

Rehabilitation nursing for people with Machado Joseph's disease: scoping review

Teresa Ferreira¹

 orcid.org/0000-0002-6170-9975

Cristina Gomes²

 orcid.org/0000-0002-6896-8654

Hugo Gomes³

 orcid.org/0000-0003-3124-0480

Hélder Teixeira⁴

 orcid.org/0000-0001-9086-4985

Jacinta Martins⁵

 orcid.org/0000-0001-8052-571X

Andreia Félix⁶

 orcid.org/0000-0001-7740-4124

¹Enfermeiro; Hospital de Braga.

²Enfermeiro; Hospital Senhora da Oliveira.

³Enfermeiro; Hospital Senhora da Oliveira.

⁴Enfermeiro; Hospital de Braga.

⁵Professor Adjunto; ESSCVP – Alto Tâmega.

⁶Professor Adjunto; ESSCVP – Alto Tâmega.

Abstract

Introduction

Machado-Joseph Disease (MJD), also called spinocerebellar ataxia type 3, is a neurodegenerative disease that follows an autosomal dominant pattern of inheritance.^{1,2} In this sense, it can affect different regions and functions of the central or peripheral nervous system, namely the areas of motor control, phonarticulation and swallowing, being manifested by changes in gait, body posture, orofacial motricity, swallowing, oculomotor system and limb coordination which maintain the cognitive function.^{1,3} In this sense, the rehabilitation intervention, with the objective of optimising functionality, reducing disability and promoting positive adaptation, plays a key role in improving the quality of life and reducing the impact of the disease, on the person, their family and caregivers.

Objective

To map the literature regarding the process of rehabilitation of the person with Machado Joseph Disease (MJD), particularly the rehabilitation nursing interventions applied to the person with this disease.

Method

Scoping review based on the principles recommended by the Joanna Briggs Institute – ScR and PRISMA-ScR, using the Medline CINAHL, Cochrane Central Register of Controlled Trials databases; Nursing and Allied Health Collection and MedicLatina, from the EBSCOhost Integrated Search platform, Scopus, LILACS and Scielo. The Participants, Concept and Context (PCC) strategy was used, in which studies with people with MJD were included. Regarding the concept, they addressed rehabilitation interventions and all intervention contexts were considered. Paired reviewers analyzed the relevance of the studies, extraction and synthesis of data.

Results

Ten studies were included in the review. Several rehabilitation interventions are reported, namely training exercises in gait, balance, strength and resistance, daily life activities, muscle toning and proprioceptive exercises.^{4,5} The results of rehabilitation interventions demonstrate their effectiveness, through evidence of improved mobility and balance, improved speed and gait condition, increased motor coordination, decreased response time to tasks and reduced risk of falling.

Discussion

Most studies show results with improved functional independence, gait, balance, and motor coordination, resulting in an improvement in quality of life, functional independence and a reduction in the risk of falling. The results presented suggest interventions and rehabilitation programmes were based on most motor limitations, but none focused on phonarticulation and orofacial motricity. It is also important to emphasize that few studies have attributed relevance to the person's ability to deal with disease progression and symptoms. Conclusion: Although the demonstrated results are in line with the aim of this ScR, there is a need for further experimental studies so more evidence is created on the impact of the

Corresponding Author:

Hélder Teixeira

E-mail: helder.teixeira@hb.min-saude.pt



aforementioned interventions with MJD.

Keywords

Rehabilitation Nursing; Machado-Joseph Disease; Spinocerebellar Ataxias.

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