Introduction

Dental anomalies in number, shape or structure are very frequent [Bulut et al., 2011; Militi et al., 2011; Laganà et al., 2011; Pippi, 2011]. However, Regional Odontodysplasia (RO) is a rare and severe anomaly of dental development [Guzman et al., 1990; Wilson and Ali, 2002]. It is not considered hereditary [Crawford and Aldred, 1989; Gerlach et al., 1998] and the aetiology is not completely known yet. Suggested aetiologic factors include somatic mutations affecting the dental lamina, vascular disorders, pharmacological treatments during pregnancy, local or systemic viral infection, activation of latent viruses, migration or differentiation failures of neural crest cells, local traumas, nutritional or metabolic deficits, radiation, hyperpyrexia, local ischaemia and Rh incompatibility [Wilson and Ali, 2002; Pandis et al., 1991; Van der Waal et al., 1993; Steiman et al., 1991; Von Arx, 1998] among others. The location includes the maxilla, mandible or both [Ozer et al., 2004; Pandis et al., 1991], but usually only one arch is affected and more commonly the upper left one [Hamdan et al., 2004]. The affected area, generally, does not cross the midline [Guzman et al., 1990; Ozer et al., 2004; Yuan et al., 1997; Hamdan et al., 2004;]. Morphology and structure are abnormal with severe mineralisation defects, rough and irregular with streaks, recesses and deep grooves [Crawford and Aldred, 1989]. Radiographically the thin layers of enamel and dentin are less radiopaque [Crawford and Aldred, 1989; Gerlach et al., 1998; Ozer et al., 2004] and they are difficult to differentiate from one another, conferring the teeth a typical radiographic appearance of "ghost teeth".

The main treatment objectives are: space maintenance, restoration of masticatory function and aesthetics, reduction of the psychological effects caused by the absence of teeth, vertical dimension maintenance [Yuan et al., 1997; Rosa et al., 2006; Hamdan et al., 2004], and jaws normal growth [Hamdan et al., 2004] to prevent facial asymmetries [Rosa et al., 2006].
The purpose of this article is to report some unusual characteristics related to gender, location and detected manifestations of severe RO in three cases.

**Case reports**

**Case 1**
Male, 7 years old boy whose parents requested dental care due to delayed eruption of several permanent teeth. The medical history revealed no data of interest, except breathing difficulties at birth, which required emergency airways management.

Parents reported that temporary teeth of the lower right hemiarch had a different aspect. At 3 years and 4 months the child presented a severe infection in that area, requiring the extraction of lateral incisor, canine and first temporary molar. Parents provided radiographs performed at the age of 5 in which it was already possible to appreciate disturbances in the primary and permanent teeth of the area involved.

The child presented a 1st phase of mixed dentition, with normal teeth except at the lower right hemiarch, where he just had a central incisor and a second temporary molar of abnormal size and structure. Occlusion was altered on the right side and he had caries in both temporary and permanent teeth.

Radiographs taken at that time showed normal germs except in the right inferior area where we could appreciate dental germs with reduced radiolucency and disrupted morphology (Fig. 1). The degree of dental impairment was higher in the teeth next to the midline, decreasing distally. Several dental tissues were unrecognisable in the incisors, canine and first premolar, and in the large pulp cavity, teeth presented pulpolites (Fig. 1, 2). The second premolar was less affected, however, the first permanent molar showed modification of crown mineralisation and a peculiar internal morphology caused by the absence of a well defined pulp chamber. The second permanent molar did not show any signs of alteration (Fig. 2). Data from the clinical history and radiographic examination allowed the diagnosis of RO located in the lower right jaw.

Caries treatment was performed and the lower right central incisor was extracted. A paediatric partial denture was placed in the affected area. Six months later, the affected central incisor showed part of the root developed, but without erupting (Fig. 3). The patient did not attend any other recall appointment.

**Case 2**
Male, 13 years old boy born by caesarean section, with uneventful medical history. The parents sought attention at the paediatric dental department due to the lack of eruption of the teeth in the lower left hemiarch. The mother recalled that temporary teeth in that area had partially erupted, presenting an atypical appearance and colour, but they had all been extracted at the age of 3, after an infectious process with cellulitis in the area.

By the time of our clinical examination, teeth eruption was completed on the right side and occlusion was within normal parameters. In the lower left hemiarch it was only possible to appreciate a permanent central incisor, of yellow colour and small size, and there was a marked reduction of the alveolar bone. In the upper left hemiarch an unerupted canine was missing and the patient showed an altered dental alignment (Fig. 4).

Panoramic and periapical radiographs, revealed teeth of normal size and shape, except for the third quadrant, where it could be observed, starting at the permanent central incisor already erupted, dental germs with very atypical morphology and structure (Fig. 5). Teeth from the lateral sector exhibited greater radiographic alterations, specially the canine, presenting an image which made it inconclusive to determine whether it was just the canine or both the canine and the first
premolar (Fig. 6), and the first permanent molar with severe mineralisation defects and a completely altered internal morphology. The second permanent molar was not radiographically affected (Fig. 7).

Furthermore, radiolucencies were visible also in the incisor apex and around the canine, premolar and molar germs. Due to preceding history and teeth radiological appearance, RO was diagnosed.

The treatment consisted of temporary rehabilitation with partial denture and orthodontic treatment. In the future it will be necessary to assess the need for extraction of affected permanent teeth, in case they are non-viable, and rehabilitation with dental implants.

Case 3
Male, 4 years and a half, Russian, adopted. At 10 months after birth he was diagnosed perinatal encephalopathy and rachitism. At the age of 2.5 years he suffered from purulent periostitis on the right side of the maxilla; two months later an airways inflammation, purulent otitis and enterobiosis were diagnosed. Five months later, after a new episode of periostitis in the same area, all temporary teeth from that hemiarch were extracted.

At the time of clinical examination, all temporary teeth were normal. In the upper right quadrant there were some dental or bone remnants. Occlusion on the left side was normal, while it was altered on the right side due of the lack of vertical and transversal growth of the jaw (Fig. 8). Extrusion and separation of mandibular teeth could be observed, as well as an altered occlusal plane and bigger diastema in the affected side.

On facial examination, erythematous areas were observed at the center and at the right side of the upper and lower lips, which were diagnosed as angiomas. The radiographs, both panoramic and intraoral, showed very low radiopacity of all permanent germs of the upper right side, which crossed the midline affecting the left permanent incisors as well (Fig. 9). Remains of atypical right incisors could also be identified. Clinical and radiological data were compatible with RO.
Root fragments from affected temporary teeth were extracted obtaining several grey fragments of small size and hard consistency. Microscopic examination revealed that they were dental root fragments with moderate lymphocytic infiltrates and fibrous reaction. The examined mucosa was lined by a squamous epithelium with normal morphology.

A removable partial denture was placed to replace the missing temporary teeth on the upper right maxilla and occlusal adjustments were made on the opposing teeth, in order to obtain better aesthetics and to stop growth disturbance in that area (Fig. 10).

After 5 years of follow-up, teeth replacement had begun and growth deficits in the right jaw persists, as well as an altered occlusal plane, although it had not worsened. Radiographs confirmed that RO affects every teeth starting on the upper right second permanent molar to the upper left permanent lateral incisor. The first permanent right molar developed better than expected, although its structure is altered (Fig. 11). Temporary upper incisors have a delayed replacement.

Both central and lateral left temporary incisors were extracted, and histologically they showed a normal structure. A partial denture was adapted freeing the right molar area in order to avoid altering the permanent molar development and eruption and also to use it as support in case, in the future, it becomes a possibility (Fig. 12).

The cases reported above show many of the clinical and radiological characteristics referred in the literature for RO, however they present multiple variations that differentiate and make them interesting for analysis. It is said that RO is more frequent in women [Wilson and Ali, 2002; Gerlach et al., 1998; Yuan et al., 1997; Van der Waal et al., 1993] and located in the maxilla [Hamdan et al, 2004], but the three cases we have studied do not follow these distribution patterns, since they are all young male subjects and, in two cases, it is the mandible that is affected.

Also, it is very rare for several quadrants to be affected [Guzman et al., 1990; Ozer et al., 2004; Rosa et al., 2006; Pandis et al., 1991]. Two of the presented cases follow this pattern, with just one quadrant affected in both dentitions; however in the third case the
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The anomaly is modified from just one quadrant affected in temporary dentition to two quadrants affected in permanent dentition. In this case the anomaly passes the midline and the left incisors, central and lateral, are both affected as those on the right side. In the majority of cases found by the authors in which the anomaly passes the midline, the central incisor was the only tooth affected, whereas in the case presented it affects both incisors with a same extent.

In Table 1, findings of the three observed cases can be found. In all of them, the anomaly is present on both dentitions, as it is most commonly seen [Wilson and Ali, 2002; Gerlach et al., 1998; Yuan et al., 1997] and, as expressed by many authors, this alteration is more likely to affect incisors and canines than posterior teeth [Guzman et al., 1990; Ozer et al., 2004; Crawford and Aldred, 1989; Van der Waal et al., 1993; Raez, 1990]. In the cases presented, both anterior and posterior teeth were affected, although in Case 2 the incisors were less affected than all the other teeth in the same hemiarch.

RO usually affects consecutive teeth, without any normal teeth between them [Crawford and Aldred, 1989; Yuan et al., 1997; Rosa et al., 2006] In Case 1 and 2 we found premolars relatively affected, among the teeth closer to the midline, as described by Gerlach et al. in 1998. The internal morphology of the altered mandibular permanent premolars, Case 1 and 2, draws special attention, as they present a very atypical pulp chamber especially in the area of the root bifurcation. We did not find any previous description of these findings. Radiographic images of the presented cases, with a reduced radiopacity, morphology and dental structures hardly recognisable, correspond to the typical image of “ghost teeth” attributed to RO.

At present aetiology remains unknown [Wilson and Ali, 2002; Pandis at al., 1991; Van der Waal et al., 1993; Yuan et al., 1997; Von Arx , 1998], despite the great variety of causes that have been mentioned. In Case 1, medical history reflects the need for urgent management during birth due to airways obstruction, therefore a possible traumatic factor is the only suspicious data. In Case 2 no possible aetiological factor was found.

In Case 3, vascular changes may be associated with the extensive history of infections in the altered side until 3 years of age. In this case RO passed the midline, affecting both incisors. As it is well known, nerve branches and blood vessels from the maxilla middle area may be extended up to the contralateral incisors, and this, together with the timing the infections occurred, gives us a possible aetiological justification of infectious transmission across the midline.

In the literature there are multiple references to the risk of presenting acute infectious processes and abscesses, even in the absence of caries [Rosa et al., 2006; Von Arx, 1998]. The three studied cases reported having suffered acute infections of the temporary dentition of the affected side at an early age, requiring some teeth extractions. Besides, in Case 2, in the apex of the recently erupted central incisor and also around canine, premolar and molar germs, one could observe radiolucencies. According to the literature, growing and swelling of fibrous tissues can be observed clinically and radiologically when teeth do not erupt. Although in this case no growth or swelling of fibrous tissues could be observed, the teeth did not erupt [Wilson and

<table>
<thead>
<tr>
<th>Case</th>
<th>Gender</th>
<th>Affected Dentition</th>
<th>Arch and Side</th>
<th>Eruption Delay</th>
<th>Intermediate not affected teeth</th>
<th>Other dental findings</th>
<th>Infection history</th>
<th>Possible aetiological factor</th>
<th>Occlusal changes</th>
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<tbody>
<tr>
<td>Case 1</td>
<td>Male</td>
<td>Temporary and Permanent</td>
<td>Mandible: Right side</td>
<td>Yes</td>
<td>2nd Premolar</td>
<td>Atypical internal morphology of permanent molar</td>
<td>Yes</td>
<td>Local trauma during birth</td>
<td>Yes</td>
</tr>
<tr>
<td>Case 2</td>
<td>Male</td>
<td>Temporary and Permanent</td>
<td>Mandible: Left side</td>
<td>Yes</td>
<td>Premolar?</td>
<td>Premolar agenesis or germs fusion. - adiolucencies - Atypical internal morphology of permanent molar</td>
<td>Yes</td>
<td>No relevant data</td>
<td>Yes</td>
</tr>
<tr>
<td>Case 3</td>
<td>Male</td>
<td>Temporary and Permanent</td>
<td>Maxilla: - Temporary teeth; right side - Permanent teeth; right and left side</td>
<td>Yes</td>
<td>No</td>
<td>Crosses midline - Second permanent molars affected</td>
<td>Yes</td>
<td>Angiomas - Local infections</td>
<td>Yes</td>
</tr>
</tbody>
</table>

**TABLE 1** The most relevant findings of the 3 RO cases reported in the present article.
Ali, 2002; Crawford and Aldred, 1989; Gerlach et al., 1998; Rosa et al., 2006).

The data reported above greatly limit treatment goals and therapeutic options. In general, the treatment aims at space maintenance [Yuan et al., 1997], restoring masticatory function, improving aesthetics, reducing psychological effects caused by teeth absence, vertical dimension maintenance [Yuan et al., 1997; Rosa et al., 2006; Hamdan et al., 2004; Bulut et al., 2011], allow jaws normal development [Hamdan et al., 2004] and prevent facial asymmetries [Rosa et al., 2006].

It is considered that if there is any possibility for the affected teeth to be preserved by means of restorative techniques, this will be the better option. Indeed, if teeth are not erupted, we have to consider the convenience of preserving the germs until the end of growth, to preserve as much alveolar bone as possible, to facilitate future implant treatment. However, when infections are severe, it is necessary to extract the germs.

Delays and failures in the eruption are common in this disease. Most publications have focused primarily on the radiographic and histological findings, without relating these dental defects with facial structure changes. Just a few authors mentioned facial asymmetry, maxillary hypoplasia and abnormal growth of the jaws [Crawford and Aldred, 1989; Pandis et al., 1991].

In all cases presented, RO coincides with altered occlusion. In Case 3 we can see occlusal plane inclination and also the width of the affected side of the maxilla seems to be reduced. According to Pandis et al. (1991), this maxillary vertical asymmetry, which produces the occlusal plane inclination, may be related with the dental absence in the right side of the maxilla and consequent lower alveolar growth and antagonists’ over-eruption.

In conclusion RO is a rare disease, causing severe dental, growing and craniofacial development anomalies. Treatment needs to be personalised, aiming at preservation of the affected teeth, but taking into account the risk of these teeth to develop severe infections. Parents should be aware of the need for an extensive follow-up of the patient.

References