

Occlusal plane orientation in Klinefelter syndrome (47,XXY males)

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SUMMARY Occlusal plane position was analysed cephalometrically in 35 Klinefelter adults (47,XXY) and compared with 60 euglycemic control males (46,XY). The significantly smaller angles between the occlusal plane and the cranial base (NSL-OLs) and between the occlusal plane and the Frankfort horizontal plane (Fr-OLs) were obtained in 47,XXY males ($P < 0.01$), while the angles between the maxillary base and the occlusal plane (NL-OLs) and between the Camper's line and the occlusal plane (Camp-OLs) were not significantly different ($P > 0.05$) from the control group. Significantly smaller angles between the occlusal plane and the cranial base (NSL-OLs) and between the occlusal plane and the Frankfort horizontal plane (Fr-OLs)

in Klinefelter males are attributed to the hereditary influence of an extra X chromosome on the smaller growth of the cranial base and the greater growth of the lower border of the mandible. Although the maxilla was also shifted forward in XXY males in relation to the cranial base it was not enough to compensate for the hereditary influence, due to the greater growth of the lower border of the mandible and the smaller cranial base in 47, XXY males, on the inclination of the occlusal plane to the Frankfort horizontal plane and the cranial base. The forward shift of the maxilla was sufficient to compensate for the inclination of the occlusal plane in 47, XXY males to the maxillary base and the Camper's line ($P > 0.05$).

Introduction

A chromosomal aneuploidy known as Klinefelter syndrome, characterized by an extra X chromosome in human cells, is manifested in numerous phenotypic differences from normal males, including the differences in craniofacial complex and tooth morphology (Gorlin, Redman & Shapiro, 1965; Darbyshire, Witkop & Cervenka, 1989; Brkić *et al.*, 1992; Brkić *et al.*, 1994a,b).

In 1942, Klinefelter *et al.* first published an article on a syndrome 'characterized by gynaecomastia, aspermatogenesis without a-Leydigism, and an increased excretion of the follicle-stimulating hormone' (Klinefelter, Reifstein & Albright, 1942). Today, the term Klinefelter's syndrome is used to designate the

characteristics associated with the male 47,XXY chromosomal constitutions.

Diagnosis of Klinefelter syndrome is rare in infancy and multiple minor anomalies are usually overlooked until puberty (Bandman & Breit, 1984). When the diagnosis has been established, usually after puberty, multiple minor anomalies in the head and face region can also be noticed, which had been neglected previously (Bandman & Breit, 1984; Varrela & Alvesalo, 1988; Brkić *et al.*, 1994b).

In addition, the effects of an extra X chromosome on the teeth shape and morphology have been reported in these patients (Gorlin *et al.*, 1965; Gardner & Girgis, 1978; Poonawalla *et al.*, 1980; Varela & Alvesalo, 1988; Witkop *et al.*, 1988; Alvesalo, Tammissalo & Townsend, 1991; Brkić *et al.*, 1992; Brkić *et al.*, 1994b).

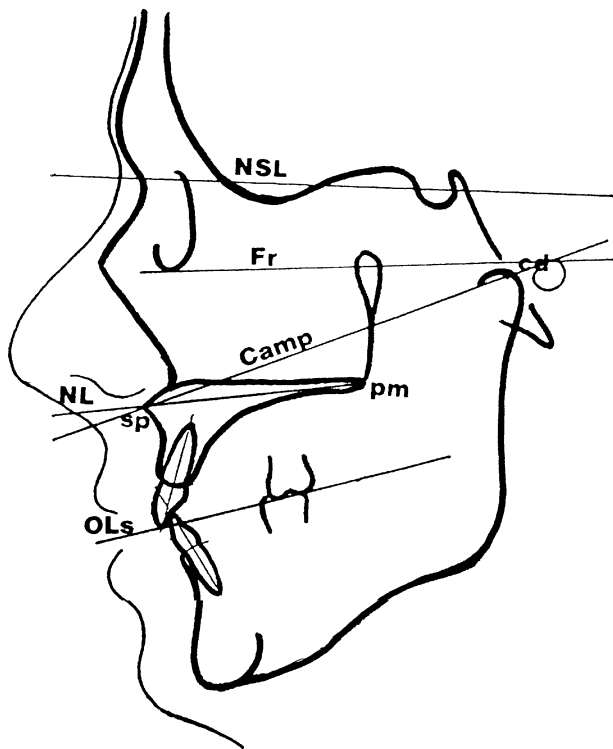


Fig. 1. Reference points and lines on the lateral radiographs. Fr, Frankfort horizontal plane; Ols, Upper occlusal line, the line through the incisal edge of the upper central incisor and the distobuccal cusp of the first upper molar; NSL, nasion-sella line, the line through *n* and *s*; Camp., Camper's line, the line connecting meatus acusticus externus and ale nasi; NL, nasal line, the line through *sp.* and *pm*; ML, mandibular line, the tangent to the lower border of the mandible through *gn*; *sp.*, spinal point, the apex of the anterior nasal spine; *pm*, pterygomaxillare, the intersection between the nasal floor and the posterior contour of the maxilla.

Cleft lip and hemifacial microtia have been reported in one case of XXXY male, but such major deformities seem to be rare (Leon *et al.*, 1959).

From the studies concerning the shape and the size of the craniofacial complex in 47,XXY males it has been reported that the cranial base and the maxilla are smaller, while the mandible is bigger in Klinefelter syndrome compared to normal 46,XY males (Fraser *et al.*, 1961; Babić *et al.*, 1991; Brkić, 1992; Brkić *et al.*, 1994a). Also, the maxilla and the mandible seem to be located forward in relation to the cranial base (Fraser *et al.*, 1961; Gorlin *et al.*, 1965; Babić *et al.*, 1991; Brkić, 1992; Brkić *et al.*, 1994a).

As there is no data in the available literature on the position of the occlusal plane in Klinefelter males, the aim of this study was to examine the orientation of the

occlusal plane in 47,XXY chromosomal constitution to find out if an extra X chromosome might have an influence on the position of the occlusal plane.

Material and methods

Thirty-five Caucasian 47,XXY males whose karyotypes had been determined at the Clinic for Gynaecology and Obstetrics, University Hospital 'Merkur', Zagreb, Croatia participated in this study, which is a part of the project 'Features of craniofacial complex in individuals with gonadal dysgenesis'. The average age of the 47,XXY participants was 27 years.

The control group consisted of 60 Caucasian (46,XY) males, aged 21–32 years who had no previous history of orthodontic treatment and had eugnath jaw relationship (Angle Class I) and all teeth in both jaws.

Lateral skull radiographs with the jaws in habitual occlusion were made* (80 KV and 5 mA ratings). The radiograms obtained were used for the cephalometric measurements suggested by Solow (1966).

The cephalometric analysis consisted of four angular measurements (Fig. 1). The points were marked in pencil on tracing paper, along with the connecting lines. Angular measurements were made with a large protractor to a precision of 0.1 mm. Measurements of the angles between the occlusal plane and the cranial base (NSL-OLS), between the occlusal plane and the Frankfort horizontal plane (Fr-OLS), between the maxillary base and the occlusal plane (NL-OLS) and between the Camper's line and the occlusal plane (Camp.-OLS) were made in all the lateral radiographs with the exception of the Fr-OLS, NSL-OLS, Camp.-OLS and NL-OLS in the 47,XXY males where the first lower molars were missing. Thus, three 47,XXY males were excluded from the measurements.

The reliability of the measurements was tested, as described in a previous study (Brkić *et al.*, 1994a). As the inter- and intra-examiner reliability was satisfactory, all the further measurements were performed by only one examiner, whose measurement results were the most consistent.

Means and standard deviations were evaluated and the significances between the differences were tested by using a *t*-test.

*Ortoceph 5: Siemens, Germany.

Table 1. Cephalometric measurements on the 47,XXY males and the controls (Student *t*-test)

Angle	47,XXY			Control males			<i>t</i>	<i>P</i>
	<i>n</i>	Mean	s.d.	<i>n</i>	Mean	s.d.		
Fr-OLs	32	9.53	3.71	60	11.53	3.38	2.52	**
NSL-OLs	32	12.25	3.91	60	15.63	4.37	2.98	**
Camp.-OLs	32	-7.50	3.69	60	-8.18	7.20	0.52	n.s.
NL-OLs	32	8.69	3.23	60	7.73	3.49	0.83	n.s.

Mean, arithmetic mean (deg); *n*, number of measurements; s.d., - standard deviation; n.s., not significant; * $P < 0.01$.

Results

Means and standard deviations for the measured variables in the both groups and the significance of the differences between the means are shown in Table 1. Statistically significant differences were found in two angular measurements between the 47,XXY and the 46,XY males. The angle between the cranial base and the occlusal plane (NSL-OLs) and the angle between the Frankfort horizontal plane and the occlusal plane (Fr-OLs) were both significantly smaller in the study group (47,XXY males) than in the control group (46,XY males) ($P < 0.01$) (Table 1).

The Camp.-OLs angle and the NL-OLs angle were not significantly different between the Klinefelter males and the normal 46,XY males ($P > 0.05$, Table 1).

Discussion

The life expectancy of 47,XXY males does not differ from the normal 46,XY males and reconstructive prosthodontic treatment might be needed in these males.

It is not an easy task to reconstruct a correct orientation of an occlusal plane in normal eugnath individuals after the partial or complete loss of the teeth, especially in cases of excessive resorption of the lower jaw (Karkazis & Polyzois, 1987; Koller *et al.*, 1992; Čelebić *et al.*, 1993, 1994a, 1995). Moreover, in a disgnath occlusal jaw relationship (Augsburger, 1953; Sinobad, 1988) or in different ethnic groups (Fletcher, 1985) the inclination of an occlusal plane differs from individuals with an eugnath jaw relationship. According to contemporary concepts, the position of the occlusal plane of prosthodontic appliances should be as close as possible to the position which was previously occupied

by the occlusal plane of the natural teeth, in order not to change the afferent proprioceptive and efferent regulatory mechanisms which ensure the normal function of the cheek, tongue and masticatory muscles (Williams, 1982; Čelebić *et al.*, 1989, 1995).

In studies of human aneuploidy a correlation between the presence of an X chromosome and mandibular retrognathism or prognathism was proved, indicating mandibular retrognathism in Turner syndrome (45,X females) (Jensen, 1974, 1985; Peltomaki, Alvesalo & Isotupa, 1989; Kaić *et al.*, 1994; Poje *et al.*, 1996) and mandibular prognathism in Klinefelter syndrome (47,XXY males) (Lyon, 1962; Babić *et al.*, 1991; Brkić *et al.*, 1992, 1994a,b; Brown, Alvesalo & Townsend, 1993).

A smaller cranial and nasal base associated with the longer mandibular base line in Klinefelter's syndrome was reported in several studies (Ingerslev & Kreiborg, 1978; Babić *et al.*, 1991; Brkić *et al.*, 1992, 1994a,b, 1995). Therefore, it was supposed that the occlusal plane orientation is also subjected to the influence of an extra X chromosome in Klinefelter's syndrome.

Therefore the NSL-OLs angle and Fr-OLs angle, as well as the NL-OLs and Camp.-OLs angle were measured on the lateral radiograms of the 47, XXY males and the control males (46,XY). Although the lateral cephalometric measurements relating the occlusal plane to some other landmarks are not very useful for the prediction of the occlusal plane inclination (Karkazis & Polyzois, 1991; Čelebić *et al.*, 1994b), they are still useful tools in studying the maxillomandibular jaw relationship and alveolar bone loss in long term studies (Tallgren, 1972; Tallgren *et al.* 1983; Tallgren, Tryde & Mizutani, 1986) and give us useful information about the inclination of the occlusal plane in different skeletal and ethnic groups (Sinobad, 1988; Čelebić *et al.*, 1994b).

The results of the cephalometric measurements in the present study show significantly smaller NSL-OLs and Fr-OLs angles ($P < 0.01$) in Klinefelter syndrome, while there is no significant difference between the NL-OLs angle ($P > 0.05$) and the Camp.-OLs angle ($P > 0.05$).

Although the nasal line (NL-OLS; maxillary base) is smaller in 47,XXY males, both the maxilla and the mandible seem to be located forward in relation to the cranial base (Brkić *et al.*, 1994a) and this is probably the reason why there was no significant difference ($P > 0.05$, Table 1) for the angle between the occlusal plane and the nasal line (NL-OLs) and the Camp.-OLs

angle. Despite the increased growth of the mandible in 47,XXY males, due to the forward shift of the maxilla the occlusal plane inclination was compensated in relation to the maxillary base (NL-OLs) and the Camper's line (Camp.-OLs) and was not different from the normal 46,XY males.

However, due to the increased growth of the mandible and the smaller cranial base in 47,XXY males (Brkić *et al.*, 1994a), the OLs-NSL and Fr-OLs angles were significantly smaller ($P < 0.01$, Table 1) than in normal males, although both, the maxilla and the mandible were located further forward in relation to the cranial base in 47,XXY males (Brkić *et al.*, 1994a). Obviously, the forward location of the maxilla in relation to the cranial base was not sufficient to compensate for the increased growth of the mandible and the smaller length of the cranial base in 47,XXY males and therefore those with Klinefelter chromosomal constitution had smaller occlusal plane inclination in relation to the Frankfort horizontal plane and the cranial base.

It is well known from the literature that growth and the development are controlled by genetic and environmental factors. The influence of these factors is also reflected in the growth and the development of the craniofacial complex. According to Scott (1967) the parts of the skull which show the greatest amount of genetic control are those which are developmentally most closely related to the chondrocranium and chondrofacial skeleton, namely the midline cranial base and the lower border of the mandible. Although the maxilla was located forward in relation to the cranial base in the study group (Brkić *et al.*, 1994a), it was not sufficient to compensate for the smaller cranial base and an increased size of the mandible. Therefore smaller inclinations of the occlusal plane to the Frankfort horizontal plane and to the cranial base in the 47,XXY chromosomal constitution were obtained in this study. The difference in occlusal plane position in 47,XXY males is attributed to the differences in the growth of the cranial base and the mandible, which are so large that the environmental factors are not able to compensate.

The data on the position of occlusal plane in Klinefelter syndrome obtained in this study are not only of anthropologic, but also of prosthodontic interest, as life expectancy in 47,XXY males does not differ from the normal chromosomal constitution. Due to the changes which might occur after the loss of some teeth and the resorption of the mandible it might be difficult

to locate the original occlusal plane position and the results obtained might be helpful in reconstructive treatment of 47,XXY males.

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