

Solitary median maxillary central incisor syndrome (SMMCI): A 4-year evaluation

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Abstract

The present report follows the case of a young boy with solitary median maxillary central incisor (SMMCI) syndrome between the ages of 4 and 7 years. This condition is characterized by the presence of one single maxillary central incisor in the midline instead of two central incisors. No other developmental abnormalities involving growth or brain function were noted at, or subsequent, to birth. This report includes a discussion of the aetiology of SMMCI syndrome and its association with birth defects such as holoprosencephaly (HPE), CHARGE and VACTERL, as well as a discussion of the long-term prognosis and associated dental and medical issues for this particular patient.

Keywords: aetiology; case report; paediatric dentistry; SMMCI syndrome; solitary median maxillary central incisor

Introduction

A solitary median maxillary central incisor (SMMCI) in the primary or permanent dentition is a rare phenomenon [1-3]. Explanations of the aetiology of the condition include fusion across the midline, conation and double tooth, fusion of two tooth buds and agenesis of a tooth germ [4-9]. The SMMCI is usually the only tooth present in both deciduous and permanent dentitions, with the incidence of cases thought to be about 1 in 50,000 [1,10].

Although SMMCI may occur as a spontaneous isolated event, it has, more importantly, been reported to occur in association with developmental congenital disorders such as holoprosencephaly (HPE), velocardiofacial syndrome, ectodermal dysplasia, growth retardation with or without growth hormone deficiency, and other mid-line developmental defects [3,8,11-17]. In addition, congenital nasal malformations, such as choanal atresia, midnasal stenosis and congenital pyriform aperture stenosis have been positively associated with SMMCI [1,10]. More recently, several studies have reported the presence of SMMCI and other dental abnormalities including ectopic eruption, submergence of primary molars and congenital absence of teeth in patients with recognized syndromes such as CHARGE (Coloboma of the eye, Heart defects, Atresia of the choanae, Retardation of growth and/or development, Genital and/or urinary abnormalities, and Ear abnormalities and deafness) or VACTERL(vertebral anomalies,, anal atresia, cardiovascular malformations, tracheo-oesophageal fistula, renal and limb anomalies) [18-21]. Most of these studies agree that, although SMMCI can occur as an isolated dental phenomenon, the presence of a single central incisor may be indicative of a wider disorder.

Case report

The male patient was first referred at 3 years of age by a local dental clinic and presented with the chief complaint of pain when eating and drinking. The medical history revealed that the patient was born after a normal gestation period and was within the expected weight range at birth. He was in hospital only as a baby and experienced a brief episode of jaundice at this age. At 3 years of age, the patient showed normal facial symmetry. Further examination and orthopantomography revealed gross caries in the lower left deciduous first and second molars which were temporarily restored. The patient was reviewed one year later with a weight of 15 kg, complaining of shooting and throbbing pain on the lower right side as well as sensitivity to hot and cold foods. Extractions were performed under general aesthetic for the removal of all deciduous first molars due to unrestorable caries. At this time it was noted that the patient had a permanent central maxillary incisor missing without a contributing trauma history. At 5 years of age, the patient continued to complain that his deciduous central incisor hurt when eating cold foods, although he was otherwise fit and well. The single central maxillary incisor was located at the midline and its coronal form was symmetrical and did not show the angle symbol and the curve symbol (Figures 1-3). A panoramic radiograph showed normal dental development of the maxillary and mandibular dentition for a child of 5 years, except for the absence of one maxillary incisor (Figures 2-4). The patient was seen again at 5 years and 8 months complaining of pain and sensitivity when eating acidic food, particularly oranges. Further examination revealed that he had a geographic tongue, a condition which was also present in his mother. At this time, his carious lesions appeared arrested and oral hygiene improved and

stabilized. He had continued to develop normally and at 6 years 6 months was 20 kg in weight and 1.21 m tall.

Further dental examinations at 7 years of age revealed that the deciduous single central incisor had exfoliated and a single median permanent incisor had erupted. The permanent incisor was completely symmetrical in shape and positioned in the midline. The tooth's mamelons were well formed in comparison to a normal incisor, in which the two proximal mamelons are normally different, with the mesial being sharper than the more rounded distal edge.

This patient was referred for urology assessment at 5 years of age following a history of intermittent urinary hesitancy and occasional dysuria. On examination, the patient was found to have normal urological findings except for the possibility of a duplex kidney.



Figure 1. Photograph of the patient at 5 years old with a solitary median maxillary central incisor (SMMC) in front (A) and profile (B) deciduous dentition view.



Figure 2. Intraoral photograph showing the presence of the solitary median maxillary central incisor (SMMC) at midline (A and B), and normal mandibular deciduous dentition (C).

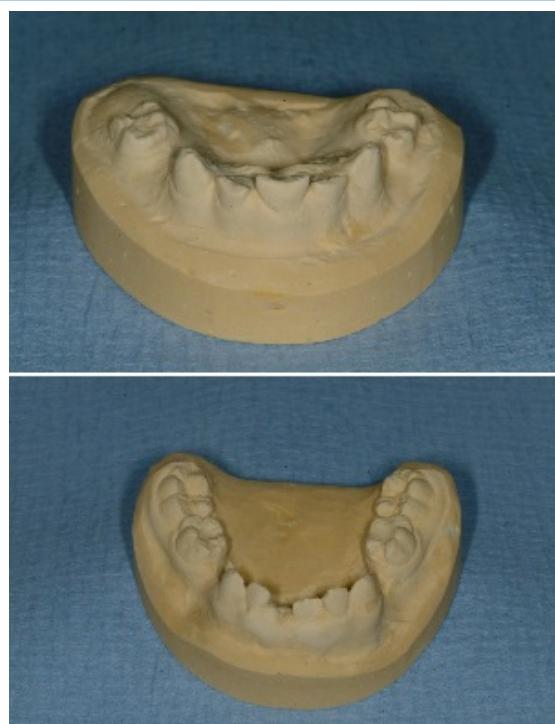


Figure 3. Model of dentition showing the maxillary and mandibular dentition.



Figure 4. Panoramic radiograph showing normal development except for the presence of a solitary median maxillary central incisor (SMMC) in the patient's deciduous dentition (A). Radiograph of the SMMC showing

Discussion

The exact midline position and symmetrical shape of the primary and permanent single central incisor in this patient is indicative of SMMC1 syndrome, rather than other conditions such as traumatic loss of one central incisor, mesiodens, fusion of a central incisor with supernumerary tooth, or developmental failure of one central incisor.

Although the disorder is a rare phenomenon, the aetiology of SMMC1 syndrome remains unknown. Several reports in the literature show that SMMC1 syndrome may occur with no other obvious abnormalities [3,10,12], though others suggest that this syndrome is a minor manifestation and presentation of other disorders [11,14,16,17,22,23]. An association between SMMC1 and HPE has been most commonly described; however, other syndromes reporting SMMC1 include CHARGE and VACTERL associations, velocardiofacial syndrome, ectodermal dysplasia, congenital nasal malformations, and other midline disorders [2,15,24-26]. These disorders generally occur as a result of defects in brain development and/or other midline structures of the body, resulting in variable degrees of abnormality in the face, skull and body organs. Symptoms range from mild to severe learning disability, congenital heart disease, choanal atresia, midnasal stenosis, congenital nasal pyriform aperture stenosis, hypothyroidism, microcephaly, hypotelorism, hypopituitarism, short stature, microcephaly, cleft lip and palate, congenital heart disease, scoliosis, SMMC1, and others [24,26-32]. Therefore, a patient presenting with SMMC1 should be assessed for signs of associated anomalies and syndromes. The young male patient presented in the current report did not exhibit any additional features diagnostic of associated syndromes which eased the family concerns.

Several specific chromosomal abnormalities have been identified in patients with SMMC1 and HPE. In particular, deletions on the sonic hedgehog (SHH) gene and other novel SHH mutations have been reported [16,31,33-36,37-39]. However, the relevance of these chromosomal abnormalities to associated syndromes and SMMC1 remains largely unknown.

There is evidence that HPE is manifest through autosomal dominant as well as X-linked autosomal recessive inheritance [25,28,31,40]. More importantly for the male in the present case, SMMC1 syndrome present in parents has been shown to be associated with the presence of more severe forms of HPE in their offspring [24,41,42]. Therefore, genetic counselling needs to be considered in discussing future management options for the child and his family.

Several possibilities exist regarding future dental management of this patient, including accepting the SMMC1 tooth in its central position and closing the spaces on either side. An alternative option, considered to be aesthetically more pleasing, could involve relocating the SMMC1 tooth orthodontically to one side, thus creating a space for a prosthetic replacement of the absent tooth. These treatment possibilities need to be assessed in the light of the underlying class III malocclusion.

In summary, this case report describes the 4-year evaluation of a male patient who presented with the presence of an SMMC1 following a normal gestation period and birth. The boy was of normal stature with no other observable abnormalities. Intraoral examination revealed moderate oral hygiene, evidence of early childhood caries and a significant class III malocclusion. Apart from the underlying malocclusion, the only abnormality observed was the presence of the SMMC1 evident in the deciduous and permanent dentition. While the presence of a duplex kidney was discovered, it was unlikely to be related to the patient's dentition.

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