

Joint Hypermobility Syndrome/Ehlers-Danlos Syndrome Hypermobility Type: a revision of the rehabilitative approach

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Abstract

Joint hypermobility syndrome (JHS) and Ehlers-Danlos syndrome, hypermobility type (EDS-HT) are two clinically overlapping heritable connective tissue disorders strongly associating with pain, fatigue and other secondary aspects. No specific treatment exist for this syndrome and rehabilitation play a role in the management of these patients. The aim of this paper is to evaluate what are the evidence in literature about rehabilitation. Research was done using database PUBMED and consist in a revision of the studies published in the last 15 years. All studies agree to the beneficial role of the rehabilitative treatment and physical therapy but it's necessary to add more further studies to establish a high quality, evidence-based physical therapy for this specific population.

Keywords: Ehlers-Danlos Syndrome, Exercise, Joint Hypermobility, Rehabilitation, Treatment.

Introduction

Joint hypermobility syndrome (JHS) is a common but largely unrecognized condition mainly characterized by joint hypermobility, joint instability, chronic pain and fatigue [1].

Recently, an international group of experts considers the JHS as clinically undistinguishable from the Ehlers-Danlos syndrome, hypermobility type (EDS-HT) that is likely one of the most common variants of EDS forms [2].

These two clinically overlapping heritable connective tissue disorders are strongly associated with pain, fatigue and other secondary aspects. Actually the JHS/EDS-HT is assessed by distinct diagnostic criteria, namely the

Villefranche [3] and Brighton [4] criteria, and still lack confirmatory instrumental/molecular tests [5]. Hence, their recognition still depends on the observer's experience, which is often hampered by the actual lack of an international consensus for the evaluation of joint hypermobility by the Beighton score [6] and for the application of available diagnostic criteria [7, 8].

The main approach able to reduce and to manage symptoms is the rehabilitative treatment; although physical therapy for people with JHS has been recommended by many authors, there are no published reports regarding the efficacy of physical therapy or any other treatment [9].

The object of this revision is to search what is present in the literature concerning the rehabilitative management of patients with JHS/EDS-HT.

Materials and methods

The primary search using the database PUBMED consist in a revision of the studies published in the last 15 years using the keywords: Hypermobility Syndrome, Joint hypermobility syndrome, Rehabilitation, Treatment, Management, Physiotherapy. Studied were included if they were full publication of randomized or non-randomized controlled trial (RCT), observational and cross sectional studies evaluating a rehabilitation program. Also review studies has been evaluated.

Studies including individuals with generic hypermobility, also associated with other connective tissue disorders, but not having a “syndrome” were excluded.

Data were independently extracted by one reviewer (FC) and verified by a second (CC) one. Main data evaluated were: age, gender, duration of symptoms, Beighton assessment, study intervention, period of follow up and clinical and functional outcome measures.

Results

A total of 13 manuscript has been selected; four has been excluded because were case reports and one because describe only professional sportsmen. Among the eight studies analyzed two are referred to pediatric approach and one about specific manipulative treatment. The results are illustrated in **Table 1**.

All the study agree to the beneficial role of the rehabilitative treatment and physical therapy is defined the mainstay of treatment [9, 10,11]; they also add the necessity to exactly decide the correct types of exercise. Regard to exercise, strengthening exercise [9] at low dose (11) and possibly in a close kinetic chain [12] may be helpful to improve muscular stability at specific joint. In particular improve trunk stability is the starting point of the rehabilitation program; improve postural stability muscles improve the postural strategy and supports the spine. Exercise needs to be associated to education [9] about ergonomic and body mechanism that is probably the most important treatment that physical therapist can provide; the association with cognitive-behavioral approach seems to be useful [13]; patients with JHS/EDS-HT have to avoid harmful postures and activities which can overstretch and strain hypermobility joints [10] but simultaneously they may be able to continue their activities (job, sports, recreation activities) at a decreased frequency or intensity [9] or even more

they may be encouraged to develop a lifelong commitment to physical activity [10].

All authors agree with an exercise program who enhance proprioception and improve stability [9,10,11,14]. There is no agreement on using splint or bracing [9, 10].

No enough evidences are present for the adequate treatment approach for infants with JHM and hypotonia [15] while in children exercises to core support stabilization, muscular tone and lax joint stabilization are indicated associated to graded exercises and increase in activity [16].

The review about management of JHS showed the useful role of the Osteopathic Manipulative treatment as an adjunctive one along with the exercise therapy. This treatment may help to reduce pain as well as improve flow and lymphatic drainage [17].

Discussion

Hypermobility syndrome is a complex, under recognized and poorly managed inherited connective tissue disorder often resulting in a great deal of pain and suffering. The management of individuals with JHS/EDS-HT can be very challenging [18].

The findings of this revision showed the importance of the rehabilitative approach in the JHS/EDS-HT management as the mainstay of treatment. It is as many evident that different types of rehabilitation programs has been used and there are not enough trials able to prove the better treatment. Furth more different phases of the syndrome has been described [19] and different level of disability characterized these patients. We think that it should be important to identify different classes of disability among the all group of patients with JHS/EDS-HT; the rehabilitation program maybe differentiated into prevention, management of the acute phases and better taking charge in the chronic phases. Also if every program of treatment should be identify on the single patient, the general guidelines are necessary [20] and moreover is absolutely considerable to know what should be not done in these patients. This is probably the topic less present in literature. No study of comparison are also present to evaluate the efficacy of physical therapies like electrotherapy, TENS, Taping application for which there are only favorable opinions. All the studies agree with the necessity of further studies to establish a high quality, evidence-based physical therapy for this specific population.

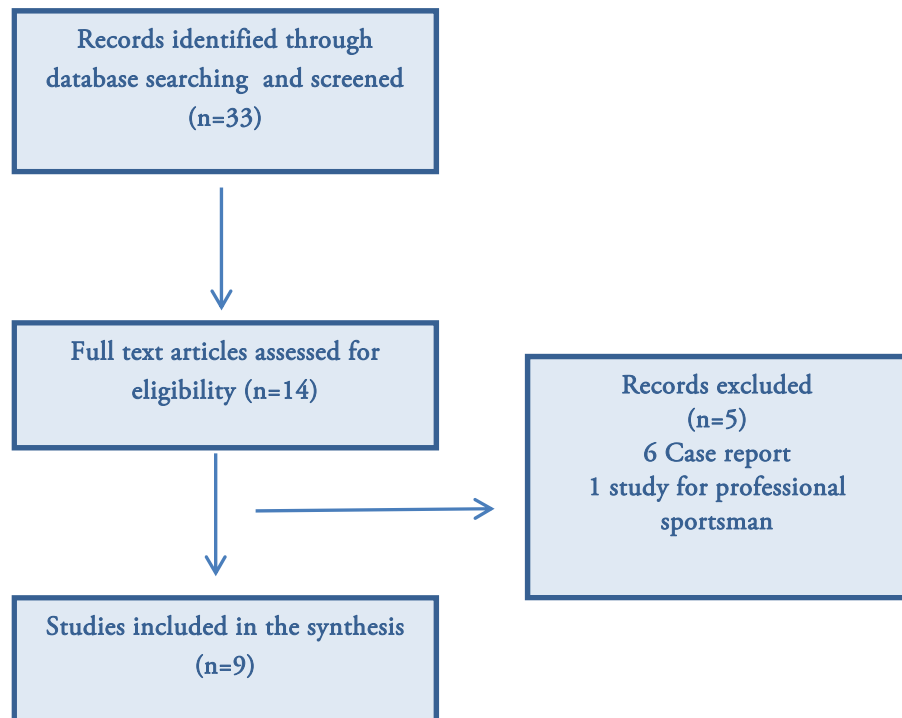
Conclusion

The role of a rehabilitation program in patients with JHS/EDS-HT is of paramount relevance and for this reason further high quality multi center trials are required.

Table 1

Author/ Year	Study design	Purpose	Setting	Subjects	Treatments
Russek LN,1999	Review	Increase awareness, understanding and discussion of HMS	Department of Physical Therapy	HMS pz	-Strengthening and proprioceptive exercises -Stretching
Ferrell,2004	Cross-sectional study	Investigate whether a home-based exercise program could produce enhancement of proprioception as well as alleviate symptoms.	Hypermobility clinic	20 pz with JHS	-Closed kinetic chain exercises -Static hamstring exercises
Simpson, 2006	Review	Looking at history, clinical presentation, diagnosis and treatment and the role of Osteopathic Manipulative Treatment in patients with hypermobility and chronic pain.	US Army Health Clinic	BJHS pz	-Strength training: closed kinetic chain exercises and open kinetic chain exercises - Proprioception exercises -Osteopathic Manipulative Treatment
Sahin, 2008		-Show if there is a disorder in proprioception in BJHS -Evaluate the proprioceptive exercise in BJHS	Physical Medicine and Rehabilitation department	40 pz with BJHS and 30 healthy individuals	-Proprioception exercises -Kinesthesia and exercise of balance
Mintz-Itkin 2009	Prospective Longitudinal Study	Discuss the effect of the frequency of physical therapy on outcome in infants referred for delayed	Child development Center	29 infants (8-12 months)	Neurodevelopment therapy approach Home program for 20 minute daily
Kemp,2010	RCT	Determine the best physiotherapy intervention in managing childhood hypermobility	Department of Pediatric Rheumatology	Fifty seven children aged 7-16	-General program: general muscular strengthening exercises and aerobic fitness -Targeted program: Exercises of motion control and proprioception
Keer R, 2011	Review	Current knowledge on the management of joint problems associated with hypermobility syndrome	Central London Physiotherapy Clinic, UK		Proprioception Closed kinetic chain exercise and dynamic balance activities
Rombaut L.,2011	Cross-sectional study	Describe medication use, surgery, and physiotherapy and effects of these treatment	Physical and rehabilitation medicine department and center for medical genetics.	Patients with HMS (N=79 ,8 men, 71 woman)	-Strengthening exercises - Joint stabilization exercises -Proprioceptive exercises -Massage
Bathen T 2013	Pilot study	To investigate the role of a multidisciplinary rehabilitation program combining physical and cognitive-behavioral therapy		12 women with EDS-HT/JHS	-Cognitive-behavioral approach -Structured exercise program to perform at home

PRISMA Flowchart presenting the results of the search strategy



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