



UNIVERSIDADE FEDERAL DE UBERLÂNDIA  
FACULDADE DE ODONTOLOGIA



GABRIELA LOURENÇO PERES

**O PAPEL DO ODONTOPEDIATRA NO  
ATENDIMENTO DE PACIENTE COM  
NEUTROPENIA CÍCLICA NA PRIMEIRA  
INFÂNCIA: RELATO DE CASO**

**THE PEDIATRIC DENTIST'S ROLE IN THE  
CARE OF PATIENT WITH CYCLIC  
NEUTROPENIA IN EARLY CHILDHOOD:  
CASE REPORT**

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Trabalho de conclusão de curso  
apresentado a Faculdade de  
Odontologia da UFU, como requisito  
parcial para obtenção do título de  
Graduado em Odontologia

Orientadora: Prof<sup>ª</sup>. Dr<sup>ª</sup>. Alessandra  
Maia de Castro Prado

UBERLÂNDIA

2017



SERVIÇO PÚBLICO FEDERAL  
MINISTÉRIO DA EDUCAÇÃO  
UNIVERSIDADE FEDERAL DE UBERLÂNDIA  
GRADUAÇÃO EM ODONTOLOGIA  
TRABALHO DE CONCLUSÃO DE CURSO

ATA DA COMISSÃO JULGADORA DA DEFESA DE TRABALHO DE CONCLUSÃO DE CURSO DO (A) DISCENTE **Gabriela Lourenço Peres** DA FACULDADE DE ODONTOLOGIA DA UNIVERSIDADE FEDERAL DE UBERLÂNDIA.

No dia quatro de julho de 2017, reuniu-se a Comissão Julgadora aprovada pelo Colegiado de Graduação da Faculdade de Odontologia da Universidade Federal de Uberlândia, para o julgamento do Trabalho de Conclusão de Curso apresentado pelo (a) aluno (a) **Gabriela Lourenço Peres**, COM O TÍTULO: "THE PEDIATRIC DENTIST'S ROLE IN THE CARE OF PATIENT WITH CYCLIC NEUTROPENIA IN EARLY CHILDHOOD: CASE REPORT". O julgamento do trabalho foi realizado em sessão pública compreendendo a exposição, seguida de arguição pelos examinadores. Encerrada a arguição, cada examinador, em sessão secreta, exarou o seu parecer. A Comissão Julgadora, após análise do Trabalho, verificou que o mesmo encontra-se em condições de ser incorporado ao banco de Trabalhos de Conclusão de Curso desta Faculdade. O competente diploma será expedido após cumprimento dos demais requisitos, conforme as normas da Graduação, legislação e regulamentação da UFU. Nada mais havendo a tratar foram encerrados os trabalhos e lavrada a presente ata, que após lida e achada conforme, foi assinada pela Banca Examinadora.

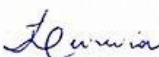
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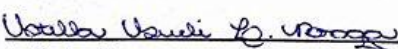
  
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THE PEDIATRIC DENTIST'S ROLE IN THE CARE OF PATIENT WITH CYCLIC  
NEUTROPENIA IN EARLY CHILDHOOD: CASE REPORT

NEUTROPENIA IN EARLY CHILDHOOD

**Keywords:**Preschool, neutropenia, etiology, diagnosis, dental  
treatment

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## **ABSTRACT**

Neutropenia is characterized by a deficiency or reduction in the number of mature neutrophils circulating in the blood and increased susceptibility of the host to acquire infections. The aim of this article is to report the case of a child diagnosed with cyclic neutropenia of autosomal dominant type caused by mutation of the gene ELANE and emphasize the importance of pediatric dentistry in dental care for the oral health, since the diagnosis, prevention and maintenance of oral health.

## **INTRODUCTION**

The term neutropenia is used to describe a group of diseases that presents a deficiency or decrease in the number of mature neutrophils circulating in the blood, therefore increasing the susceptibility of the host to acquire infections.<sup>1</sup> Neutrophils are white blood cells, also known as differentiated leukocytes which form the first line of defense of the organism,<sup>2,3</sup> making the most abundant population of leukocytes in the blood and participating in the first stages of the inflammatory response in the body.<sup>4</sup>

In order to undergo the process of maturation and reach the bloodstream, neutrophils require nine to eleven days in physiological conditions.<sup>4,5</sup> After the maturation process, they start to circulate in the blood for a period of approximately

six to ten hours, and survive for up to 48 hours in inflamed tissues. Within that period, if they are not recruited to an inflamed site, they suffer apoptosis and phagocytosis by macrophages. During the inflammatory response of tissues, when they are recruited, neutrophils cluster on the surface of endothelial cells and reach the area of the inflamed tissue, causing the destruction of pathogenic agents.<sup>6</sup>

Clinically, an individual affected by neutropenia may have recurrent infections, which are often fatal. The frequency and severity of infections depend on the count and speed of decline in the number of neutrophils, abnormalities of the phagocytic function, the deficits of adaptive immune function, host conditions and specific germ.<sup>7</sup>

The neutropenia can be acquired or congenital (table 1) and regarding the severity, may be classified as mild (1,000 to 1,500 neutrophils/mm<sup>3</sup>), moderate (500-1,000 neutrophils/mm<sup>3</sup>) or severe (< 500 neutrophils/mm<sup>3</sup>).<sup>4</sup>

The cyclic neutropenia was first described by Leale in 1910, as autosomal dominant, a result from the mutation of the gene ELANE of the character position 13.3 of the short arm of chromosome 19 that codes the neutrophil elastase.<sup>9</sup> Its prevalence is of 1-9 per 1,000,000.<sup>10</sup> The disease is characterized by periods of neutropenia (< 1,000 neutrophils/mm<sup>3</sup>) with a duration of three to ten days that are repeated at intervals of 21 days,<sup>11</sup> and may vary from 14-36 days. During neutropenia it can be observed fever, malaise,

periodontal disease, ulceration of the oral mucosa, impetigo and lymphadenopathy, common symptoms in children and adolescents. Whereas adults have mild-to-moderate neutropenia without well-defined cycles. Some complications such as cellulitis, necrotizing enterocolitis and bacteremia are severe and can be fatal.<sup>12</sup>

Oral manifestations, such as ulcers, periodontal disease, tooth mobility and early tooth loss are common signs in patients with congenital neutropenia.<sup>13</sup> The diagnosis of congenital neutropenia is suspected in children with recurrent oral infection and persistent bacterial or fungal infection.<sup>14</sup>

For the diagnosis of cyclic neutropenia, it is necessary to perform complete blood count (twice a week for six weeks) and bone marrow examination.<sup>14</sup>

In the 1980s, the granulocyte colony-stimulating factor (rHUG-CSF) was introduced in the treatment of neutropenic patients, increasing the number of circulating neutrophils and stimulating the proliferation, differentiation and maturation of precursor cells of neutrophils, reducing the frequency and severity of infections, with a prognosis resulting in a better quality of life for the treatment of patients with neutropenia.<sup>15,16</sup>

Another form of treatment is transplantation of hematopoietic system cells. In addition to these treatments, the prophylaxis, which consists of the use of broad spectrum antibiotics, has been recommended.<sup>17</sup> However, many patients

still suffer due to the disease disorders.<sup>18</sup> The objective of this work is to report the case of a child with cyclic neutropenia, addressing etiology, diagnosis, clinical aspects, oral manifestations and treatment, as well as dental care in the treatment of these patients.

## **CASE REPORT**

Patient, female, caucasian, attended the Clinic for babies from Pediatric Dentistry from the School of Dentistry of Federal University of Uberlândia, accompanied by her mother, who consented to have the case was described in this report. At the first child's dental care service, she was 13 months old. In the anamnesis, the evaluation of the medical history revealed that there were no complications during pregnancy, it was cesarean, the child was born at term, with 5,76 lbs and 46 cm, without any change originating from complications during the labor work. Up to 12 months, the child had never been hospitalized, but had already presented respiratory problems (bronchiolitis) at three months and frequent cases of tonsillitis, with use of antibiotics. Regarding the food habits, the child was breastfed just in the first month, used with some frequency pasty feed and tooth brushing was performed by parents on average twice a day, with some difficulty, due to the child's behavior.

During extra-oral clinical examination, no alterations were found and the intra-oral revealed eruption of deciduous teeth that were compatible with the age of the child (upper and lower erupted incisors), ankyloglossia and absence of caries lesions. In the follow-up, after four months, the mother reported as main complaint bleeding during brushing. During the clinical examination, it was noted gingival edema, inflamed marginal gingiva, presence of dental visible biofilm, calculus on the proximal faces of the lower anterior teeth (Figure 1), and presence of ulceration on the oral mucosa, suggesting the presence of canker sore (Figure 2).

Professional prophylaxis with Robinson toothbrush and toothpaste was performed and the child was scheduled to return monthly for dental biofilm control and gingivitis (Figure 3). For the removal of calculus, found in the anterior teeth proximal, exploratory probe was used and strip of sandpaper for resin finishing between the teeth to complete the calculus removal. However, in subsequent follow-ups there was no improvement in gingival bleeding and even after new sessions of prophylaxis, the gum was still reddish in color, with the presence of gingival bleeding during brushing even with a reduction of the biofilm.

Thus, before the clinical conditions, medical opinion was requested and the child was referred for examination at the medical clinic, at 24 months old. According to medical report, the child had neutropenia in laboratory examinations and

during investigation was observed severe neutropenia (below 500) at certain times. In addition, usually the child is affected by recurrent infections characterized by tonsillitis, boils in the genital region and gingivostomatitis, with frequent use of antibiotics (approximately twice or more per month). Due to the fact aforementioned, prophylactic antibiotic of wide spectrum was started in an attempt to maintain the child free from infectious processes, but, even so, she showed persistent infection signs. Other immunological parameters were within normal levels for her age. In this case for the diagnosis of the type of neutropenia, complete blood counts were performed (twice a week, on Tuesdays and Fridays for six weeks) and bone marrow examination that allowed to define the diagnosis of cyclic neutropenia at three years of age.

Thus, for the dental treatment, the child is being accompanied at the pediatric dentistry clinic once a month for mechanical control of dental biofilm, and now with evident improvement of the gingival condition and continues without any tooth decay.

## **DISCUSSION**

The dental surgeons play an important role in early detection of diseases in the oral cavity, due to initial signs that can be observed in the clinical examination.<sup>19</sup>

During the early childhood, it is important that the pediatric dentistry learns to diagnose and treat oral manifestations whenever necessary, because the infants exhibit unique and transitory anatomical structures typical of this phase. It should be emphasized that children with systemic changes are 17.4 times more likely to have oral manifestations compared to those without systemic changes.<sup>20</sup> And although changes in the mucosa are relatively common in children, oral severe disorders are associated with medical conditions.<sup>21</sup> So in the case described, the persistence of the gingivitis, even with professional control of biofilm was the aspect that attracted the attention to investigate the child's systemic conditions. In another case report 6-year-old child, changes in the oral cavity as recurrent oral ulcers, gingival recession, gingivitis, tooth mobility were the signs that led to the diagnosis of cyclic neutropenia.<sup>22</sup>

Periodontal disease is an inflammatory process of infectious origin which affects the gingival tissues and/or the tissue supporting the teeth,<sup>23</sup> occurring as a result of inflammatory reactions and immune responses in periodontal tissues induced by micro-organisms of the biofilm, damaging the connective tissue and alveolar bone.<sup>24,25</sup> The deficiency of host response in individuals with congenital neutropenia can obstruct the interaction between the host and the microbial flora. Thus, the deficiency of neutrophils increases the risk of infections, as well as periodontal disease.<sup>26</sup> Although the

periodontal disease appears as a response of the gums to the formation of biofilm and calculus, it is also recognized that this response can be modified by systemic factors. The persistent gingivitis exhibited by the child in this case can be attributed to the reduction of neutrophils, because even with the frequent sessions of professional prophylaxis improvement was not found in the bleeding and redness of the gum tissue of the patient.<sup>27</sup>

The periodontal manifestations found are related to the degree of neutropenia. Mild cases may lead to the appearance of changes in gingival tissues, with the appearance of ulcerative gingivitis, characterized by the presence of redness, bleeding, ulcers and pain.<sup>28</sup> In severe cases, it can affect the tissues supporting the teeth, bone loss and bag formation, characterized as an aggressive periodontitis.<sup>29</sup>

The main objective of the periodontal treatment for patients with neutropenia, must be the prevention through control of biofilm and odontological follow-up.<sup>30</sup> Therefore, the fact that the mother has sought dental care to thirteen months of age and participate actively in the child's oral hygiene, was fundamental so that the gingivitis did not worsen. And after the systemic treatment, it was also possible to observe the improvement in the gingivitis condition' simultaneously. Unlike the report presented of a child diagnosed at 6 years old, in which despite the systemic treatment used, the irregularity of periodontal conditions

maintenance, led to the persistence of gingival inflammation and deep periodontal pockets in the permanent dentition.<sup>22</sup>

Thus, periodic evaluations with the dental surgeon, along with a patient motivated to perform a good oral hygiene, are the best way to prevent diseases of the oral cavity.<sup>31</sup> In addition, regular visits to the dental office for monitoring and control of dental biofilm, contribute to improving both the oral health, as the patient's systemic condition.<sup>30,32</sup>

Some preventive strategies and treatment were developed for neutropenic patients who suffer with some changes in the oral cavity and according to Olvera et al. (2015), consultations with a pediatrician for checking the current status of the patient and recent counts of defense cells to check the number, variety, concentration and quality of blood cells should be performed, because it is important to know the normal values of groups of cells to know when they can make an oral rehabilitation with safety and without complications. And to avoid complications, according to Olvera et al. (2015) it is important to follow a prophylactic scheme with antibacterial basis, amoxicillin 50mg/kg or clindamycin 100mg/kg in the case of allergy, to avoid infectious processes, using it as a therapeutic regimen (every eight hours for seven days) when the consultations are consecutively or as a prophylactic scheme (one hour before beginning the dental treatment, in a single dose) when treating emergency or isolated consultations. Check the total number of neutrophils before

and after the treatment is a good alternative to monitor the condition of the patient's neutropenia.

Therefore, preventive strategies for control of biofilm by brushing after every meal, implement the use of dental floss, adequate nutrition, regular consultations with the pediatrician and follow-up by means of exams for knowledge of the count of neutrophils in the blood, applying the stimulating factor of colonies, mouth wash with chlorhexidine twice per day, systemic antibiotics and regular consultations with the surgeon-dentist to better oral hygiene, are important measures in order to improve the oral condition of these patients.<sup>9</sup>

In the case of the patient, who at the time of the first dental examination was 13 months old, the mouth wash with chlorhexidine was not recommended, and in subsequent consultations when there was an improvement in the gingivitis conditions, the application of chlorhexidine was performed with sterile gauze on the tooth surfaces and gingival region by the dental surgeon on the day of the dental care. Although the susceptibility of an individual to periodontal disease is influenced by many factors, such as systemic and genetic diseases, the evidence indicates that prevention of gingival inflammation by controlling the dental biofilm reduces the severity of such conditions in this group of patients.<sup>27</sup>

The majority of patients with cyclic neutropenia during the outbreaks of neutropenia and associated infections are

treated with antibiotics or immunoglobulin for intravenous administration (IVIG). In severe cases, the treatment consists of the use of Granulocyte colony-stimulating factor (rHUG-CSF) in symptomatic patients,<sup>12</sup> increasing the neutrophil count for more than 1,000 neutrophils/mm<sup>3</sup>. Although most patients do not require rHUG-CSF, in serious cases of non-responsive Cyclic neutropenia to therapy with rHUG-CSF the bone marrow transplant is indicated.<sup>33</sup> In the case of the patient, the option of treatment proposed by physicians was the injectable medication (Filgrastim 300mcg three times a week), which is a Granulocyte colony-stimulating factor, with the purpose of inducing the production of neutrophils, thereby improving the children' life quality and the morbimortality. In a case of an 8-year-old patient, also treated with Filgrastim, the authors reported that there was an improvement in the child's overall health, but the damage caused to the periodontal tissues could not be re-established, however, the progression of periodontal disease was interrupted, and it was possible the oral health maintenance by means of periodic follow-ups and use of chlorhexidine 0.2% in episodes of neutropenia.<sup>34</sup>

According to the medical guidance, the child will make use of the medication until 10 years old, because during this period of puberty there may be a natural increase in the numbers of neutrophils, no longer being necessary the use of medication. Therefore, there will be a follow-up to determine

the interruption of medication or a new treatment strategy, if the increase in neutrophils do not happen as expected.

Thus, the dental surgeons who diagnose gingival inflammation and periodontal disease in children apparently healthy should direct these patients to a pediatric clinic for evaluation.

The odontological prevention and follow-up of neutropenia patients should be emphasized, it is important to carry out mechanical control of dental biofilm, removal of calculus, prevention of dental caries, orientation of oral hygiene in addition to adequate nutrition. Parents should be encouraged to actively participate in these preventive activities.

## **CONCLUSION**

It was possible to confirm the importance of the role of the pediatric dentist since the diagnosis, prevention and maintenance of oral health in patients with cyclic neutropenia diagnosed during childhood.

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**TABLE**Table 1: Classification of neutropenias.<sup>8</sup>

| Congenital Neutropenias                                                            | Acquired Neutropenias                                                     |
|------------------------------------------------------------------------------------|---------------------------------------------------------------------------|
| Isolated congenital Neutropenias                                                   | Neutropenia as isolated or dominant citopenia                             |
| Syndrome of Kostmann and congenital severe neutropenia                             | Infections: viruses, bacteria, fungi, rickettsia and protozoal infections |
| Cyclic neutropenia                                                                 | Drugs                                                                     |
| Benign family Neutropenia                                                          | Immune                                                                    |
| Neutropenia in the context of syndromes of congenital medullary insufficiency      | Infancy primary autoimmune Neutropenia                                    |
| (Syndrome of Shwachman - Diamond, Fanconi's anemia, congenital dyskeratosis, etc.) | Autoimmune secondary neutropenia                                          |
| Congenital Neutropenia as part of multisystem disorder                             | Nutrition: anorexia nervosa, copper deficiency                            |
| Barth's syndrome                                                                   | Idiopathic chronic Neutropenia                                            |
| Pearson's syndrome                                                                 | Neutropenia associated with                                               |

|                                                                      |                                                                             |
|----------------------------------------------------------------------|-----------------------------------------------------------------------------|
|                                                                      | complement activation                                                       |
| Glycogenosis type Ib                                                 | Neutropenia as part of pancytopenia                                         |
| Congenital Neutropenia in association with adaptive immunity defects | Acquired bone marrow failure (Aplastic anemia, medullar infiltration, etc.) |
| Severe combined immunodeficiency (SCID) and reticular dysgenesis     | Nutritional deficiency (deficit of vitamin B12 and folic acid)              |
| Hypogammaglobinemia associated with X                                | Splenic sequestration                                                       |
| Syndrome of Hyper-IgM                                                |                                                                             |
| Syndrome of WHIM and mielocataxe                                     |                                                                             |

**FIGURE 1****FIGURE 2**

**FIGURE 3**

**FIGURE LEGENDS**

**Figure 1.** Intraoral clinical examination showing inflamed marginal gingiva and presence of dental calculus on the proximal faces of the lower anterior teeth.

**Figure 2.** Intraoral clinical examinationshowing the presence of ulceration on the oral mucosa.

**Figure 3.** Professional prophylaxis.

## **APPENDIX**

### **SPECIAL CARE IN DENTISTRY**

#### **Author Guidelines**

The mission of Special Care in Dentistry (SCD) is to provide a forum for research findings, case reports, clinical techniques and scholarly discussion relevant to the oral health and oral health care of "special care patients." The designation of the special care patient is not limited to hospitalized, disabled or older individuals, but includes all patients for whom oral health and oral health care are complicated by physical, emotional, financial and/or access factors.

Effective with the 2015 volume, this journal will be published in an online-only format.

#### **Manuscripts**

Original articles are considered and accepted for publication on the condition that they have not been published or are not simultaneously submitted for publication elsewhere. A letter signed by all authors stating that the submission is an original article, not previously published or simultaneously being considered for publication elsewhere, must accompany the submission.

All manuscripts should be submitted through the online submission system at <http://mc.manuscriptcentral.com/scid>.

The manuscript should be submitted with all material doublespaced, flush left (preferably in Courier typeface),

with at least a 1" margin all around. All pages should be systematically numbered. The editor reserves the right to edit manuscripts to fit available space and to ensure conciseness, clarity and stylistic consistency.

### **Title page**

Titles of articles should be descriptive but concise. Long titles discourage reading, present typographic and layout problems and create difficulties in indexing.

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