Amelogenesis imperfecta in a child with cerebral palsy

Simone Helena Ferreira
Camilla de Moraes Pasini
Priscila Humbert Rodrigues
Moisés Zacarias Cardoso
Tássia Silvana Borges

ABSTRACT

Amelogenesis Imperfecta (AI) is an inherited alteration that affects the enamel of primary and permanent teeth, with no systemic manifestations. Cerebral Palsy (CP) is a congenital condition that affects the central nervous system before the age of two, influencing the performance of activities usually conducted by children with normal development. The objective of this study was to describe the dental treatment performed in a 10-year-old child with both amelogenesis imperfecta and cerebral palsy. The family of the child sought the office of the extension project "Gaining Health: Dental Care for Patients with Disabilities" of the Lutheran University of Brazil, located in the city of Canoas, in search of aesthetic rehabilitation. Upon physical and radiographic examination, the AI diagnosis was confirmed. The use of physical restraint with the consent of the parents was necessary to perform dental care. The treatment began with orientations on oral hygiene and eating habits, in addition to supragingival scaling. Next, it was decided to cover the posterior teeth with glass ionomer and to reconstruct the anterior teeth with resin-based composite, using acetate crowns. In the follow-up of the case, an improvement in oral health and aesthetics of the patient was observed. Moreover, this report demonstrates that the complex dental care of disabled patients can be performed in an outpatient environment.

Keywords: Cerebral Palsy; Amelogenesis Imperfecta; Dental Care.

Amelogenese imperfecta em uma criança com paralisia cerebral

RESUMO

A Amelogênese Imperfeita (AI) é uma alteração hereditária que afeta o esmalte dentário dos dentes decíduos e permanentes, com ausência de manifestações sistêmicas. A Paralisia

Simone Helena Ferreira – MSc in Public Health from ULBRA, Canoas, RS, Brazil, and an associate professor at the School of Dentistry, ULBRA, Canoas, RS, Brazil.
Camilla de Moraes Pasini – DDS from the School of Dentistry, Universidade Luterana do Brasil (ULBRA), Canoas, RS, Brazil.
Priscila Humbert Rodrigues – PhD in Dentistry from School of Dentistry, ULBRA, Canoas, RS, Brazil, and an associate professor at the School of Dentistry, ULBRA, Canoas, RS, Brazil.
Moisés Zacarias Cardoso – MSc in Pediatric Dentistry from School of Dentistry, ULBRA, Canoas, RS, Brazil.
Tássia Silvana Borges – MSc in Health Promotion from Universidade de Santa Cruz do Sul (UNISC), Santa Cruz do Sul, RS, Brazil, and a PhD candidate at the School of Dentistry, ULBRA, Canoas, RS, Brazil.
Correspondence: Tássia Silvana Borges, Rua Joana Machado, 103/302, Camobi, CEP 97105-180, Santa Maria, RS, Brasil.
Tel.: +55 (55) 9914.7698. E-mail: tassia.s.borges@hotmail.com

Cerebral (PC) é uma alteração congênita que afeta o sistema nervoso central antes dos dois anos de idade, dificultando o desempenho de atividades frequentemente realizadas por crianças com desenvolvimento normal. O objetivo deste estudo foi descrever o tratamento odontológico realizado em criança com 10 anos de idade com Amelogênese Imperfeita e Paralisia Cerebral. A família procurou a clínica do Projeto de Extensão "Conquistando Saúde: Atendimento odontológico à pacientes com deficiência" da Universidade Luterana do Brasil Canoas/RS em busca de reabilitação estética. Ao exame físico e radiográfico confirmou-se o diagnóstico de AI. A utilização de estabilização física com a concordância dos pais foi necessária para a realização do tratamento odontológico. O tratamento iniciou com orientações de higiene bucal, dieta e raspagens supra gengivais. Na sequência, optou-se pelo recobrimento com ionômero de vidro dos dentes posteriores e reconstrução dos anteriores com resina composta utilizando-se coroas de acetato. No acompanhamento do caso observou-se melhora na saúde bucal e estética da paciente. Além disso, este relato demonstra que o atendimento odontológico complexo em pacientes com deficiência pode ser realizado em ambiente ambulatorial.

Palavras-chave: Paralisia Cerebral; Amelogênese Imperfeita; Assistência Odontológica.

INTRODUCTION

Amelogenesis Imperfecta (AI) is an inherited alteration that affects the enamel of primary and permanent teeth, with no systemic manifestations. The gene can be transmitted either as an autosomal dominant or autosomal recessive trait, or it can be X-linked. The enamel may have formation disorders or even defects in its mineral and protein content (1,2).

The clinical presentation involves dark-brown colored teeth due to the transparency of the adjacent dentin, in addition to roughness alterations on the enamel surface, favoring greater plaque accumulation and consequent greater chances of developing caries (1,3). These teeth are more prone to abrasion, which causes loss of vertical dimension, with aesthetic impairment and dentin exposure, possibly leading to sensitivity to temperature changes. The radiographic analysis shows the enamel is poorly defined.

Cerebral Palsy (CP), also called Non-Progressive Chronic Encephalopathy, is a result of a static lesion occurred in prenatal, perinatal, or postnatal period, affecting the central nervous system during the brain's structural and functional development. It is predominantly a sensorimotor dysfunction, involving disorders in muscle tonus, posture, and voluntary movement (4). Some characteristics of CP are lack of movement control and adaptive changes in muscle length, which in some cases can result in bone deformities, interfering in the function and in the performance of children's regular activities (5).

The information found in the Literature is conflicting in regards to the occurrence of oral diseases in patients affected by CP (6). No intraoral anomalies are exclusive to people with cerebral palsy, but they can be more severe than in the general population. The dental treatment of CP patients is highly complex. Since their spastic muscles are in a state of continuous contraction, it is difficult for them to be properly seated in the chair. The dental caregiver must be aware of speech disorders and also lack of correspondence between the chronological age of the patient and his actions. Therefore, manual skills cannot be requested, and involuntary reflexes should not be interpreted as rejection to

the treatment (3). Based on that, this study aims at describing the clinical case report of the dental treatment of a 10-year-old patient affected by amelogenesis imperfecta and cerebral palsy.

CLINICAL CASE REPORT

A 10-year-old, white, female patient, came to the dental office of an extension project called "Gaining Health: Dental Care for Patients with Disabilities" of the Lutheran University of Brazil, located in the city of Canoas. The patient was joined by her parents, who reported that the child had cerebral palsy and aesthetically impaired teeth (Figure 1).

FIGURE 1 – Patient AJS, 10 years old, at the first dental appointment.

The current medical history of the patient do not present further systemic alterations, and she is not under any medication. Moreover, the patient presents adequate weight and height according to the physical growth curve of her age group. As a result of the abuse inflicted by her biological parents, the child suffered pulmonary bleeding and respiratory arrest, and was sent to a shelter when she was sixteen months old. These episodes may be related to the development of the cerebral palsy. The patient was adopted when she was three years old. According to her adoptive parents, until the age of two the child was only given liquid diet through a syringe, presenting severe malnutrition. The parents say that the patient currently eats regular meals; however, all food is chopped or has pasty

consistency. The parents do not observe sensitivity to foods and the patient does not demonstrate any pain symptoms.

The child attends an APAE school (Association of Parents and Friends of Handicapped People), where she engages in physiotherapy, speech therapy, and psycho-pedagogical activities. The patient has a finger-sucking habit of her left thumb, both in stressful and in pleasant situations, causing anterior open bite. Alterations can also be observed on the sucked finger (Figure 2).

FIGURE 2 – Patient sucking the left thumb; clinical aspect of the sucked fingers.

For the conduction of the physical examination, the patient had to be physically restrained due to her psychomotor alterations (Figure 3A). During the examination it was observed that the patient was in the phase of mixed-dentition and both dentitions presented partial or total absence of enamel, color alteration (yellow-brown), rough surface, and first stages of loss of vertical dimension, thus characterizing amelogenesis imperfecta (Figure 3B). At the physical examination, a panoramic radiograph was requested; it showed presence of all permanent teeth, little enamel contrast and order of eruption compatible with her age group (Figure 3C).

FIGURE 3 – A) Restraint of the patient by the dental team and family members; B) Clinical aspect of the dentition; C) Panoramic radiograph.

After the physical examination, a treatment plan that covered both functional and aesthetic aspects was drawn up. The treatment plan took into account the challenges related to the dental care of CP patients. The first step was giving oral hygiene instruction to the parents, highlighting the importance of using fluoride toothpaste and a soft toothbrush. The parents were extremely cooperative, but reported difficulties to perform tooth brushing. To ease the process, the use of mouth openers (PVC pipes or a PET bottle top) was recommended.

In the course of the following appointments, the primary teeth with advanced root resorption – which prevented good oral hygiene – were extracted. All of the procedures were preceded by fluoride tooth brushing. The extractions of teeth 53, 63 and 74 were conducted under topical anesthesia (5% Xylocaine) and infiltration anesthesia (2% Lidocaine).

Over the next sessions, supragingival scaling was conducted to decrease gingival bleeding and to improve the overall oral hygiene. The following step was to start restorative procedures in the posterior teeth. A VITREMER® glass ionomer cement (GIC) was chosen to cover the occlusal surface of teeth 16, 26, 36, 46, 14, 15, 24 and 25, using the standard protocols for this material (Figure 4).

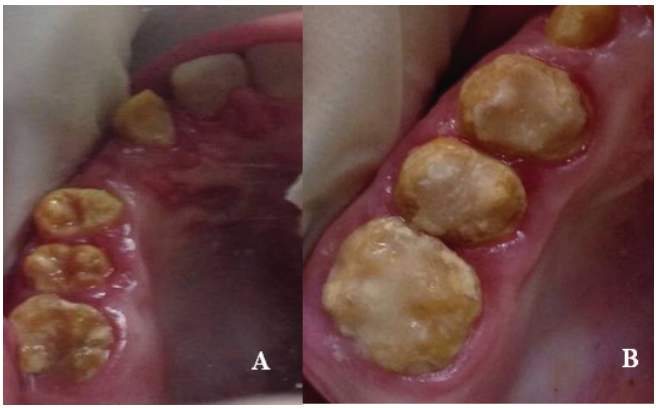

FIGURE 4 – A) Posterior teeth; B) Posterior teeth covered with GIC.

For the clinical procedures, mouth-openers made with wooden tongue depressors and gauze or a PVC pipe were used, in addition to continuous use of an aspirator tube (Figure 5 A and B).

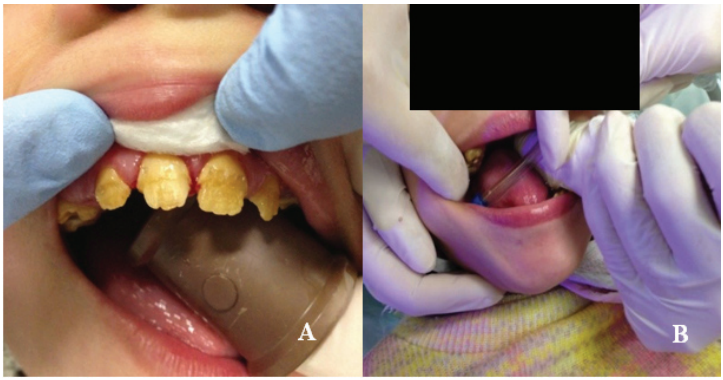

FIGURE 5 – A) PVC pipe; B) Aspirator use.

After covering the posterior teeth with GIC, aesthetic restorative procedures of the anterior teeth started. Such procedures were also conducted with the help of physical restraint and mouth-opener. For these restorations, prefabricated acetate crowns were selected. The crown had to be adapted by cutting its cervical border with the aid of curved scissors and scalpel (Figure 6).

FIGURE 6 – A) Initial clinical aspect; B) Adapting the acetate crown; C) Restoration of teeth 11 and 21.

After adapting the acetate crown, additional retentions were created in the vestibular surface of teeth 11 and 21 using a drill. Once the teeth were prepared, the color of the resin was selected. The resin selected was the CHARISMA® A3. All the principles that guide resin restorations were followed, using relative isolation, dental aspirator and adhesive system. After the restorations were made, finishing and polishing was conducted using diamond tips, flexible discs, silicone tips, as well as polishing brushes.

Teeth 12, 22, 31, 32, 41 and 42 were also restored following the same abovementioned steps. Even after the restorative procedures, the anterior open bite could be seen (Figure 7).

FIGURE 7 – Superior and inferior incisors after restorative treatment.

After undergoing rehabilitation treatment, the patient is considered clinically healthy. She was directed for preventive and periodic maintenance appointments. In these appointments, the conditions of the GIC restorations of the posterior teeth will be evaluated. If necessary, new applications will be made. Moreover, control of the aesthetic restorations and oral biofilms, in addition to hygiene and diet orientation, should be reinforced to maintain the patient's oral health.

DISCUSSION

The dental care of patients affected by disabilities is often out of the usual routine, requiring specialized knowledge, greater sensitivity, increased attention and adequate environment (7).

Cerebral palsy, which is the condition affecting the previously mentioned patient, is a non-hereditary lesion that affects the nervous system before the age of two, during pre-, peri- or postnatal period. It leads to poor or abnormal motor development. This disorder can be isolated or combined with other issues – cognitive, psychic, sensory, and/or language disorders, depending on the affected area and the extension of the lesion (4). CP patients can present disorders in speech development because of the alterations on expressive-motor aspects of language; nevertheless, comprehension is preserved in some cases (8).

In this clinical case, the patient presents intellectual disability, no speech and comprehension, and increased muscle tonus, in addition to compromised lower limbs, causing the patient to use a wheelchair. The symptoms of the patient prevent her understanding of the dental treatment and, consequently, cause her to resist any procedures.

The oral diseases that affect CP subjects are the same as for the general population. However, they are usually more frequent in CP patients because of factors such as: poor oral hygiene, type and consistency of the food, medication use, increased muscle tonus, lack of information and lack of access to dental services (6,9).

Techniques of behavioral control proved to be inefficient due to the limitations of the patient. The option was to use aversive techniques that restricted the movements of the patient. These techniques range from the use of mouth-openers to the total restriction of movement. Such measures were only taken with the consent and help of the parents and without using any pharmacological agents (10). The parents were cooperative and understanding at all times during restraining procedures. The restraint of the subject is precisely indicated for cases in which neuromotor disorders lead the patient to make involuntary movements. It aims at the comfort of the patient without harm to the dental caregiver, with the objective of providing a highly safe treatment for the dental surgeon, the dental team and the patient (9).

In addition to the cerebral palsy systemic frame, this patient presented amelogenesis imperfecta, a malformation of the dental enamel that affects both primary and permanent dentition (11). No direct relation between the two conditions shown by the patient can be found in the Literature. According to Tahmassebi et al., (2003) (12) teeth affected by AI have altered size and shape, yellowish color and rough surface. Gingivitis is commonly present due to the bacterial plaque accumulation caused by grooves on the enamel. Through radiographs, it is possible to confirm the condition on both dentitions, which agrees with other reports on amelogenesis imperfecta (12).

The diagnosis of Hypoplastic Amelogenesis Imperfecta was confirmed based on the anamnesis and physical and radiographic examinations, since the enamel presented reduced thickness, with furrows and pits. Linear pits covered all the surface of the enamel and the color was brown-yellowish (13).

The parents were unable to confirm the presence of the condition on other family-members, since the child was adopted. It is important to emphasize that a precise and

early diagnosis helps minimizing sequelae and planning the best possible treatment for such cases (13).

There is a large array of possibilities for treating AI, with a variety of restorative materials and methods; it is up to the dental caregiver to select the most appropriate ones (14). Besides choosing a technique, further aspects that must be taken into consideration during the design of a treatment plan are the desires and expectations of the patient or parents, the socioeconomic status, the AI type and severity, the age of the patient, the overall oral health and the cooperation of the patient for the performance of the procedures (15).

Some AI patients can be bullied due to the aesthetic compromise caused by this condition. It can intervene in socializing, lower self-steem, and eventually affect the quality of life of the subject and the family. The awareness of the possible implications allows the dental surgeon to effectively conduct different treatments, improving the aesthetic appearance of the affected teeth (16).

In the reported clinical case, the patient is a 10-year-old girl, with low socioeconomic status, hygiene difficulties, and little or no cooperation for the performance of the treatment due to her systemic frame. Given that, the aim of the dental treatment was to minimize sensitivity, to improve the aesthetic factor (the parent's desire) and to improve oral health.

Therefore, covering the posterior teeth with glass ionomer cement was the option. The GIC is an important material in the minimally invasive dental practice, and it stands out by its adhesion to the dental structure, linear thermal expansion coefficient similar to the tooth, biocompatibility, fluoride release, and easy technique (17). The application in a single increment makes the procedure faster, which should be taken into consideration when treating patients with disabilities.

The aesthetic restorations of the anterior teeth were made using Charisma® resin-based composite and prefabricated acetate crowns for those teeth that were severely damaged, fractured, discolored, or malformed, allowing the resin to cover most of the remaining structure (18). A 30-seconds acid conditioning and a Prime & Bond® adhesive system preceded the restoring technique to provide a surface resistant to fracture (19). The treatment using acetate crowns allows for little etching of the dental surface because it is performed directly on it, in addition to having stable color, enabling normal occlusion, and presenting good resistance and durability, thus leading to satisfactory results (20-22). Furthermore, this could protect the tooth from biofilm formation and from consequent caries development (23).

However, it must be taken into consideration that the process of adhesion of the dental structure can be an issue for AI tooth restorations. The Literature shows conflicting results, since some authors report no differences between sound dental structures and those affected by AI (19), only presenting relation to uncertain patterns of acid conditioning and adhesive system. On the other hand, other authors have found adhesion problems in AI teeth, blaming it on the morphologic differences of exposed dentin (24). In the

reported case, the restorative material did not show any adhesion problems to the dental structures up to this moment.

The performance of dental care in disabled patients faces multiple challenges, starting with their specific needs. Moreover, the difficult access to dental services and the fact that those services are often precarious, added to the lack of qualified personnel (dental surgeons and dental assistants) can be mentioned as problems related to this matter. However, the issues linked to the dental caregivers are usually not technical, but rather related to human factors, such as moral, philosophical and psychological aspects (25).

FINAL CONSIDERATIONS

The presented case achieved clinical success with an improvement in oral health and aesthetic conditions of the patient. It is imperative that she takes part in a program of periodic and preventive maintenance. The case shows that it is possible to perform complex dental care of disabled patients in an outpatient environment.

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