



## **Pediatric Dentistry and Oral Health Features of Children with Certain Genetic Diseases**

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### **Authors' contributions**

*This work was carried out in collaboration among all authors. Authors UAK and ADP designed the study, performed the statistical analysis, wrote the protocol and wrote the first draft of the manuscript.*

*Authors YD and DRP managed the analyses of the study. Authors SBB and VVG managed the literature searches. All authors read and approved the final manuscript.*

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### **ABSTRACT**

Pediatric dentistry provides primary and comprehensive preventive and therapeutic dental care for infants and children in adolescence, as well as provides special medical needs. This specialty includes many skills, disciplines, procedures, and techniques that share a common origin with other dental specialties, but they have been modified and reformed to meet the special requirements of infants, children, adolescents, and special medical needs. However, pediatric dentistry is mainly

focused on healthy children, while quite often there are cases when such assistance is required for children with congenital genetic diseases. The prevalence of rare diseases is approximately 65 cases per 100,000 people. This estimate reflects the existence of 7,000 different types of rare diseases, most of which are genetic in nature. Rare diseases of genetic origin are often associated with other comorbidities, which are often characterized by debilitating and chronically degenerative conditions associated with phenotypes. Patients with rare diseases may sometimes exhibit challenging behaviors and often experience physical, mental, and sensory impairments that can affect functioning and participation in daily life. The purpose of the work is to consider the features of pediatric dentistry and the features of oral health of children with certain genetic diseases.

*Keywords: Pediatric dentistry; oral cavity condition; children; congenital genetic diseases; developmental features; osteogenesis imperfect; mucopolysaccharidosis; restorative therapy.*

## 1. INTRODUCTION

Oral health is a fundamental part of the overall health and well-being of every infant, child, and adolescent, as such diseases affect function, development, and quality of life. Children's oral care should be safe, consistent, diverse, easily accessible, cost-effective, of high quality, and respectful to each child and their family. This aid prevents and eliminates diseases, pain and infections; restores the function and shape of the dentition and corrects obvious facial dysfunction or deformity [1].

The practice of dentistry is undergoing a new paradigm shift with the advent and use of new technologies. New imaging devices, restorative procedures, the Internet and powerful electronic devices, laser dentistry, and new materials are examples of advances affecting dentistry [2,3,4,5,6]. Although there are not many new methods of treatment in pediatric dentistry, however, their practice has improved significantly in recent years.

It is necessary to consider current practices and advances in paediatric dentistry, based on evidence, and directs the provision of primary prevention, early intervention, and reparative care for primary and permanent malocclusion.

In addition to healthy children, children with congenital genetic diseases, such as mucopolysaccharidosis (MPS) and osteogenesis imperfecta (NO), also need dental care [7]. These are two rare genetic diseases. MPS is a group of inherited metabolic diseases caused by the absence or disruption of certain specific lysosomal enzymes in the body that are necessary for the breakdown of glycosaminoglycans. The absence or improper functioning of these enzymes means that over

time, glycosaminoglycans (GAGS) accumulate in cells, blood, brain and spinal cord tissues. The accumulation of GAG can lead to severe morbidity and shortened life expectancy. While the symptoms of MPS may be similar, they can also vary from person to person, making early diagnosis difficult. The overall incidence of MS is estimated to be between 1:20,000 and 1:25,000. MPS can affect appearance, skeletal development, physical abilities, the functioning of organs and systems, and, in most cases, cognitive development.

Osteogenesis imperfecta is a genetic disorder that is present from birth. It is characterized by a reduction in connective tissue, which may be of poor quality or absent altogether. The reduction or absence of connective tissue can cause hypermobility of the joints, brittle bones that can break without cause, and the growth of the jaws and teeth can be disrupted. Imperfect dentinogenesis (DI) is often associated with OI and leads to dentin dysplasia. According to recent estimates, OI affects one in 10,000 to 20,000 children. OI can be divided into four types according to the phenotype and genotype characteristics: type I (mild), type II (lethal), type III (severe), and type IV (moderate) [8].

People with MPS or NO usually have a poor oral condition. Genetic changes associated with MPS and OI also affect the stomatognathic system. People with these specific genetic disorders usually often suffer from malocclusion, microdontia, tooth rotation, and tooth agenesis. Enamel development defects are common in people with MPS, whereas DI is common in people with NO. Motor coordination limitations often accompany these clinical conditions, making it difficult to perform oral hygiene procedures such as brushing teeth, while limited access to oral hygiene education, dental

services, and barriers to knowledge for health professionals may contribute to greater exposure to dental caries in patients with MPS and OI [9].

The purpose of the work is to consider the features of pediatric dentistry and the features of oral health of children with certain genetic diseases.

## 2. MATERIALS AND METHODS

In the process of writing the study, the literature was studied, which dealt with the peculiarities of performing dental manipulations for young patients in general and children with disabilities in particular. In this regard, various innovative approaches to the organization of dental treatment of children were studied and the peculiarities of applying various dental manipulations to children suffering from chronic somatic diseases were noted. The leading research method was the analytical method, as this tool was used to review the sources in the study.

## 3. RESULTS

Children's dentistry today is a complete system of dental care for young patients. It takes into account the age of the patient, his development and psychosocial status in order to strengthen health, prevent oral diseases.

The dentist who provides dental care examines, diagnoses oral conditions and makes a treatment plan, including preventive services, and all services are performed under the supervision of the dentist together with other dental staff. Collaboration with other health care providers, such as pediatricians, pediatric nurses, family doctors, who most often see children in the first years of life, improves the health of the child's oral cavity.

Prevention plays an important role in pediatric dentistry. Professional prevention is useful for familiarizing a young child with dental procedures, facilitating the examination of hard and soft tissues, teaching parents and a child or teenager proper methods of oral hygiene and the removal of plaque, tartar [10].

Also in modern pediatric dentistry, internal or external whitening of individual teeth of milk and young permanent teeth is used, which is indicated for discoloration caused by trauma (darkening with devitalization, calcifying

metamorphosis), color anomalies of the enamel of the permanent tooth due to trauma or infection of the milk tooth, or internal color change.

One of the main roles in pediatric dentistry belongs to the control of pain. In children, pain is difficult to measure because of its subjectivity. Children experience pain and show variability in the expression of pain due to differences in their cognitive and emotional development, health status. To assess pain in children with special needs, observations such as vocalization, facial expressions, and body movements are necessary.

Pain in pediatric dentistry can be effectively controlled and relieved with both non-drug and pharmacological pain management strategies prior to surgery. Non-drug behavioral therapy includes guided imagery, distraction, play therapy, and behavioral demonstration techniques. Pharmacological therapy may consist of a hierarchical structure in terms of adequate local anesthesia, moderate sedation, or deep sedation regimens [11].

Inadequate pain management can have significant physical and psychological consequences, so it is extremely important to recognize and assess pain and record it in the patient's chart in order to determine the clinical diagnosis, plan treatment, and determine the patient's need for analgesics. Familiarity with the medical history helps to avoid prescribing contraindicated drugs and to understand the consequences, morbidity and toxicity associated with the use of specific therapeutic agents [12].

The choice of an analgesic depends on the individual patient, the degree of treatment, the duration of the procedure, psychological factors, the patient's medical history, and physiological factors such as blood clotting disorders, liver or kidney problems.

Analgesics should initially be prescribed on a regular schedule if moderate or severe pain is considered likely within the first 36-48 hours, rather than as needed to create stable plasma levels of analgesics and reduce the likelihood of sudden pain. It is necessary to consider the use of non-opioid analgesics, for example, non-steroidal anti-inflammatory drugs (NSAIDs), acetaminophen as first-line agents for the treatment of acute mild and postoperative pain. The combination of NSAIDs with acetaminophen provides a greater analgesic effect than one

agent alone. Opioid analgesics in combination with NSAIDs can be used to treat moderate to severe postoperative pain in children and adolescents [13].

In the modern period, various restorative techniques are also used in pediatric dentistry. There are many options for restoration treatment for both milk and permanent teeth. These include dental amalgams, glass ionomers, both resin-modified and conventional, alternative restoration methods (ART), temporary therapeutic restorations ( ITR), resin infiltration, dental composites, ionomer sealants, resin-based composites, dental composites, stainless steel crowns, primary molars, finished metal crowns, strip crowns, pre-lined crowns, and aesthetic restorations [14].

Modern treatment of dental caries involves identifying the individual risk of developing caries, understanding the disease process, and active surveillance to assess the progression of the disease and treatment with appropriate preventive services, complemented by the most appropriate restorative therapy as indicated. Decisions about when to repair carious lesions include clinical criteria for visual detection of enamel cavitation , visual detection of enamel shading, or radiographic recognition of increased lesions over time.

The benefits of restorative therapy include the removal of cavitation or defects to eliminate areas prone to tooth decay; stopping the progression of tooth demineralization; restoring the integrity of the tooth structure; preventing the spread of infection to the tooth pulp; preventing tooth displacement and loss of space due to loss of tooth structure.

The risks of restorative therapy include reducing the longevity of the teeth, making them more susceptible to fractures, recurrent damage, restoration failures, pulp exposure during caries removal, future pulp complications, and iatrogenic damage to adjacent teeth.

Baby teeth may be more susceptible to restoration failures than permanent teeth. In addition, before restoring baby teeth, it is necessary to take into account the time of exfoliation of baby teeth.

Step-by-step caries removal is a two-step caries removal process in which the carious dentin is partially removed at the first visit, leaving the

caries on the pulp with the installation of a temporary filling. At the second appointment, all remaining carious dentin is removed and the final restoration is established. With partial or one-stage caries removal, part of the carious dentin is removed, but the caries remains on the pulp, and then the foundation and final restoration are established.

Resin infiltration is an innovative approach designed primarily to stop the progression of non-cavitated interdental caries. The purpose of the resin infiltration method is to ensure the penetration of low-viscosity resin into the porous body of the lesion in enamel caries. An additional use of resin infiltration has been proposed to restore white spots formed during orthodontic treatment.

Dental amalgam has been the most commonly used material for the restoration of lateral teeth for over 150 years and is still widely used worldwide. The amalgam contains a mixture of metals such as silver, copper, and tin, in addition to about 50% mercury. The use of dental amalgam has declined over the past decade, possibly due to controversy over the perceived health effects of mercury vapor, environmental concerns over its mercury content, and increased demand for aesthetic alternatives. There is strong evidence that dental amalgam is effective in the restoration of cavities of classes I and II of milk and permanent teeth [15].

Composites consist of a polymer matrix and chemically bonded fillers. They are classified according to the size of the filler, as the size of the filler affects the polishability and aesthetics, the depth of polymerization, the shrinkage during polymerization, and the physical properties. Hybrid resins combine a mixture of particle sizes to increase strength while maintaining aesthetics.

The smaller size of the filler particles provides greater polishability and aesthetics, while the larger size provides durability. Fluid resins have a lower volume percentage of filler than hybrid resins. Bonding for enamel and dentin is able to reduce edge staining and detectable fields for various types of composites. In the case of milk molars, strong evidence has been obtained in randomized controlled trials that composite polymers are successful when used for Class I restorations and Class II lesions on milk teeth.

Resin-modified glass ionomer cements (RMGIC) with acid-base polymerization, supplemented by

light-curing polymerization of the second resin, are effective for baby teeth.

Pre-made metal crowns (also known as stainless steel crowns) are pre-made metal crowns that adapt to individual teeth and cement with a biocompatible fixing agent. They are indicated for the restoration of milk and permanent teeth with extensive caries, when other available restoration materials may be ineffective (for example, interdental caries that goes beyond the right angles, bruxism, etc.). Restoration of the front teeth is a difficult task due to: the small size of the teeth; the immediate proximity of the pulp to the surface of the tooth; relatively thin enamel; no surface for communication and behavior of the child. Treatment options for milk front teeth include composites for Class III and V restorations and permanent dentition, as well as pre-lined stainless steel crowns, pre-molded stainless steel crowns, and open stainless steel crowns for full-coverage restorations.

Also, the children's dentist must know the techniques of anesthesia of the patient. Pain relief is the reduction or elimination of pain and anxiety in a conscious patient. The patient responds to verbal commands, all vital signs are stable, without significant risk of loss of protective reflexes and with a safe return to preoperative mobility. Pain relief can speed up the execution of procedures that do not cause much discomfort, but require the patient to minimize movement.

For the purpose of anesthesia in pediatric dentistry, nitrous oxide inhalation is used. This method is a predictable pharmacological approach to reduce or relieve anxiety, discomfort or pain, it improves communication with the patient and cooperation, increases the pain threshold, increases the tolerance of long visits, reduces vomiting, increases sedation and helps in the treatment of special needs children. This method also has a number of contraindications: COPD, emotional disorders, coblamine deficiency, etc.

During treatment, it is important to continue visual monitoring of the patient's respiratory rate and level of consciousness. The effects of nitrous oxide depend largely on psychological confidence. Therefore, it is important to continue the traditional methods of behavior management during treatment. After stopping the nitrous oxide supply, 100% oxygen must be supplied within five minutes. Before leaving the dental office, the

patient should return to the reactivity that was before the treatment. Nitrous oxide supply equipment must be checked for proper gas supply [16].

Important attention should be paid to the features of prevention with antibiotics.

After invasive dental procedures, the appearance of bacteria in the bloodstream is possible. Thus, infectious endocarditis is a rare, but life-threatening complication that occurs as a result of bacteremia. Many cases of bacteremia can be the result of everyday activities, such as brushing your teeth, using dental floss, and chewing. However, prevention with antibiotics is recommended for certain dental procedures.

An effective antibiotic treatment regimen should be directed against the most likely infecting organism, with antibiotics prescribed shortly before the procedure. Antibiotics are recommended for high-risk cardiac patients (in the presence of a heart transplant or valve transplant, with congenital heart disease, weakened immunity, autoimmune diseases, diabetes, etc. [17].

#### 4. DISCUSSION

Innovative methods of pediatric dentistry are especially relevant when providing dental care to children with rare genetic diseases.

Children and adolescents with rare genetic diseases that affect the development of the skeleton, according to studies, have increased levels of caries, dental anomalies and malocclusion compared to children without rare genetic diseases. An analysis of the literature has shown that rare genetic diseases that affect the development of the skeleton are directly related to caries, dental anomalies and malocclusion. This finding highlights the differences in oral health associated with rare genetic diseases affecting skeletal development among children and adolescents [18].

Children and adolescents with rare genetic diseases that affect skeletal development also showed poor oral hygiene, poor dental attendance, and shorter duration of breastfeeding than children without rare genetic diseases.

A study of different approaches to dental care and oral hygiene in children suffering from rare

genetic diseases that affect the development of the skeleton, as well as an analysis of the number of cases of dental caries in such children, revealed possible ways in which rare genetic diseases that affect the development of the skeleton can affect dental caries. In addition, it was noted that the number of dental visits had an indirect effect on the relationship between rare genetic diseases that affect the development of the skeleton and malocclusion.

The authors believe that rare genetic diseases that affect the development of the skeleton can affect the health of the oral cavity in children and adolescents. In particular, there is a high tendency of children with rare diseases to caries, dental anomalies and malocclusion [19].

Defects in the development of enamel and DI in patients with OI are the result of genetic mutations involved in the signaling pathway of tooth tissue formation. In addition, malformations of enamel and DI are associated with an increase in dental caries. A higher prevalence of malocclusion in patients with MPS occurs as a result of damage to the upper respiratory tract, macroglossia, and the life-giving palate [20]. These characteristics predispose to anterior open bite and cross bite, which are the most common signs of malocclusion. Patients with BUT experience a growth deficit at the base of the skull, which limits the growth and displacement of the upper jaw in the anterior-lower direction. Thus, the prevalence of posterior crossbite and class II malocclusion is high among individuals with OI [21].

Research shows that parents and caregivers face challenges in oral care, and there are additional barriers to accessing adequate dental care for children with disabilities. This means that caregivers usually seek dental care for their children in an emergency, rather than for preventative reasons. Other barriers to access are the lack of dentists with appropriate training to treat children with disabilities, as well as competing priorities for comorbidities and hospitalizations that affect oral health and care [22].

## 5. CONCLUSIONS

The peculiarity of pediatric dentistry is that, given the changing scope of practice, the doctor was always aware of current trends, based on evidence, collaborated with other disciplines and provided high-quality dental care adapted to the

specific needs of each child. Applying innovative methods of dental treatment to healthy children, the dentist should pay special attention to children with rare genetic diseases, while prevention should play an important role here.

Therefore, there is a need to develop public health policies to improve oral health in children and adolescents with rare genetic diseases that affect skeletal development. It is necessary to take into account that such children are in urgent need of preventive dental measures, as often an appeal to the dentist is carried out when the child needs treatment. Parents need education and support regarding their child's oral health to ensure that non-communicable diseases such as dental caries do not become the norm for these groups of children and adolescents.

## CONSENT

As per international standard, parental written consent has been collected and preserved by the author(s).

## ETHICAL APPROVAL

As per international standard or university standard written ethical approval has been collected and preserved by the author(s).

## COMPETING INTERESTS

Authors have declared that no competing interests exist.

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