# Use of Marsupialization as a Definitive Treatment for Large-sized Dentigerous Cysts in a Patient with Mucopolysaccharidosis Type I

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**Abstract:** The correct diagnosis is fundamental for the appropriate treatment to be employed in a particular pathology. The best treatment is not the one that solves only local problems, fragmenting the patient, and therefore, it is necessary to integrate the entire systemic condition of the individual before initiating any local treatment. This context inevitably requires dentistry to participate in a multidisciplinary approach, where the role of the dentist is expanded in concepts that encompass ethics, human dignity, and professional valorization. This article describes a clinical case of a patient with mucopolysaccharidosis type I, whose treatment of cystic lesions present in the mandible was exclusively performed through marsupialisation. The objective of this study is to demonstrate, within the complexity of this rare syndrome, the difficulties of diagnosis and the need for evaluation of the patient beyond the limits of the oral cavity, as well as to report two cases of large dentigerous cysts, surgically treated conservatively through marsupialisation, without the need for re-approach for enucleation and without recurrences over a 20-year period.

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## Introduction

The triad of pathology-semiology-diagnosis is true and indispensable, given that the goal of diagnosis can only be achieved through it (Castro et al., 1995). Thus, the work carried out by the professional with the patient in collecting clinical data that will compose not only the symptomatic picture of the disease but also its entire physical and psychological panorama, past and present, needs to be done in a detailed manner. In dentistry, for the diagnosis of oral clinical alterations, it is no different; semiotic maneuvers are necessary and indispensable (Tommasi, 1989).

Thus, the diagnosis should include a systematic and organized general assessment of the patient. In this context, it is important to remember that the oral cavity and the maxillofacial region can be the initial site of manifestation of rare diseases (Tommasi, 1989). Tommasi (1989) affirm that the dentist has the ethical responsibility to base their professional practice on continually updated knowledge, which allows them to better interpret the pathologies they encounter and to provide effective prevention and appropriate treatment for them.

Mucopolysaccharidoses (MPS) represent a typical example of the complexity of the human body and the challenge that professionals face regarding a comprehensive and systematic patient assessment (Zhou et al., 2020). According to Murahovschi (2013), MPS are a group of diseases characterised by excessive urinary excretion of acid mucopolysaccharides and accumulation of tissue material. Clinically, it can be recognized as a disease with abnormal phenotype, classified into 7 types and 13 subtypes based on their genetic, clinical, and biochemical characteristics (Nagpal et al., 2022). The first case of MPS was described by Charles Hunter in 1917, and two years later, MPS I was described by the same author. The prevalence of different types of MPS varies across continents, suggesting an association with geographical regions and ethnicity (Zhou et al., 2020). The estimated occurrence of this syndrome is 1 in 25,000 births (Hampe et al., 2020). It is an autosomal recessive disease, except for MPS II, which is transmitted as an X-linked recessive disorder (Ribeiro et al., 2015; Nagpal et al., 2022).

The metabolic alteration through lysosomal degradation of mucopolysaccharides present in this rare genetic disease is reflected in significant changes in many tissues of the body, including bones, cartilage, and connective tissues. In general, MPS is caused by the deficiency or absence of a specific enzyme necessary to break down mucopolysaccharides, leading to the accumulation of these molecules in body tissues. The symptoms and severity of MPS can vary widely depending on the type of the disease and the degree of enzyme deficiency (Hampe et al., 2020; Zhou et al., 2020; Nagpal et al., 2022).

The etiology of mucopolysaccharidosis type I is related to the lack or relative deficiency of the enzyme α-L-iduronidase, and the symptoms mainly include alterations in physical appearance, growth delay, bone deformities, short stature, facial abnormalities, and umbilical and inguinal hernias. Additionally, there are

typically associated health problems such as respiratory infections, cardiac and hepatic issues, neurological problems, and delayed cognitive development (Ribeiro et al., 2015; Hampe et al., 2020; Zhou et al., 2020).

Regarding diagnosis, unfortunately, in most cases, it is established when irreversible characteristics of the disease are already present. The treatment of MPS mainly involves managing symptoms and complications, and the prognosis depends on the type and severity of the disease. In general, the most severe forms of MPS can lead to a reduced life expectancy, while milder forms may not alter the life expectancy of affected patients. However, most people with MPS face significant challenges and require ongoing medical support and care throughout their lives (Turra and Schwartz, 2009; Hampe et al., 2020).

Regarding the buccomaxillofacial complex, MPS can cause gingival alterations such as fibrosis and hyperplasia, as well as malocclusions, dental retentions, and the development of cysts and tumours (Sharma et al., 2012; Sabry et al., 2018; Torres et al., 2018; Zhou et al., 2020). This paper describes a clinical case of a patient with MPS type I with significant physical impairment and multiple maxillofacial manifestations. Among them, dental retentions and large dentigerous cysts were observed. Marsupialization was successfully instituted as the sole treatment for the patient's dentigerous cysts, and no recurrence was observed over a period of 20 years.

## Case report

A 13-year-old male patient of Caucasian descent, presented to the surgery clinic at UNIFAL-MG with a complaint of swelling on the face that had started 3 years ago. Initially, it was possible to observe physical growth deficiency, lip incompetence, stiffness in the hand joints, and skeletal and thoracic deformity (Figure 1A and B). The patient did not present mental retardation. Corneal opacification was already observed. In the medical history, bronchitis, asthma, and adenoid hypertrophy were recorded.

During the extraoral clinical examination, no lymph node enlargement in the head and neck region was observed, with only volumetric increase on the left side of the face, near the mandibular body and ramus. Additionally, widened head and nose, dilated nostrils, prominent supraorbital ridges, round cheeks, and thick lips were observed (Figure 1C). Upon intraoral clinical examination, gingival hyperplasia, open bite, delayed eruption, and ectopic positioning of some teeth were detected (Figure 2). Panoramic and occlusal radiographs revealed extensive unilocular radiolucent lesions causing expansion in the basal cortical bone of the left mandible, involvement of retained teeth in the right mandible, displaced tooth germs (17, 18, 27, 28, 37, 38, 47, and 48), presence of a supernumerary tooth located in the maxillary midline, maxillary sinuses with irregular contours due to tooth displacements, and significant condylar resorption (Figure 3).

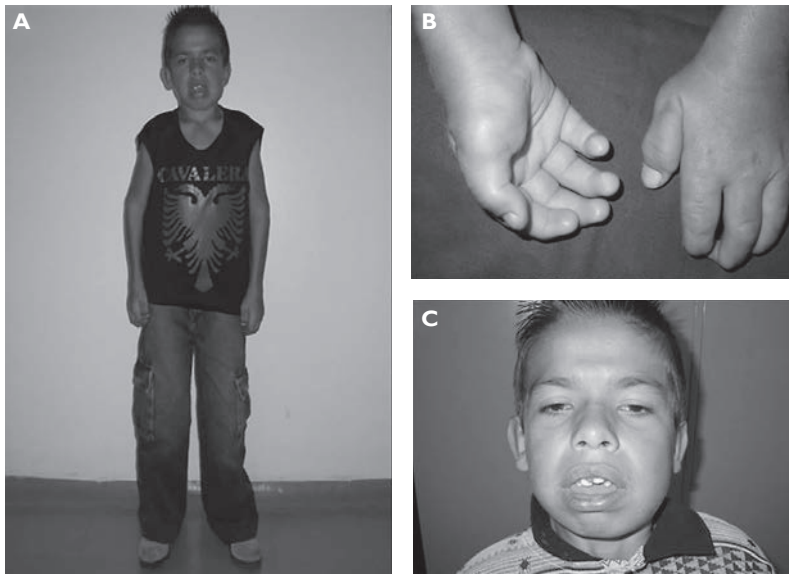


*Figure 1 – Physical growth deficiency, skeletal and chest deformity, changes in the joints of hands (A and B); lip incompetence, enlarged head and nose, dilated nostrils, prominent supraorbital region, thick cheeks and lips (C).*

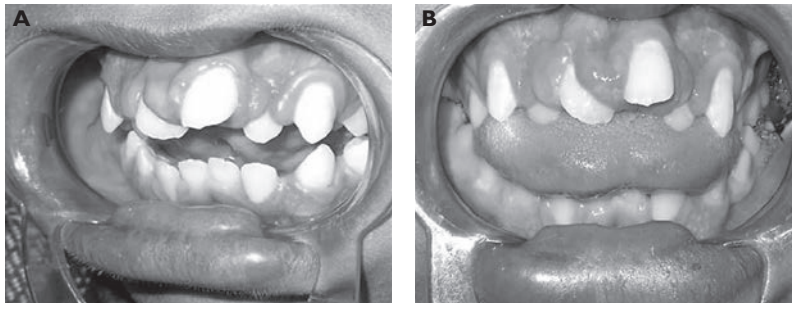


*Figure 2 – Intraoral photograph showing gingival hyperplasia, anterior open bite with tongue interposition, macroglossia, delayed eruption, and ectopic positioning of dental elements (A and B).*

Based on the clinical examination, medical history, and radiographic findings, three diagnostic hypotheses were suggested: cleidocranial dysostosis, Gorlin-Goltz syndrome, and MPS. The latter was supported by medical documentation provided by the patient, indicating the possibility of this pathology.

The treatment plan for the oral lesions involved marsupialization due to the possibility of pathological fracture of the mandible. Initially, the left side was addressed. After local anesthesia, a puncture was performed, with positive aspiration of a transparent yellow fluid. Subsequently, an elliptical incision was made in the area

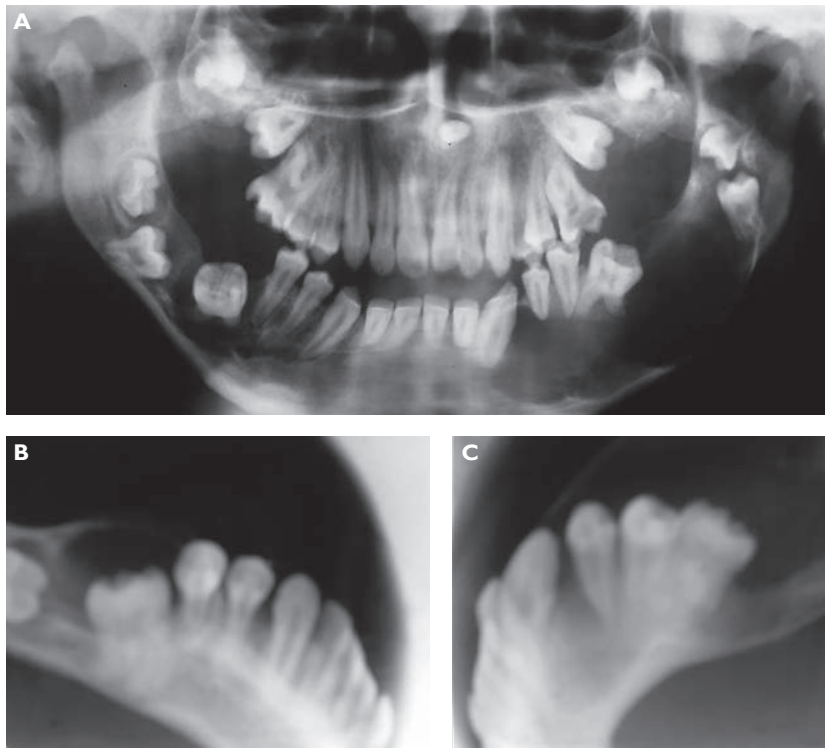


*Figure 3 – Panoramic radiographs (A) showing extensive unilocular radiolucent lesions in the mandible bilaterally, involving tooth 46 on the right, retained teeth, supernumerary teeth in the midline, and marked condylar resorption (A). Occlusal radiographs showing expansion caused by the cystic lesion in the basal cortical bone of the right mandible (B) and left mandible (C).*

in question, followed by tissue dissection that allowed direct access to the cystic cavity. The specimen was sent for histopathological examination, and a semi-rigid latex drain was installed (Figure 4). The patient and their guardian were instructed to irrigate with saline solution three times a day. After 10 days, with epithelialization of the fistulous tract observed, the latex drain was removed, and the patient and guardian were instructed to maintain cavity hygiene as previously described. The histopathological result revealed the presence of a dentigerous odontogenic cyst. Surgery on the right side of the mandible was then scheduled. The incision of the fibrous gingiva allowed access to the radiolucent lesion in the region of tooth 46 for drain installation to permit decompression of the cystic cavity. The patient and guardian were again instructed on the same postoperative cavity hygiene care as in the previous procedure. The drain was removed after 10 days.

The patient attended clinical and radiographic follow-up until the postoperative period of 1 year and 6 months, during which significant bone formation was observed in the region of the cystic lesions (Figure 5). After this period, the patient

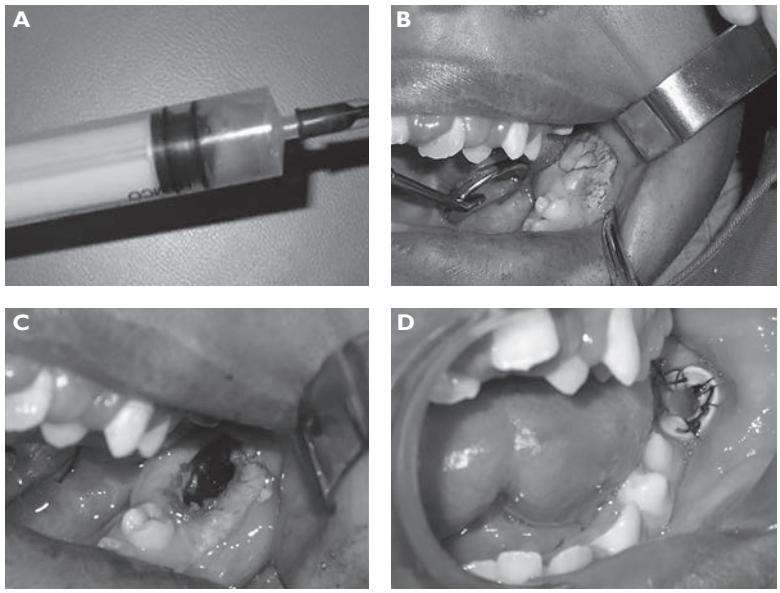


*Figure 4 – Images showing the surgical technique of marsupialization. Positive aspiration of the lesion with return of a transparent yellow fluid (A); elliptical incision over the mandibular ridge (B); excision of part of the cystic capsule for incisional biopsy and tissue dissection to access the cystic cavity (C); semi-rigid latex drain in position (D).*

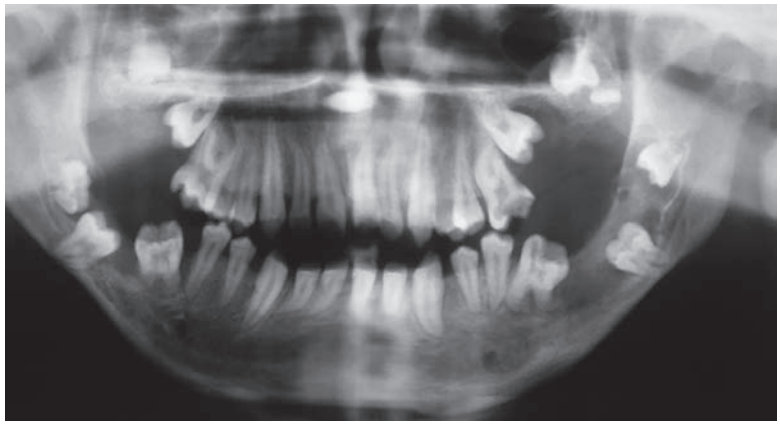

*Figure 5 – Panoramic radiograph taken 18 months postoperatively showing significant bone formation in the region of the cystic lesions.*

did not attend further follow-up appointments, returning recently, after 20 years, with the main complaint of malocclusion. Upon extraoral clinical examination, in addition to the evolution of the body changes already observed in childhood, significant opacification associated with blindness in the right eye and reduced

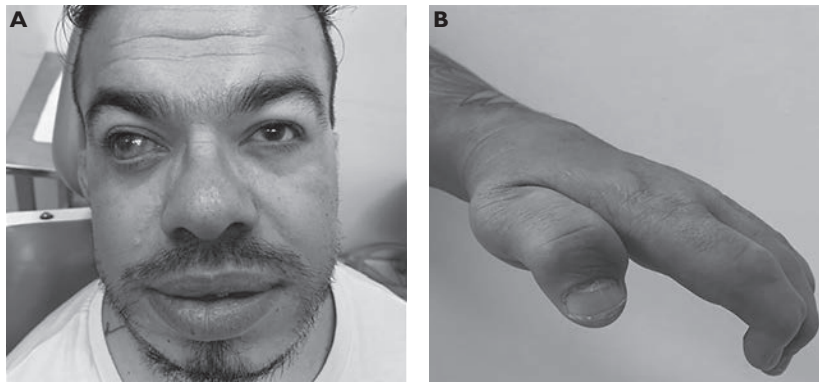


*Figure 6 – Extrabuccal photos showing the evolution of the body changes already observed in childhood (A and B); ocular involvement with significant corneal opacity associated with blindness in the right eye (A).*

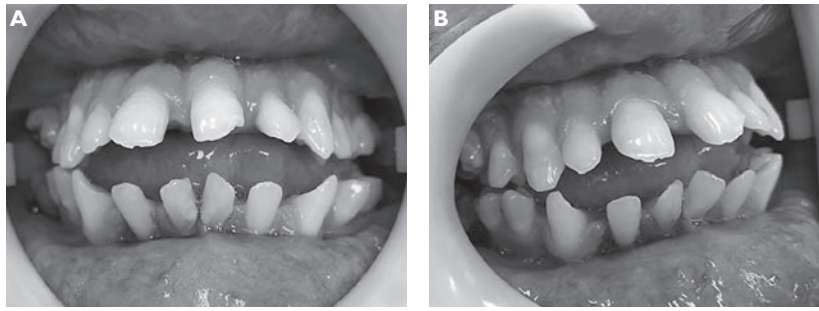


*Figure 7 – Intraoral photos showing anterior open bite with significant vestibular inclination of the anterior teeth, macroglossia, and tartar accumulation (A and B).*

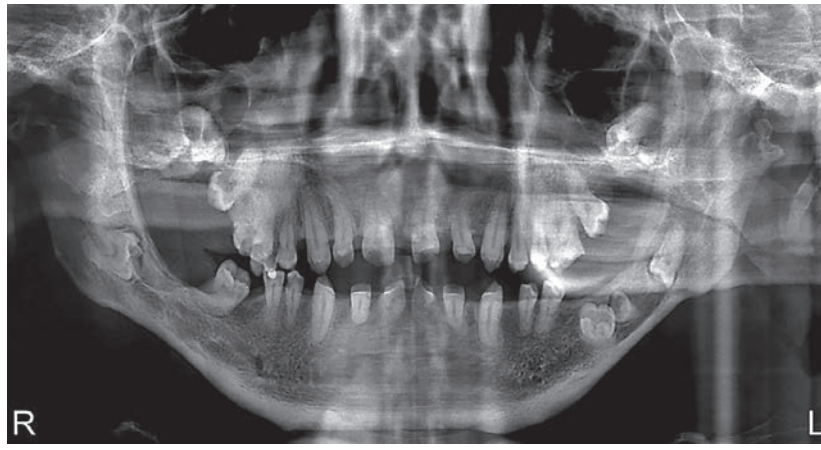


*Figure 8 – Panoramic radiograph after 20 years from the first visit showing retained teeth and absence of recurrent cystic lesions.*

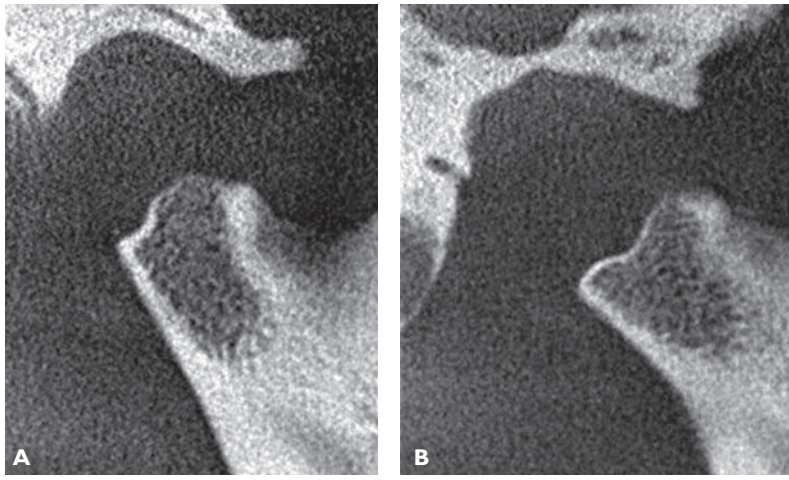


*Figure 9 – Sagittal section of computed tomography scan of the right (A) and left (B) TMJ (temporomandibular joints) showing severe reabsorption.*

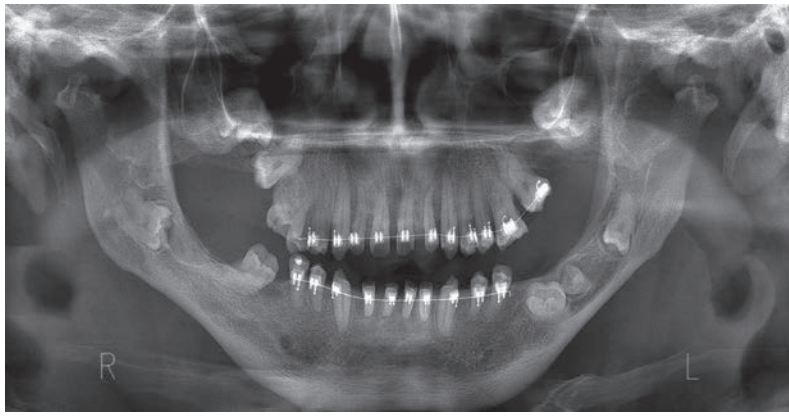


*Figure 10 – Panoramic radiograph after 20 years from the first visit showing retained teeth, absence of recurrent cystic lesions and orthodontic treatment in progress.*

left visual acuity was observed (Figure 6). Upon intraoral physical examination, anterior open bite with significant anterior teeth vestibular inclination, macroglossia, and tartar accumulation were observed (Figure 7). Current radiographs show dental retentions but the absence of associated cystic lesions (Figure 8) and in the computerized tomography of the TMJs (temporomandibular joints), severe resorption oh the condyles was observed (Figure 9). This situation demonstrates that isolated marsupialization was, in this case, sufficient as the definitive treatment for two large dentigerous cysts. The patient underwent periodontal treatment and is undergoing orthodontic treatment for malocclusion correction (Figure 10: panoramic

radiograph with the orhodontic treatment in progress). Regarding this, dental inclination is largely due to macroglossia. It is known that macroglossia is preferably treated through partial glossectomy. However, the patient in question has a history of serious complications due to intubation for general anesthesia due to tracheal stenosis. Therefore, the application of botulinum toxin was proposed to reduce tongue muscle tone and consequently muscular force. This procedure is being planned and will be performed soon.

## Discussion

The lack or relative deficiency of the α-L-iduronidase enzyme in MPS I is caused by a mutation in the IDUA gene located on chromosome 4p.16 (Bay et al., 2021; Nagpal et al., 2022). This enzymatic deficiency alters the metabolism of mucopolysaccharides, resulting in the accumulation of glycosaminoglycans (GAGs) and dermatan and heparan sulfates in organs and tissues. GAGs constitute the main component of the extracellular matrix and are responsible for cell-cell and cell-matrix adhesion. Additionally, they provide structural stability for cartilaginous structures such as heart valves and joints (Tatapudi et al., 2011; Hampe et al., 2020). The incidence of MPS I varies from 1 to 3 in 100,000 live births (Carvajal Gavilanes et al., 2023). Patients with a severe phenotype of the disease, if left untreated, have a low life expectancy, with death commonly occurring in the first decade of life. Prevention of severe cardiac and respiratory compromise, as well as cognitive impairment, can only be achieved through early diagnosis. However, early diagnosis is challenging as the initial manifestations of the disease are nonspecific (Hampe et al., 2020; Zhou et al., 2020; Bay et al., 2021).

Three subtypes of MPS I have been proposed, differing in the severity of the disease, ranging from mild (Scheie syndrome), moderate (Hurler-Scheie syndrome), to severe (Hurler syndrome or MPS-IH). However, in 2008, the guidelines on MPS I were revised and updated, so that now the disease is classified into two broader groups. These are severe MPS I (Hurler syndrome) and attenuated MPS I (Scheie syndrome and Hurler-Scheie syndrome) (Grupo de Trabajo de Enfermedades poco frecuentes, 2008). The severe form of the disease is caused by the absence or extremely low levels of the α-L-iduronidase enzyme, associated with genotypes with deletions and nonsense mutations (Hampe et al., 2020). This paper describes a clinical case of a patient with attenuated MPS I, whose characteristics and symptoms are suggestive of Hurler-Scheie syndrome.

Diagnosing MPS in general unfortunately occurs when irreversible characteristics of the disease are already present (Zhou et al., 2020). In countries like Brazil, the diagnosis is often delayed (Ribeiro et al., 2015). According to Vieira et al. (2008), there is an average of 4.8 years between the onset of symptoms and the diagnosis of the disease. Thus, methods that can achieve early diagnosis, in the asymptomatic

phase, are important to improve the quality of life and survival of patients affected by the disease. Radiographs, for example, can reveal kyphosis of the lumbar vertebrae, which is the first sign of the disease, especially in MPS IV. Additionally, computed tomography (CT) and magnetic resonance imaging (MRI) of the skull can show early neurological changes characteristic of MPS types I, II, and III. For definitive diagnosis and determination of the MPS subtype, new approaches involve urine and blood tests for GAGs, genetic testing, and enzymatic assays (Zhou et al., 2020; Bay et al., 2021). However, initially, the most important aspect is the observation of the family medical history, as it is a hereditary disease. A positive family history, the presence of signs and symptoms, as well as the order of their appearance, are important for establishing the diagnostic hypothesis of MPS (Torres et al., 2018; Zhou et al., 2020). Torres et al. (2018) described a clinical case of a patient diagnosed with MPS II even during the intrauterine period. Early diagnosis was possible due to a family history of MPS with severe phenotype. For the patient in question, initiating treatment in the first months of life managed to attenuate the manifestation of the disease (Torres et al., 2018).

In this context, it is important for healthcare professionals to be aware of the characteristics, signs, and symptoms of MPS in order to facilitate early diagnosis. Early symptoms observed before six months of age include frequent rhinitis, hernias, hepatosplenomegaly, thoracolumbar kyphosis, thin and non-elastic skin, and early closure of facial sutures. Additionally, specific facial characteristics are observed, including enlarged head and nose, dilated nostrils, prominent supraorbital ridges, round cheeks, thick lips, macroglossia, and dental alterations (Ribeiro et al., 2015; Torres et al., 2018; Hampe et al., 2020). In the patient of the present clinical case, all the facial characteristics listed above were identified, along with malocclusion, delayed eruption of some teeth, dental retentions, and mandibular cysts. Torres et al. (2018), described a case of a patient with MPS II who also presented multiple maxillofacial manifestations, including delayed tooth eruption, dental retentions, and macroglossia. Sharma et al. (2012), reported a clinical case of severe MPS I (Hurler syndrome), in which the patient also had mandibular cysts, similar to the present clinical case.

The skeletal and soft tissue alterations present in patients with MPS I are due to the accumulation of GAGs in these tissues and may predispose to upper airway obstruction. Regarding skeletal changes, nasal dysmorphism, shortened neck, abnormalities in cervical vertebrae, and mandibular hypoplasia can be mentioned. As for soft tissue alterations, enlargement of the tongue, adenoids, and tonsils can narrow the airway passage. This situation leads to noisy breathing, apnea, and recurrent respiratory infections (Ribeiro et al., 2015; Torres et al., 2018; Hampe et al., 2020). In the case of the present clinical case, the patient has a medical history of adenoid hypertrophy and respiratory diseases (bronchitis and asthma). It is worth noting that initially, respiratory symptoms are more pronounced in the upper airways, but as the disease progresses, bronchotracheal involvement may occur. This

is because GAG deposition causes deformity and narrowing of the tracheal lumen. In severe cases, this situation can lead to death (Hampe et al., 2020). In the present clinical case, there is a history of difficulties in intubating the patient for general anesthesia due to tracheal stenosis. Therefore, partial glossectomy for the treatment of macroglossia was not performed, as it requires intubation and general anesthesia. Instead, the injection of botulinum toxin into the tongue was proposed to reduce tongue tone and volume and thus control tongue protrusion. This procedure is being scheduled. In this context, it is important to note that the application of botulinum toxin to the tongue has been reported by several authors in the treatment of involuntary tongue protrusion resulting from oromandibular dystonia (Charles et al., 1997; Yoshida, 2019; Hassell and Charles, 2020).

After 6 months of age, other symptoms of MPS may become evident, such as visual and hearing impairment, cardiac alterations, more pronounced musculoskeletal defects, and psychomotor retardation (Ribeiro et al., 2015; Hampe et al., 2020). In the initial contact with the patient in the present study, at the age of 13, there was slight corneal opacification, but no impairment of visual acuity. However, when he returned as an adult, after 20 years, marked corneal opacification associated with blindness in the right eye and decreased visual acuity in the left eye was observed. The ocular changes resulting from MPS I are caused by the accumulation of GAGs in ocular tissues and include glaucoma, abnormalities in eye movement, changes in the retina, optic nerve atrophy, exophthalmos, and corneal opacification (Hampe et al., 2020; Nagpal et al., 2022). The latter is the most common ocular alteration in MPS (Nagpal et al., 2022).

Among all manifestations of MPS I, skeletal changes, collectively referred to as multiple dysostosis, are the most frequent and disabling alteration. The most prominent skeletal changes include thoracolumbar kyphosis and growth restriction. However, other skeletal alterations may also be present, such as: enlarged skull, flattening of vertebral bodies, hypoplasia of the odontoid process in the C2 vertebra, paddle-shaped ribs, short and widened clavicles, bullet-shaped phalanges, enlarged skull, and J-shaped sella turcica. In addition to structural skeletal changes, MPS I can cause decreased bone density predisposing to fractures (Hampe et al., 2020). The patient in the present clinical case already presented many skeletal alterations since the first appointment. He had growth restriction, enlarged skull, changes in the hands with stiffening of the joints, malformation of the thorax, and condylar resorption. The bone changes progressed as the patient aged but without significant impairment of his ambulation and mobility.

After the first year of life, cognitive changes may appear, especially in severe MPS I (Hurler syndrome). In these patients, cognitive development slows down at 6–9 months of age, seems to plateau in the second year of life, and may progress to decline thereafter if appropriate treatment is not instituted (Hampe et al., 2020). The patient in the present clinical case did not show signs of cognitive impairment in adolescence or adulthood. Therefore, the MPS I in this patient is certainly the

attenuated form of the disease. This is because most patients with Hurler syndrome die in the first decade of life due to progressive neurological impairment associated with cardiorespiratory failure (Tatapudi et al., 2011). In contrast, patients with the attenuated disease form can reach adulthood, although they also present systemic changes. Patients with Scheie syndrome have a higher life expectancy than those with Hurler-Scheie syndrome and Hurler syndrome (Zhou et al., 2020).

In the attenuated forms of MPS I, the clinical presentation may be limited to growth restriction, corneal opacification, mild facial features, hepatosplenomegaly, and micrognathia. In this attenuated variant of the disease, skeletal changes in the hands and spine may not be severe. However, due to airway narrowing and macroglossia, respiratory infections are almost always recurrent (Ribeiro et al., 2015; Hampe et al., 2020). The described condition is observed in the patient of the current article. However, it is important to note that in the attenuated form, although milder, the symptoms of the disease appear later, making early diagnosis more challenging than in the severe form of the disease. Assadeck et al. (2019) reported a series of three cases of attenuated MPS I (Hurler-Scheie syndrome) from the same family, in which babies were considered normal at birth. The first signs appeared slowly after the second year of life, and the diagnosis was only established when the patients were 12 years old, the same age range as the patient in the present clinical case. The timing of diagnosis in these patients suggests, once again, an attenuated form of MPS I, characterized by late symptoms, a longer life expectancy, and mild or absent neurological changes (Assadeck et al., 2019).

Regarding the treatment of MPS, it mainly involves managing the symptoms and complications of the disease to slow its progression and improve the patient's quality of life. Enzyme replacement therapy and hematopoietic stem cell transplantation are the mainstays of treatment, but it is worth noting that they are not effective for all cases (Sharma et al., 2012; Torres et al., 2018; Zhou et al., 2020). Therefore, treatment also includes palliative care, orthopedic surgeries to correct bone deformities, respiratory therapy, and physiotherapy. Overall, therapies are aimed at improving quality of life and reducing disease complications (Hampe et al., 2020). Torres et al. (2018) describe a clinical case of a 7-year-old patient who was diagnosed with MPS II in utero and was treated with hematopoietic stem cell transplantation at 70 days of age. This study highlights the importance of early diagnosis and treatment in attenuating the disease manifestation (Torres et al., 2018). It is worth noting that there is a promising treatment option still in development called gene therapy, which is targeted at treating the corneal opacification associated with MPS. However, knowledge about gene therapy is limited to animal studies, so more human studies are needed for a better understanding of its effects and applications (Nagpal et al., 2022).

Regarding the facial characteristics, in the present article, the patient exhibited coarse facial features such as cranial enlargement, prominent supraorbital ridges, widened nose, rounded cheeks, thick lips, and incompetent lips. Upon intraoral

examination, malocclusion with anterior open bite associated with macroglossia was observed. Tatapudi et al. (2011) described a case of a 15-year-old adolescent with MPS I (Hurler-Scheie syndrome), who also had anterior open bite associated with macroglossia, among other facial characteristics observed in the present clinical case. According to a cross-sectional study conducted by Turra and Schwartz (2009), in which 78 patients with different types of MPS were evaluated for alterations in the structures and function of the stomatognathic system, teeth and the tongue were the most affected structures; while swallowing and chewing were the most compromised functions. The authors of the mentioned study emphasize that the high frequency of alterations in the structures of the stomatognathic system in MPS leads to changes in its functions. These, in turn, can exacerbate structural alterations (Turra and Schwartz, 2009).

Among the dental alterations observed in patients with MPS I, abscesses, cavities, enamel defects, retained teeth, agenesis, and cysts can be mentioned (Tavares et al., 2004; Sharma et al., 2012; Ribeiro et al., 2015; Sabry et al., 2018; Torres et al., 2018; Carneiro et al., 2021). Regarding dental development, according to Tavares et al. (2004), delayed tooth eruption, inclusion of permanent teeth, hyperplasia of dental follicles, and several teeth associated with a single follicle, leading to a rosette-like radiographic image, can be observed. This characteristic is only reported in cases of MPS. Additionally, other authors, as in the present study, also highlight the association of macroglossia with anterior open bite. This results from the excessive pressure of the enlarged tongue due to the deposition of GAGS on the anterior teeth (Tatapudi et al., 2011; Sharma et al., 2012). However, in the case of MPS I, it is important to remember that mandibular hypoplasia associated with pronounced condylar resorption is also frequently present and influences the development of anterior open bites (Koehne et al., 2018). In the present study, pronounced condylar resorption was observed, with condyles already showing signs of flattening and concavities.

Ribeiro et al. (2015) conducted a multicenter study in which they evaluated the orofacial manifestations of 26 patients with different types of MPS. The authors observed that facial manifestations such as deficiency of the midface, anterior open bite, convex profile, macroglossia, gingival hyperplasia, and diastemas are frequently observed characteristics in MPS in general and do not allow differentiation between its subtypes. However, enamel hypoplasia was significantly more associated with MPS IV ($p=0.043$) (Ribeiro et al., 2015). On the other hand, another study evaluated maxillomandibular structural differences (width and height) in patients with different types of MPS and found that the mandible in MPS I is smaller than in MPS II and III and compared to the disease-free population, and such a finding is due to the progressive condylar resorption present in MPS I patients (Koehne et al., 2018). In the present clinical case, at the age of 13, during the initial assessment, significant condylar resorption was already observable, which worsened over time as shown in the panoramic radiograph taken at 33 years old, where little condylar remains with a

concave surface. Such radiographic characteristic was quite common in MPS I patients in a previous study (Koehne et al., 2018). Another cross-sectional study evaluated the craniofacial characteristics of MPS patients through anthropometric and lateral cephalometric measurements and observed a prevalence of the dolichofacial profile, as well as an increase in the lower anterior face, pronounced incisor inclination, reduced upper airway space, and lip incompetence ($p<0.05$) (Carneiro et al., 2023). It is important to note that facial abnormalities in MPS not only affect aesthetics and social life but can also alter vital functions such as eating and breathing. The latter is directly affected by maxillomandibular hypoplasia (Koehne et al., 2018).

Carneiro et al. (2021) conducted research on dental and maxillofacial alterations in MPS patients by evaluating panoramic radiographs and observed that various conditions such as supernumerary teeth, conoid teeth, retained teeth, root dilaceration, periapical and furcation lesions, condylar hypoplasia, and dentigerous cysts are more prevalent in MPS patients compared to the general population. However, in the literature review conducted in this study, few reports of dentigerous cysts in MPS I patients were observed. Sabry et al. (2018), on the other hand, described a situation similar to the present clinical case, in which a dentigerous cyst associated with the impacted tooth 37 was treated exclusively by marsupialization in an MPS II patient. In this work, as in the present study, enucleation was not necessary, as complete resolution of the cyst was achieved after 12 months of follow-up (Sabry et al., 2018). This scenario, of a syndrome with so many maxillofacial manifestations, poses a significant challenge for the dentist and highlights the importance of understanding MPS characteristics. This is because the contribution of the dentist can be crucial for the diagnosis of the disease. Furthermore, most alterations of the stomatognathic system in MPS require monitoring and treatment (Sharma et al., 2012; Ribeiro et al., 2015; Carneiro et al., 2021).

In the case of the patient in the present study, the presence of extensive radiolucent lesions involving the angle and body of the left mandible and tooth 46 on the right side, as well as the possibility of a pathological fracture of the mandible, determined the choice of appropriate surgical treatment, with marsupialization of the cystic lesions being proposed and performed. The marsupialization procedure involves creating a surgical window in the cyst wall, emptying the cystic content, maintaining continuity between the cyst and the oral cavity to decrease intracystic pressure, reduce the size of the lesion, and promote bone filling (Ellis, 1996; Sharma et al., 2012). Normally, future surgical intervention is needed for enucleation. However, in the present clinical case, despite the large extent of the cysts, isolated marsupialization allowed for the eruption of tooth 46 and the disappearance of the cysts without any sign of recurrence after 20 years postoperatively. Sharma et al. (2012) and Sabry et al. (2018) also employed marsupialization alone as the technique of choice for the treatment of extensive mandibular dentigerous cysts

involving the lower first molars in an MPS patient. This is because the technique is more conservative, allows for cyst regression and gradual bone tissue formation, and the teeth involved in the lesion can be preserved (Sharma et al., 2012; Sabry et al., 2018). The disadvantage of marsupialization is the need to educate the patient and/or caregiver about the importance of hygiene care and follow-up visits to keep the drain patent, and after its removal, the fistula, so that cystic decompression occurs effectively, and the technique is successful. The main complications associated with marsupialization are recurrent infections, especially due to poor hygiene, and early closure of the fistula, which requires re-intervention (Sabry et al., 2018). In the present clinical case, the patient and their caregivers were committed to hygiene care and follow-up visits for a period of 18 months, ensuring good progress in the case.

In dentistry, regardless of the type of lesion present, it is crucial to integrate local treatment, essential for improving the patient's quality of life, with their systemic conditions, achieved through a comprehensive clinical examination, refined knowledge, and a multidisciplinary approach (Tommasi, 1989). In the reported case, the patient not only presented with retained teeth, malocclusion, macroglossia, and intraosseous cystic lesions. He is an individual with a rare syndrome, still incurable, which typically brings significant systemic complications and may require modifications in the treatment plan. However, it is possible to treat these oral conditions, providing the patient with better oral health and survival conditions.

## Conclusion

For a correct diagnosis in dentistry, it is essential to consider the patient as a whole and, whenever necessary, resort to a multidisciplinary approach to institute appropriate and comprehensive treatment for the patient and promote health. MPS is a rare syndrome with a difficult diagnosis that causes significant systemic alterations. Therefore, it requires multidisciplinary and intensive patient care, including the need for a dentist on the team due to the pronounced maxillofacial involvement. In this article, the clinical case of a patient with the attenuated form of MPS I was described, yet still presenting multiple manifestations of the disease. Among these, two large dentigerous cysts in the mandible were successfully treated exclusively by marsupialization, without the need for a second intervention for enucleation.

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