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Recommendations for evaluation and management of pain in patients with mucopolysaccharidosis in Latin America

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Summary

The mucopolysaccharidosis (MPS) constitute a heterogeneous group of rare genetic disorders caused by enzymatic deficiencies that lead to the accumulation of glycosaminoglycans (GAGs). Several types of MPS are described, historically numbered from I to IX. Clinical observations strongly suggest the presence of chronic pain in patients with all types of MPS. There are few data in the literature on the evaluation and management of pain in these patients, a fact which can compromise the quality of life even more. Professionals with extensive experience in the care of patients with MPS held a meeting in April 2017 to discuss and propose recommendations for the evaluation and management of pain in patients with MPS in Latin America. This article summarizes the content of the discussions and presents the recommendations produced at the meeting. Patients with MPS present joint, bone, and muscle pain, as well as entrapment syndromes (spinal, optic nerve, carpal tunnel). The panel suggests the use of the following instruments for pain assessment: Face, Legs, Activity, Cry and Consolability (FLACC) scale for children of up to 4 years of age and patients unable to communicate their pain; Child Health Assessment Questionnaire (CHAQ) scale, Facial Pain Scale and Numerical Pain Scale (FPS; NPS) for patients of 5 to <18 years of age; Brief Pain Inventory (BPI) and Short Form Health Survey 36 (SF36) scales for patients of 18 years of age or older. Based on the scores verified in these scales, the panel proposes pharmacological interventions for pain relief in this population of patients.

Abbreviations:

- FLACC: Face, Legs, Activity, Cry and Consolability;
- CHAQ: Child Health Assessment Questionnaire;
- FPS: Facial Pain Scale;
- NPS: Numerical Pain Scale
- BPI: Brief Pain Inventory
- SF36: Short Form Health Survey 36

Introduction

The mucopolysaccharidosis (MPS) are a group of inborn errors of metabolism (IEM) caused by specific lysosomal enzymes deficiencies that affect the degradation of glycosaminoglycans
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