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ARTICLE REVIEW

Diligence in patients with mucopolysaccharidosis

Diligencia en pacientes con mucopolisacaridosis

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ABSTRACT

Introduction: mucopolysaccharidosis type I or Hurler syndrome is a rare genetic metabolic disorder caused by deficiency of the enzyme alpha-L-iduronidase required to break down mucopolysaccharides. This deficiency leads to progressive accumulation of glycosaminoglycans in lysosomes, resulting in multisystem dysfunction.
Objective: to determine the behavior of mucopolysaccharidosis type I.
Methods: the methodology used is the systematic review based on components of the PRISMA 2020 method, following diligence guidelines in patients with MPS type I that involved the application of a rigorous approach in the identification and selection of relevant studies, extraction and synthesis of data in a methodological manner from digital platforms of scientific database.
Results: the review conducted according to the PRISMA 2020 methodology, highlights that since MPS type I is a rare, enzymatic, chronic and irreversible disease, it occurs in apparently healthy children whose family history may or may not have the condition. Treatment consists of two modalities: ERT with laronidase and hematopoietic cell transplantation (HCT) with limited benefits and potential risks, hence the need for ongoing evaluations and multidisciplinary patient follow-up.
Conclusions: these findings support the importance of timely diagnosis to avoid underdiagnosis, therapeutic intervention and nursing care to prevent serious complications and improve quality of life. As a rare disease, continued research is needed to improve the understanding and management of MPS.

Keywords: Mucopolysaccharidosis I; Systematic Review; Multidisciplinary Monitoring.

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RESUMEN

Introducción: la mucopolisacaridosis tipo I o Síndrome de Hurler es un trastorno metabólico genético poco común, causada por la deficiencia de la enzima alfa-L-iduronidasa necesaria para descomponer los mucopolisacáridos. Esta deficiencia conduce a la acumulación progresiva de glucosaminoglicanos en los lisosomas, lo que resulta en disfunción multisistémica.
Objetivo: determinar el comportamiento de la mucopolisacaridosis tipo I.
Métodos: La metodología utilizada es la revisión sistemática basada en componentes del método PRISMA 2020, siguiendo directrices de la diligencia en pacientes con MPS tipo I que implicó la aplicación de un enfoque riguroso en la identificación y selección de estudios pertinentes, la extracción y síntesis de datos de manera metodológica de plataformas digitales de base de datos científicos.
Resultados: la revisión realizada según la metodología PRISMA 2020, resalta que al ser la MPS tipo I una enfermedad rara, enzimática, crónica e irreversible, se presenta en niños aparentemente saludables cuyos antecedentes familiares pueden o no tener la condición. El tratamiento consiste en dos modalidades: TRE con laronidasa y el trasplante de células hematopoyéticas (TCH) con beneficios limitados y riesgos potenciales, de ahí la necesidad de evaluaciones permanentes y de un seguimiento multidisciplinario del paciente.
Conclusiones: Estos hallazgos respaldan la importancia del diagnóstico oportuno para evitar que sea subdiagnosticada, la intervención terapéutica y el cuidado de enfermería para prevenir complicaciones graves y mejorar la calidad de vida. Al ser una enfermedad rara es necesario investigaciones continuas para mejorar la comprensión y el manejo de la MPS.

Palabras clave: Mucopolisacaridosis Tipo I; Revisión Sistemática; Seguimiento Multidisciplinario.

INTRODUCTION

Mucopolysaccharidosis type I (MPS I) is a rare genetic disease caused by a lack of an enzyme called alpha-L-iduronidase. This deficiency causes a buildup of glycosaminoglycans (GAGs) in cells, leading to problems in several body systems.[1] People with MPS I do not produce enough of one of the 11 enzymes needed to break down these sugar chains because of mutations in the alpha-L-iduronidase gene (4p 16,3), resulting in either a complete lack of the MPS-1H enzyme or a partially functioning MPS 1-S enzyme. This causes dermatan sulfate (DS) and heparan sulfate (HS) to build up in the lysosomes of connective tissue cells such as cartilage and bone.[2]

Previous European research on the prevalence of different types of MPS revealed significant differences in various populations. The lowest incidence rate was reported in a Swedish study by Malm.[3] In Sweden with 1,75 cases per 100,000 births while the highest incidence was recorded in the Netherlands by Poorthuis et al.,[4] with 4,5 cases per 100,000 births. Despite these variations, the overall prevalence is estimated to be between 3,4 and 4,5 per 100,000 births.[5]

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MPS I in the United States is estimated to affect 1 in every 25,000 live births. The incidence of the disease varies by region and population; it is one of a group of 4,000 rare genetic disorders.[6] The literature reviewed indicates that MPS I is diagnosed through a blood test or a skin biopsy.[1] There is no cure, but early treatment with enzyme replacement therapy can help improve symptoms and quality of life.[7]It develops during early childhood, but in adulthood it develops more pronounced and characteristic forms such as short stature, blunt facial features, a short trunk, stiff joints in the hands, and walking on tiptoes.[8]

In Spain, Ureña,[9] It highlights the need to adapt care to the needs of patients with MPS during hospital stays, in order to stabilize the patients' condition, following the multiple complications associated with this degenerative and progressive disease for which there is little quality clinical evidence and even less specific nursing care.

In Latin America, Gomez,[5] It reports that MPS I is the most common type of MPS in an analysis of patients with suspected inborn errors of metabolism, equivalent to 25 % of all MPS cases; MPS VI is in second place, accounting for 18 % of all MPS cases; while in Colombia, the information available on this disease is scarce.

There are no epidemiological studies in the Ecuadorian population. According to Jimbo, in 2019, patients with MPS with a prevalence of type II were identified in Manabí; in Puyo and Ibarra, with type I; in Pastaza province, 19 children with MPS have been diagnosed who are not receiving enzyme replacement therapy. Laboratory testing is not performed in the country due to the lack of an enzyme-associated laboratory, in addition to a lack of qualified personnel to diagnose the disease and provide the necessary care for these patients.[6]

According to Suarez,[1] Different types of MPS have been identified, the clinical manifestations of which are related by mutations in specific genes encoding enzymes responsible for GAG degradation. Deficiency or dysfunction of these enzymes causes the accumulation of GAGs in cells and tissues, resulting in characteristic symptoms for each type of MPS, whose specific clinical features are related, such as developmental delay, bone abnormalities, joint stiffness, corneal opacity, coarse facies, enlarged liver, and heart disease.

There are also differences between the various types of MPS, as each type of enzyme deficiency determines the accumulation of a particular type of GAG. Furthermore, the literature reviewed indicates that some types of MPS are more severe and have a more rapid course, while others are milder and may progress more slowly. The most severe and rapidly progressing types of MPS are:

- **MPS I (Hurler Syndrome):** It is the most severe form of MPS. It is characterized by severely impaired physical and mental development; most children do not survive beyond adolescence.[5]
- **MPS II (Hunter Syndrome):** It mainly affects men, life expectancy is variable but is generally lower than normal.
- **MPS VI (Maroteaux-Lamy Syndrome):** According to Masia, the life expectancy of patients depends on the severity of the symptoms; without proper treatment, affected individuals survive until late childhood or adolescence.[10]

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The mildest and slowest progressing MPS:

- **MPS III (Sanfilippo Syndrome):** There are four subtypes with varying degrees of severity. They are characterized by progressive cognitive decline, behavioral problems, and intellectual disability. Life expectancy varies depending on the subtype but generally reaches adolescence and rarely adulthood.[11]
- **MPS IV (Morquio Syndrome):** It has two subtypes and the life expectancy of affected individuals is 40 years.
- **MPS VII (SLY Syndrome):** Characterized by heart problems, accumulation of GAGs in cells, and intellectual disability, life expectancy depends on the symptoms; some affected individuals may live into adolescence and even adulthood.[1]

The factors that influence disease severity and course vary depending on the type of MPS. As mentioned above, some MPS are inherently more serious than others. Added to this is the risk of enzyme deficiency. Early diagnosis and access to timely treatment, enzyme replacement therapy (ERT), and other treatments can improve quality of life and life expectancy. Another very important factor is family support and access to quality medical care, which can influence the course of the disease.[12]

Nursing care for MPS type 1 includes key aspects for providing comprehensive care to patients affected by this genetic disease. Specific care measures include:

- Design of a specific care plan that covers the biopsychosocial needs of the pediatric patient.
- Knowledge of the disease and available treatment to provide appropriate patient care.
- Provide specific and up-to-date care for the altered needs of patients.
- Improve care for caregivers to provide discharge recommendations that facilitate the continued care of the patient, collaborating between specialized care and primary care to improve the quality of life of the patient and their environment.[13]
- Interdisciplinary collaboration is essential to provide a coordinated multidisciplinary approach, with nursing staff playing a crucial role in providing care, information, and emotional support to parents throughout the process.

These nursing care options aim to ensure comprehensive and specialized care for patients with MPS type 1, contribute to improving quality of life, and effectively manage the complications associated with this genetic disease. Based on the above, the following question arises: What is the behavior of mucopolysaccharidosis type 1?

Aim: Determine the behavior of mucopolysaccharidosis type I

METHODS

The PRISMA methodology is used

1. Inclusion and Exclusion Criteria

Inclusion criteria include:

- Documents submitted between the period 2015-2023
- Articles with access to their summary or the entire content.
- Documents written in Spanish or English
- Documents focused on diagnosis and treatment
- The type of documents included are care guides and case studies.

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Exclusion criteria

- Studies that do not allow access to their summary and content.
- Articles published outside the 2015-2023 period.
- Non-Nursing Items

2. Selection of studies

- Review of titles, abstracts and full texts to identify relevant studies.

DEVELOPMENT

PRISMA 2020 Methods

1. **Review protocol**: A detailed table was developed describing objectives, inclusion criteria, search strategies and analysis methods.
2. **Data extraction**: of relevant information from each study, selecting the sources of information, sample, synthesis of results, limitations of evidence.
3. **Quality assessment**: use risk and bias methods from individual studies.
4. **Analysis and Synthesis:** perform a systematic analysis of the extracted data and qualitatively synthesize the results in a coherent manner.
5. **Final report:** Write a detailed report including introduction, methods, results, discussion, and conclusions

The raw method used in the research is shown in theFlowchart figure 1

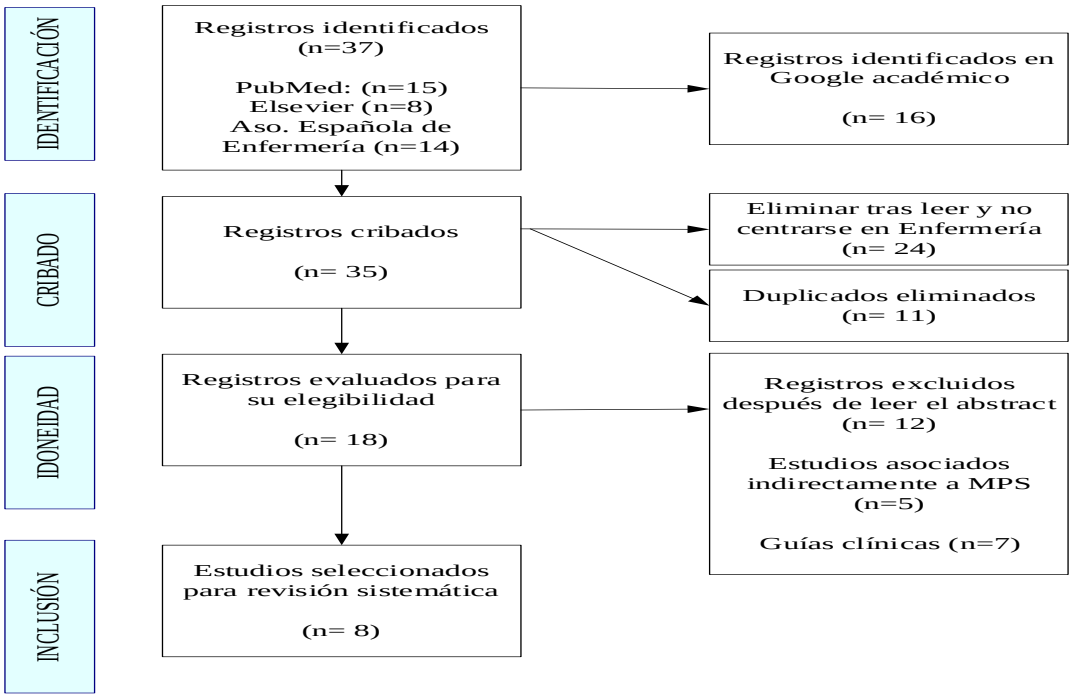

Fig. 1 Flowchart according to PRISMA 2020.

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Description of the articles used for the research:

1. E CareNursing in the multidisciplinary care of pediatric mucopolysaccharidosis: a clinical case report.[14]

Aim: Improving the quality of life of a child with mucopolysaccharidosis
Source of Information: Health Research Journal
Risk and bias of individual studies: Selection
Methodology: Case study, qualitative
Sample: 11-year-old boy with Hunter syndrome or mucopolysaccharidosis type II
Summary of results: The visit is used to identify the needs of the patient and family and to provide emotional support.
Limitations of the evidence: Administration of home treatment of weekly medication.
Implication: Disease management is through the care of a multidisciplinary team.

2. Mobile application to guide treatment in patients with Mucopolysaccharidosis Type I.[6]

Aim: Propose a guide for the application of treatment in patients with mucopolysaccharidosis type I implemented in an App
Source of Information: Dialnet
Risk and bias of individual studies: of selection
Methodology: Bibliographic review
Sample: 18 cases without treatment.
Summary of results: The APK is a more specific guideline for this disease that provides comprehensive care when applying enzyme replacement therapy.
Limitations of the evidence: This type of guidance cannot be implemented in pediatric patients.
Implication:It consists of applying the care plan and discharge recommendations in clinical practice, adapting them to the individual characteristics of each patient and their family, and coordinating care with the multidisciplinary team.

3. Nursing Care Plan and Discharge Recommendations for Pediatric Patients with Mucopolysaccharidosis.[13]

Aim:Propose a care plan and recommendations upon discharge for pediatric patients with Mucopolysaccharidosis that covers their biopsychosocial care needs.
Source of Information: University of Valladolid
Risk and bias of individual studies: Of realization
Methodology: Bibliographic review
Sample: 20 interviews with adolescents and health personnel.
Summary of results: The care plan is individualized and tailored to each patient's unique characteristics, taking into account the specific type of mucopolysaccharidosis, age, overall health, and specific needs.
Limitations of the evidence: The care needs of pediatric patients with mucopolysaccharidosis can be highly variable.
Implication: Apply a care plan and recommendations upon discharge in clinical practice, adapting them to the patient's characteristics.

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4. Nursing in mucopolysaccharidosis: More than just hospital treatment.[15]

Aim: To know the perception of the patients treated regarding nursing care during hospital admission for the infusion of treatment
Source of Information: Spanish Association of Nursing in Neurosciences
Risk and bias of individual studies: of generalization
Methodology: Case study
Sample: seven nurses and three nursing assistants
Summary of results: Patients positively value the nursing care they receive, highlighting the professionalism, humanization, information and health education.
Limitations of the evidence: The care needs of pediatric patients with mucopolysaccharidosis can be highly variable.
Implication: Disseminate the study results among healthcare professionals, patient associations, and health authorities to promote improved quality of care and raise awareness about these rare diseases.

5. Mucopolysaccharidosis type I. Report of a case**.**[16]

Aim: To present the case of a 10-year-old boy with Hurler Syndrome or MPS type I
Source of Information: Argentine Journal of Orthopedics and Traumatology
Risk and bias of individual studies: Observational
Methodology: Case study
Sample: a patient
Summary of results: Multidisciplinary monitoring of the patient is imperative, as is knowledge of the risk of spinal cord compression and its timely surgical treatment.
Limitations of the evidence: High anesthetic risk, myelopathy, and progressive symptoms indicate surgical decompression**.**
Implication: disseminate the case to promote awareness, early diagnosis, and access to treatment for MPS that require comprehensive care.

6. Nursing care plan for a pediatric patient with mucopolysaccharidosis**.**[17]

Aim: Perform a nursing care process in a pediatric patient diagnosed with mucopolysaccharidosis to control their alterations.
Source of Information: Google Scholar
Risk and bias of individual studies: Observation and interpretation
Methodology: Evidence-based nursing
Sample: a patient
Summary of results: Individualized nursing care was used
Limitations of the evidence: Given the debilitating nature of the symptoms and the complexity of the disease, pediatric patients with MPS require comprehensive care that encompasses both the patient and their family environment.
Implication: Implement a care plan in clinical practice, adapting it to the individual characteristics of each patient and family to achieve coordinated care by the multidisciplinary team.

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7. Experiences of Parents of Children with Mucopolysaccharidosis in Türkiye: A Qualitative Study.[18]

Aim: Examine parents' experiences regarding their child's illness process
Source of Information: Elsevier
Risk and bias of individual studies: of selection
Methodology: Qualitative study
Sample: 10 parents who had a child with MPS for six months
Summary of results: This study provides evidence on parents' experiences related to psychosocial effects, difficulties and needs, and coping resources. Living with MPS is almost equivalent to uncertainty for families.
Limitations of the evidence: Parents faced challenges and needs related to caring for their child, accessing treatment, information about the disease, emotional support, and adapting to changes.
Implication: The study provides evidence on parents' experiences related to MPS and suggests implications for comprehensive nursing practice that is planned according to the needs of children with MPS.

8. Clinical case: Hunter syndrome (mucopolysaccharidosis type II).[19]

Aim: To present the clinical case of a patient with Hunter syndrome, a rare genetic disease that primarily affects men, and to describe his clinical course, his treatment with enzyme replacement therapy, and his nursing assessment.
Source of Information: Health Research Journal
Risk and bias of individual studies: of wear
Methodology: case study
Sample: a patient
Summary of results: Hunter Syndrome is a rare disease that requires early diagnosis and appropriate treatment to prevent and delay complications.
Limitations of the evidence: It is based on a single case, which makes it difficult to generalize the results to other patients with the same disease. The study lacks a control group or a randomized design, which reduces its internal and external validity.
Implication: Disseminate the case among health professionals, authorities, and associations to promote early diagnosis and access to treatment for MPS.

Results of the systematic review on diligence in patients with mucopolysaccharidosis, it is necessary to point out that being a rare, enzymatic, chronic, irreversible disease,[6]that is presented by an apparently healthy child, whose family history may or may not have the condition,[14]There are few nursing studies that address nursing care, the inclusion criteria were expanded to include works from 2015 to 2023 in order to cover the largest number of relevant studies on this topic;To facilitate the understanding and integration of the results, a detailed analysis of the studies selected for systematic review is carried out below, most of them case studies and bibliographic reviews.

The literature reviewed and included in the systematic review indicates that the diagnosis of MPS I is based on clinical suspicion, determination of enzyme activity, measurement of GAGs in urine, and genetic study of the IDUA gene.[1]Treatment consists of two modalities with specific indications: ERT with laronidase and hematopoietic stem cell transplantation (HCT); both therapies have limited benefits and potential risks, hence the need for multidisciplinary evaluation and follow-up of patients.

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The studies reviewed systematically agree on the need for a timely diagnosis to initiate treatment, enabling a multidisciplinary and comprehensive approach to improving the quality of life of patients with MPS and their families.[19]

The reviewed studies emphasize the need for multidisciplinary nursing work in caring for patients with MPS,[15]administering enzyme replacement therapy, monitoring patient needs, providing emotional support and a care plan based on NANDA diagnoses, NIC interventions, and NOC outcomes, focusing on managing diarrhea, risk of fluid volume deficit, impaired skin integrity, and risk of infection.[13]

Another aspect that should be noted is the classification and genetics of MPS, which are classified into seven types according to the deficient enzyme and the affected gene Guerrero.[14] MPS are inherited in an autosomal recessive manner, except for type II or Hunter syndrome, which is X-linked. MPS are characterized by the accumulation of glycosaminoglycans (GAGs) in tissues, resulting in multisystemic alterations of varying severity; the most common symptoms are coarse facial features, macrocephaly, hypertrichosis, osteoarticular alterations, visceromegaly, and neurological involvement. Diagnosis is based on the detection of GAGs in urine, measurement of enzyme activity, and genetic analysis.[20]

Regarding the treatment and management of MPS, it includes symptomatic and supportive treatment, enzyme replacement therapy, hematopoietic progenitor cell transplantation, substrate reduction, and gene therapy.[1] The management of MPS requires multidisciplinary and personalized care,[18,19,20.21]which includes education and support for families, home and palliative care, and periodic monitoring of complications.

In summary, the studies subjected to systematic review capture the experiences of patients and healthcare professionals in addressing MPS, as well as recommendations for improving quality of life, access to diagnosis and treatment, and coordination between different levels of care.[17]

The reviewed results provide a detailed overview of the different variants of MPS. Their classification is crucial for early diagnosis and appropriate selection of specific treatments. Regular follow-up is therefore essential to assess disease progression and tailor clinical and care management to the patient's specific needs.[6,14,19]

Guerrero,[14] García,[15] Besse,[16] and Roncales,[19] in their case study work, agree on the need to disseminate the results of the study among health professionals and health authorities in order to promote improved quality of care and raise awareness about these rare and serious diseases, which require comprehensive and multidisciplinary care. They also emphasize the need for early diagnosis to access appropriate and timely treatment for MPS.

In the bibliographic reviews carried out by Jimbo,[6] It is based on technological applications that contribute to adherence to MPS treatment, with the limitation of not being able to implement it in pediatric patients; while Ureña addresses the main clinical, diagnostic and therapeutic characteristics of MPS and with the help of a care plan based on the Virginia Henderson model and the NANDA-NIC-NOC taxonomy addresses the main alterations and problems of patients during hospitalization that the nurse must be aware of when providing care in order to support both the patient and the family.[13]

These studies offer a broad overview of the technological and technical tools available to patients and nursing staff that contribute to the management of patients with MPS.

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Arpaci, for its part, determined among its results the social and emotional impact on parents and relatives of children with MPS, pointing out the discrimination they feel and the lack of social and health support in these cases;[18]These effects are embedded in the reviewed results because a large part of the considerations made by nursing staff as part of the care of patients with MPS is to offer emotional support and information about the disease to family members and parents of patients with MPS.[6,13,19]

CONCLUSION

The incidence of MPS worldwide varies according to the specific type. As a rare genetic disease, MPS presents specific clinical characteristics and manifestations that affect different body systems due to the accumulation of mucopolysaccharides. Diligence and timely treatment in monitoring patients with this disease are crucial to improve their quality of life and prevent serious complications. The importance of a multidisciplinary approach that includes specialized physicians, occupational and physical therapists, as well as regular follow-up and timely access to specific treatments for this disease, could be emphasized.

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