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The primary maxillary central incisor in the Solitary Median Maxillary Central Incisor syndrome

**ABSTRACT**

**Aim** Solitary Median Maxillary Central Incisor (SMMCI) is a developmental anomaly in the permanent dentition with one single central incisor in the maxilla, positioned exactly in the midline. This condition has been associated with extra- and intraoral malformations in the frontal suture in the cranial segment of the cranium and face. It is not known whether the centrally located permanent incisor is always preceded by a centrally located primary incisor. The aim was to analyse whether a permanent single central incisor in SMMCI is always preceded by a primary single central incisor and to study extra- and intraoral phenotypic traits of the condition.

**Methods** Study design: cross-sectional radiographic study of 11 children, visual analysis of photos and dental and panoramic radiographs.

**Results** Nine of the 11 cases exhibited a primary SMMCI with one symmetrical crown and root. Two cases exhibited two separate primary central incisor crowns with fused roots. The phenotypical traits (indistinct philtrum, lack of normal upper lip contour, missing superior labial frenulum and distinct mid-palatal ridge) were findings observed in young children with a primary SMMCI.

**Conclusion** The present study concludes and stresses the necessity of diagnosing the SMMCI condition early in life. Furthermore, paediatric dentists are recommended to be aware of the condition and to refer these patients to interdisciplinary diagnostics and treatment.

**Keywords** Centrally located incisor; Developmental anomalies; Phenotypic traits.

**Introduction**

The occurrence of a single maxillary central incisor, a condition designated as Solitary Median Maxillary Central Incisor (SMMCI), in the human dentition is considered very rare, its prevalence being approximately 1 in every 50,000 live births [Hall, 2006]. SMMCI is a developmental anomaly, described in the permanent dentition and characterised by a single central incisor in the maxilla, positioned exactly in the midline [Cohen et al., 1971; Siebert et al., 1990; Hall, 2006]. SMMCI is a malformation in the frontal suture in the cranial segment of the cranium and face occurring around day 35-38 of the gestational period [Cohen et al., 1971; Siebert et al., 1990; Hall, 2006; Kjær et al., 1997; 2001]. This has led to the suggestion that a permanent SMMCI is always preceded by a primary SMMCI. However, this has never been clearly investigated or confirmed.

Many SMMCI patients suffer from orbital hypotelorism in varying degrees. The narrow interorbital space is associated with a narrow tubular nose. The nasal cavity is concomitantly narrow and the nasal septum is often asymmetrical [Siebert et al., 1990; Kjær et al., 2001]. An arch-shaped upper lip contour and an indistinct philtrum are also external characteristics of SMMCI [Kjær et al., 2001; 2002]. SMMCI patients may exhibit an abnormal sella turcica, e.g. sella bridge, oblique anterior wall or diminished sella volume [Kjær et al., 2001]. SMMCI patients often suffer from endocrine deficiencies and short body height [Kjær et al., 2010]. Compared to control subjects, the cephalometric nasion-sella distance is short and both the maxillary prognathia and inclination are diminished [Kjær et al., 2001; 2002]. The absence of the interincisal part of the intermaxillary suture is a characteristic trait of the condition, as is the missing foramen incisivum [Kjær et al., 2002; Becktor et al., 2001]. Intraorally, the palate is narrow and V-shaped with a midline ridge extending to the posterior border of the hard palate. The papilla incisiva is missing [Hall, 2006; Kjær et al., 1997], as well as the superior labial frenulum [Kjær et al., 2001; 2002].

Since SMMCI can be present as a mild form of holoprosencephaly (HPE) [Nanni et al., 2001; DiBiase et al., 2006], it has been suggested that the genetic defect is to be found in HPE associated genes. Some of the chromosome deletions that appear to have an association with cases of SMMCI, are those of chromosome 7 (7q36.1) and chromosome 18 (18p11.3) [DiBiase et al., 2006]. Certain interest has been focused on SHH and SIX3 [Nanni et al., 2001; White and Pharoah; 2004; DiBiase et al., 2006]. Studies have shown that SHH can act as a positional signal in vertebrate CNS development [Hehr et al., 2004], whereas SIX3 acts as a direct regulator of the sonic hedgehog expression in the vertebral forebrain [Nanni et al., 2001; Lacbawan et al., 2009].

The purpose of this study is to analyse whether a single permanent central incisor in the SMMCI condition is...
always preceded by a single primary central incisor, and to study the extra- and intra-oral phenotypic traits of the condition.

Material and methods

The total material comprised data from patient records, radiographs and photos from 27 cases of SMMCI. The material was sent to the Department of Orthodontics, Institute of Odontology, University of Copenhagen, Denmark, from dentists and orthodontists in Denmark, Sweden, Germany and USA for diagnostics and possible treatment guidance. Of the 27 cases information on primary SMMCI was available in 11 cases. Two of these 11 cases were male and 9 were female; their ethnicity was not disclosed. Of these 11 cases the following material was available.

- Panoramic radiographs of the primary dentition (3 cases).
- Intraoral radiographs of the maxillary incisor region (7 cases).
- Clinical photos (5 cases).
- Panoramic radiographs of the permanent dentition (8 cases).

The 11 cases containing radiographs and clinical photographs of the primary maxillary central incisor were subjected to further visual analysis:

- assessment of crown and root morphology of the primary SMMCI;
- assessment of intra- and extraoral phenotypic traits.

Results

All 11 cases exhibited a permanent SMMCI mid-axially located. Nine of the 11 cases exhibited a primary SMMCI with one symmetrical crown and root (Fig. 1). Two cases exhibited two separate primary central incisor crowns with fused roots (Fig. 2). The phenotypical traits (indistinct philtrum, lack of normal upper lip contour, missing superior labial frenulum and distinct mid-palatal ridge) were findings observed also in young children with a primary SMMCI dentition (Fig. 3-5).

Discussion

In the present study two SMMCI cases showed abnormal morphology of the primary central incisor. In these two cases two central incisor crowns with fused roots were observed. The permanent incisor in these two cases exhibited a large mesiodistal width. The present study thus includes two cases where the single central incisor has been preceded by a tooth consisting of two separate crowns with fused roots and separate pulp chambers. The explanation might be found in the embryonic development of the primary tooth buds. One can speculate whether there is a fusion of two tooth buds or whether the central incisor tooth bud from the very start develops abnormally.
malformations also occur in some cases [Kjær et al., 2010]. Therefore, referral to a neurologist is important. The present study concludes and urges the necessity of diagnosing patients with the SMMCII condition early in life. The study furthermore recommends paediatric dentists to be aware of the condition and to refer these patients to interdisciplinary diagnostics and treatment.

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References

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