

## ODONTODISPLASIA – CASO FAMILIAR RARO

### *ODONTODYSPLASIA – RARE FAMILY REPORT*

Thásia Luiz Dias Ferreira\*  
Jurandyr Panella\*\* (In Memoriam)  
Claudio Fróes de Freitas\*\*\*

#### ABSTRACT

Odontodysplasia is considered a rare dental development anomaly, which leads to enamel and dentin hypoplasia of the affected teeth, whose etiology is still uncertain and broadly discussed. At the clinical exam, the teeth with odontodysplasia are usually shorter than those of normality standards, with irregular crown shape and form, with hypoplastic, yellow or pigmented external surface. The radiographic aspect depends on the stage the anomalous tooth is x-rayed, given the different evolutionary stages of the mineralization process; however, in general, there is a significant reduction in the radiopacity of all mineralized structures, with no distinction between enamel and dentin, with abnormally wide pulp chambers and, at times, unshapely. In this paper the authors present a case of three sisters with odontodysplasia and the main clinical and radiographic characteristics of this development anomaly, based on a literature review. The work at issue is of clinical relevance as it shows three odontodysplasia cases in the same family, suggesting genetic inheritance, which opposes to the researched papers.

DESCRIPTORS: Odontodysplasia – Tooth abnormalities

#### RESUMO

A odontodisplasia é considerada um distúrbio de desenvolvimento dentário raro, que resulta na hipoplasia de esmalte e dentina dos dentes afetados, tendo sua etiologia ainda incerta e muito discutida. Ao exame clínico, os dentes com odontodisplasia usualmente se apresentam com o tamanho menor que os padrões de normalidade, o contorno e a forma das coroas alteradas, com a superfície externa hipoplásica, amarelada ou pigmentada. O aspecto radiográfico depende da fase em que o dente anômalo é radiografado, devidos aos diferentes estágios evolutivos do processo de mineralização; contudo, em geral, há marcante redução na radiopacidade de todas as estruturas mineralizadas, não se distinguindo esmalte de dentina, surgindo câmaras pulpares anormalmente grandes e, por vezes, disformes. No presente trabalho os autores apresentam um caso de três irmãs portadoras de odontodisplasia e as principais características clínicas e radiográficas dessa anomalia de desenvolvimento, embasados em uma revisão da literatura. O trabalho em questão é de relevância clínica por se tratar de três casos de odontodisplasia na mesma família, sugerindo herança genética, o que vem de encontro aos trabalhos pesquisados.

DESCRITORES: Odontodisplasia - Anormalidades dentárias

\* DS, Msc and PhD from Dental School of University of São Paulo – Brazil - Email: thasia@usp.br

\*\* Full Professor of the Radiology Dental School of University of São Paulo – Brazil.

\*\*\* Associated Professor of the Radiology Dental School of University of São Paulo – Brazil.

## INTRODUCTION

It is attributed to Mc Call & Wald (1947) the first description of this anomaly and to Zegarelli *et al.* (1963) the term odontodysplasia. Odontodysplasia is a rare anomaly in which the dental structures are affected (follicle, enamel, dentin, cementum and pulp), and it may involve both primary and permanent dentitions (Gerlach *et al.*<sup>1</sup>, 1998). Only one tooth may be compromised or a group of teeth or all teeth (Sadeghi e Ashrafi<sup>2</sup>, 1981).

The anomaly is more frequent in the maxilla than in the mandible, and there are more occurrences in the anterior region than in the posterior region Gomes *et al.*<sup>3</sup>, (1997) with no distinction in regards to ethnic groups (Gerlach *et al.*<sup>1</sup>, 1998, Gomes *et al.*<sup>3</sup>, 1997). In those cases where the anomaly would cross the midline, the term Odontodysplasia generalized was suggested (Ansari *et al.*<sup>4</sup>, 1997).

The etiology still remains uncertain, although there are some hypotheses, among them, one suggested that somatic mutations occurs in the early stages of tooth development, interrupting the normal sequence of odontogenesis Witkop<sup>5</sup>, (1975), but the most accepted theory refers to an ischemia caused by vascular disorders Gomes *et al.*<sup>3</sup> (1997), which also caused changes in the odontogenesis process Raez<sup>6</sup>, (1990), this theory is supported by the large number of haemangiomas in regions opposite to the teeth affected by this anomaly (Gomes *et al.*<sup>3</sup>, 1997, Fanibunda e Soames<sup>7</sup>, 1996). Other possible etiological factors are: failure in the migration of cells from neural crest, viral infection, trauma and drug use (medications) during pregnancy (Fanibunda e Soames<sup>7</sup>, 1996). However, no references favorable to the hereditary factor were found in the literature researched.

At the clinical exam the lack of teeth is a common finding, because usually the affected teeth do not erupt in oral cavity and when they do, in general, they are delayed Gerlach *et al.*<sup>1</sup> (1998), Gomes *et al.*<sup>3</sup> (1997); these teeth are usually shorter than usual, with irregular crown shape and form, with hypoplastic, yellow or pigmented external surface (Gerlach *et al.*<sup>1</sup>,

1998). Recurring infections and abscesses are common findings, as a result of the structural frailty of the affected teeth (Gerlach *et al.*<sup>1</sup>, 1998).

In general, the gum near the teeth presents edemas, hyperemia, fibrosis and it usually has fistulas (Gomes *et al.*<sup>3</sup>, 1997).

The radiographic aspect depends on the stage the anomalous tooth is x-rayed, given the different evolutionary stages of the mineralization process. In general, there is a significant reduction in the radiopacity of all mineralized structures, with no distinction between enamel and dentin, with abnormally wide pulp chambers and, at times, unshapely (Gibson *et al.*<sup>8</sup>, 2003). The deformity extends with more or less severity to the entire crown region, where enamel and dentin are irregular, with open apices and short roots (Gerlach *et al.*<sup>1</sup>, 1998). Multiple pulp nodules and/or pulp dystrophic calcifications can be found (Gomes *et al.*<sup>3</sup>, 1997). Given the reduction in the radiopacity of mineralized tissues, the lines that surround the dental tissues are weakly seen in radiographies; that is why affected teeth are commonly referred to as "ghost teeth" (Gerlach *et al.*<sup>1</sup>, 1998, Gomes *et al.*<sup>3</sup>, 1997).

This work presents the clinical and radiographic aspects of three sisters with odontodysplasia, suggesting genetic inheritance.

## CLINICAL CASE

Patient RCS, female, 17 years old, attended the radiology center for an extra-oral panoramic radiography. At the extra-oral clinical exam, the patient presented discrete facial asymmetry towards the left side (Fig. 1); in the intraoral exam, the patient had no posterior superior teeth as well as central incisors. Some primary teeth were still present in the oral cavity, but with changes in shape and color (Fig. 2).

Through a panoramic radiography, it was verified the presence of multiple teeth that have not erupted (Fig. 3). From permanent teething, the only teeth that erupted in the oral cavity were the first inferior molars; those not erupted presented radiopacity reduction, suggesting enamel and dentin hypoplasia, mainly in the coronal

FERREIRA TLD

PANELLA J

FREITAS CF

ODONTODISPLASIA

- CASO FAMILIAR

RARO



FERREIRA TLD  
 PANELLA J  
 FREITAS CF

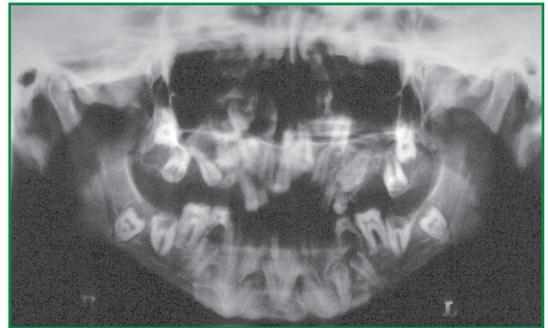
ODONTODISPLASIA  
 - CASO FAMILIAR  
 RARO



**Fig. 1** Ao exame clínico extraoral nota-se discreta assimetria facial para o lado



**Fig. 2** – Ao exame clínico intraoral notam-se vários dentes com alteração na forma e na coloração.



**Fig. 3** – Radiografia panorâmica apresentando múltiplos dentes não irrompidos e diminuição da radiopacidade dos tecidos mineralizados dos elementos dentários.

•• 280 ••

area, except for the third molars, which were within the normal radiographic standards; however, they presented an increase in the pericoronal flap; the teeth 5, 6, 10, 16, 17, 22, 27, and 32 presented root anomaly (dilacerations); the second superior pre-molar on the left side was missing. Primary teeth also presented changes in radiopacity and shape (short and conic), suggesting odontodysplasia in all teeth, both primary and permanent. Intraoral periapical radiographies were taken for more detailed observation (Fig. 4).

At the end of the exams the patient reported that the teeth of her two sisters were equal to hers, so her sisters were called for an assessment. Her middle sister was 20 years old and at the intraoral exam, the gum presented fibrous appearance with edemas, with primary teeth and some permanent teeth, all of them with yellow crowns, shorter and with altered shapes (Fig. 5); besides presenting precarious periodontal health. Through panoramic and periapical radiographic exams, it was verified the presence of the other permanent teeth, not erupted (Fig. 6).



**Fig. 4** - Radiografia periapical mostrando maior detalhe.

Except for teeth 4, 5, 13, 16, 22, 30, and 32, the other dental elements presented radiopacity reduction and difficulty in the delimitation of dental crowns, which characterized enamel and dentin hypoplasia. In the periapical radiography, for the region of the left side inferior canine, it was noticed the broad pulp chamber of the primary canine with the open apices persistence (Fig. 7). Another common fin-





Fig.5 – Exame clínico extraoral sem alteração da normalidade



Fig.6 – Ao exame intracanal evidencia-se coloração das coroas amareladas, menores e com as formas alteradas.

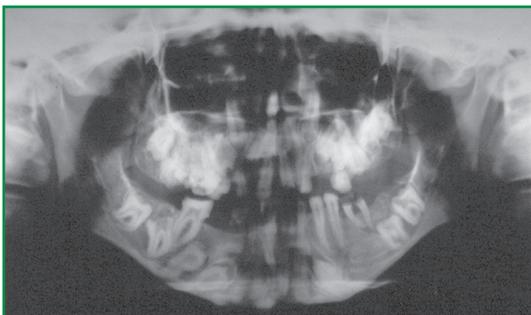


Fig.7 - Radiografia panorâmica apresentando múltiplos dentes não irrompidos e diminuição da radiopacidade dos tecidos mineralizados dos elementos dentários.



Fig.8 – Radiografia periapical mostrando ampla câmara pulpar do canino decíduo com a persistência do ápice aberto.



Fig.9 – Por meio da radiografia periapical nota-se destruição coronária, em decorrência da fragilidade estrutural da coroa, e rarefação óssea periapical difusa assoada ao dente 36.

ding in some teeth was dilaceration. As a result of the structure frailty of the crowns, the teeth are more susceptible to caries, as in tooth 36, which led to pulp compromising and formation of diffuse periapical bone rarefaction (Fig. 8). The clinical aspects associated to the radiographic findings suggested odontodysplasia.

FERREIRA TLD  
PANELLA J  
FREITAS CF  
ODONTODISPLASIA  
- CASO FAMILIAR  
RARO





**Fig.10** – Ao exame clínico extraoral a paciente não apresenta nenhuma alteração digna de nota.



**Fig.11** – Ao exame clínico intraoral observa-se a anatomia curta das coroas dentárias.

The oldest sister (22 years old) was advised to go to the radiology center for an assessment, once her sisters had odontodysplasia. At the extraoral clinical exam, the patient did not present any alterations worthy mentioning. At the intraoral exam, the short anatomy of dental crowns stood out, in addition to the yellow color and the crossbite in almost all teeth, except for the left superior central incise and the right superior primary canine.

Through the panoramic radiography, it was noticed the presence of the four cani-



**Fig.12** – Por meio da radiografia panorâmica constatou-se poucos dentes com a imagem radiográfica compatível com odontodysplasia.

nes, not erupted, of the inferior pre molars (with an increase in the pericoronal flap), of the left side superior pre molar, the second molars on the right side and the four third molars; the only missing tooth was 18. Few teeth presented radiographic image compatible with odontodysplasia, despite the majority presents disfigured dental anatomy (Fig. 9). Among the three sisters, this is the one who presented the less aggressive odontodysplasia.

#### DISCUSSION AND CONCLUSIONS

Odontodysplasia is considered as a rare dental development anomaly, whose etiology is still uncertain and broadly discussed. In the literature studied, many hypotheses have been suggested, but none alluded to heredity; some authors even disregard this assumption, which opposes to the work presented, as the clinical cases herein described correspond to three sisters, suggesting a family compromising. Some authors Gerlach *et al.*<sup>1</sup> (1998), Gomes *et al.*<sup>3</sup> (1997) agree that the female genre is the most affected genre; our clinical cases confirm and increase this statistics, as it deals with three female patients.

In regards to clinical aspects, in the literature studied, the authors report that most teeth affected did not erupt in the oral cavity (Gerlach *et al.*<sup>1</sup>, 1998, Gomes *et al.*<sup>3</sup>, 1997). The same thing occurred in our cases, except for the primary teeth, which, even compromised, erupted in the oral cavity; another characteristic, that has also been confirmed, was the delay in the eruption chronology (Gerlach *et al.*<sup>1</sup>, 1998). The three sisters, being the oldest



22 years old, still have primary teeth in oral cavity. Gum changes have also been ratified.

The radiographic standards found were the same described by most authors Gerlach *et al.*<sup>1</sup> (1998), Gomes *et al.*<sup>3</sup>, (1997), who reported that as a result of enamel and/or dentin hypoplasia, the dental limits, in special the crowns', were poorly seen, providing the aspect of ghost tee-

th (Gerlach *et al.*<sup>1</sup>, 1998, Gomes *et al.*<sup>3</sup>, 1997). The two patients who developed the most aggressive odontodysplasia presented multiple dilacerations, but this root anomaly was not reported in any of the articles studied. The root shortening, the increased volume of pulp chambers and open apices were also some of the characteristics found.

#### REFERENCES

1. Gerlach RF, Jorge J, Jr., de Almeida OP, Coletta RD, Zaia AA. Regional odontodysplasia. Report of two cases. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 1998 Mar;85(3):308-13.
2. Sadeghi EM, Ashrafi MH. Regional odontodysplasia: clinical, pathologic, and therapeutic considerations. *J Am Dent Assoc* 1981 Mar;102(3):336-9.
3. Gomes A, Pinto AG, Valle M, Perez E. Odontodysplasia regional: aspecto clínico, radiográfico e histológico *Rev Odontopediatria* 1997 out.-dez.;5(4):147-53.
4. Ansari G, Reid JS, Fung DE, Creanor SL. Regional odontodysplasia: report of four cases. *Int J Paediatr Dent* 1997 Jun;7(2):107-13.
5. Witkop CJ, Jr. Hereditary defects of dentin. *Dent Clin North Am* 1975 Jan;19(1):25-45.
6. Raez AG. Unilateral regional odontodysplasia with ipsilateral mandibular malformation. *Oral Surg Oral Med Oral Pathol* 1990 Jun;69(6):720-2.
7. Fanibunda KB, Soames JV. Odontodysplasia, gingival manifestations, and accompanying abnormalities. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 1996 Jan;81(1):84-8.
8. Gibson T, Kelsch R, Sokoloff S, Pillar C. Regional odontodysplasia: a review of the literature and a report of 3 cases. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 2003 96(3):294.

Recebido em: 16/08/2011

Aceito em: 12/09/2011

