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OSTEOGENIC DISTRACTION TO TREAT SOLITARY MEDIAN MAXILLARY CENTRAL INCISOR SYNDROME: A CASE REPORT

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Trabalho apresentado à Universidade Fernando Pessoa

como parte dos requisitos para obtenção do grau de

Mestre em Medicina Dentária

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RESUMO

A síndrome do incisivo central superior médio solitário (SICSMS) é uma doença de desenvolvimento rara caracterizada por um único incisivo central superior simétrico. A literatura tem relatado poucos casos com tratamento dentário abrangente. Nenhum tratamento cirúrgico foi proposto até o momento.

Relatamos o caso de uma menina de oito anos de idade inicialmente tratada com expansão rápida da maxila. Após o fracasso de duas expansões rápidas da maxila, a paciente foi tratada com sucesso com distração osteogénica aos 12 anos de idade. A distracção, associada a um tratamento ortodôntico fixo multi-brackeado, criou espaço suficiente para colocar um incisivo central protético sem extracções dentárias.

A distracção osteogénica é uma opção interessante para tratar pacientes com SICSMS.

Palavras-chave: Síndrome dos incisivos centrais superiores solitários, distração osteogénica, agenesia dentária, relato de caso

ABSTRACT

Solitary median maxillary central incisor syndrome (SMMCI) is a rare developmental disorder characterized by a single symmetrical maxillary central incisor. The literature has reported few cases with comprehensive dental treatment. No surgical treatment has been proposed before.
We report the case of an eight-year-old girl initially treated with rapid maxillary expansion. After the failure of two rapid maxillary expansions, the patient was successfully treated with osteogenic distraction at the age of 12 years old. The distraction, associated with multi-bracketed fixed orthodontic treatment, created enough space to place a prosthetic central incisor without dental extractions.
Osteogenic distraction is an interesting option to treat patients with SMMCI.

Keywords: Solitary median maxillary central incisor syndrome, osteogenic distraction, dental agenesis, case report

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FIGURE INDEX :

1. I - INTRODUCTION

Solitary median maxillary central incisor (SMMCI) syndrome is a complex disorder and a rare dental anomaly, estimated to occur in approximately 1 in 50,000 live births (Hall, 2006). It results from an in utero midline developmental defect occurring at about the 35th to 38th day from conception. The etiology is unknown. A missense mutation in the *SHH* gene (I111F) at 7q36 may be associated with SMMCI (Nanni *et al.*, 2001). The most characteristic feature in this condition is a symmetrical single maxillary central incisor tooth located exactly in the midline of the maxillary arch.

As demonstrated by Hall (2006), the phenotypic spectrum is highly variable. In addition to the characteristic tooth, the following characteristics are typically associated: preterm birth and low birth weight (37%), potentially life-threatening congenital nasal airway obstruction (choanal atresia, mid nasal stenosis or nasal pyriform aperture stenosis; over 90%), short stature (50%), small head circumference (33%), hypotelorism (45%), intellectual disability varying from slow learning to severe disability (50%), deviant sella turcica and pituitary gland morphologies (10–50%), and possibly epilepsy and attention deficit hyperactivity disorder (Hall, 2006; Hall *et al.*, 1997).

Few authors have reported cases with comprehensive dental treatment. Two proposed maxillary expansion (Bolan *et al.*, 2010; Lygidakis *et al.*, 2013), and one proposed a premolar transplantation (Pseiner, 2014).

This article aims to describe an alternative treatment: osteogenic distraction associated with orthodontic therapy.

2. II - CLINICAL CASE

An eight-year-old girl first consulted for a maxillary dental eruption disorder. The patient presented a single median permanent central maxillary incisor, an absence of upper lip frenulum, and a medio-palatal ridge (*Figure 1*). Intraoral examination showed an Angle Class II occlusion associated with a maxillary transverse deficiency (but without crossbite). Extraoral examination revealed a deviated septum. However, the patient did not present other symptoms of SMMCI syndrome.

Figure 1: intraoral photograph of the patient at the beginning of the treatment

The diagnosis was confirmed by orthopantomogram (*Figure 2*). A Cone beam Computed Tomography (CBCT) was performed to complete the dental diagnosis, revealing a partial fusion of the midpalatal suture (*Figure 3*). Rapid maxillary expansion (RME) was the primary treatment plan. The desired effect was correction of the maxillary transverse deficiency and enlargement of the dental arch to place a prosthetic central incisor without premolar extraction.

A first RME was initiated at the age of eight. However, the treatment was unsuccessful. A V-shaped maxillary arch persisted, and the enlargement was insufficient. A second RME with a fan-shaped maxillary expander was performed when the patient was 10 years old, which also failed indicating that the midpalatal suture was fusionned. After a multidisciplinary consultation, a surgical option was proposed. Performing a bipartite maxillary osteotomy (Le Fort I with asymmetric intermaxillary disjunction for osteogenic distraction) was decided. The patient was 12 years old at the moment. The distraction aimed to enlarge the dental arch to place a prosthetic central incisor. In our case, an asymmetrical distraction was performed to lateralize the incisor and correct the axis of the incisal root.

Figure 2: orthopantomogram of the patient. Only one central incisor is present.

Figure 3: CBCT displaying the midpalatal suture fusion

During the procedure, in association with the Le Fort I osteotomy, an additional asymmetrical intermaxillary osteotomy line was made, starting from tooth 22 and joining the midline approximately 2 cm back (*Figure 4*). Finally, an upper lip lengthening plasty was performed before closure. The internal distraction device was activated at 2 mm intraoperatively, following the protocol of the department. Then, activation of 0.25 mm per day for two weeks was implemented. No short term complication (bleeding, excessive pain or infection) occurred.

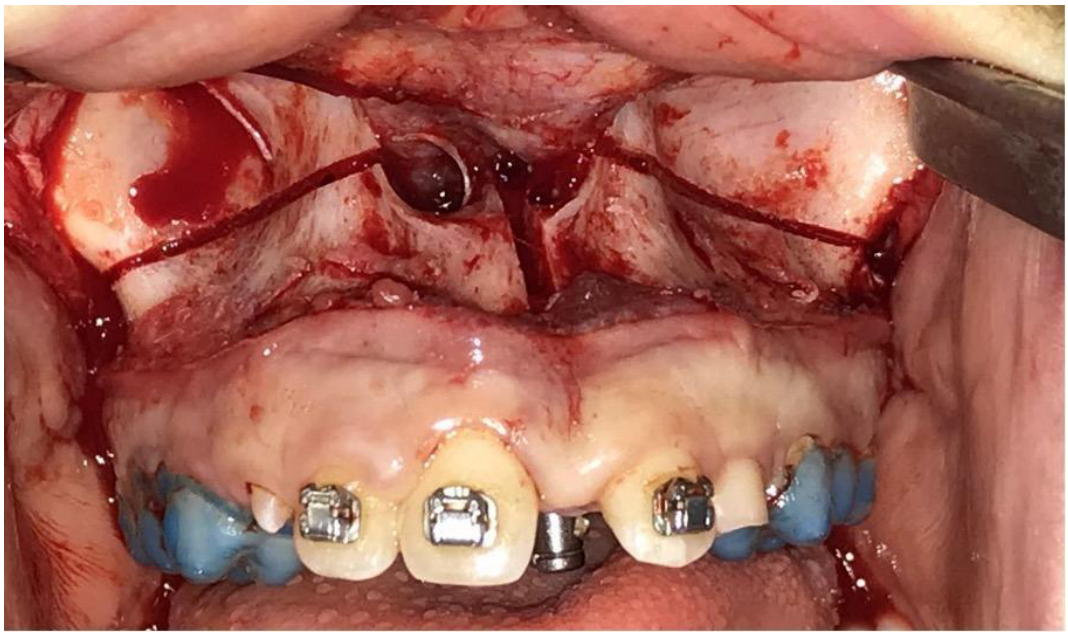

Figure 4: P er operative photograph of the osteotomy lines

After eight months of postoperative surgery, the benefits were clear: the arch was harmonious and rounded. Considerable space was gained on the dental arch, facilitating the opening of the space for the second central incisor. The following step was multi-bracketed fixed orthodontic treatment to align and open the space for the second central incisor. Thereunto, fixed appliances were used, involving .022x.028 self-ligating brackets. First, the dental arches were levelled and aligned with sequential .014, .016, .018, .020 nickel-titanium wires. Then, molar distalization on the maxillary arch was performed with springs and .016x.022. nickel-titanium wires. The following step was the lateralisation of the existing incisor to the right side and the opening of the space for a prosthetic left central incisor (tooth 21). Intermaxillary elastics were used to complete the interdigitation. When the multi-bracketed fixed orthodontic treatment was complete, the brackets were removed and the teeth polished. Immediately after debonding, the patient started to wear a clear plastic retainer every night to maintain the space of the missing incisor. The final step of the treatment plan was the replacement of the missing tooth #21 by a bonded bridge. A coronoplasty was performed on the single incisor (tooth 11) to convert the anatomy of the respective tooth into a more natural central incisor size and form (*Figure 5*). A second clear plastic retainer was produced to maintain the final result and was worn as a retainer. When the patient reaches 18 years old and her facial growth will probably be complete, she could benefit from a possible implant rehabilitation.

Figure 5: 1 year after debonding intraoral photograph: tooth 21 is missing, replaced by a bonded bridge

3. III - DISCUSSION

SMMCI syndrome is a rare developmental disorder characterized by a single symmetrical maxillary central incisor. Additional features have been reported: a pseudo-notched or arch-shaped appearance of the upper lip with an indistinct philtrum, the absence of a labial frenulum, and a V-shaped palate with an unusual narrow ridge along the midpalatal suture (Hall, 2006).

Few cases of SMMCI have been reported. Most authors have described the clinical features or discussed treatment alternatives without providing clinical results (McNamara *et al.*, 1999 ; Negi *et al.*, 2020). To our knowledge, only three authors have reported cases with comprehensive dental treatment. Pseiner (2014) proposed a premolar transplantation associated with the extraction of three remaining premolars in an 11-year-old girl. Lygidakis *et al.* (2013) presented the case of a patient with SMMCI associated with congenital nasal pyriform aperture stenosis. They chose to enlarge the maxilla using a slow maxillary expansion. Bolan *et al.* (2010) described the case of a six-year-old boy. They proposed rapid maxillary expansion (RME) to enlarge the maxillary arch and treat the posterior crossbite. They performed CBCT to evaluate the impact of RME on the suture. After RME, the crossbite was corrected. However, the CBCT showed no suture opening after the activation of the expander screw.

In our case, RME therapy was attempted twice. However, it failed due to the partial fusion of the midpalatal suture. After a multidisciplinary consultation, performing a bipartite maxillary osteotomy (Le Fort I with asymmetric intermaxillary disjunction for osteogenic distraction) was decided to enlarge the maxilla and gain space for a prosthetic incisor without dental extraction. Osteogenic distraction is a method of increasing bone length with gradual increments of bone separations (Ilizarov., 1988). It is mainly used in craniofacial surgery to correct maxillary hypoplasia, such as in cleft palate (Yu *et al.*, 2012). This option presents numerous advantages: in addition to the gained space, nasorespiratory function is improved (Saito *et al.*, 2006). The morbidity associated with distraction osteogenesis is very low (Kloukos *et al.*, 2018). The main disadvantage is the risks of hemorrhage, loss of upper lip, maxillary tooth sensitivity and infection (Cheung *et al.*, 2006). None of these complications occurred in our case.

Another treatment option was to extract premolars. However, we chose to reject this possibility. Premolar extraction constricts the dental arch, as demonstrated by Cotrin *et al*. (2020). Their study compared the maxillary and mandibular arch dimensions in two groups of patients, with and without premolar extraction. At the end of the treatment, the dental arch dimensions were significantly smaller in the extraction group, except for the maxillary intercanine width (Cotrin *et al.*, 2020). In our case, the patient already had small maxillary dimensions, and premolar extraction would have worsened the maxillary transverse deficiency and respiratory prognosis.

Relatively to the prosthesis rehabilitation, the age of the patient was a limitator for a definitive treatment intervention. Although specific clinical cases (Bohner *et al.,* 2019) support the use of dental implants in early age, the patient did not have finished her complete facial growth and an implant installation would have aesthetical and bone limitations at the long term. The installation of mini-implants vertically or a wire-band-tooth were also considered but were declined.

Therefore, our treatment plan was very satisfactory to address the goal we expected and the patient and her family were very satisfied with the results, including the respiratory benefit achieved.

4. IV - CONCLUSION

Osteogenic distraction is an interesting option to treat patients with SMMCI without dental extraction.

This therapy allows to keep the maxillary and the pyriform orifices wide so as not to compromise the normal ventilatory function.

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VI – ANNEXES

Universidade Fernando Pessoa

Exma. Senhora
Prof. Doutora Sandra Gavinha
Directora da FCS

Nº	Data
FCS/MED – 199/21	13 de Julho de 2021

Exma. Senhora Professor Doutora,

A Comissão de Ética, depois de apreciado o projeto de investigação apresentado por Jean-Baptiste Pierre Marcel Gilbert Kerbrat, intitulado "Osteogenic distraction to treat a solitary median maxillary central incisor syndrome: a case report", a realizar no âmbito do Mestrado Integrado em Medicina Dentária, considera o estudo muito pertinente. O objectivo do estudo é efectuar uma revisão da literatura científica sobre o síndrome do incisivo central maxilar solitário, a partir da apresentação de um interessante caso clínico a fim de comparar o diagnóstico e o tratamento usado com o que é relatado na literatura científica.
A autoria deste caso clínico é da inteira responsabilidade do investigador principal que é o director clínico da Clínica onde este paciente foi tratado. Por ser menor de idade, o consentimento informado de autorização de participação voluntária foi assinado pela responsável pela menor, concordando que os dados clínicos, incluindo fotografias e radiografias, podem ser usados para fins científicos. Não está em causa o risco e o bem-estar do participante interveniente, nem qualquer alteração do plano de tratamento previsto.
A Comissão de Ética nada não tem a opor à realização do estudo.

Com os melhores cumprimentos.

A Presidente da
Comissão de Ética da UFP

Teresa Toldy