Bilateral hypodontia is more common than unilateral hypodontia in children with Down syndrome: a prospective population-based study

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SUMMARY
BACKGROUND: Hypodontia is defined as the congenital absence of one or a few teeth (Pindborg, 1970) and is the most common developmental anomaly in humans (Thind et al., 2005; De Coster et al., 2009). Hypodontia of permanent teeth occurs most often among a few specific teeth, namely the mandibular second premolars (about 45 per cent), the maxillary second premolars (about 19 per cent) lateral incisors (about 15 per cent) and third molars (about 20 per cent) (Jensen et al., 1973; Aasheim and Ogaard, 1993; Kjaer et al., 1994; Russell and Kjaer, 1995; Mostowska et al., 2012). Hypodontia of the third molar is common among children with DS, with a rate of 74 per cent compared with 16.4 per cent among individuals in the general population (Shapira et al., 2000; Acerbi et al., 2001; Lomholt et al., 2002). DS has also been associated with the absence of permanent teeth other than the third molars.

MATERIALS AND METHODS: This population-based cross-sectional study was part of a national prospective study evaluating upper airway function, hearing, dental, and craniofacial characteristics in a cohort of children with DS born in 2002. The cohort consisted of 29 children with DS and represented 57 per cent of all children born with DS in Norway in 2002. Hypodontia was assessed using panoramic and/or dental radiographs. Data were collected prospectively at TAKO-Centre, National Resource Centre for Oral Health in Rare Medical Conditions, Lovisenberg Diakonale Hospital, Oslo, Norway.

RESULTS: Hypodontia of permanent teeth, excluding third molars, was found in 61.5 per cent of the 26 children included in the final sample. Among the 16 children with hypodontia, 75.0 per cent were missing two or more permanent teeth. Two children (7.7 per cent) had severe hypodontia (oligodontia). The teeth most often missing were the maxillary lateral incisors, followed by the mandibular second premolars and maxillary second premolars. Most (68.9 per cent) cases of hypodontia occurred bilaterally.

CONCLUSIONS: The majority of the children with DS were missing one or more permanent teeth. Unlike in the general population, bilateral hypodontia was more common than unilateral hypodontia in this sample of children with DS.
third molar (van Trotsenburg et al., 2005; Suri et al., 2011) although the research in this area is limited. Excluding third molars, the reported hypodontia rate in the general population ranges from 3.2 to 7.6 per cent, depending on methods of sampling, examination, demographic, and geographic profiles (Ingervall et al., 1972; Mattheeuws et al., 2004; Polder et al., 2004; De Coster et al., 2009; Harris et al., 2011). The occurrence of hypodontia in DS resembles that of the general population with respect to type and localization, but it is considerably more frequent among individuals with DS (Jensen et al., 1973; Russell and Kjaer, 1995; Kumasaka et al., 1997; Shapira et al., 2000; Acerbi et al., 2001; de Moraes et al., 2007; Reuland-Bosma et al., 2010; Suri et al., 2011). Numerous researchers have reported that hypodontia typically occurs unilaterally even though the body’s genetic information is presumed to be identical on both sides (Arte, 2001; Harris et al., 2011). However, symmetry patterns of hypodontia have not been adequately described among children with DS.

Hypodontia is a complex trait and is regarded as a multifactorial condition involving genes, environmental factors, and their interaction, with respect to both the aetiology and developmental pathogenesis (Maguire et al., 1987; Näsman et al., 1997; Arte, 2001; Vieira 2003; Mostowska et al., 2012). The observed familial recurrence pattern in hypodontia provides strong evidence of a genetic contribution, probably caused by several independent genes, acting alone or in combination with other genes (Grahnén, 1956; Mostowska et al., 2012; van den Boogaard et al., 2012). Hypodontia can also occur as part of a syndrome involving multiple organs (Mostowska et al., 2012; van den Boogaard et al., 2012). Syndromic hypodontia is likely due to chromosome anomalies, teratogenes, or uncategorized syndromes, the causes of which remain undetected (Ingervall et al., 1972; Mattheeuws et al., 2004; Polder et al., 2004; De Coster et al., 2009; Harris et al., 2011). Prior studies have focused on diverse samples, but studies are also needed to determine whether specific syndromes, such as DS, have specific patterns of hypodontia. Such studies will not only provide more detailed knowledge of the dental aspects of DS but also increase our understanding of the specific manifestations of hypodontia in different genetic syndromes, such as DS. Thus, the aim of the present study was to describe the prevalence and patterns of hypodontia in a population of 8- to 9-year-old children with DS.

Materials and methods

This study was a prospective population-based study conducted among Norwegian children with DS. It was part of a national study evaluating upper airway function, hearing, dental, and craniofacial characteristics in a complete age cohort of children with DS in a public health care system. The study population comprised all children with DS born in 2002 in the health region of the Southern and Eastern Norway Regional Health Authority. The inclusion criteria were a confirmed genetic diagnosis of DS and a date of birth in 2002. The initial cohort consisted of 29 children and they comprised 91 per cent of the age cohort with DS in this health region. These 29 children also represented 57 per cent of all children with DS born in Norway in the year 2002 with at least one parent of Norwegian ancestry. The children were examined clinically and their parents were interviewed at the TAKO-centre by two of the authors (EMA, SA). At the same visit, intra- and extra-oral photographs, dental impressions, panoramic radiographs, and/or dental radiographs were taken. Hypodontia of permanent teeth, excluding third molars, was assessed using the panoramic radiographs or intraoral radiographs. Five of the children (18.5 per cent) had intraoral radiographs taken under general anaesthesia for another dental and/or medical procedure. Permanent teeth were considered to be congenitally missing when no mineralization of the crown could be seen. Descriptive statistics were used to summarize patterns of hypodontia.

The study design and protocol was approved by The Regional Committee for Medical and Health Research Ethics within the health region. All patients and their parents gave informed consent in accordance with the regulations of the Biomedical Ethics Committee to participate in this study (reference number of approval letter 2010/1950).

Results

Of the 29 children included in the cohort, three of the children were not able to complete the panoramic and/or dental radiographs due to behaviour issues and/or anxiety. After excluding these three children, the final sample comprised 26 children (15 girls and 11 boys). The mean age was 8.9 years (SD ± 0.4; range: 8.2 – 9.6 years) at the time the radiographs were taken.

Hypodontia was found in 16 of the 26 children (61.5 per cent). Hypodontia was more common in females (56.2 per cent) than males (43.8 per cent). Of the 16 children with hypodontia, 4 (25.0 per cent) presented with a single missing tooth and 12 (75.0 per cent) were missing two or more permanent teeth. Oligodontia was found in two children (7.7 per cent of the sample), one male and one female.

The occurrence of hypodontia for each tooth type in relation to gender is presented in Table 1. The tooth types most commonly absent were maxillary lateral incisors, followed by the second upper premolar and second lower premolar. Maxillary hypodontia was more prevalent (62.7 per cent) than mandibular hypodontia (37.3 per cent). Hypodontia was approximately equal on the left (51.0 per cent) and right (49.0 per cent) sides.

In Table 2, all participants with hypodontia are listed according to missing teeth. The data show that hypodontia
Table 1 Occurrence of hypodontia (excluding third molars) in relation to tooth and gender in the 16 children with hypodontia out of 26 children with Down syndrome (M = male; F = female).

<table>
<thead>
<tr>
<th>Right maxilla, n = 14 missing teeth</th>
<th>Left maxilla, n = 18 missing teeth</th>
</tr>
</thead>
<tbody>
<tr>
<td>M</td>
<td>M</td>
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<tr>
<td>M</td>
<td>M</td>
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<td>M</td>
<td>M</td>
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<td>M</td>
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<tr>
<td>Right mandible, n = 11 missing teeth</td>
<td>Left mandible, n = 8 missing teeth</td>
</tr>
</tbody>
</table>

Table 2 Occurrence of hypodontia (excluding third molars) in relation to tooth and combination of teeth in the 16 children with hypodontia out of 26 children with Down syndrome.

<table>
<thead>
<tr>
<th>Gender</th>
<th>Number of missing teeth</th>
<th>Missing teeth location(s)</th>
<th>Uni/bilateral</th>
</tr>
</thead>
<tbody>
<tr>
<td>Female (n = 9/15)</td>
<td></td>
<td></td>
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<tr>
<td>F</td>
<td>8</td>
<td>12, 15, 17, 22, 25, 27, 35, 45</td>
<td>Bilateral</td>
</tr>
<tr>
<td>F</td>
<td>5</td>
<td>15, 25, 35, 45, 41</td>
<td>Bilateral</td>
</tr>
<tr>
<td>F</td>
<td>4</td>
<td>12, 22, 27, 35</td>
<td>Bilateral</td>
</tr>
<tr>
<td>F</td>
<td>3</td>
<td>15, 25, 27</td>
<td>Bilateral</td>
</tr>
<tr>
<td>F</td>
<td>3</td>
<td>15, 25, 45</td>
<td>Bilateral</td>
</tr>
<tr>
<td>F</td>
<td>2</td>
<td>12, 22</td>
<td>Bilateral</td>
</tr>
<tr>
<td>F</td>
<td>2</td>
<td>12, 22</td>
<td>Bilateral</td>
</tr>
<tr>
<td>F</td>
<td>2</td>
<td>22, 42</td>
<td>Unilateral</td>
</tr>
<tr>
<td>F</td>
<td>1</td>
<td>22</td>
<td>Unilateral</td>
</tr>
<tr>
<td>Male (n = 7/11)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>M</td>
<td>8</td>
<td>12, 15, 22, 25, 31, 35, 45</td>
<td>Bilateral</td>
</tr>
<tr>
<td>M</td>
<td>4</td>
<td>15, 25, 35, 45</td>
<td>Bilateral</td>
</tr>
<tr>
<td>M</td>
<td>4</td>
<td>31, 37, 41, 47</td>
<td>Bilateral</td>
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<tr>
<td>M</td>
<td>1</td>
<td>12, 22</td>
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<tr>
<td>M</td>
<td>1</td>
<td>42</td>
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<tr>
<td>M</td>
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<td>22</td>
<td>Unilateral</td>
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<tr>
<td>M</td>
<td>1</td>
<td>15</td>
<td>Unilateral</td>
</tr>
</tbody>
</table>

most commonly affected both teeth in a pair and seldom uni-
laterally. When hypodontia occurred, the maxillary second
premolar was symmetric in 85.7 per cent of the cases, the
maxillary lateral incisor was symmetric in 66.7 per cent of
the cases, and the mandibular second premolar was sym-
metric in 66.7 per cent of the cases. Collectively over all tooth
types, agenesis was bilateral in 68.8 per cent of the children.

Discussion

This study included 26 children with DS with complete
dental records. The sample came from an age cohort of 29
children with DS, which represented 91 per cent of all 8–
to 9-year-old children with DS in the health region of the
Southern and Eastern Norway Regional Health Authority.
The present study was conducted in a genetically homog-
enous group as all children had trisomy 21. Although the
sample was somewhat small and thus limited our ability to
describe less common patterns of hypodontia, the results
are representative of this age group in children with DS.

Our findings of hypodontia of permanent teeth in 61.5
per cent of the children is consistent with the prevalence
of hypodontia of permanent teeth (53.5–63.0 per cent)
reported in previous studies of DS populations in other
countries (Jensen et al., 1973; Russell and Kjaer, 1995;
Kumasaka et al., 1997; Shapiro et al., 2000; Acerbi et al.,
2001; de Moraes et al., 2007; Reuland-Bosma et al., 2010;
Suri et al., 2011). Compared with normally developing chil-
dren, the occurrence of agenesis in DS has been reported to
be about 10 times higher, particularly in the incisor region
of both jaws (Russell and Kjaer, 1995), which is consistent
with the findings of this study.

Among the 26 children in this study, only two were clas-
sified with oligodontia, indicating that mild hypodontia was
the most common agenesis pattern in DS. Mild hypodontia
most commonly occurs unilaterally in the general popula-
tion (Harris et al., 2011), but in our study, symmetrical pat-
terns of hypodontia were more frequently observed. One
pattern involved the simultaneous absence of both maxil-
lar permanent lateral incisors, and another pattern showed
agenesis of both second permanent premolars in both jaws.
Kumasaka et al. (1997) reported the incidence of multiple
missing teeth (two or more) was higher in subjects with DS
than in the control subjects. In our sample 75.0 per cent of
the children with hypodontia were missing two or more
teeth. Their data indicated that congenitally missing per-
manent teeth occur in a characteristic pattern. Russell and
Kjaer (1995) reported a finding of the higher incidence of
tooth agenesis on the left side, although in this study, the left
and right sides had similar rates of agenesis. In the present
study, we observed that hypodontia was more common in
the maxilla than in the mandible in children with DS, which
is in accordance with Suri et al. (2011). However, in the
general population hypodontia is more common in the man-
dible (Aasheim and Øgaard, 1993; Bäckman and Wahlin,
2001; Polder et al., 2004).

Suri et al. (2011) examined hypodontia in children with DS
and observed that the average number of missing teeth
per affected subject was 2.8 teeth, and a similar but slightly
higher number (3.2 teeth) was observed in this sample. They
also observed that hypodontia was more prevalent and severe
in females, with the most frequently missing teeth being
the maxillary lateral incisors followed by the mandibular
second premolars, the mandibular incisors, and the maxillary second premolars. Similarly, in our study, females were more likely to have hypodontia and the most commonly missing permanent tooth was the maxillary lateral incisors, followed by the second upper premolars and second lower premolars. This agenesis pattern differs from studies in the general population, where the mandibular second premolars are the most frequent missing tooth (Asheim and Ogaard, 1993; Bäckman and Wahlin, 2001; Polder et al., 2004). This gender difference is also observed in the general population (Asheim and Ogaard, 1993).

The general pattern of nerve branching is critical for the location of hypodontia, and agenesis of permanent teeth is related to the fact that they are the latest developing ones in the segment. Thus, agenesis is most commonly observed in the lateral incisors in the frontonasal developmental segment, second premolars in the lateral maxillary developmental segment and third molar in the palatal developmental segment (Russell and Kjaer, 1995; Lomholt et al., 2002; Kieser et al., 2003).

In our study, the assessment of hypodontia was performed on a group of young children 8–9 years of age with DS. Late mineralization of second premolars may occur, especially in boys, which might give a false-positive diagnosis of hypodontia in radiographs. This was demonstrated by Wisath et al. (1974) as the prevalence of hypodontia decreased with age when their sample of 7-year-old males was re-examined 2 years later. Asheim and Ogaard (1993) reported that approximately 11 per cent of the boys showed late mineralization of second premolars between the ages of 9 and 12 years. According to Schalk-van der Weide et al. (1992), delayed tooth mineralization increases with the number of missing teeth. However, Bäckman and Wahlin (2001) reported that in their study of 739 children that only one tooth showed later mineralization when their original sample of 7-year-old children was re-examined at 14 years of age.

Hypodontia is frequently associated with other common oral anomalies, including delayed tooth development, abnormal tooth morphology, such as reduced crown and root size (microdontia), conical-crown shape, enamel hypoplasia, taurodontism, and concomitant occurrence of supernumerary teeth (Schalk-van der Weide et al., 1993; Shapira et al., 2000; Acerbi et al., 2001). The same oral anomalies have been reported in DS at even higher frequencies (Brin et al., 1986; Endo et al., 2006). Studies have also reported increased levels of plaque and calculus among children with DS, which are related to a lowered immune response (Barr-Agholme et al., 1998; Pilcher, 1998). Children with DS are also more likely to have delayed eruption of deciduous teeth (Ondarza et al., 1997), have a 10-fold increased occurrence (15 per cent) of impacted maxillary canines (Shapira et al., 2000), and have a 15 per cent prevalence of premolar transposition, compared with a rate of 0.03 per cent in the general child population (Peck et al., 1993). In addition, there is evidence of increased risk for periodontitis, with reported prevalence rates as high as 30–40 per cent among teenagers with DS and close to 100 per cent by the age of 30. Elevated risk of periodontitis may be related to a 57 per cent reduction in stimulated whole salivary flow rates (ReuLand-Bosma and van Dijk, 1986; Barr-Agholme et al., 1998; Siqueira and Nicolau, 2002) and a lack of ‘self-cleansing’ due to reduced motor control of the tongue and lips (Siqueira and Nicolau, 2002).

Thus, individuals with DS often present with complex dental and oral needs, in addition to their medical, behavioural, cognitive, and social needs (Hattori et al., 2000). The increased risk of hypodontia among individuals with DS described in this study typically occurs along with higher risks for a number of other dental anomalies as well as increased prevalence and severity of periodontal disease. These combined risks further complicate dental care and treatment for children and adults with DS (Peck et al., 1993; Schalk-van der Weide et al., 1993; Kumasaka et al., 1997; Ondarza et al., 1997; Shapira et al., 2000; Acerbi et al., 2001). Considering these combined risks as part of an extended phenotype of DS may be useful for guiding future research, increasing our understanding of syndromic hypodontia, and improving dental care for individuals with DS.

Conclusions

Hypodontia of permanent teeth was found in 62 per cent of the children with DS, which is approximately 10 times higher than among children without DS. Hypodontia of permanent teeth in DS occurred bilaterally in 69 per cent of the cases where teeth were missing, whereas unilateral hypodontia is more common in the general population. The results of this study, in conjunction with prior research, indicate that dental anomalies should be considered in an extended phenotype for DS. Furthermore, it is important for multidisciplinary dental teams to be familiar with the different medical, oral and cognitive aspects of DS, as well as the potential behavioural issues, in treatment planning and habilitation for children with DS.

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References


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