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Original Research Article

How Benign Is Benign Hypermobility Syndrome [BHMS]

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Abstract: Benign joint hyper mobility syndrome (BHMS) is primarily seen in children and younger adolescents. It consists of joint hyper mobility (joints easily move beyond their normal expected range) associated with chronic exercise-related pain. Other commonly associated features include marked fatigability, tiredness, back pain, joint subluxation and soft tissue injuries. There is significant overlap between this condition and Ehlers-Danlos syndrome type 3 (hypermobility type). BHMS is now perceived as a heritable disorder of connective tissue (HDCT) which shares some features with Marfan's syndrome and Ehlers-Danlos syndrome type three. Aim of the study is to assess the muscloskeletal complaints in rheumatic clinic in south part of Chennai. The study was conducted from January 2016-may 2016. Totally 60 young adults of age range between 17-34 who has attended the rheumatic clinic with polyarthralgia and low back pain along with various other symptoms are included in the study. Detailed clinical history such as type of pain, duration of pain, muscloskeletal pain on various regions after daily activities of daily living are montoried and presence of BHMS was recorded using Modified Beighton Criteria. Totally 50 patients were included in the study. Among the 50 patients. 23 were male and 27 were female. The age was around 17-35 yrs. Duration of symptoms ranged between1-3 years. 44 patients had both knee pain and back pain. 18 patients have mechanical back pain. Elbow pain was observed in 31 patients. Neck pain was observed in 14 patients. Easy fatigability was observed in 37 patients. Among 50 patients 39 patients had reduced pain threshold on overnight rest. 11 patients didn't have a complete relief of pain after overnight complete rest. Benign joint hypermobility syndrome has a strong genetic component with an autosomal dominant pattern. First–degree relatives with the disorder can be identified in as many as50% of cases. The syndrome appears to be due to an abnormality in collagen or the ratio of collagen subtypes. Mutations in the fibrillin gene have also been identified in families with BHMS.

Keywords: Benign joint hyper mobility syndrome (BHMS), Polyarthrology, Beightons Criteria

INTRODUCTION:

Joint hyper mobility syndrome (JHS) is very common in musculoskeletal disease clinics, but the diagnosis is often missed, and the actual prevalence of JHS is not known Joint hyper mobility is very common in the general population, affecting 20-30% of individuals to some degree either in isolated joints or more generalized [1]. It is most common in childhood and adolescence, in females, and in Asian and Afro-Caribbean races. Joint hyper mobility tends to lessen with ageing. In one large survey in the United Kingdom, the combination of joint hyper mobility (JHM) and chronic widespread pain, which is typical of many patients with JHS, was found in 3 percent of a general population [2]. There has been a lack of general population studies or other studies of sufficient sample size to accurately estimate the prevalence of JHS JHS is considered by many experts in rheumatology and in clinical genetics to be indistinguishable from, if not identical to, the most common variant of Ehlers-Danlos syndrome (EDS), EDS-hyper mobility type (EDS-HM), but the precise relationship between EDS-HM and JHS remains uncertain. Joint Hyper mobility Syndrome (JHS) is a complex spectrum of signs and symptoms of varying degrees and combination [3]. It is considered synonymous with the hyper mobility variant of Ehlers-Danlos Syndrome. Joint hyper mobility is a two-edged sword, as flexibility is more often an advantage than a disadvantage [4]. Extra flexibility confers increased mobility and agility and is common amongst successful sportsmen and dancers. In general, hyper mobility should be regarded as an advantage, provided the child

is strong and fit. However, there are some adverse effects of tissue laxity, of which soft tissue injury and pain are the most significant. More chronic difficulties occur when the body has become weak and deconditioned.Only a proportion of patients with hyper mobile joints develops exercise-related pain. However, when they do they tend to avoid exercise. A worsening spiral is set up, with decreasing fitness and muscle reconditioning leading to worsening fatigue, lowered exercise tolerance and, eventually, exercise avoidance [5]. This can be associated with heightened pain sensitivity. Additionally, many have pain and fatigability when writing by hand and experience posture-related muscular pains, generalized tiredness and altered sleep patterns and concentration - all of which can affect school performance.Extra-articular symptoms may include abdominal pain, headaches and mood disturbance [6]. Average age at onset of symptoms was 6.2 years and age at diagnosis 9.0 years The major presenting complaint was arthralgia in 74%, abnormal gait in 10%, apparent joint deformity in 10% and back pain in 6%.Mean age at first walking was 15.0 months; 48% were considered 'clumsy' and 36% as having poor co-ordination in early childhood.12% had 'clicky' hips at birth and 4% actual congenital dislocatable hip. Urinary tract infections were present in 13% and 6% of the female and male cases, respectively. 14% had speech and learning difficulties diagnosed [7]. History of recurrent joint sprains was seen in 20%.Actual subluxation/dislocation of joints was seen in 10%.40% had problems with handwriting tasks.48% had major limitations of school-based physical education activities.67% had major limitations of other physical activities.41% had missed significant periods of schooling because of symptoms [8]. 43% described easy bruising.94% scored > or =4/9 on the Beighton scale for generalized hyper mobility, with knees (92%), elbows (87%), wrists (82%), hand metacarpophalangeal joints (79%) and ankles (75%) being most frequently involved [9].

Symptoms of BHMS [BENIGN HYPERMOBLITY SYNDROME] [10, 11]

- Pain and stiffness in the joints and muscles
- Clicking joints
- Joints that dislocate (come out of the correct position) easily
- Fatigue (extreme tiredness)
- Recurrent injuries – such as sprains
- Digestive problems – such as constipation and irritable bowel syndrome (IBS)
- dizziness and fainting
- Thin or stretchy skin.

MATERIALS AND METHODS:

The study was conducted from January 2016-may 2016. Totally 50 young adults of age range between 17-34 who has attended the rheumatic clinic with polyarthralgia, and low back pain along with combined symptoms are included in the study. Detailed clinical history such as type of pain, duration of pain, muscle skeleton pain on various regions on daily living activities are monitored and with the help of Modified Beightons Criteria. Patients with BJHS may have a family history of “double jointe dness”or recurrent dislocations. Other presentations include easy bruising, ligament or tendon rupture, congenital hip dysplasia, and temporo mandibular joint dysfunction. Findings of the physical examination vary based on the joint affected. Pain in response to manipulation of the joint is Common. Mild effusions are not common but may be present .Clinically significant tenderness along with redness, swelling, fever, or warmth suggests inflammation and is not present in patients with BHMS

Clinical Presentation:

The signs and symptoms of BJHS are variable. Most commonly, the initial complaint in a hyper mobile patient is joint pain, which may affect one or multiple joints and may be generalized or symmetric. Primary care physicians can use the five simple questions to aid in recognizing hyper mobility. The onset of symptoms can occur at any age, and many patients have been referred to specialists in orthopedics. Rheumatology or physiatry. Typically, children have self-limited pain in multiple joints; however, pain can last for a prolonged time and may become constant in adulthood. Pain may involve any joint but most commonly involves the knee and ankle, presumably because they are weight–bearing joints. Physical activity or repetitive use of the affected joint often exacerbates the pain. Consequently, pain usually occurs later in the day and morning stiffness is uncommon. Less common symptoms are joint stiffness, myalgia, muscle cramps, and non articular limb pain. Patients with BJHS often say that they are “double-jointed” or that they can contort their bodies into strange shapes (volitionary subluxation) or do the splits. Such an admission, however, is not necessary for including BJHS in the differential diagnosis .Patients with BHM S may also have a history of shoulder or patellar dislocation.

RESULTS:

Joint hyper mobility, defined as a more-than-normal range of movement (ROM) in a joint, is either localized (increased ROM of a single joint) or generalized. Joint hyper mobility depends on age, gender, family and ethnic background. A score of 5/9 or greater defines hyper mobility.

Table: 1 Shows the Beightons Criteria Score Among Patients

Patients Number	Score Of Patients On Physical Examination
28	4/9
12	6/9
6	7/9
2	8/9

Table: 2 Shows the Clincal Presentation of Muscloskeleton Complaints in Rheumatic Clinic

Physical examination pain perception sites	In male patients [n=23]	In female patients [27]	Total [50]
Both knee pain	21	23	44
Mechanical back pain	9	9	18
Elbow pain	16	15	31
Elbow pain with wrist pain	7	9	16
Neck pain	6	8	14
Easy fatigability	17	20	37
Pain subside on overnight rest	19	20	39
Stable pain after overnight rest	4	7	11
One or two symptoms existence	17	12	29
Poly arthritis symptoms	13	17	30

Totally 50 patients were included in the study. Among the 50 patients. 23 were male and 27 were female. The age was around 17-35 yrs. Duration of raised symptoms were observed in the patients between 1-3 years. 44 patients had both knee pain and back pain. 18 patients have mechanical back pain. Elbow pain was observed in 31 patients. Neck pain was observed in 14 patients. Easy fatigability was observed in 37 patients. Among 50 patients 391 patients had reduced pain threshold on overnight rest. 11 patients don't have a complete pain relief after overnight complete rest.

DISCUSSION:

When considering a diagnosis of BJHS, other genetic disorders, inflammatory, infectious and autoimmune disorders need to be excluded as they can present with joint hyper mobility and warrant unique or urgent management [12]. Genetic disorders such as Marfan syndrome, Ehlers –Danlos syndrome (except type III), Osteogenesis Imperfecta, Down's syndrome and autoimmune disorders such as Rheumatoid arthritis (RA) can present with joint hypermobility. The common characteristic signs or symptoms of these genetic disorders or other conditions such as RA were not present in each of our patient's history or though observation and palpation and subsequently these conditions were not likely to be present. Other more "benign" functional neuromusculoskeletal deficits were accessed through a focused neuromusculoskeletal examination including the Brighton criteria [13]. Children inherently have a greater range of joint motion than adults, the prevalence of hypermobility, as defined by several criteria, varying in different populations from 5 to 30%. This variation probably represents ethnic differences, but also the different ages at which joint examinations were undertaken and the different populations chosen for study [14]. The frequency of musculoskeletal disorders (MSDs) arising from such hyper mobility in childhood is quite variable, both across populations and within individuals. Some studies have suggested a definite causal link between hyper mobility of joints and MSDs, but others have not found such a link [15]. In pediatric services, and in particular pediatric rheumatology services, a referral bias is undoubted, and studies including control groups of healthy asymptomatic individuals with and without hypermobility are essential to fully establish such links. Nonetheless, the body of evidence suggests that hyper mobility is associated with significant MSDs in childhood that lead to consultation with medical practitioners and health-care providers [16]. We believe that manifestations may occur at any age, with a spectrum of potential disorders ranging from congenital dislocation of the hip at birth, through growing pains in early school age children to back pain and occasionally spondylolysis in teenagers. A score of 4/9 is taken as a positive marker of joint hypermobility [17]. A general physician can easily apply these criteria to diagnose patients with generalized joint laxity. On this subset of patients, Brighton's criteria are applied to diagnose patients of BJHS. Brighton's criteria, which incorporate Beighton 'scoring system, help distinguish patients of BJHS from other connective tissue disorders [18].

CONCULSION:

In summary, the BJHS is a common cause of musculoskeletal complaints in general practice, with quite well-described patterns of symptoms and signs,

and early recognition is likely to improve the outcome. Further understanding of the genetics of connective tissue integrity and laxity is likely to yield better understanding of the physiological and pathological variants that no doubt underlie the variable modes of presentation and phenotype seen in the children [19, 20]. Benign hyper mobility is a benign condition but causes considerable morbidity.

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