Craniofacial morphology of the frontonasal segment in patients with one or two macrodontic maxillary central incisors

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SUMMARY
The purpose was to describe the craniofacial morphology of the frontonasal segment in patients with one or two macrodontic central incisors.

Latero-lateral head radiographs from 21 patients were analyzed. Cephalometric analyses were performed with focus on the morphology of the frontonasal segment of the cranium, including nasal bone, maxilla, thickness of the frontal bone, sella turcica, and the anterior cranial fossa. The macrodontic incisors are located in this frontonasal segment.

Increased values compared with normal were observed in the following: the length of the nasal bone (P = 0.038), the thickness of the frontal bone (P = 0.003), the length of the sella turcica (P = 0.006), the cranial base angle (P = 0.036), the length of the anterior cranial fossa (P = 0.002), and the height (P = 0.042) and length (P = 0.011) of the maxilla. The prognathia of the maxilla was significantly decreased (P = 0.003). The depth of the sella turcica and the inclination of the maxilla were normal.

This study is a new example of how the morphology of the dentition and the underlying jaw (the frontonasal segment) are interrelated. It is also an example of how the maxillary incisors and jaws are interrelated with the anterior cranial base, including the sella turcica morphology.

Introduction
Macrodontic permanent incisors have several designations, e.g. double teeth or connect teeth. A macrodontic central incisor does not result from a fusion of the central and lateral incisor. When the mesiodistal width of the crown of the permanent central incisor is larger than 9.99 mm for males (8.83 mm + 2 SD) and larger than 9.68 mm for females (8.85 mm + 2 SD), then the term macrodontia is applicable (Moyers, 1973).

The prevalence of macrodontic central incisors in the permanent dentition is between 0.02 and 2.1%, most studies reporting prevalence between 0.1 and 0.2% (Grahnén and Granath, 1961; Brook, 1974; Ruprecht et al., 1985; Duncan and Helpin, 1987; Hamasha and Al-Khateeb, 2004; Ezoddini et al., 2007). Differences between ethnic groups are seemingly not described. Gender differences demonstrate that the prevalence is highest for males (Grahnén and Granath, 1961).

The macrodontic tooth appears with an enlarged crown, often with a bifid crown shape, sometimes with a notch on the incisal edge and a vertical ridge or depression on the facial surface (Pindborg, 1970; McDonald, 1974; Grover and Lorton, 1985; Spuller and Harrington, 1986; Neville et al., 1995; Crawford et al., 2006). The tooth has a wide root, most often with a single root canal (McDonald, 1974; Kelly, 1978; Neville et al., 1995; Crawford et al., 2006) and one pulp chamber (Spuller and Harrington, 1986). Some studies describe the incisor crown as two crown halves mirroring each other (Kelly, 1978) resulting in one symmetrical tooth (McDonald, 1974). The aetiology is still unclear.

In experimental studies, exencephaly was observed in mice that were given overdoses of vitamin A and riboflavin and also tooth fusions were observed (Knudsen, 1965).

Macrodontia has mainly been described in case reports focusing on diagnostics and treatment (Duncan and Helpin, 1987; Gazit and Liberman, 1991). The condition has also been described in epidemiological studies on the prevalence of deviations in the dentition (Brook, 1974; Ruprecht et al., 1985).

The macrodontic incisor is often seen as an isolated feature not associated with syndromes (Hattab and Hazza’a, 2001). Still, macrodontia is a phenotypic trait in severe syndromes (Smithson et al., 2000). The KBG Syndrome (named after the initials of the first three patients described) is one example where macrodontic incisors and mental retardation must be present in order to diagnose this condition (Brancati et al., 2006; Skjei et al., 2007; Kumar et al., 2009).

Another example is single median maxillary central incisor (SMMC). In these cases, there is only one central incisor, often macrodontic. An association between this deviation and fusions of the frontal lobes of the brain in the midaxial plane has been described in SMMC (Kjær et al., 2009). Also, osseous malformations are reported in SMMC. These are all located within the frontonasal developmental field or segment, extending from the frontal aspect of the...
sella turcica to the medial structures of the eyes, including the nasal bone, the lower part of the frontal bone, the sella turcica, the anterior cranial base, the maxilla, the nasal septum, and the incisors (Kjær et al., 2009). This field is illustrated on latero-lateral head radiographs by Kjær (2010). The frontonasal field develops from a group of neural crest cells located anterior at the neural tube. The composition of the frontonasal field is supposed to be genetically different from the maxillary field, which arises from other neural crest cells, located more posterior at the neural tube (Kjær et al., 2009).

The hypothesis of this study is that the morphology of the frontonasal segment of the cranium, which includes the macrodontic central incisor/s, is different from normal morphology.

Materials and methods

Lateo-lateral head radiographs from 21 patients, 4 females and 17 males (aged 7 years and 6 months to 16 years and 6 months), were included in the study. Ten patients had one macrodontic central incisor (1 in the right side and 9 in the left) and 11 had two macrodontic central incisors. Macrodontic central incisors are illustrated in Figure 1. Of the unilateral macrodontia cases, three dentitions had supernumerary central maxillary incisors and three dentitions had agenesis of one or two maxillary lateral incisors. In the bilateral cases, ageneses or supernumerary incisors were not observed.

Anamnestic information was not complete in all records. No information was given regarding body growth, but information was available in 11 records regarding minor mental deviation, ranging from concentration problems to specific diagnoses in few cases [e.g. attention deficit/hyperactivity disorder (ADHD) and deficit in attention, motor control and perception (DAMP)].

The radiographic material was referred to the Department of Orthodontics, Institute of Odontology, University of Copenhagen from municipal dental clinics in Denmark for diagnostics and treatment advice. As the material originated from different clinics, the magnification factors differed in some cases. These differences were known in all cases except two and have been taken into account in the analyses. For the two cases in which the magnification factor was not known, length measurements were not included in the cephalometric analyses. No corrections were made regarding head posture.

Control material

For comparison, standard values from a normal material from the Oslo University Growth Archive were used. This archive contains a series of latero-lateral head radiographs taken every third year of patients at the ages of 6, 9, 12, 15, and 18 years. The control material was collected between 1972 and 1992 and was described by el-Batouti et al. (1994) and Axelsson et al. (2003). All participants were Caucasians with class I occlusion, i.e. normal sagittal, transverse, and vertical dimensions without ageneses or extraction. Only minor deviations in the form of tooth rotations and/or spacing (less than 1 mm) were observed at the age of 18 years. No facial disharmony was observed and none of the participants had undergone orthodontic treatment. The group was considered appropriate for comparison with the group of patients in this study, even though minor deviations were observed in the dentition. Similarities in age and ethnicity existed between the two population groups and the standard material included both genders. Furthermore, the cephalometric methods used in the analyses and comparisons were the same. The results of the groups were compared according to age and gender.

Lateo-lateral head radiographs

Cephalometric analyses were performed on latero-lateral head radiographs according to Björk (1960) and Axelsson et al. (2003, 2004). The analysis focused on the frontonasal field including the nasal bone, the thickness of the lower part of the frontal bone, the sella turcica, the cranial base, and the maxilla, illustrated in Figure 2.

Lateo-lateral head radiographs were available from 21 patients: 1 female and 4 males in the age group 7½–10½ years (compared with control group 9 years), 3 females and 11 males in the age group 10½–13½ years (compared with control group 12 years), and 2 males in the age group 13½–16½ years (compared with control group 15 years).

The following landmarks were used:

- s Sella. The centre of the sella turcica. The upper limit of the sella turcica is defined as the line joining the tuberculum and dorsum sellae.
- n Nasion. The most anterior part of the frontonasal suture.
- ba Basion. The most postero-inferior point on the clivus.
- ss Subspinale. The most posterior point on the anterior contour of the upper alveolar arch.
- 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.
- st Tuberculum sellae. The frontal upper limit of the sella turcica.
- sd Dorsum sellae. The posterior upper limit of the sella turcica.
- sf Sella floor. The deepest point of the sella turcica.
- na Nasal apex. The tip of the nasal bone.
- br Bregma. The intersection between the sagittal and coronal sutures on the surfaces of the cranial vault.
- fo Frontale (in this study, renamed fo = frontal bone outer border). A point on the surface of the frontal bone, determined by the median normal to a line from nasion to bregma.
- fi Frontal bone inner border. The inner limit of the frontal bone determined by the median normal to a line from nasion to bregma.
The following lines and angles were registered:

- The length of the nasal bone: n-na (Figure 3).
- The thickness of the frontal bone: fo-fi. The thickness is measured perpendicular to the midpoint of the n-br line (Figure 3).
- The length of the sella was measured as the distance from the tuberculum sella (st) to the tip of the dorsum sella (sd; Figure 3).
- The depth of the sella was measured perpendicular to this line to the deepest point on the floor (sf; Figure 3).
- The cranial base angle: s-n-ba (Figure 4).
- The antero-posterior length of the anterior cranial fossa: n-s (Figure 4).
- The maxillary prognathia: s-n-ss (Figure 4).
- The anterior height of the maxilla: n-sp.
- The length of the maxilla: sp-pm (Figure 4).
- The inclination of the maxilla: the angle between the line NSL (connecting s and n) and the line NL (connecting sp and pm; Figure 4).

The lengths and angular values were statistically compared with normal standards (el-Batouti et al., 1994; Axelsson et al., 2003, 2004; including unpublished data). Length measurements were only performed on material of which the magnification factor was known, while angular measurements were performed on all laterolateral head radiographs as the angle is independent of the magnification.

**Method error**

Measurements of 10 cephalometric variables were performed by the same observer at three different times. Between the first and second measurements, 5 months elapsed. Between the second and third measurements, 5 days elapsed.

The assessment of the measurement variation was done according to a method of Dahlberg (1940; Table 1) and presented as the coefficient of variation.

The conclusion is that the measurements are reproducible.

**Statistics**

The cephalometric measurements were compared with the reference values by calculating the difference for each variable matched by age and gender, and the hypothesis that the mean difference is zero was tested by a one-sample t-test. In addition, 95% confidence intervals (CI) for the mean difference were calculated. Z-scores were calculated for the cephalometric measurement where the standard deviations (SD) from the control material were available, i.e. the Z-scores are the differences for each variable normalized by the reference standard deviation (Ward et al., 2000). Differences between those with one or two macrodontic incisors were done using a linear model. *P* values less than 5% were considered significant. All calculations were done using SAS (v 9.1, SAS Institute, Cary, North Carolina, USA).

**Results**

Significantly increased values were found in the distances s-n, sp-pm, and n-sp; the cranial base angle; thickness of the frontal bone; nasal bone length; and the length of.
The maxillary prognathia (s-n-ss) was significantly reduced. Results with 95% CI and Z-scores are shown in Table 2. The only significant difference seen when comparing unilateral and bilateral macrodontia is the length of the anterior cranial fossa (s-n), i.e. cases with bilateral macrodontia have significantly higher values than those with unilateral macrodontia in the right side ($P = 0.009$) but not those with unilateral macrodontia in the left side ($P = 0.27$). However, this result is not significant when corrected for multiple testing. The same comparison of the other cephalometric measurements could not demonstrate significant differences ($P$ values from 0.12 to 0.94).

Discussion

This study on macrodontic permanent incisors is the first cephalometric study based on a large material of macrodontic central incisors. Even though the material comprises only 21 cases, the material is considered large because of the very low prevalence of this condition (approximately $0.1–0.2\%$; Grahnén and Granath, 1961; Brook, 1974; Ruprecht et al., 1985; Duncan et al., 1987; Ezoddini et al., 2007).

This study demonstrates an interrelationship between the incisor morphology and the bony structures in the frontonasal segment extending from the incisors to the sella turcica. The central incisor belongs to the frontonasal segment developed from the anterior aspect of the neural crest. It is presumed that a large tooth in this segment can be associated with larger dimensions within the field. A similar interrelation has previously been described in SMMC1 (Becktor et al., 2001; Kjær et al., 2001; Tabataia et al., 2008). In that case, the single incisor was associated with smaller dimensions in the frontonasal segment and even with fused frontal hemispheres (Kjær et al., 2009). This interrelationship between maxillary incisors, the anterior part of the cranium, and the hemispheres indicates that there may be a neurological explanation for the mental deviation reported in 11 cases in this study. This calls for collaboration with neurologists for exact brain analyses.

A connection between other types of deviations in the dentition and the craniofacial morphology has been demonstrated previously. Nodal et al. (1994) found that patients with multiple agenesis had maxillary retrognathia,

Table 1  Assessment of measurement error.

<table>
<thead>
<tr>
<th>Variables</th>
<th>Coefficient of variation (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Maxilla inclination (NSL/NL)</td>
<td>4.59</td>
</tr>
<tr>
<td>Frontal bone thickness (fo-fi)</td>
<td>2.20</td>
</tr>
<tr>
<td>Nasal bone length (n-na)</td>
<td>1.48</td>
</tr>
<tr>
<td>Cranial base angle (n-s-ba)</td>
<td>0.35</td>
</tr>
<tr>
<td>Maxilla anterior height (n-sp)</td>
<td>0.42</td>
</tr>
<tr>
<td>Length of the anterior cranial fossa (s-n)</td>
<td>0.23</td>
</tr>
<tr>
<td>Maxilla prognathia (s-n-ss)</td>
<td>0.24</td>
</tr>
<tr>
<td>Sella turcica depth (s perpendicular to st-sd)</td>
<td>2.05</td>
</tr>
<tr>
<td>Length of the maxilla (sp-pm)</td>
<td>0.54</td>
</tr>
<tr>
<td>Sella turcica length (st-sd)</td>
<td>2.50</td>
</tr>
</tbody>
</table>
diminished cranial base angle, and anterior inclination of the mandible compared with controls. Ben-Bassat and Brin (2003) also demonstrated a characteristic skeletal pattern in patients with congenitally missing teeth. Vedtofte et al. (1999) found that patients with arrested eruption of the lower second permanent molars had jaw retrusion. In a longitudinal study on subjects with syndromic ectodermal dysplasia, Bondarets et al. (2002) found that the subjects had a tendency to anterior growth rotation and increased class III development over time. Additionally, it has been shown that patients with arrested eruption of the maxillary canines have a lower and broader maxilla than normal (Larsen et al., 2010). Also SMMC cases have craniofacial deviations, specifically located to the frontonasal field (Tabatabaei et al., 2008). The interrelationship between tooth development and jaw development observed in this study could be due to alterations in the ectomesenchymal coordination associated with the migration of neural crest cells. It is well known that the neural crest cells forming the maxilla arise from different regions on the neural crest and that the frontonasal jaw segment, in which the macrodontic incisor is located, arises from the most anterior aspect of the neural crest (Kjær, 1998). It could be presumed that a regional frontonasal alteration exists in the ectomesenchyme, while such an alteration is not seen in the other maxillary segments. This has to be studied further.

Interestingly, this study shows that when both central incisors are macrodontic and when the left maxillary central incisor is macrodontic, the length of the anterior cranial fossa is significantly enlarged. When the right maxillary central incisor is macrodontic, the interrelationship is not significant. This calls for investigations on an even larger material regarding the bilateral difference. In this study, only one right and nine left macrodontic incisors were observed in the unilateral group. This may have influenced the results. Meanwhile, a cephalometric study of SMMC cases with one central incisor by Tabatabaei et al. (2008) showed that the cranial base was significantly short compared with a normal control group. For the orthodontic treatment, these diagnostic observations in the frontonasal segment are highly important.

The material in this study came from different dentists and specialists in orthodontics, and the quality of the radiographic material varies. As a macrodontic incisor is a rare deviation, it is necessary to collect the material from different dentists in order to obtain a large material. With the current sample size (n = 17), a one-sample t-test will have approximately 80% power to detect a significant difference at the 5% level if the coefficient of variation of the variable in question is 140%.

It would be interesting to study gender differences and differences between patients with one or two macrodontic incisors on a larger material. It can also be questioned whether a secular variation exists between the material studied and the standard material, which originated from the years 1972 to 1992. This cannot be decided in this study. When comparisons are performed between two groups of patients, it is important to note that differences between published reference standards may exist (Dibbets and Nolte, 2002).

It was not possible in this material to divide the patients into subgroups according to age and gender due to the small number of patients as the subgroups would be too small. As a statistical difference between the cephalometric measurements was not found in the different age groups, the total population was pooled into one group and tracings of the patients’ ages and genders based on subgroups were accordingly not performed.

This study is a new example of how the morphology of the dentition and the underlying jaw (the frontonasal segment) are interrelated. It is also an example of how the maxillary incisors and jaws are interrelated with the anterior cranial base, including the sella turcica morphology. This seems to be relevant for orthodontic diagnostic and treatment planning. The question regarding choice of treatment in these cases is always whether to extract the macrodontic tooth. Information on deviations in the craniofacial profile must be taken into consideration.

### Table 2: Difference between the cephalometric variables measured on latero-lateral head radiographs of patients with one or two macrodontic maxillary central incisors and matched value from the control material, with mean Z-scores also shown.

<table>
<thead>
<tr>
<th>Cephalometric variables</th>
<th>N</th>
<th>Mean difference</th>
<th>Lower 95% CL for mean</th>
<th>Upper 95% CL for mean</th>
<th>Mean Z-score</th>
<th>Pr &gt;</th>
<th>t</th>
</tr>
</thead>
<tbody>
<tr>
<td>Nasal length (n-na)</td>
<td>17</td>
<td>1.87 mm</td>
<td>0.12</td>
<td>3.61</td>
<td>0.68</td>
<td>0.038</td>
<td></td>
</tr>
<tr>
<td>Sellar turcica length (st-sd)</td>
<td>14</td>
<td>1.20 mm</td>
<td>0.49</td>
<td>1.92</td>
<td>1.39</td>
<td>0.003</td>
<td></td>
</tr>
<tr>
<td>Sella turcica depth</td>
<td>19</td>
<td>1.75 mm</td>
<td>0.58</td>
<td>2.92</td>
<td>1.30</td>
<td>0.006</td>
<td></td>
</tr>
<tr>
<td>Cranial base angle (n-s-ba)</td>
<td>21</td>
<td>-0.93 mm</td>
<td>-2.46</td>
<td>0.61</td>
<td>-0.90</td>
<td>0.22</td>
<td></td>
</tr>
<tr>
<td>Anterior cranial fossa</td>
<td>19</td>
<td>3.01*</td>
<td>0.22</td>
<td>5.79</td>
<td>0.65</td>
<td>0.036</td>
<td></td>
</tr>
<tr>
<td>Maxilla anterior height</td>
<td>19</td>
<td>2.75 mm</td>
<td>1.16</td>
<td>4.35</td>
<td>1.12</td>
<td>0.002</td>
<td></td>
</tr>
<tr>
<td>Maxilla prognathia</td>
<td>21</td>
<td>-2.75*</td>
<td>-4.42</td>
<td>-1.08</td>
<td>-0.96</td>
<td>0.003</td>
<td></td>
</tr>
<tr>
<td>Maxilla inclination</td>
<td>21</td>
<td>1.00*</td>
<td>-0.55</td>
<td>2.55</td>
<td>0.41</td>
<td>0.20</td>
<td></td>
</tr>
<tr>
<td>Length of the maxilla</td>
<td>19</td>
<td>2.07 mm</td>
<td>0.54</td>
<td>3.60</td>
<td>NA</td>
<td>0.011</td>
<td></td>
</tr>
</tbody>
</table>

Patient numbers vary because not all measurements could be performed on all radiographs. This was due to radiographic protective shielding, resulting in reduced image field, or missing information regarding magnification factor. NA, not available.
Acknowledgements

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