Solitary median maxillary central incisor in two healthy siblings: case report

Roberta Barcelos DDS MSD PhD¹, Patricia Nivoloni Tannure DDS MSD², João Alfredo Farinhas DDS MSD², Evelyn Kahn MD MS³ and Rogerio Gleiser DDS MSD PhD²

¹Department of Specific Formation, Federal Fluminense University, Nova Friburgo, Brazil; ²Department of Pediatric Dentistry and Orthodontics, Federal University of Rio de Janeiro, Rio de Janeiro, Brazil; ³Genetics Ambulatory, Martagão Gesteira Pediatric and Puericulture Institute, Federal University of Rio de Janeiro, Rio de Janeiro, Brazil and Pediatric Genetics Ambulatory, Antônio Pedro University Hospital, Fluminense Federal University, Niterói, Brazil

Abstract

A solitary median maxillary central incisor is an unusual dental finding and may be associated with short stature, growth hormone deficiencies, syndromes and chromosomal abnormalities. This paper describes two cases of the absence of a maxillary central incisor affecting both dentitions in two healthy siblings. No reports were found in the literature similar to this rare situation. This anomaly can be considered a predictor of holoprosencephaly in the next generation and affected individuals may require a long term evaluation by a multidisciplinary team.

Key words: Tooth abnormalities, hypodontia, malformations, children, holoprosencephaly

Introduction

Tooth agenesis is the most common congenital anomaly in humans and has a prevalence of between 2.6% (Altug-Atac and Erdem, 2007) and 7.2% (Backman and Wahlin, 2001) excluding third molars. In contrast, the presence of a solitary median maxillary central incisor (SMMCI) at the maxillary midline in both dentitions is a rare dental finding and is considered a syndrome, occurring in the ratio of approximately 1:50,000 people (Hall, 2006). This syndrome is a complex disorder consisting mainly of multiple midline defects resulting from unknown events that took place in the uterus between the 35th and 38th day after conception.

The SMMCI tooth differs from the normal central incisor, in that the shape of the crown is symmetric; it develops and erupts precisely at the midline of the maxillary dental arch in both primary and permanent dentitions (Hall, 2006). In some cases, only one central incisor tooth may be present but this does not represent a SMMCI, if for example the following occur:

- When one central incisor failed to proceed beyond the cellular developmental stage and the remaining normal tooth in spite of developing on one side of the midline, erupts near or on the midline
- Loss of a central incisor due to trauma
- Fusion of a primary and/or permanent tooth with a supernumerary tooth

SMMCI has been reported as an isolated trait (Cho and Drummond, 2006; Bolan et al., 2009) or associated with holoprosencephaly, pituitary dysfunction, congenital heart disease, malformations of the sella turcica, hypotelorism and abnormalities of the nasal cavity (Aughton et al., 1991; Hall et al., 1997; Nanni et al., 2001). As the SMMCI syndrome should be considered a predictor of various anomalies, especially for holoprosencephaly, a detailed paediatric, ear-nose-throat and genetic examination is recommended. Some features should be considered as diagnostic criteria, such as: preterm birth and low birth weight, cleft lip and palate, choanal atresia, short stature, small head circumference, hypotelorism, holoprosencephaly spectrum, intellectual disability, deviant sella turcica, pituitary gland morphological abnormalities and also familial history of one of these characteristics. The SMMC diagnosis is possible with ultrasound at 18–22 weeks or, possibly, on genetic testing in familial cases, but is rarely made prenatally. The SMMCI tooth can be detected by radiographic exam prior to its eruption.

The aetiology of SMMCI is uncertain. Nanni et al. (2001) reported an autosomal dominant family with SMMCI and suggested that missense mutation in the Sonic Hedgehog gene (I111F) at 7q36 may be associated with this anomaly (Nanni et al., 2001). SMMCI can be found in association with other chromosomal abnormalities such as deletion of 22q11.2 (Hall et al., 1997). Letsirovirakul and Hall reported an association with the oromandibular-limb
hypogenesis syndrome (Lertsirivorakul and Hall, 2008). This case report presents two siblings with SMMCI with no other abnormalities or congenital malformations.

**Case Report**

Two Afro-American siblings, a 12-year-old female and a 7-year-old male, came to the Pediatric Dental clinic at the Federal University of Rio de Janeiro, Brazil with a complaint that an upper central incisor was absent. Their medical histories were unremarkable, and the pregnancies and term deliveries of both children were normal. There was no family history of congenitally missing teeth or other craniofacial anomalies.

The sister’s intraoral examination revealed a healthy permanent dentition and soft tissues. The absence of an upper labial frenulum and a solitary maxillary central incisor (Figure 1) was observed. A periapical radiograph confirmed the SMMCI. The mother confirmed no history of previous trauma and related a single tooth in this region in the deciduous dentition. The patient had a Class II molar relationship, and was referred for orthodontic treatment. The boy presented healthy soft tissues and no upper labial frenulum. An enamel fracture in the incisal edge was observed in the solitary incisor. He was at the mixed dentition stage with an Angle Class I molar relationship (Figure 2). A periapical radiograph taken when the boy was 2-years-old confirmed the SMMCI in the primary dentition (Figure 3). Radiographic exam revealed a SMMCI in the permanent dentition with a single pulp chamber and root canal (Figure 4). The immediate treatment plan consisted of restoration of the isolated fractured incisor with composite resin and installation of an acrylic removable appliance with a Hawley clamp to improve axial inclination and aesthetics.

In both cases, the isolated tooth was situated on the midline and presented a morphologically symmetrical structure with identical mesial and distal surfaces. The siblings were referred to their paediatrician for further evaluations. The patients’ heights and weights were within normal limits and considered compatible with their chronological ages. Both children were in the tenth percentile of the growth curve. The patients were evaluated in the genetics clinic ambulatory to investigate other minor malformations. No abnormalities were diagnosed and a MRI examination and CT scan were not indicated by the physicians. Their intellectual development was considered normal and both children had a good performance at school.

**Discussion**

The management of a child with SMMCI involves a complex approach. A child with SMMCI should visit the dentist at an early age concerning the non-eruption of one maxillary central incisor. The compilation of an extended family pedigree may have brought to light previously unrecognised associated anomalies in family members and ancestors. Implications for future pregnancies of parents (and later of the child) can be explained (Hall, 2006). In our case, two healthy siblings of the same family were affected by this anomaly. No reports were found in the literature similar to this rare situation.

Owing to the absence of one maxillary central incisor, besides the aesthetic point of view, there is a space loss in the maxillary inter-canine region, which may subsequently cause crowding in the mandibular incisor region. It has also been stated that this condition is ideally managed by combined orthodontic and prosthetic rehabilitation (Bolan et al., 2010) or alternatively, it can be left untreated (Hall, 2006). Several possible dental treatments have been suggested for this anomaly. One option described was to move the SMMCI distally and install a temporary adhesive prosthesis until the time for definitive treatment, involving orthodontics and implants (Bolan et al., 2010). Maxillary expansion in patients with SMMCI is limited. This implies that rapid expansion in patients with SMMCI syndrome will not open the inter-maxillary suture because of the probable fusion of the anterior sutural region where the single tooth is located (Bolan et al., 2010). The orthodontic
procedures, in these cases, vary depending on the degree of involvement of the maxillary bone structures, occlusion and mainly of the midpalatal suture. The girl was referred for orthodontic treatment, although her dental alignment was considered satisfactory by her parents.

For the boy, in our case study, the immediate treatment proposed was correction of the SMMCI axial inclination with a removable appliance. The patient and his parents were aware that this was a temporary solution and that he should be assessed for future orthodontic and prosthetic or implant treatments. In most SMMCI cases the dentist is the first health professional to evaluate this condition and it is important to understand the diagnostic options and the required treatment (Bolan et al., 2009). In conclusion, SMMCI is a risk factor for holoprosencephaly in offspring and can be associated with many disorders. Affected individuals may require a long term evaluation by a multidisciplinary health team, including a speech therapist, psychologists and a dental team.

References


Barcelos et al.: Solitary median maxillary central incisor

Figure 3. Periapical radiograph of male patient showing a single tooth localised on the midline in the primary dentition

Figure 4. Periapical radiograph of male patient showing a single tooth localised on the midline in the permanent dentition

Hall RK. Solitary median maxillary central incisor (SMMCI) syndrome. Orphanet J Rare Dis 2006; 1: 12.

Address for correspondence:
Roberta Barcelos
Rua Silvio Henrique Braune 22
Nova Friburgo – RJ – Brasil
CEP 28625-650
Email: rbarcelos@vm.uff.br