# Craniofacial Morphology in Klinefelter Syndrome A Roentgencephalometric Investigation

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The purpose of the present investigation was to compare the *craniofacial morphology* in a group of subjects with *Klinefelter syndrome* (XXY) with a control group in order to discover whether the abnormal *chromosome* constitution combined with the related endocrinologic aberrations had influenced craniofacial morphogenesis.

The sample was comprised of 37 adult subjects with true Klinefelter syndrome. The control group consisted of 102 adult male students. For each subject, *roentgencephalometric films* in the lateral and frontal projections were obtained. Twenty-nine angular and 27 linear dimensions were measured directly on each film.

The *calvarium* was smaller, the *cranial base* angle was smaller, and the *gonion angle* was larger in the Klinefelter than in the control group. Furthermore, the subjects with Klinefelter syndrome exhibited increased maxillary and mandibular *prognathism* which could be related to the altered shape of the cranial base.

## Introduction

In 1942 Klinefelter, Reifenstein, and Albright described a syndrome characterized by gynaecomastia, aspermatogenesis without reduced function of the Leydig cells, and increased secretion of follicle stimulating hormone. These anomalies had been reported earlier, but they had not before been considered to be related in their occurrence and thus had not been regarded as a syndrome.

In 1956, it was demonstrated that many patients with Klinefelter syndrome were chromatin positive (Bradbury et al., 1956; Jackson et al., 1956; Plunkett and Barr, 1956; Riis, Johnsen, and Mosbech, 1956), and Nelson (1956) introduced the term "true" Klinefelter syndrome for the chromatin-positive patients with the characteristic features of the syndrome. Jacobs and Strong (1959) showed that chromatin-positive patients with Klinefelter syndrome had 47 chromosomes, the extra one being a sex chromosome X. It was thus established that patients with true Klinefelter syndrome have the chromosome constitution 47, XXY.

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Klinefelter syndrome has been found to occur in 0.2–0.3 per cent of the male population. About 25 per cent of the affected persons are mentally retarded (Mosier, Scott, and Cotter, 1960; Gustavson and Åkesson, 1961; and MacLean and Mitchell, 1962). Oral anomalies seem to be rare, although cleft lip and palate have been reported in these patients (Leon et al., 1959). Gorlin, Redman, and Shapiro (1965) suggested that the palate was shallow and the mandible prognathic in these patients.

Variants of Klinefelter syndrome exhibiting the sex-chromosome constitutions XXXY and XXXXY have been observed. These patients have usually more severe clinical aberrations than those with true Klinefelter syndrome. Fraser et al. (1961) and Scherz and Roeckel (1963) reported severe mental retardation and a high frequency of cleft lip and palate among patients with these variants, while Albright, Smith, and Fraser (1942), Day et al. (1963), Schade, Schoeller, and Toeberg (1963), and Gorlin, Redman, and Shapiro (1965) described mandibular prognathism and Albright, Smith, and Fraser (1942) congenital absence of maxillary central incisors.

The purpose of the present investigation was to describe in detail the craniofacial mor-

phology in adults with true Klinefelter syndrome (XXY) in order to determine whether the altered chromosome constitution, combined with the related endocrinologic aberrations (Frøland, 1968), had influenced the craniofacial morphogenesis. Since the subjects are phenotypically men, a male control group was used.

## Material

Thirty-seven adult subjects with true Klinefelter syndrome (XXY) were examined. These patients had previously been reported from cytogenetic, endocrinologic, psychiatric, and psychologic points of view by Nielsen (1969). In the present investigation, two individuals were excluded from the original sample, one because the sex-chromosome constitution was XXXY and the other because of lack of cooperation. Seventeen (46 per cent) of the 37 subjects examined had or had previously had mental illness. None had cleft lip or palate or displayed congenital absence of maxillary central incisors.

The control group consisted of 102 male dental students. Cephalometric radiographs of these subjects were collected and analysed by Solow (1966). The roentgencephalometric films were remeasured by one of the authors (C.H.I.), and his measurements served as control data in the present study. All measurements in the two groups were performed by the same investigator (C.H.I.) in order to reduce the possibility of systematic errors.

## Method

For each subject, roentgencephalometric films in the lateral and postero-anterior projections and a radiograph of the right forearm were obtained. The roentgencephalometric technique has been described by Björk (1968) and Ingerslev and Solow (1975). For the lateral projection, the enlargement of the midsagittal plane was 5.6 per cent. No corrections were made for the enlargement. The reference points and lines are indicated in Figure 1. For definitions see Björk (1960), Solow (1966), and Kisling (1966).

Twenty-nine angular and 27 linear dimensions describing the craniofacial morphology were measured directly on each film. This set of variables was similar to the variables employed by Ingerslev and Solow (1975) in a study on sex differences in craniofacial morphology. Measurements requiring occlusion of the dental arches were not performed in cases with unstable occlusion resulting from loss of teeth. In order to relate the craniofacial dimensions to other body dimensions, five linear measurements were obtained from the film of the right forearm.

Angles were read to the nearest half degree and linear dimensions to the nearest half millimeter, except for the transverse measurements of the radius and ulna, which were read to the nearest tenth of a millimeter.

The significance of the differences between the variances was tested by Snedecor's F-test

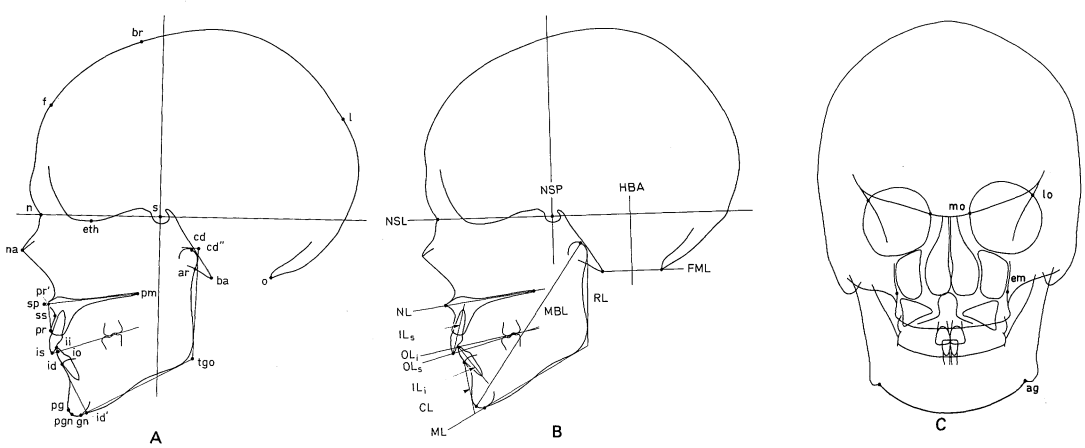


FIGURE 1A. Reference points on the profile cephalometric radiograph.
Figure 1B. Reference lines on the profile cephalometric radiograph.
Figure 1C. Reference points on the postero-anterior cephalometric radiograph. For definitions see Björk (1960), Solow (1966) and Kisling (1966).

and the significance of the differences between the means by the Student t-test. The five per cent and one per cent levels of significance are indicated in the tables by one and two asterisks. In this study, only differences significant at the one per cent level will be considered.

The statistical calculations were carried out at NEUCC, the Northern Europe University Computing Center, Copenhagen.

## Results

### Statistics

The statistical description of the distributions and the differences between the means for the syndrome group and the control group are presented in Table 1. Departures from normal distribution were few and differed for the two groups. The magnitudes of the departures were moderate. There was no agreement between the occurrence of skewness and kurtosis within the groups.

For 19 of the 29 angular measurements and for 11 of the 32 linear dimensions, the differences between the means in the two groups were significant at the one per cent level.

The differences in mean craniofacial morphology are visualized in Figure 2. The tracings are based on the mean dimensions for the two groups.

### Morphology

*Calvarium.* The linear dimensions, s-f, s-br, and l-ba, were all significantly smaller in the Klinefelter group than in the control group. The frontal bone was less prominent, f-n-s, in the Klinefelter group.

*Cranial base.* The cranial base length, n-s, was significantly smaller in the Klinefelter than in the control group. The cranial base angles, n-s-ba, n-s-cd, and n-s-ar, were all found to be smaller in the Klinefelter group than in the controls. There was, however, no significant difference in the eth-s-ba angle or in the antero-posterior diameter of the foramen magnum, ba-o. The inclination of the foramen magnum, represented by the angle NSL/FMP, was somewhat smaller in the Klinefelter than in the control group.

*Pharynx.* The pharyngeal depth, ba-pm, and the pharyngeal angle, pm-s-ba, did not differ significantly in the two groups.

*Nasal bone.* The nasal bone had almost the same length, n-na, in the two groups, but was more protruded in the Klinefelter, s-n-na.

*Orbits.* The inner orbital distance, mo-mo, did not differ significantly in the two groups, whereas the outer orbital distance, lo-lo, was smaller for the Klinefelter than for the control group.

*Maxilla.* The length of the maxillary base, sp-pm and ss-pm, did not differ significantly in the two groups, but the width of the maxilla, em-em, was smaller for the Klinefelter group than for the control group. The maxilla was more prognathic, s-n-sp and s-n-ss, in the syndrome group than in the controls, and the posterior border of the maxilla was located further forward in the Klinefelter subjects, n-s-pm. The upper anterior face height, n-sp, was smaller in the syndrome group, but the upper posterior face height, s-pm, did not differ in the two groups. The maxilla showed a smaller inclination in relation to the anterior cranial base, NSL/NL, in the syndrome group. Neither the inclination of the upper incisors, $IL_s$/NL, nor the inclination of the upper occlusal plane, $OL_s$/NL, differed significantly from the control group.

*Mandible.* The length, pgn-cd, and width, ag-ag, of the mandible did not differ significantly in the two groups. The gonion angle, ML/RL, was larger and the mandible was more prognathic, s-n-pg, in the Klinefelter group than in the control group. The posterior

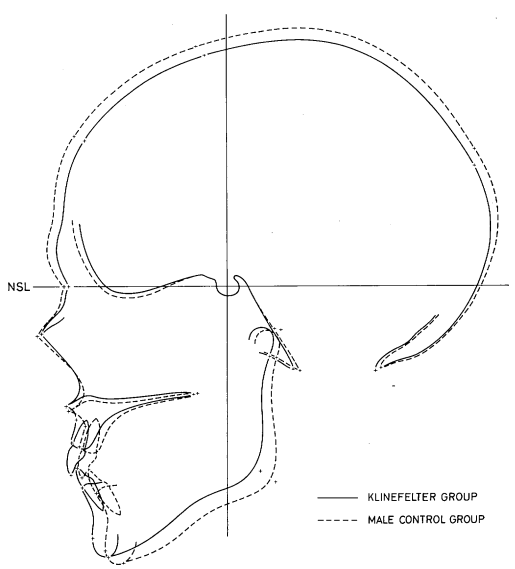


FIGURE 2. Tracings based on the mean X-ray cephalometric dimensions for the two groups. The tracings are superimposed on the nasion-sella-line (NSL) and registered on the sella-point.

vertical dimension, cd-tgo, was smaller in the Klinefelter subjects, whereas the lower anterior face height, sp-gn, did not differ significantly in the two groups. The inclination of the mandible, NSL/ML, in relation to the anterior cranial base, did not differ in the two groups. There was no significant difference in the alveolar prognathism in the mandible, CL/ML, but the inclination of the lower incisors, $IL_i$/ML, was smaller in the syndrome group than in the control group.

*Inter-maxillary relationship.* The sagittal and vertical jaw relationships, ss-n-pg and NL/ML, were significantly greater in the Klinefelter group than in the control group, whereas the interincisal angle, the overjet and the overbite did not differ significantly. None of the Klinefelter patients exhibited mandibular overjet.

*Right forearm.* The distal breadth of the radial head, RDW, was smaller in the Klinefelter group than in the control group. None of the four other measurements of the right forearm showed significant differences at the one per cent level.

## Discussion

In the interpretation of the findings, it should be taken into account that the control group cannot be considered to be representative of the Danish male population. Dental students would be expected to differ physically, educationally, and socioeconomically from corresponding age groups of the population as a whole (Solow, 1966; Dahl, 1970). However, the mean standing height of the Klinefelter group, 176.9 cm, differed only a little and not significantly from the mean height, 178.7 cm, of the control group. Only one of the five measurements on the radiograph of the right forearm was significantly smaller in the Klinefelter group. On the other hand, although the "somatic parameters" did not reveal differences significant at the one per cent level, the mean values were all smaller in the Klinefelter than in the control group. This finding might also be a factor with respect to the smaller dimension of the brain case in the Klinefelter group.

Craniofacial morphology differed significantly in the two groups. The calvarium was located further back and downwards in relation to the anterior cranial base in the Klinefelter group than in the control group, and at the same time, the cranial base angle was smaller. This is consistent with the relationship between the form of the calvarium and the flexion of the cranial base found in a normal sample by Björk (1955). However, the cranial base angle was only significantly smaller in the Klinefelter group when measured to nasion, whereas the eth-s-ba angle did not differ significantly in the two groups. To obtain a visual impression of the possible influence of the location of the nasion-sella line on the craniofacial shape evaluation, the two mean tracings were superimposed on the eth-s-line (ESL) and registered on sella (Figure 3). Analyzed in this way, it would appear that part of the differences in craniofacial shape found in the present study could be explained by a different location of nasion in the two groups, i.e. the shape of the calvarium, the inclination of the nasal bone, the inclination of the maxilla, and the increased prognathism of the jaws etc.

The larger gonion angle and the smaller posterior mandibular height in the Klinefelter subjects seemed to indicate an underdevelopment of the gonion region which again might suggest an altered muscular function (Møller, 1966). The smaller inclination of the mandibular incisors could also be explained by the deviant shape of the mandible.

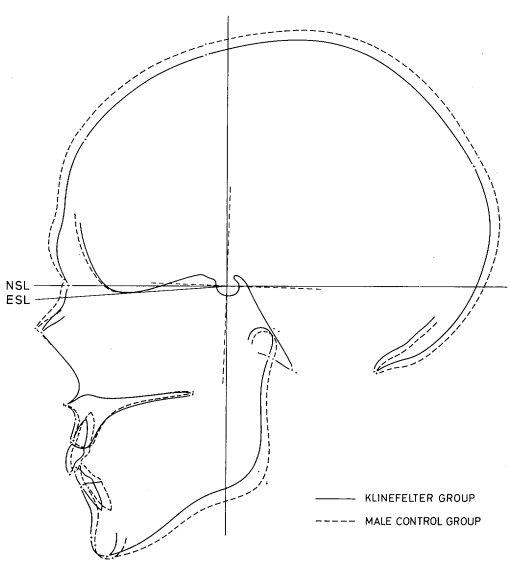


FIGURE 3. Tracings based on the mean X-ray cephalometric dimensions for the two groups. The tracings are superimposed on the eth-sella-line (ESL) and registered on the sella point.

TABLE 1. Statistical data for Klinefelter Group (1) and control group (2)

| Variable | Group | N | min. | max. | $\bar{x}$ | S.E. | S.D. | Skewiness $\sqrt{b1}$ | Kurtosis b2 | Kurtosis a | Diff. |
|---|---|---|---|---|---|---|---|---|---|---|---|
| 1. f-n-s | 1 | 37 | 77.0 | 94.0 | 83.70 | 0.593 | 3.61 | 0.423 | 3.27 | 0.7967 | −1.92** |
| | 2 | 102 | 74.5 | 93.0 | 85.62 | 0.350 | 3.53 | −0.370 | 3.45 | 0.7777 | |
| 2. n-s-ba | 1 | 37 | 118.0 | 140.5 | 128.20 | 0.929 | 5.65 | 0.129 | 2.49 | 0.7991 | −3.31** |
| | 2 | 102 | 118.0 | 145.0 | 131.51 | 0.534 | 5.39 | 0.205 | 2.64 | 0.8349* | |
| 3. eth-s-ba | 1 | 35 | 115.5 | 138.5 | 123.61 | 0.920 | 5.44 | 0.571 | 2.93 | 0.8317 | −1.59 |
| | 2 | 102 | 111.0 | 137.0 | 125.20 | 0.555 | 5.60 | 0.058 | 2.37 | 0.8429* | |
| 4. NSL/FMP | 1 | 37 | 77.0 | 101.5 | 89.23 | 0.944 | 5.74 | 0.469 | 2.79 | 0.7899 | −1.87* |
| | 2 | 102 | 74.5 | 101.0 | 91.10 | 0.448 | 4.52 | −0.729** | 4.27* | 0.7725 | |
| 5. n-s-cd | 1 | 33 | 120.0 | 142.0 | 129.64 | 1.067 | 6.13 | 0.529 | 2.49 | 0.7956 | −4.65** |
| | 2 | 102 | 116.5 | 119.5 | 134.29 | 0.592 | 5.98 | 0.030 | 3.06 | 0.7957 | |
| 6. n-s-ar | 1 | 33 | 111.5 | 130.0 | 120.42 | 0.763 | 4.38 | 0.378 | 2.61 | 0.7814 | −3.77** |
| | 2 | 102 | 112.5 | 136.5 | 124.19 | 0.196 | 5.00 | 0.117 | 2.50 | 0.8323 | |
| 7. n-s-pm | 1 | 36 | 66.0 | 78.5 | 72.06 | 0.528 | 3.17 | 0.058 | 2.23 | 0.8358 | −2.20** |
| | 2 | 102 | 67.0 | 82.5 | 74.26 | 0.302 | 3.05 | 0.044 | 2.72 | 0.8182 | |
| 8. pm-s-ba | 1 | 37 | 46.0 | 68.0 | 56.19 | 0.839 | 5.10 | 0.156 | 2.46 | 0.8352 | −0.20 |
| | 2 | 102 | 46.0 | 69.0 | 56.39 | 0.501 | 5.06 | 0.279 | 3.07 | 0.7714 | |
| 9. s-n-na | 1 | 37 | 102.0 | 139.0 | 121.20 | 1.142 | 6.95 | −0.383 | 4.24 | 0.7468 | 3.83** |
| | 2 | 102 | 99.0 | 135.5 | 117.37 | 0.608 | 6.14 | −0.378 | 3.52 | 0.7838 | |
| 10. s-n-sp | 1 | 36 | 82.5 | 99.5 | 91.06 | 0.653 | 3.92 | −0.278 | 2.77 | 0.7809 | 3.32** |
| | 2 | 102 | 81.0 | 96.5 | 87.74 | 0.328 | 3.31 | 0.229 | 2.66 | 0.8027 | |
| 11. s-n-ss | 1 | 33 | 80.5 | 91.5 | 86.47 | 0.542 | 3.11 | −0.035 | 1.85 | 0.8710* | 5.09** |
| | 2 | 102 | 75.0 | 90.0 | 81.38 | 0.311 | 3.14 | 0.369 | 2.78 | 0.7888 | |
| 12. NSL/NL | 1 | 37 | −3.0 | 13.0 | 5.00 | 0.603 | 3.67 | −0.086 | 2.99 | 0.7689 | −2.62** |
| | 2 | 102 | 2.5 | 16.5 | 7.62 | 0.292 | 2.95 | 0.486* | 3.08 | 0.8168 | |
| 13. s-n-pr | 1 | 32 | 83.0 | 95.0 | 88.92 | 0.532 | 3.01 | 0.185 | 2.32 | 0.8100 | 4.47** |
| | 2 | 102 | 78.5 | 92.0 | 84.45 | 0.301 | 3.04 | 0.444* | 2.68 | 0.8059 | |
| 14. $IL_s$/NL | 1 | 32 | 96.5 | 136.0 | 111.55 | 1.534 | 8.68 | 0.645* | 3.33 | 0.8034 | 0.70 |
| | 2 | 102 | 93.0 | 128.0 | 110.85 | 0.644 | 6.51 | −0.326 | 3.01 | 0.7953 | |
| 15. $OL_s$/NL | 1 | 31 | −0.5 | 15.5 | 8.73 | 0.744 | 4.14 | 0.231 | 2.32 | 0.8404 | 1.22 |
| | 2 | 102 | −2.5 | 18.0 | 7.51 | 0.340 | 3.44 | 0.190 | 3.98* | 0.7670 | |
| 16. NSL/$OL_s$ | 1 | 31 | 3.0 | 20.5 | 12.89 | 0.777 | 4.33 | −0.359 | 2.89 | 0.7851 | −2.55** |
| | 2 | 102 | 5.0 | 27.5 | 15.44 | 0.411 | 4.15 | 0.155 | 3.40 | 0.7671 | |
| 17. s-n-is | 1 | 32 | 84.5 | 96.0 | 89.47 | 0.536 | 3.03 | 0.165 | 2.34 | 0.8287 | 4.66** |
| | 2 | 102 | 76.5 | 93.0 | 84.81 | 0.315 | 3.18 | 0.208 | 2.86 | 0.7923 | |
| 18. s-n-pg | 1 | 33 | 78.0 | 91.5 | 84.64 | 0.611 | 3.51 | −0.270 | 2.43 | 0.7987 | 3.67** |

| | | | | | | | | | | | |
|---|---|---|---|---|---|---|---|---|---|---|---|
| | 2 | 102 | 73.0 | 90.0 | 80.97 | 0.318 | 3.21 | 0.155 | 3.09 | 0.7914 | |
| 19. s-n-id | 1 | 33 | 80.5 | 92.0 | 86.12 | 0.547 | 3.14 | −0.066 | 2.43 | 0.7896 | 4.71** |
| | 2 | 102 | 72.5 | 88.5 | 81.41 | 0.295 | 2.97 | 0.094 | 2.97 | 0.8076 | |
| 20. n-s-tgo | 1 | 33 | 94.0 | 109.0 | 99.97 | 0.689 | 3.96 | 0.354 | 2.29 | 0.8480 | −3.47** |
| | 2 | 102 | 95.5 | 112.0 | 103.44 | 0.360 | 3.63 | 0.336 | 2.62 | 0.8108 | |
| 21. NSL/ML | 1 | 33 | 20.0 | 41.0 | 29.52 | 0.886 | 5.09 | 0.117 | 2.61 | 0.8076 | 1.52 |
| | 2 | 102 | 13.5 | 46.5 | 28.00 | 0.583 | 5.89 | 0.071 | 3.19 | 0.7903 | |
| 22. NSL/MBL | 1 | 33 | 46.0 | 63.5 | 53.30 | 0.698 | 4.01 | 0.403 | 2.94 | 0.7975 | −1.47* |
| | 2 | 102 | 46.0 | 67.5 | 54.77 | 0.387 | 3.91 | 0.362 | 3.31 | 0.7825 | |
| 23. ML/RL | 1 | 37 | 111.0 | 140.0 | 125.08 | 1.051 | 6.39 | 0.017 | 2.62 | 0.8360 | 4.94** |
| | 2 | 102 | 103.5 | 134.5 | 120.14 | 0.627 | 6.33 | −0.369 | 2.99 | 0.7776 | |
| 24. CL/ML | 1 | 34 | 63.0 | 81.5 | 70.88 | 0.804 | 4.69 | 0.506 | 2.60 | 0.8250 | 0.41 |
| | 2 | 102 | 55.0 | 87.0 | 70.47 | 0.667 | 6.74 | 0.138 | 2.75 | 0.7951 | |
| 25. $IL_i$/ML | 1 | 34 | 78.0 | 107.0 | 92.24 | 1.191 | 6.94 | 0.012 | 2.51 | 0.8251 | −6.23** |
| | 2 | 102 | 80.5 | 119.0 | 98.47 | 0.719 | 7.26 | 0.042 | 2.90 | 0.7928 | |
| 26. $OL_i$/ML | 1 | 31 | 11.0 | 29.5 | 19.11 | 0.800 | 4.46 | 0.800* | 3.23 | 0.7894 | 1.71* |
| | 2 | 102 | 6.5 | 29.5 | 17.40 | 0.411 | 4.15 | 0.235 | 3.12 | 0.7995 | |
| 27. ss-n-pg | 1 | 32 | −3.5 | 6.0 | 1.91 | 0.414 | 2.34 | 0.115 | 2.45 | 0.8130 | 1.68** |
| | 2 | 102 | −7.0 | 8.5 | 0.23 | 0.299 | 3.01 | 0.270 | 3.31 | 0.7856 | |
| 28. NL/ML | 1 | 33 | 15.0 | 35.5 | 24.76 | 0.881 | 5.06 | 0.313 | 2.91 | 0.7740 | 4.42** |
| | 2 | 102 | 7.5 | 35.5 | 20.34 | 0.570 | 5.75 | 0.338 | 2.99 | 0.8084 | |
| 29. $IL_s/IL_i$ | 1 | 32 | 93.5 | 162.0 | 130.09 | 2.429 | 13.74 | −0.170 | 3.30 | 0.8253 | −1.64 |
| | 2 | 102 | 107.5 | 160.0 | 131.73 | 1.087 | 10.98 | −0.014 | 2.85 | 0.8010 | |
| 30. s-f | 1 | 37 | 83.5 | 102.0 | 92.20 | 0.674 | 4.10 | 0.022 | 3.26 | 0.7331* | −4.01** |
| | 2 | 102 | 86.5 | 108.0 | 96.21 | 0.443 | 4.47 | 0.136 | 2.81 | 0.8069 | |
| 31. s-br | 1 | 37 | 94.0 | 116.0 | 104.12 | 0.773 | 4.70 | 0.231 | 3.05 | 0.7872 | −4.84** |
| | 2 | 102 | 94.0 | 120.5 | 108.96 | 0.476 | 4.80 | −0.036 | 3.54 | 0.7569* | |
| 32. br-1 | 1 | 33 | 104.0 | 149.5 | 130.23 | 1.690 | 9.71 | −0.469 | 3.47 | 0.7581 | −2.30 |
| | 2 | 97 | 118.0 | 152.5 | 132.53 | 0.624 | 6.15 | 0.275 | 3.19 | 0.7953 | |
| 33. 1-ba | 1 | 34 | 113.0 | 138.0 | 123.01 | 1.137 | 6.63 | 0.357 | 2.01 | 0.8801** | −4.24** |
| | 2 | 96 | 113.0 | 140.5 | 127.25 | 0.534 | 5.23 | −0.009 | 2.97 | 0.8036 | |
| 34. n-s | 1 | 37 | 65.5 | 78.5 | 71.58 | 0.445 | 2.70 | 0.330 | 3.25 | 0.7640 | −1.80** |
| | 2 | 102 | 67.0 | 81.0 | 73.38 | 0.308 | 3.11 | 0.191 | 2.40 | 0.8270 | |
| 35. s-ba | 1 | 37 | 40.5 | 53.5 | 47.62 | 0.526 | 3.20 | −0.064 | 2.51 | 0.8145 | −1.31* |
| | 2 | 102 | 42.5 | 55.0 | 48.93 | 0.283 | 2.86 | −0.085 | 2.36 | 0.8336 | |
| 36. ba-o | 1 | 37 | 31.5 | 42.5 | 36.91 | 0.486 | 2.96 | −0.177 | 2.23 | 0.8335 | 0.12 |
| | 2 | 102 | 30.0 | 45.5 | 36.79 | 0.325 | 3.28 | 0.133 | 2.55 | 0.8270 | |
| 37. ba-pm | 1 | 37 | 38.5 | 55.5 | 47.09 | 0.658 | 4.00 | 0.042 | 2.27 | 0.8251 | 0.99 |
| | 2 | 102 | 38.5 | 54.5 | 46.10 | 0.318 | 3.21 | 0.054 | 2.74 | 0.8109 | |
| 38. n-na | 1 | 37 | 17.5 | 33.5 | 24.55 | 0.627 | 3.81 | 0.340 | 2.80 | 0.7720 | −0.47 |

TABLE 1. *continued*

| Variable | Group | N | min. | max. | $\bar{x}$ | S.E. | S.D. | Skewness $\sqrt{b1}$ | Kurtosis b2 | Kurtosis a | Diff. |
|---|---|---|---|---|---|---|---|---|---|---|---|
| | 2 | 102 | 18.0 | 33.5 | 25.02 | 0.295 | 2.98 | 0.195 | 3.02 | 0.8053 | |
| 39. mo-mo | 1 | 37 | 22.0 | 30.5 | 26.70 | 0.356 | 2.17 | 0.149 | 2.37 | 0.8327 | 0.55 |
| | 2 | 102 | 21.5 | 32.5 | 26.15 | 0.229 | 2.31 | −0.122 | 2.50 | 0.8307 | |
| 40. lo-lo | 1 | 37 | 87.0 | 104.0 | 93.58 | 0.694 | 4.22 | 0.465 | 2.63 | 0.8254 | −3.19** |
| | 2 | 102 | 89.5 | 104.5 | 96.77 | 0.300 | 3.03 | −0.069 | 2.94 | 0.7939 | |
| 41. sp-pm | 1 | 36 | 49.0 | 65.0 | 57.64 | 0.636 | 3.82 | −0.193 | 2.87 | 0.7894 | −0.52 |
| | 2 | 102 | 51.0 | 65.0 | 58.16 | 0.281 | 2.83 | −0.000 | 2.65 | 0.8062 | |
| 42. ss-pm | 1 | 33 | 47.0 | 60.5 | 53.94 | 0.594 | 3.41 | 0.006 | 2.90 | 0.7471 | 1.02* |
| | 2 | 102 | 46.5 | 60.0 | 52.92 | 0.238 | 2.41 | 0.117 | 3.15 | 0.7934 | |
| 43. em-em | 1 | 33 | 54.0 | 72.5 | 63.58 | 0.630 | 3.62 | 0.141 | 3.60 | 0.7864 | −3.43** |
| | 2 | 102 | 60.5 | 74.5 | 67.01 | 0.313 | 3.17 | 0.094 | 2.60 | 0.8032 | |
| 44. n-sp | 1 | 37 | 46.5 | 63.5 | 53.50 | 0.592 | 3.60 | 0.588 | 3.78 | 0.7375* | −2.16** |
| | 2 | 102 | 48.5 | 64.0 | 55.66 | 0.304 | 3.07 | 0.013 | 2.85 | 0.7865 | |
| 45. s-pm | 1 | 37 | 44.0 | 58.0 | 51.03 | 0.513 | 3.12 | 0.161 | 2.51 | 0.8360 | 0.81 |
| | 2 | 102 | 44.5 | 58.5 | 50.22 | 0.286 | 2.88 | 0.131 | 2.63 | 0.8294 | |
| 46. sp-is | 1 | 32 | 26.0 | 37.5 | 30.67 | 0.560 | 3.17 | 0.443 | 2.28 | 0.8407 | −0.03 |
| | 2 | 102 | 23.5 | 37.0 | 30.70 | 0.271 | 2.74 | −0.121 | 2.66 | 0.7985 | |
| 47. pr-pr′ | 1 | 32 | 12.0 | 21.0 | 16.39 | 0.434 | 2.46 | 0.089 | 2.27 | 0.8139 | −0.29 |
| | 2 | 102 | 11.5 | 23.0 | 16.68 | 0.238 | 2.41 | 0.028 | 2.87 | 0.7799 | |
| 48. pgn-cd | 1 | 37 | 115.5 | 139.0 | 126.05 | 0.947 | 5.76 | 0.119 | 2.25 | 0.8601 | 0.20 |
| | 2 | 102 | 114.5 | 141.0 | 125.85 | 0.492 | 4.97 | 0.276 | 3.45 | 0.7768 | |
| 49. ag-ag | 1 | 37 | 81.0 | 98.0 | 89.14 | 0.778 | 4.73 | 0.458 | 2.16 | 0.8247 | −1.02 |
| | 2 | 102 | 79.0 | 102.0 | 90.16 | 0.436 | 4.40 | 0.186 | 2.89 | 0.8119 | |
| 50. sp-gn | 1 | 33 | 62.0 | 84.5 | 71.86 | 0.903 | 5.19 | 0.467 | 3.26 | 0.7633 | −0.38 |
| | 2 | 102 | 58.5 | 86.5 | 72.24 | 0.519 | 5.24 | 0.083 | 2.96 | 0.7980 | |
| 51. s-ar | 1 | 33 | 29.0 | 42.5 | 36.08 | 0.539 | 3.10 | 0.033 | 2.73 | 0.8119 | −2.72** |
| | 2 | 102 | 32.0 | 47.5 | 38.80 | 0.331 | 3.35 | 0.446* | 2.87 | 0.7926 | |
| 52. s-tgo | 1 | 33 | 74.5 | 94.5 | 84.44 | 0.874 | 5.02 | −0.212 | 2.59 | 0.7983 | −5.45** |
| | 2 | 102 | 75.5 | 101.5 | 89.89 | 0.558 | 5.63 | −0.298 | 2.90 | 0.7857 | |
| 53. cd″-tgo | 1 | 37 | 52.5 | 72.5 | 64.41 | 0.733 | 4.46 | −0.429 | 3.22 | 0.7626 | −3.94** |
| | 2 | 102 | 57.5 | 82.0 | 68.35 | 0.462 | 4.67 | −0.110 | 2.77 | 0.8038 | |
| 54. id-id′ | 1 | 34 | 28.5 | 40.0 | 33.04 | 0.458 | 2.67 | 0.613 | 3.47 | 0.7458 | 0.26 |
| | 2 | 102 | 27.5 | 41.5 | 32.78 | 0.264 | 2.66 | 0.374 | 3.32 | 0.7961 | |
| 55. is-io | 1 | 31 | 1.0 | 9.5 | 3.55 | 0.318 | 1.77 | 1.445** | 5.71 | 0.7373* | 0.34 |
| | 2 | 102 | −1.0 | 9.5 | 3.21 | 0.149 | 1.50 | 1.179** | 6.51** | 0.7246** | |

| | | | | | | | | | | | |
|---|---|---|---|---|---|---|---|---|---|---|---|
| 56. ii-io | 1 | 31 | 0.0 | 7.0 | 2.34 | 0.294 | 1.63 | 1.019** | 3.96 | 0.7660 | −0.38 |
| | 2 | 102 | −1.0 | 7.5 | 2.72 | 0.159 | 1.60 | 0.042 | 2.83 | 0.8179 | |
| 57. RML | 1 | 37 | 233.0 | 290.0 | 262.22 | 2.107 | 12.82 | 0.111 | 2.59 | 0.8311 | 0.40 |
| | 2 | 102 | 235.5 | 290.0 | 261.82 | 1.218 | 12.30 | −0.041 | 2.10** | 0.8562** | |
| 58. RHW | 1 | 37 | 20.5 | 29.6 | 24.75 | 0.335 | 2.04 | 0.551 | 3.24 | 0.7620 | 0.60* |
| | 2 | 102 | 21.5 | 27.0 | 24.15 | 0.120 | 1.21 | 0.035 | 2.69 | 0.8129 | |
| 59. RDW | 1 | 37 | 29.6 | 44.0 | 34.99 | 0.397 | 2.41 | 1.158** | 6.97 | 0.6658** | −0.92** |
| | 2 | 102 | 32.2 | 40.3 | 35.91 | 0.179 | 1.81 | 0.160 | 2.36 | 0.8256 | |
| 60. UML | 1 | 37 | 247.0 | 312.0 | 280.49 | 2.178 | 13.25 | −0.096 | 2.98 | 0.8221 | −1.35 |
| | 2 | 102 | 249.5 | 305.5 | 281.84 | 1.221 | 12.34 | −0.161 | 2.34* | 0.8398* | |
| 61. UHW | 1 | 37 | 16.3 | 21.7 | 18.44 | 0.239 | 1.46 | 0.357 | 2.17 | 0.8726 | 0.03 |
| | 1 | 102 | 14.7 | 20.9 | 18.41 | 0.116 | 1.17 | −0.286 | 3.17 | 0.7900 | |

* Sig. at the one per cent level of confidence.
** Sig. at the five per cent level of confidence.

Thus, the presence of an extra X-chromosome, combined with the related endocrinologic aberrations in Klinefelter syndrome (Frøland, 1969), seem to influence craniofacial morphogenesis. The major craniofacial deviations were found in the shape of the cranial base and the mandible. Gorlin, Redman, and Shapiro (1965) suggested, on the basis of a roentgencephalometric investigation of one patient with Klinefelter syndrome, four with Turner syndrome (XO), and a control group, that the loss or addition of an X-chromosome influences mandibular and possibly maxillary prognathism (s-n-sm and s-n-ss). Thus, according to Gorlin, Redman, and Shapiro, patients with Klinefelter syndrome should exhibit increased facial prognathism whereas patients with Turner syndrome should exhibit facial retrognathism.

The presence of increased facial prognathism in Klinefelter syndrome is supported by the present investigation, but the increased prognathism seemed to be due to the altered size and shape of the cranial base rather than to differences in the size and position of the jaws.

A recent roentgencephalometric investigation on 30 adult patients with Turner syndrome (Jensen, 1974) revealed total facial




FIGURE 4. The mean tracing of the Klinefelter group superimposed on a mean tracing of a female control group. The tracings are superimposed on the nasion-sella-line (NSL) and registered at the sella-point.

retrognathism in these patients and a flattening of the cranial base. The finding of facial retrognathism confirms Gorlin, Redman, and Shapiro's hypothesis (1965). However, the retrognathism in Turner syndrome is probably also related to the altered shape of the cranial base, and it might be hypothesized that loss or addition of an X-chromosome influences the shape of the cranial base and thereby the measurement of facial prognathism.

One might further speculate that Klinefelter patients resemble females more than males since they are chromatin-positive and have two X-chromosomes. In Figure 4, the mean tracing of the Klinefelter group is superimposed on a mean tracing of normal adult females (Ingerslev & Solow, 1975). It appears that the size of the calvarium is almost identical in the two groups whereas the size of the jaws are larger in the Klinefelter than in the female control group. The differences in craniofacial shape between the Klinefelter subjects and the females are of the same nature as those described between the Klinefelter and the male control group.

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