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Generalized Joint Hypermobility: A Review

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ABSTRACT

Generalized Joint Hypermobility (GJH) is a condition where most of an individual's synovial joints are found to have a range of motion (ROM) beyond their normal limits. Like most human traits, joint hypermobility is multifactorial resulting from a combination of environmental factors (eg. age, trauma, injury, conditioning, infection, inflammation) and multiple genetic factors, each contributing a small amount to the total phenotype. Clinical features of JHS are mainly associated to the connective tissue and can either be articular or extra-articular. The Beighton 9-point scoring system – also referred to as the modified or revised Beighton score – is a widely accepted method used to define GJH. Management of Generalized Joint Hypermobility is multidisciplinary which requires a team of physicians, physiotherapists, occupational therapists and podiatrists, among others. Physical therapy rehabilitation comprising of core stabilizing, joint stabilizing, strengthening and proprioception enhancing exercise coupled with general fitness program form the basis of GJH management.

Key words: Generalized Joint Hypermobility, Beighton Score, Joint Hypermobility

INTRODUCTION

Generalized Joint Hypermobility (GJH) is a condition where most of an individual's synovial joints are found to have a range of motion (ROM) beyond their normal limits.[1] A hypermobile joint exceeds the normal for that individual, taking into consideration age, sex and ethnicity.[2] The maximal range of movement that a joint can move is determined by the tightness or otherwise of the restraining ligaments. Thus, the primary cause that leads to hypermobility is ligamentous laxity. This is determined by their fibrous protein genes and is inherent in a person's make up.[2]

The term generalised joint hypermobility (GJH) is reserved for use when multiple joints are involved and a particular threshold for defining GJH is reached.[3] If only a small number of joints is hypermobile, i.e., the Beighton score is <5 for adult men and women up to the age of 50, the condition may be called localized joint hypermobility (LJH). LJH usually affects one or two smaller or larger joints only, and may be bilateral, such as in bilateral genu recurvatum.[63] GJH is most often assessed by the Beighton score, which assesses hypermobility at nine joints.[4-6] Joint hypermobility syndrome (JHS) is a hereditary, systemic connective tissue disorder associated with GJH and widespread joint pain.[7-9] JHS is also known as hypermobility syndrome (HMS)[8] and benign joint hypermobility syndrome (BJHS).[5,7,10] The term "benign" is used so as to distinguish JHS from more serious conditions like Ehler-Danlos syndrome (classical or vascular types), Marfan syndrome, and osteogenesis imperfecta that present with joint hypermobility too and which should be ruled out in the diagnosis of JHS.[11-13]

GJH is more often a congenital, possibly an inherited trait, unlike LJH. Acquired forms of GJH also exist which

include widespread inflammatory or degenerative diseases of the joints, musculoskeletal tissues and nerves, hypothyroidism and other endocrine disorders.[14]

CAUSES-

Like most human traits, joint hypermobility is multifactorial resulting from a combination of environmental factors (eg. age, trauma, injury, conditioning, infection, inflammation) and multiple genetic factors, each contributing a small amount to the total phenotype.[15]

Genetic Factors:

The role of genetics in the development of joint hypermobility syndrome is evolving and controversial with the rapid adoption of genetic testing in research and clinical settings.[17] The vast majority of cases appear not to be linked to any identifiable mutation, although pedigree studies have shown a weak autosomal dominant inheritance pattern with variable penetrance.[18] In fewer than 10% of cases, a mutation in the gene TNXB, coding for the extracellular matrix glycoprotein Tenascin X, is identified. These patients tend to have more pronounced dermatologic signs, including easy bruising, skin hyperextensibility and velvety skin.[19]

In addition to the genetic predisposition due to presumed difference in collagen structure, certain theories emphasize the importance of localized biomechanical overloading and chronic soft tissue injury due to joint laxity and instability.[17]

Environmental factors:

Several studies found a significant decrease in hypermobility with increasing age, which corresponded well even with those of earlier observations. The reduction in joint mobility with advancing age is attributed to increased intra-fibrillar cross-links in collagen, the collagen in flexor tendons and volar plates getting stiffer in the elderly and also because of progressive biochemical changes in collagen structures which result in stiffening of connective tissue components of joints.[16,20,21]

Joint hypermobility sometimes can be developed, for example by gymnasts and athletes, through the training exercises they do. Yoga can also make the joints more supple by stretching the muscles.[23]

Some studies reported a high prevalence of GJH for certain disciplines such as dancing and ballet[22-24] possibly due to the great requirements of flexibility in these activities.[25]

OUTCOME MEASURES-

The Hospital del Mar criteria have been shown to be valid tool in the diagnosis of JH. Hospital Del Mar criteria consists of one point/item (overall range zero to ten) including evaluation of apposition of the thumb, metacarpophalangeal, elbow, external shoulder rotation, hip abduction, rotular (patella) hypermobility, ankle and feet hypermobility, metatarsophalangeal, knee hyperflexion and ecchymoses.[26]

Contompasis score is a semi-quantitative modification of the Beighton score and provides a more refined grading of joint hypermobility. It assesses each of the nine Beighton criteria, as well as hindfoot eversion at the calcaneus. Instead of providing a single positive or negative response, it gives a graded response between two and eight points for each criterion. A Contompasis score of greater than 20 is considered as indicative of GJH. However, it is more time consuming and may exhibit greater measurement variability, increasing the potential for error.[27]

The Lower Limb Assessment Score (LLAS) may be a useful score for health professionals specifically interested in lower limb hypermobility. It comprises of 12 bilateral tests of mobility of the hip, knee, tibiofibular, ankle and foot joints in all 3 planes of motion. The cut-off score for the LLAS has been established as $\geq$7/12 unilaterally, in both adults and children.[28]

The 12-item Upper Limb Hypermobility Assessment Tool (ULHAT) is a reliable and valid tool for identifying upper limb hypermobility and generalized joint hypermobility. It was designed as an upper limb analogue of the 12 composite tests of the LLAS and measures mobility of multiple upper limb joints in all movement planes. A cut-off of ≥7/12 unilaterally is established.[29]

BEIGHTON CRITERIA-

The Beighton 9-point scoring system – also referred to as the modified or revised Beighton score – is a widely accepted method used to define GJH.[12,16] It consists of a series of nine dichotomous joint extensibility tests, where a joint is either tested as hypermobile (score = 1) or not hypermobile (score = 0). Therefore, the total score (Beighton score) lies between 0 and 9, with higher scores indicating greater joint laxity.[30]

In the past, a Beighton score of ≥4 was often used to indicate GJH in adults, although certain studies used cut-off scores of ≥3, ≥5 or ≥6.[16,26,31,32] Because joint laxity is found to be greatest in infants and then gradually decreases during childhood and adolescence, a higher threshold was advocated for use in children. In 2017, the International Consortium on the Ehlers-Danlos syndromes (EDSs) proposed the use of following cut-off Beighton scores for the diagnosis of GJH: ≥6 for pre-pubertal children and adolescents, ≥5 for pubertal men and women up to the age of 50, and ≥4 for those >50 years of age.[33]

Many individuals with GJH continue to remain asymptomatic throughout their lives.[20] Sometimes they even take advantage of their hypermobile condition to excel in sports such as ballet[34] or dancing[24]. But, they also may be at an increased risk for musculoskeletal injuries, for example, sports-related injuries to the ankle, knee and shoulder joints.[35,36,37]

ASSOCIATED RISKS-

Clinical features of JHS are mainly associated to the connective tissue and can either be articular or extra-articular.[38]

The most common musculoskeletal manifestation is chronic and generalized pain.[9]Joint pain in JHS is thought to be caused by excessive movement which increase stress on joint surfaces, ligaments and neighbouring structures.[39] Pain causes muscle inhibition, leading to atrophy and reduced joint control.[40] Proprioceptive acuity is adversely affected perhaps due to joint mechanoreceptor damage.[43] The inability to acknowledge extreme joint ranges creates an even more unstable joint by further stretching supporting structures.

A wide spectrum of extra-articular clinical manifestations has been recognized in association with musculoskeletal symptoms[45] such as predisposition to ecchymosis, poor wound healing, early onset of osteoarthritis, valvulopathy, vesicoureteral reflux, inguinal hernia, an increased incidence of cardiac complications (Mitral Valve Prolapse, high elasticity of the aortic wall)[38] and changes in intestinal motility.[2] It is suggested that a subsequent natural history of hypermobility leads to traumatic synovitis and later to osteoarthrosis, normally in the fourth or fifth decades.[20]

There are also other recognized manifestations, such as fatigue, anxiety, and depression, negatively affecting social function and well-beingthereby having a substantial impact on individuals.[44]

There is evidence that hypermobility syndrome is multisystemic, incorporating three main components: chronic pain, autonomic dysfunction, and dysfunction of gastrointestinal motility.[2]

PHYSIOTHERAPY MANAGEMENT-

Management is multidisciplinary which requires a team of physicians, physiotherapists, occupational therapists and podiatrists, among others. Patients often face a delay before receiving their definitive diagnosis and are sometimes dismissed as malingerers.[5]

Acute pain exacerbation may be managed using taping, bracing or splinting.[40] However, education[32] and therapeutic exercise[46] remain the mainstays of long term management. Encouragement of an active lifestyle may improve function and enhance quality of life.[9] Available evidence suggests that patients who receive exercise intervention improve over time.[47]

Physical rehabilitation comprises of core stabilizing, joint stabilizing and proprioception enhancing exercise coupled with general fitness program.[48]. A randomized controlled trial by Celenay et al. carried out a lumbar spinal stabilization exercise program for 8 weeks in patients with BJHS, the study found that the spinal stabilization exercise program was more effective in improving pain intensity, trunk muscle endurance, and postural stability compared to controls.[49]

Stretching exercises are limited to gentle stretching to avoid any risks of subluxations or dislocations. Techniques that are used in treating pain include manual therapy for overactive muscles, trunk stabilization, posture re-education, joint awareness using biofeedback, joint mobilization with muscle release.[9]

Strengthening exercises targeting stabilising muscles around hypermobile joints enhance joint support throughout movement and reduce pain.[50,51] Closed chain exercises reduce strain on injured ligaments [52], enhance proprioceptive feedback, and optimise muscle action. [53] Ferrell et al. found that therapeutic exercise (knee strengthening, knee proprioception and balance exercises) enhanced proprioceptive acuity, balance and strength; reduced pain VAS scores; and improved the physical functioning and mental health components of the SF-36.[54] Coordination and balance exercises such as wobble board training improve proprioception.[55,56] Neural pathways and movement patterns consisting of muscle pair co-contractions are reinforced.[57] This can encourage compensation reactions, preventing joints moving into extreme ranges and avoiding further injury.[39]

A generalised exercise approach, in contrast to specific muscle training also can be taken, addressing cardio-respiratory, musculoskeletal and neurological aspects of movement [9] and reducing general deconditioning. [58] The cohort study by Barton and Bird investigated a general exercise programme. They found significant improvements in the maximum distance walked and pain on movement (in both the most affected joint and in all joints in general). [59]

Kemp at al compared a 6-week generalized program, improving muscular strength and fitness, with a targeted program aimed at correcting the motion control of symptomatic joints. They reported significant and sustained reduction in pain with both interventions.[60] One trial has been conducted assessing the effectiveness of a generalised versus a targeted physiotherapy exercise-based intervention for children with BJHS, two month follow-up results revealed no difference between the interventions.[60]

Hydrotherapy can be a successful medium to perform such exercises [9], challenging balance and core strength within a supportive environment, with water resistance and buoyancy increasing exercise variability.[61] Aquatic mode of therapy should be encouraged as the combination of buoyancy, support, and warmth makes it a suitable environment to treat JHS patients with the possibility to enable them to exercise safer and longer without exacerbating symptoms.[9]

Sahin et al. conducted assessment of the effectiveness of proprioceptive-based exercises for people diagnosed with BJHS a statistically significant improvement in VAS pain on resting and motion and AIMS-2 questionnaire (Arthritis Impact Measurement Scale) score was reported from baseline to eight-week follow-up in those allocated to receive exercises.[62]

CONCLUSION

Joint hypermobility syndrome is a relatively common cause of chronic pain in the general population but because its diagnosis requires a high level of clinical suspicion and the performance of specific physical examination maneuvers, it is often overlooked by providers. In addition to explaining patients about their disease process, patients with joint hypermobility syndrome can also be counseled about ways to protect their joints and to undergo lifestyle modification in order to prevent further damage.

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