Craniofacial development in children, young adults and adults with Prader-Willi syndrome

Background

Prader-Willi syndrome (PWS) is a rare disorder with distinct genetic and clinical features. Among other clinical symptoms, PWS is characterized by infantile hypotonia with feeding problems, followed by obesity and hyperphagia.

PWS is associated with facial dysmorphology, orofacial dysfunction, oral anomalies, low salivary flow and severe tooth wear. Little is known about the craniofacial development of PWS with growth.

Purpose of the study

The purpose of this study was to assess the craniofacial development in children, young adults and adults with PWS, using a cephalometric analysis of lateral cephalograms.

Material and methods

Forty-two individuals with PWS were divided in three groups by age:

<12 years (children, n=12), 12-20 years (young adults, n=12) and > 20 years (adults, n=20) and compared with a matching age and gender control group. The cephalograms were analysed.

References

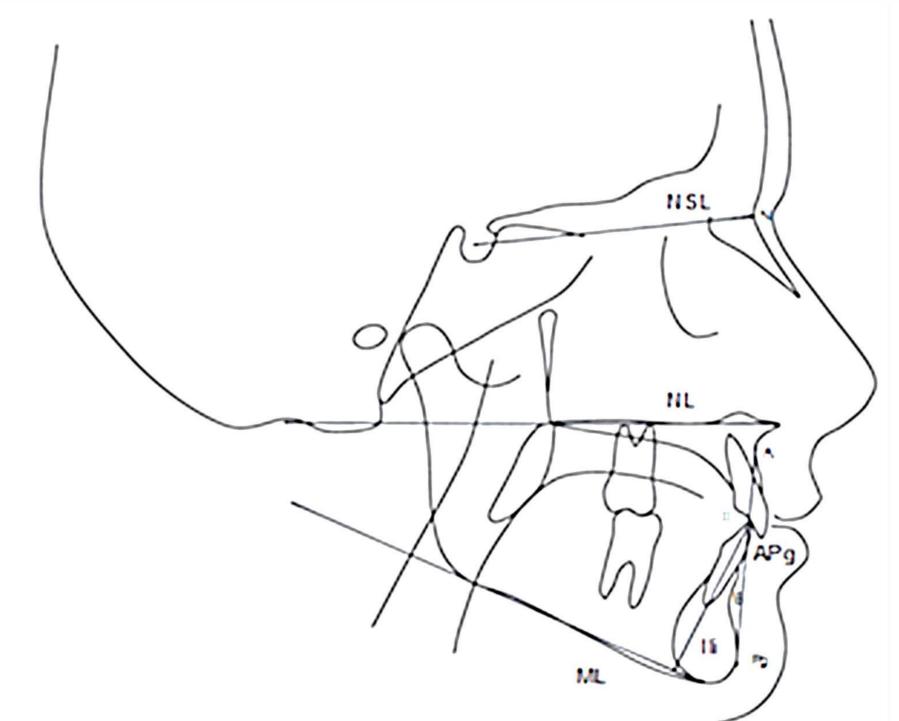
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Results

Sagittal relationships: A-N-B is larger in PWS children and reduced in PWS adults than the control.

Vertical dimensions: NL/NSL is larger in PWS children and decreased in PWS young adults than the control. ML/NSL is larger in the PWS children and young adults than the control. N-Sp and Sp-Gn were higher in PWS children and young adults compared with control.

Growth direction: FA/NBa is higher in PWS children and lower in PWS adults than the control.

Dental occlusion: IIs/NSL and IIs/APg were larger in the PWS adults than the control. IIi/ML and IIi/APg were reduced in the PWS adults than the control.

Conclusions

It seems that, with growth, the craniofacial vertical dimensions in PWS is reduced. The mandibula moves to an anterior position relative to the maxilla. The growth direction changes to a horizontal and anterior position. The maxillary incisors procline and the mandibular incisors retrocline as a dental compensation.

This study may contribute to a better understanding of the craniofacial development in PWS, and may be of clinical interest when setting up a treatment plan.