Solitary Median Maxillary Central Incisor

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ABSTRACT

Solitary median maxillary central incisor syndrome (**SMMCIS**) is a rare abnormality characterized by the presence of a central incisor positioned at the maxillary mid-axis. This morphologic defect also can be associated with other diseases. The purpose of this paper was to present a case report of a 4-year-old twin child with SMMCIS. The patient showed a symmetrical primary maxillary central incisor located at the midline, with an absence of labial frenulum, an indistinct philtrum, and an incisive papilla. Radiographic examination confirmed the presence of only a maxillary central incisor in both dentitions. The patient was referred for a genetic and otolaryngological assessment, however, no other abnormality than the ones reported were detected.

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Solitary median maxillary central incisor syndrome (**SMMCIS**) is characterized by the presence of just 1 central incisor positioned at the maxillary midline in both dentitions. This anomaly is considered rare, with a prevalence of approximately 1:50,000 live births. Moreover, it may involve structures of the body's midline, including craniofacial bones, the nasal cavity, and the brain.[1]

SMMCI's etiology is not completely understood, but it may be related to a disruption in the development of the maxilla, which occurs intrauterinely around days 35 to 38. During that period, research suggests that the lack of space in the dental arch and the deficiency in the maxilla's lateral growth and other structures of the mid-axis region could interact to bring about a premature fusion of the dental lamina at the midline, with no normal formation of tooth germs, alveolar bone, or soft tissue.[2,3]

SMMCIS involves a central incisor with symmetric morphology, an absence of labial frenulum and incisive papilla, an absence of intermaxillary suture in front of the incisive fossa, and a prominent midpalatal ridge.[4]

The solitary central incisor can appear as an isolated feature or be associated with other systemic alterations, such as deficiency of growth hormone, premature puberty, micropenis, short stature, mental retardation, holoprosencephaly, pituitary dysfunction, malformations of the sella turcica, hypotelorism, and some abnormalities of the nasal cavity such as choanal atresia, midnasal stenosis, and nasal pyriform aperture stenosis.[1,5,6]

The purpose of this paper was to report a case of a patient with a solitary median maxillary central incisor.

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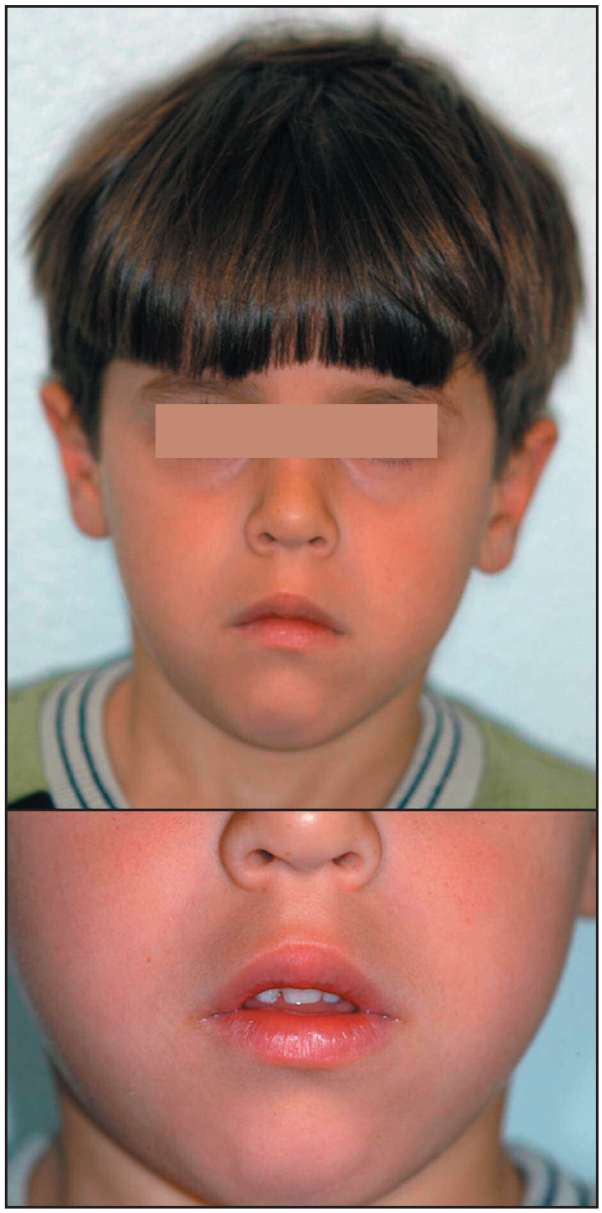

Figure 1. Facial appearance showing the prolabium of the upper lip without curved contours and the indistinct philtrum.

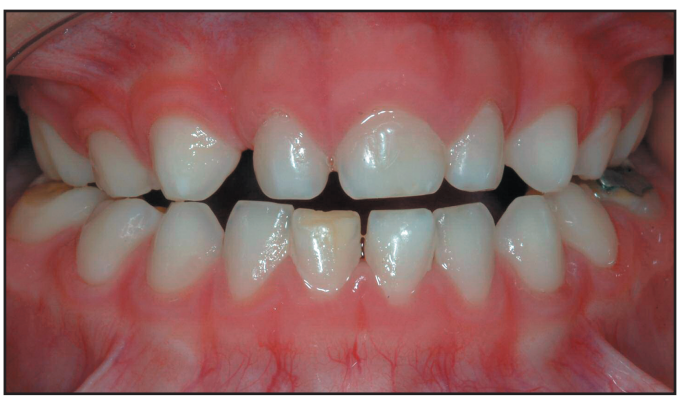

Figure 2. Frontal view showing a symmetrical solitary maxillary primary central incisor and the absence of the labial frenulum.

CASE REPORT

A male 4-year-old patient presented the Pediatric Dentistry Clinic at the Federal University of Santa Catarina, Florianópolis, Brazil, with his mother. She reported the strange appearance of the child's maxillary anterior teeth, which embarrassed him in front of his friends.

The patient was a twin, and his brother did not show any similar feature. There was no report of any hereditary diseases in the family.

At physical examination, he weighed 19 kg, had a height of 1.12 m, and measured 18 cm for cephalic circumference. His brother weighed 20 kg, had a height of 1.14 m, and had a cephalic circumference of 20 cm.

The patient's mother reported that, at birth, he showed an "open trachea" and had breathing difficulties, a congenital stiff neck and hypoglycemia, causing him to be admitted to the hospital for 10 days. At 4 months, he had bronchiolitis.

During the dental examination it was observed that the patient has only 1 maxillary central incisor located precisely at the mid-axis. The crown of that tooth was symmetric, making it impossible to distinguish its medial and distal angles. The patient also showed maxillary atresia and an absence of labial frenulum, an indistinct philtrum, an incisive papilla, and a prominent midpalatal ridge. No other facial abnormalities were observed (Figures 1-3).

The patient was in the primary dentition stage, and enamel hypoplasia in the mandibular second molars as well as caries in teeth L, K and T were noted. There were no dental avulsions or tooth extractions at the incisors region.

Periapical radiographs showed that the incisor had a root with 1 canal and only 1 permanent maxillary central incisor germ, which was confirmed by a computed tomography (**CT**) scan. An ortopantomogram showed that all the other permanent tooth germs were developing normally (Figure 4). Based on all these features, the patient was diagnosed with SMMCIS. The contour of the sella turcica was analyzed and considered normal, as described by Kjaer et al[7] (Figure 5).

The patient was referred for genetic, cardiologic, and otolaryngological assessment. The medical data confirmed that the patient did not present any malformations in addition to the solitary incisor and the morphologic alterations of the upper lip and palate. A CT scan of the cranium requested by the otolaryngologist revealed normal structures with no brain malformations. Genetic assessment based on DNA from a blood specimen did not show the mutations associated with the syndrome. The patient is still under genetic consultation.

All the decayed teeth were treated in the prevention program, which includes tooth-brushing, professional tooth cleaning, and topical application of fluoride. Recently, when the child was 6 years and 10 months old, the permanent central incisor erupted. The malocclusion treatment is being performed by a multidisciplinary team that includes an orthodontist, a pediatric dentist, and a radiologist. The patient is also receiving psychiatric follow-up care.

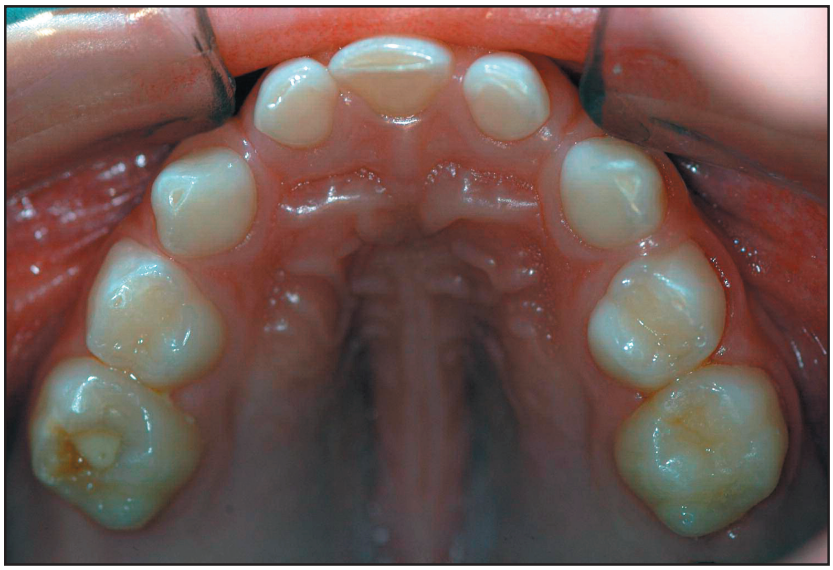

Figure 3. Occlusal view showing the prominent midpalatal ridge and the absence of incisive papilla.

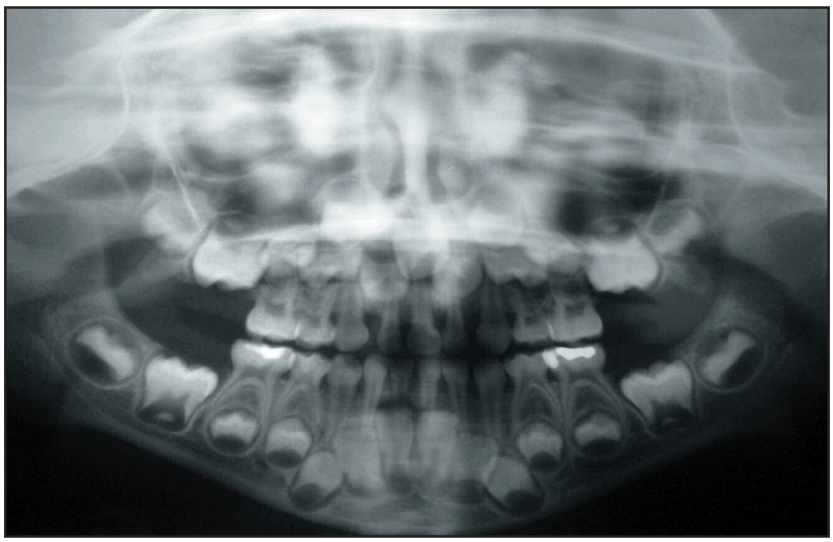

Figure 4. Orthopantomogram confirming the existence of a solitary maxillary central incisor in both dentitions. No other dental anomalies were observed.

DISCUSSION

The presence of a solitary incisor at the midline can be an isolated feature or a mild manifestation of larger malformations of the neural system holoprosencephaly (**HPE**)—a developmental defect affecting the proencephalon and the face. In most cases, HPE is incompatible with life.[7] All HPE patients have SMMCIS, but not all who have SMMCIS have been diagnosed as holoprosencephalic. Currently, mutations in the human sonic hedgehog gene are the most frequent cause of autosomal dominant inherited HPE.[8] The HPE spectrum can demonstrate incomplete penetrance, and it is estimated that only 70% of obligate carriers show some clinical feature of this condition.[9] Therefore, SMMCIS' presence must be considered a risk factor for HPE, even in the absence of any other clinical signs. For this reason, in cases where SMMCIS cannot be explained on the basis of the clinical history, it is suggested that the subject be referred for further genetic analysis.[10]

Yassin and El-Tal[11] reported a case of SMMCIS in a twin patient. Neither his brother nor any of his relatives, however, had characteristics of the syndrome as described in the present paper. The DNA analysis showed no genetic mutations.

There is a large variation in the reported anomalies associated with the presence of a solitary incisor. These anomalies include: "CHARGE" (an acronym for ocular coloboma, heart defects, choanal atresia, retarded growth and development, genital anomalies, and ear anomaly); "VACTERL" (an acronym for vertebral anomalies, anal atresia, cardiac malformations, tracheoesophageal fistula, and nasal and labial anomalies); Goldenhar's syndrome; hypothalamic harmatoma; triple-X syndrome; ectodermal dysplasia; hypotelorism; and mid-axis defects in craniofacial and brain structures.[3,4,12] The literature also implicates chromosomal deletion in some SMMCIS cases, such as those in chromosomes 7, 18, and 22.[1,8]

There are reports of SMMCIS associated with short stature and growth hormone deficiency.[1,6,13] In this report, however, the patient's stature and skeletal development[14] were considered compatible with his chronological age. The same has been observed by other authors,[13,15-19] most recently Cho and Drummond[20] in a report of 3 SMMCIS cases. Stanhope et al,[21] however, emphasized that a child with midline defects can have endocrinopathy that may appear at a later time. Hall et al[1] suggest that children with SMMCIS and at least 2 standard deviations below the mean height for their age and gender should be referred to an endocrinologist for monitoring and regular follow-up for grow and development. The same authors described 2 SMMCIS patients with sella turcica abnormal morphology. Even severe anatomic change, however, does not always mean pathophysiological function, nor does disturbed pituitary function necessarily indicate an abnormal sella turcica. The patient in the present study had a sella turcica with a normal contour.

Learning deficiencies and intellectual retardation were described in SMMCIS patients,[6] but they were not verified in this case. The patient was considered intellectually normal, and his mother reported that he attends the same class as his brother.

Some defects in the midline of the nasal cavity in SMMCIS patients can result in congenital nasal obstruction, such as nasal stenosis, choanal atresia, and the nasal pyriform

opening stenosis.[1] Brown et al[22] reported that the latter is a major cause of neonatal nasal obstruction. Cho and Drummond[20] described the same feature in their patients at birth, although without breathing blockage during childhood. In the case reported by Velasco et al,[23] the patient showed severe asphyxia at birth. Hall et al[1] and Kjaer et al[7] also observed neonatal nasal obstruction in SMMCIS patients, although the nasal blockage was considered clinically expressive just in some of them. The authors suggest that patients who have breathing difficulties at birth have a less severe nasal deformity.

According to Hall et al,[1] nasal cavity defects are not commonly detected on presentation. This is because the cause of a neonatal nasal obstruction must be treated with surgery immediately at the neonatal period before the eruption of the incisors, at the second half of the first year after birth. Therefore, once SMMCIS is diagnosed, it is important to make a referral to an otolaryngologist. According to the mother's report, at birth, the patient showed breathing difficulties because of an "open trachea" that was controlled and not requiring surgery. It was not possible to contact the patient's pediatrician at birth to get information on the treatment. State-of-the-art ortopantomograms and a CT scan did not show pathology in the nasal cavity or any degree of nasal obstruction.

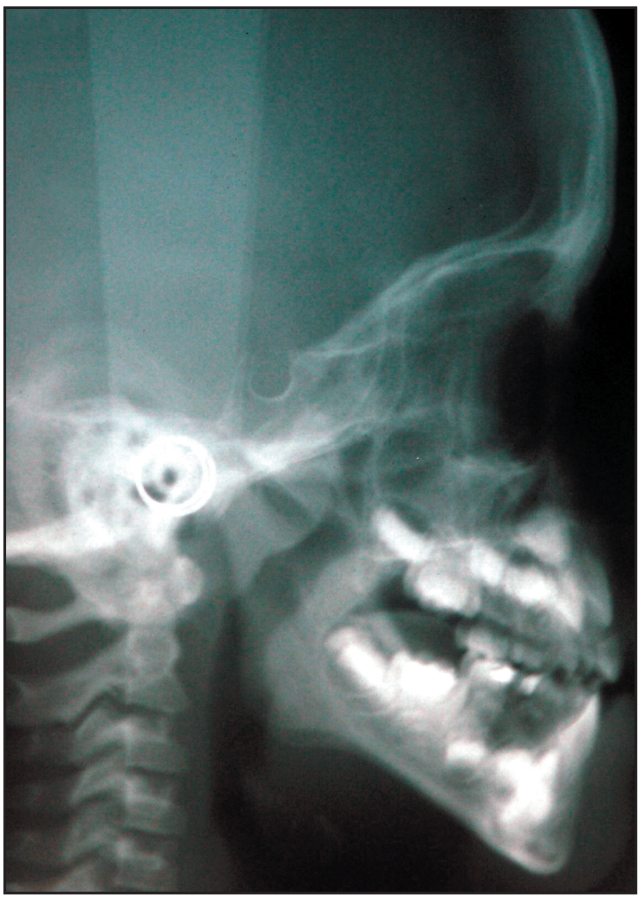

Figure 5. Profile radiograph illustrates the normal outline of sella turcica.

Posterior crossbite is another feature found in some SMMCIS cases.[1,6,18] Beckor et al[24] analyzed ortopantomograms of 11 SMMCIS patients and concluded that, while the vertical and horizontal maxillary growth were normal, the transversal growth might be altered. Maxillary atresia can be explained in cases of nasal obstruction and oral breathing associated with reduction in arch perimeter by the absence of a tooth. The patient's occlusion was borderline in the centric relationship. Because of the occlusion repositioning provoked by contact with a canine tooth, he dislocated the mandible to the right side.

Some authors recommend that the dental treatment should be initiated just at the permanent dentition using orthodontic appliances to gain space for implant-prosthodontic rehabilitation.[3,4,13]

This anomaly can alert the clinician to the possibility of other systemic problems related to SMMCIS that require special care.[3] Because in most cases the dentist is the first professional to evaluate a patient with this condition, it is important to know about SMMCIS and the required treatment and diagnostic options. SMMCIS treatment should be performed by a multidisciplinary team, including a dentist and specialized professionals, such as geneticist, otolaryngologist, endocrinologist, and psychiatrist (indispensable for long-term follow-up of the patient's normal growth and development).

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