the maxilla being more retrusive and with reduced posterior height. Mandibular overjet is a dominant symptom in severe OI.