Regional odontodysplasia: An analysis of 161 cases from 1953 to 2017

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Abstract

Background and objective: Regional odontodysplasia (RO) is a localized developmental anomaly involving both dentitions. The characteristic clinical findings are soft discolored several contiguous teeth with atypical morphology associated with swelling or abscess formation. Radiographic images of the involved teeth show a ghost-like appearance because enamel and dentin are hypoplastic and hypocalcified. Although RO is known for more than seven decades, the literature on RO is mostly limited to case reports.

The objective of this investigation was to provide an updated review of the literature with regards to clinical presentation and epidemiology of RO. Because of its rarity, retrospective analysis of the published reports across the world is considered one of the most appropriate available methods to analyze epidemiological and clinical data of RO. This review was reported and conducted according to the PRISMA guidelines.

Results: Between 1953 and 2017, 161 cases were reported in literatures written in English. At the time of diagnosis, the age of the patients ranged between 1 and 25 years. There was slight female predilection with a ratio of 1.37:1. When examined collectively, the maxilla was commonly affected compared to the mandible. RO occurred in both dentitions in 75 reported cases. While it affects the deciduous teeth alone in 19.3% and the permanent teeth in 34.2% of the cases. Failure of tooth eruption was the most common presenting finding (41.6%) followed by swelling of the affected area (32.3%).

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Conclusion: Although RO was first described almost 80 years ago and the total subsequent case reports were over 160 cases, there is no consensus on its pathogenesis and how it should be treated. The descriptive nature of the examined case reports reveals the limited information on this condition. Further experimental and genetic studies are needed.

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1. Introduction
Developmental anomalies are interesting findings, which raise the curiosity of clinicians to seek more knowledge about their origin and nature. Regional odontodysplasia (RO) is a sporadic developmental anomaly that is still ambiguous yet it provokes questions regarding its pathogenesis. It is defined as a localized non-hereditary developmental anomaly with adverse effects on the formation of dental tissues (Gardner and Sapp, 1973; Lustmann et al., 1975).

Hitchin (1934) was the first to describe the condition, however it was not given a specific term at that time. Then, various terminologies were designated such as “arrested tooth development” (McCall JO, 1947), “shell teeth” (Rushton, 1954), “unilateral dental malformation” (Bergman et al., 1963), and “ghost teeth” (Rushton, 1965). Zegarelli et al. (1963) first introduced the most widely used term “odontodysplasia”.

Later, Pindborg (1970) added the term ‘regional’ emphasizing the localized nature of this lesion.

The etiology of this disease remains unknown although several theories have been suggested. Local trauma, infection, teratogenic drug exposure, local circulatory disorders, Rhesus incompatibility, irradiation, neural damage, hyperpyrexia, vitamin deficiencies, metabolic and nutritional disorders have all been assigned as predisposing factors to RO (Bergman et al., 1963; Sibley and Zimmermann, 1962; Walton et al., 1978; Zegarelli et al., 1963). Activation of latent viruses in the odontogenic epithelium, and disorders of neural crest cell migration also have been suggested (Dahllöf et al., 1987; Nagai et al., 1963; Sabah et al., 1992). All authors agree that this is a non-hereditary condition, however the distribution of affected teeth in all four quadrants in some cases makes some authors suggest that somatic mutation may be the causative factor (Grover, 2014; Matsuyama et al., 2014; Sabah et al., 1992).

Clinically, the affected teeth are usually small with atypical morphology with yellow or brown discoloration (Farne et al., 1986; Marques et al., 1999). The age at the time of diagnosis ranges from 1 to 23 years. RO occurs in both dentitions and both jaws; however, it is usually present in one jaw and typically in one quadrant (Crawford and Aldred, 1989; Tervonen et al., 2004). Radiographic images of the involved teeth show a ghost-like appearance. Affected teeth regularly show enlarged pulp chambers and root canals with open apices (Rushton, 1965). Histologically, almost all dental tissues are altered. Enamel and dentin appear as hypoplastic and hypocalcified and the pulp is larger than normal (Carlos et al., 2008). Treatment of RO needs to be individualized and requires a multidisciplinary approach (Cahuana et al., 2005; Mukhopadhyay et al., 2016).

Although RO is known for more than seven decades, the literature on RO is mostly limited to case reports. The aim of this article was to assess reported cases of RO in the English literature and to assemble epidemiological and clinical data.
software (SPSS version 22.0) was utilized for data analysis. This review was reported and conducted according to the PRISMA guidelines.

3. Results

Between 1953 and 2017, 161 cases were reported in the English literature. Results are summarized in Table 1. At the time of diagnosis, the age of all reported patients ranged between 1 and 25 years. The review of the age distribution showed bimodal peaks; 2 to 5 years and 8 to 12 years (Fig. 1). There was a slight female predilection with a ratio of (1.37: 1). Around 42% of the cases occurred in males, and 57.8% occurred in females. When examined collectively, the maxilla was commonly affected compared to the mandible with 55.9% and 33.5% of the cases respectively.

In each patient, one single quadrant, bilateral quadrants, ipsilateral quadrants, or contralateral quadrants were affected. Seven cases showed generalized odontodysplasia (Chaudhry et al., 1961; Herman and Moss, 1976; Ibrahim Mostafa et al., 2013; Lowe and Duperon, 1984; Russell and Yakobi, 1993; Sabah et al., 1992; Walton et al., 1978) and two cases reported one single tooth affected (Gurunathan et al., 2011; Pullon and Miller, 1984). When single quadrant was solely affected, the order of occurrence was as follows; left maxilla; right maxilla; left mandible; and right mandible with percentages of 24.8%, 25.5%, 13%, 11.2%, respectively.

RO occurred in both dentitions in 75 reported cases. While it affects the deciduous teeth alone in 19.3% and the permanent teeth in 34.2%. The distribution of affected teeth according to their type is summarized in Table 2. The deciduous teeth were almost equally affected. In the permanent dentition, anterior teeth are the most frequently affected while the least affected tooth was the third molar.

Regarding the reported clinical signs and symptoms, failure of tooth eruption was the most common presenting findings of

| Table 1  | Clinical data of reported cases of RO. |
|----------|----------------------------------------|
| Clinical data | No. (%) |
| **Age (years)**      |            |
| Mean               | 7.9 years  |
| Range              | 1–25 years |
| **Gender**         |            |
| Male               | 68 (42.2)  |
| Female             | 93 (57.8)  |
| **Jaw**            |            |
| Maxilla            | 90 (55.9)  |
| Mandible           | 54 (33.5)  |
| Both               | 17 (10.6)  |
| **Dentition**      |            |
| Deciduous          | 31 (19.3)  |
| Permanent          | 55 (34.2)  |
| Both               | 75 (46.6)  |
| **Site**           |            |
| Right maxilla      | 40 (24.8)  |
| Left maxilla       | 41 (25.5)  |
| Left mandible      | 21 (13)    |
| Right mandible     | 18 (11.2)  |
| Bilateral maxilla  | 7 (4.3)    |
| Bilateral mandible | 15 (9.3)   |
| Generalized        | 7 (4.3)    |
| Single tooth       | 2 (1.2)    |
| Ipsilateral or contralateral quadrants | 10 (6.1) |
| **Signs and symptoms** |            |
| Failure of tooth eruption | 67 (41.6) |
| Swelling of the affected area | 52 (32.3) |
| Local pain or tenderness | 39 (24.2) |
| Periapical inflammation | 19 (11.8) |
| Associated vascular nevus | 15 (9.3) |
| Hyperplastic gingival tissues | 14 (8.7) |
| History of trauma  | 6 (3.7)    |

![Fig. 1](image-url) Age of patients at time of diagnosis.
Table 2  RO of teeth as reported in the dental literature between 1953 and 2017.

| Tooth                  | Number of cases | Percentage of total |
|------------------------|-----------------|---------------------|
| Deciduous teeth        |                 |                     |
| Central incisor        | 71              | 44.1                |
| Lateral Incisor        | 68              | 42.2                |
| Canine                 | 72              | 44.7                |
| First molar            | 58              | 36.0                |
| Second molar           | 63              | 39.1                |
| Permanent teeth        |                 |                     |
| Central incisor        | 99              | 61.5                |
| Lateral Incisor        | 95              | 59.0                |
| Canine                 | 88              | 54.7                |
| First Premolar         | 71              | 44.1                |
| Second premolar        | 63              | 39.1                |
| First molar            | 76              | 47.2                |
| Second molar           | 49              | 30.4                |
| Third molar            | 19              | 11.8                |

41.6% followed by swelling of the affected area due to abscess formation (32.3%). Local pain or tenderness was reported in 24.2% of patients, periapical inflammation in 11.8%, and hyperplastic gingival tissues overlying the affected unerupted teeth in 8.7%. RO was found to be associated with vascular nevus in 9.3% of cases. History of trauma was reported in 3.7% of the patients.

4. Discussion

Many authors have considered RO to be an uncommon condition (Gibbard et al., 1973; Lucas and Eveson, 2012). Review of the literature reveals that most publications are case reports and only two retrospective studies which were presented by Crawford and Aldred (1989) and Tervonen et al., (2004). The purpose of this investigation was to provide an updated review of the English-language literature with regards to clinical presentation and epidemiology of RO. Because of its rarity, retrospective analysis of the published reports across the world is considered one of the most appropriate available methods to analyze epidemiological and clinical data of RO.

The present review showed bimodal peaks of occurrence; 2 to 5 years and 8 to 12 years which correlate with the anticipated time of eruption of primary and permanent dentitions, respectively. Similar observation was reported by the previous reviews (Crawford and Aldred, 1989; Tervonen et al., 2004). The present review showed a 1.37:1 ratio of females to males, which is somewhat similar to the findings of Crawford and Aldred, and slightly lower when compared with the findings of Tervonen et al. The literature review revealed that, of all reported cases of RO, about 55.9% were found in the maxilla, and 34.2% in the mandible. This predilection was also previously observed (Crawford and Aldred, 1989; Tervonen et al., 2004).

Generally, the reported cases exhibited common clinical features such as irregular surface contour of the teeth with pitting and groves, and discoloration. Similarly, all of the cases showed “ghost teeth” appearance radiographically. Nonetheless, they had varied variations with regard to the degree of involvement. Classically, the condition is a regional disorder affects a focal area of jaw, wherein few contiguous teeth are involved. In this review, one single quadrant was affected in 74.5%. It is hard to explain why one specific quadrant is involved while the others are not. However, the segmental nature of occurrence of these cases excludes the systemic causes. Sometimes, ipsilateral, contralateral or bilateral involvements in the same jaw have been described which account for 19.7%. On the other hand, the generalized involvement is rare which was reported in seven cases. In 1961, Chaudhry et al. (1961) was the first to describe a case where three quadrants where affected in a 7 years old girl. Later on, other authors (Sabah et al., 1992; Herman and Moss, 1976; Lowe and Duperon, 1984; Russell and Yacobi, 1993; Ibrahim Mostafa et al., 2013; Walton et al., 1978) reported generalized odontodysplasia with involvement of all quadrants. However, the diagnosis of some cases has been questioned due to poor radiographs (Lowe and Duperon, 1984; Russell and Yacobi, 1993). The differences of the locations and type of involvement of the cases may indicate that there is no single etiological factor and that each patient might have different cause than the other. Thus, no single etiological factor can reasonably explain the condition.

The anomaly affected both dentitions in the same region in about half of the cases. This finding suggests that although the exact etiology is unknown, the damaging factor must be active during the period of teeth development in both deciduous and permanent dentitions.

Regarding the reported clinical signs and symptoms, details of the cases history were not always presented. According to the current review, the most common clinical finding is failure of tooth eruption followed by swelling of the affected area. The affected teeth are irregular with malformed roots which prevent tooth eruption. Usually in RO the teeth are more prone to dental caries due to their defective structure and consequently periapical infection and swelling (Al-tuwirqi et al., 2014). Sometimes the associated swelling is attributed to non-inflammatory gingival enlargement in the affected area (Marques et al., 1999).

The associated sign and symptoms of this condition are very important to a point where possible etiological factors can be proposed and anticipated. Unfortunately, details of sign and symptoms were not always reported. One of the remarkable findings is the presence of vascular nevus which supports the possible evidence that vascular pathogenesis can lead to such lesion. Moreover, experimental studies showed that cutting the blood supply of various head and neck arteries resulted in similar changes of the dentition (Kraus et al., 1969). Walton et al. (1978) proposed that local disturbance of vascular supply produces a localized ischemia that disturbs odontogenesis. Another interesting finding was reported in a 5-year-old girl which presented with generalized enamel defects in the primary dentition in addition to regional odontodysplasia (Al-Mullahi and Toumba, 2016). Although generalized enamel defects in primary teeth of healthy children are relatively common (Robles et al., 2013), the authors did not present any investigations to correlate between those concomitant conditions.

In conclusion, regional odontodysplasia is a rare condition; however, it causes severe dental disturbances. Although it was first described almost 80 years ago and the total subsequent case reports were over 160 cases, there is no consensus on its pathogenesis and how it should be treated. The descriptive
nature of the examined case reports reveals the limited information on this condition. Further experimental and genetic studies are needed.

**Ethical Statement**

The manuscript is a Review Article of published cases and does not require an ethical approval.

**Declaration of Competing Interest**

The authors have no conflict of interest to declare.

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