ORIGINAL ARTICLE

RELATIONSHIP OF JOINT HYPERMOBILITY AND MUSCULOSKELETAL PROBLEMS AND FREQUENCY OF BENIGN JOINT HYPERMOBILITY SYNDROME IN CHILDREN

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Background: The majority of individuals with joint hypermobility remain asymptomatic. However, those associated with Benign Joint Hypermobility Syndrome (BJHS), develop a number of systemic manifestations. Our objective was to determine the relationship between joint hypermobility and musculoskeletal problems, and frequency of BJHS in children and adolescents. **Method:** This cross-sectional observational descriptive study was conducted at Outpatient Department, The Children's Hospital, Lahore, Pakistan. A total of 872 individuals (4–18 year) were examined for hypermobile joints using Beighton score ≥4. A questionnaire was implied to get data regarding demographic profile, musculoskeletal and extra-articular complaints, family history of joint problems and daily activity. Brighton's criteria were implied for diagnosis of BJHS. **Results:** The frequency of joint hypermobility was 37.0%; male 39.5%, and female 34.2% (p=0.1). There was a gradual decline in mean Beighton score with age. The female population showed increase in mean Beighton score around 16–17 year age. Arthralgias and back pains 7.7% vs. 1.6%, (p<0.001), and hernias 2.5% (p=0.03) were significantly higher in individuals with joint hypermobility. History of joint problems in the family was also significantly higher in children with joint hypermobility (p=0.01). BJHS was detected in 4.8% children (male 3.6% and female 6.3%, p=0.06). Arthralgias (51.0%), hernias (16.3%), joint dislocations (8.2%) and varicose veins (8.2%) were the most common presentations. **Conclusion:** BJHS is common among children. Arthralgias, back pains and hernias are significantly higher in these individuals.

Keywords: Joint hypermobility, Benign Joint Hypermobility Syndrome, musculoskeletal problems, Children

INTRODUCTION

Joint Hypermobility is defined as an abnormal increased range of joint movement beyond the norms for those joints. Benign Joint Hypermobility Syndrome (BJHS) is characterized by the occurrence of musculoskeletal symptoms in hypermobile individual in the absence of any demonstrable systemic rheumatologic disease.[1]

The majority of individuals with joint hypermobility remain asymptomatic. However, those associated with BJHS, develop a number of systemic manifestations like Marfanoid habitus, skin hyperextensibility, sciatica, Reynaud's phenomenon, varicose veins, abdominal and inguinal hernias. This condition arises from naturally lax connective tissue resulting in excessive stress over joints and periarticular structures. Repeated stress is considered to cause the musculoskeletal complaints including frequent sprains and arthralgias. This laxity has been attributed to Tenaxin B gene causing qualitative defect in collagen type-I.[1] Early recognition prevents unnecessary investigations. Patients with BJHS can be reassured about a relatively benign course requiring a few simple preventive measures. There are marked ethnic, age and gender differences in articular hypermobility. South Asian population has been considered more hypermobile than caucasians. There is scarce data regarding the actual prevalence of joint hypermobility. Its effect on musculoskeletal symptoms in this subgroup during childhood and adolescence is poorly understood.

In this study, we tried to determine the frequency of joint hypermobility and BJHS in pediatric and adolescent population in Lahore, Pakistan and assess the role of joint hypermobility as cause of musculoskeletal symptoms in this population subgroup.[2]

MATERIAL AND METHODS

This cross-sectional, observational, descriptive study was conducted at the outpatient department of The Children's Hospital and Institute of Child Health, Lahore from June 2006 to May 2007. Purposive non-probability sampling technique was used to collect the data. Healthy children between ages of 4 and 18 years accompanying patients were included in the study after acquiring a verbal consent. Any individual with acute ailment, trauma or taking medication known to influence joint mobility like corticosteroids, hormone replacement therapy or anti-hyperlipidaemics was excluded from the study to remove any confounding effect. Dysmorphic children, individuals with connective tissue disorders like Ehler Danlos syndrome, Marfan syndrome or children with neurological deficit were excluded from the study.

Demographic profile was recorded including the age, gender and ethnic group. Children were asked in detail regarding the clinical features of musculoskeletal problems including arthralgias, backache, dislocations, and joint swellings as well as for drooping eyelids, Marfanoid habitus, varicose veins and skin striae. Family history of joint problems was sought. Details of severity and time of onset of these clinical findings were also collected. Medical examination was conducted by two examiners to minimise the observer bias. The joint hypermobility was assessed according to Beighton score which includes evaluation of nine joints for hypermobility.[3] GJHS was diagnosed on the basis of Brighton criteria.

Data was analysed using SPSS-14.0. Student's *t*-test and Chi-squared test were used for test of significance where applicable and $p<0.05$ was considered significant.

Box-1: Beighton scoring system for joint hypermobility

For a positive Beighton Score, 4 or more points out of 9 are required:

1. Hyperextension of the elbows more than 10º
2. Passively touch the forearm with thumb while flexing the wrist.
3. Passive extension of the fingers or a 90º or more extension of the fifth finger (Gorling's sign). This is used as a 'Screen Test'.
4. Knees hyperextension greater than 10º (genu-recurvatum)
5. Touching the floor with the palms of the hands when reaching down without bending the knees. This is possible as a result of the hypermobility of the hips, and not of the spine as it is commonly believed

➢ One point is given for all manoeuvres with separate score for right and left side of body for first 4 joint movements.

Box-2: Brighton's criteria for diagnosis of BJHS

Major Criteria

1. A Beighton score of 4/9 or greater (either currently or historically)
2. Arthralgia for longer than 3 months in 4 or more joints

Minor Criteria

1. A Beighton score of 1, 2 or 3/9 (0, 1, 2 or 3 if aged 50+)
2. Arthralgia (>3 months) in one to three joints or back pain (>3 months), spondylosis, spondylolysis/spondylolisthesis.
3. Dislocation/subluxation in more than one joint, or in one joint on more than one occasion.
4. Soft tissue rheumatism. >3 lesions (e.g., epicondylitis, tenosynovitis, bursitis).
5. Marfanoid habitus (tall, slim, span/height ratio >1.03, upper: lower segment ratio less than 0.89, arachnodactily, positive Steinberg/wrist signs).
6. Abnormal skin: striae, hyperextensibility, thin skin, papyraceous scarring.
7. Eye signs: drooping eyelids or myopia or antimongoloid slant.
8. Varicose veins or hernia or uterine/rectal prolapse.

➢ The Joint Hypermobility Syndrome (JHS) is diagnosed in the presence two major criteria, or one major and two minor criteria, or four minor criteria. Two minor criteria will suffice where there is an unequivocally affected first-degree relative.

RESULTS

The evaluation was conducted on 900 healthy children between ages of 4–18 years. Twenty-eight children were excluded from the study based on previously set exclusion criteria. The data regarding a total of 872 children were analysed. The individuals studied 474 (54.4%) males and 398 (45.6%) females. The mean age was 12.85±3.9 years, in male population it was slightly lower (12.64±3.7 years) than females (13.1±4.1 years). Most (86.9%) of the children were students, while a few elder children were involved in farming, manual labour or office clerkship.

The frequency of Joint Hypermobility was 37% (n=323/872) with no significant difference between male (39.5%, n=187/474) and female (34.2%, n=136/398) population, (p=0.1). A positive family history of musculoskeletal symptoms was found in 13.1% (n=118/872). People with joint hypermobility had a significant history of joint problems in other family members, i.e., 17.3% (n=56/323) vs. 11.3% (n=62/549) in children without joint hypermobility, (p=0.01).

The frequency of arthralgias and back pain was 3.9% (n=34/872), hernias 1.3% (n=11/872) while the rest of symptoms were 0.1%–0.8% in overall individuals. Arthralgias and back pain were significantly higher in children with joint hypermobility; 7.7% (n=25/323) compared to 1.6% (n=9/549) in children without joint hypermobility, (p<0.001). Hernias were also found to be significantly higher in children with joint hypermobility, 2.5% (n=8/323) vs. 0.5% (n=3/549) in children without hypermobility, (p=0.03). Similarly, joint subluxation and dislocations were more frequent in people with joint hypermobility, 1.2% (n=4/323) compared to 0.2% (n=1/549) but did not achieve significant difference, (p=0.13). The frequency of the rest of symptoms studied including skin and eye signs, rectal or uterine prolapse and marfanoid features, were also not significantly different (Table-1).

The frequency of BJHS was found to be 4.8% (n=42/872), 6.3% (n=25/398) in female population and 3.6% (n=17/474) in males, p=0.06. In all the children studied, there were 57 events of systemic or musculoskeletal problems. There was gradual decrease in Beighton score in males with increase in age. In female children, there was an increase in Beighton score around 16 and 17 years of age. The frequencies for individual joint hypermobility showed highest frequency for left little finger 60.7% and right little finger 58.5%. The difference in frequencies of individual hypermobile joints in people with and without joint hypermobility was highly significant for all joints, (p<0.001) (Table-2). Knees had the highest positive predictive value for having generalised joint hypermobility (Table-3).

Table-1: Distribution of symptoms among BJHS and children without joint hypermobility

Symptom/sign	Children with GJHS (n=49)	Children without joint hypermobility (n=25)
Arthralgias, Backache	25 (51%)	9 (36%)
Joint subluxation, sprains	4 (8.2%)	1 (4%)
Soft tissue rheumatism	2 (4.1%)	3 (12.0%)
Hernias, rectal prolapse	8 (16.3%)	4 (16.0%)
Varicose veins	4 (8.2%)	3 (12.0%)
Marfanoid habitués	1 (2.0%)	3 (12.0%)
Skin signs	4 (8.2%)	4 (16.0%)
Eye signs	1 (2.0%)	1 (4.0%)

Table-2: Comparison of the frequency of individual hypermobile joints in children with and without joint hypermobility

Joints evaluated	Hypermobility in Children with joint hypermobility %(n) N=323	Hypermobility in Children without joint hypermobility %(n) N=549	*p*
Right little finger	89.2 (288)	40.4 (222)	$p \leq 0.001$
Left little finger	90.4 (292)	43.2 (237)	
Right thumb	52.9 (171)	13.1 (72)	
Left thumb	53.3 (172)	20.0 (110)	
Right elbow	29.4 (95)	4.6 (25)	
Left elbow	31.0 (100)	6.6 (36)	
Right knee	69.7 (225)	7.8 (43)	
Left knee	70.6 (228)	7.3 (40)	
Palm touching floor	18.6 (60)	6.0 (33)	

Table-3: Individual Joint Hypermobility for detecting joint hypermobility

Joint Hypermobility	Sensitivity	Specificity	Positive predictive value	Negative predictive value
Right little finger	89.16	59.56	56.47	90.33
Left little finger	90.4	56.68	55.2	90.96
Right thumb	52.94	86.89	70.37	75.83
Left thumb	53.25	79.96	60.99	74.41
Right elbow	29.41	95.45	79.17	69.68
Left elbow	30.96	93.44	73.53	69.7
Right knee	69.66	92.71	83.96	83.77
Left knee	70.59	92.71	85.08	84.27
Palm touching floor	18.58	93.99	64.52	66.24

DISCUSSION

BJHS is a major cause of non-specific musculoskeletal complaints with quantitative defect in underlying connective tissue. The symptoms can be controlled with limited intervention. The condition is well known to rheumatologists and orthopaedics but primary care physicians and paediatricians fail to identify this condition resulting in undue investigations and unjustified interventions. It can induce unnecessary anxiety on part of children and their parents. Unrecognised condition may even lead to long term disability.

The frequency of joint hypermobility was found to be present in 37% individuals. The frequency was similar to prevalence reported from Scandinavian and Caucasian studies (Iceland 40.5%, Italy 34%, Houston 34%).[4] Studies from South America, Spain and African region had much higher prevalence (Sao Paulo 64.6%, Nigeria 43%, Spain (55%).[5] On the other hand, a few studies from Middle East showed much lower prevalence (Israel 13%, Egypt 16.1%). New Zealand (4.0–6.2%) and Singapore (17%) also had much lower prevalence of BJHS.[6] While comparing all these data, the criteria used for the diagnosis should be kept in mind as well as the cut off point used within each criterion.[7]

The frequency of joint hypermobility has a dynamic character, and while evaluating joint hypermobility, age has a significant implication. The male population showed a gradual decrease with increase in age. The female population showed increased hypermobility around 16–18 years age. This peculiar rise in female joint hypermobility around 15–18 years was also reported by Jansson and co-workers.[8] The exact reason for this rise in frequency of joint hypermobility in 15 year old females remains unclear. Hormonal interplay with their interaction has been theoretically postulated but Beynnon *et al* showed no cyclic change in joint hypermobility with cyclic change in oestrogen and progesterone levels.[9] Relaxin may play a role but no study has been conducted to study the effects. The prevalence of joint hypermobility decreases with age, the rate of decline however, has been variable in different studies.[10]

A number of studies favour female population showing higher frequency of hypermobility. This difference is most likely attributed to difference in age groups studied. Jansson documented higher prevalence of joint hypermobility in female 15 years old Swedish population.[8] Study in 12 years old Icelandic school children also showed females being more hypermobile.[4] The gender difference was more pronounced in studies conducted on elder populations. The results from most studies on younger paediatric population had no gender difference. In our study, joint hypermobility was more common in adolescent (13–18 years) females in our study but the difference was not significant (p=0.1). These results were in harmony with various studies on British, Dutch, Italian, Finnish and Egyptian school children.[11] The increase in frequency of joint hypermobility in females with increase age may point towards hormonal interplay with structural joint laxity. The exact causative factor remains unknown as yet.

Musculoskeletal symptoms were significantly high in people with joint hypermobility. The most common complaint was low back pain and arthralgias (p<0.001). el-Shahaly *et al* also reported similar findings.[12] Gedalia *et al* documented increased frequency of joint pain in knees, wrist and fingers.[11] These musculoskeletal pains have been attributed to recurrent injuries and elevated GH, IGF-1.[13] Up to 74%

children with joint hypermobility have arthralgias.[14] Recurrent sprains and dislocations were reported as high as 20%. The frequency was lower in our study but still significantly higher than those without joint hypermobility ($p<0.01$). Knees, back and wrist are more commonly involved.[15] Hypermobility has been established as an important factor in low back pain in pregnant females.[16] Decreased proprioception has been postulated also contributing to recurrent tissue injury and musculoskeletal symptoms.[17] Secondary problems associated with recurrent subluxations and sprains have shown to cause osteoarthritic changes in both ankles in joint hypermobility and chondrocalcinosis in knee joints.[18] Extra-articular manifestations like varicose veins were not common in studied population. It was also in harmony with observations by Biro and Zapata *et al*.[19] Hernias were the only extra articular manifestation significantly more common in children with joint hypermobility due to connective tissue weakness ($p=0.03$). It also attributes to enhanced nociception and visceral pain due lax suspension and supported tissue for the viscera. The weak connective tissue has serious implication during tendon or ligament surgery where this inherent condition leads to over correction. No other symptoms were found to be significantly associated with joint hypermobility. Few studies have shown a high incidence of genital prolapsed. It is presumed that genital prolapse occurs more commonly after child birth. As a result of recurrent prolapse, chance of genuine stress incontinence increases.[20] Skoumal *et al* also found extra articular manifestations less frequent than musculoskeletal problems especially in children.[21]

The children with BJHS did not have any typical morphological features. These findings were against the popular belief that tall lean people have higher prevalence of joint hypermobility than short people. Similarly, Remvig *et al* also found no extra articular manifestation significantly associated with joint hypermobility and BJHS.[1]

The hypermobility was found to be higher in upper limbs and in small joints. The observation supports the fact that connected tissue is more important in small joint stability which is weak in BJHS. Left and right little fingers were found to be most hypermobile in children studied for hypermobility, 60.7% and 58.7% respectively. These were followed by wrists and elbows. The figures were in harmony with studies involving Egyptian and Brazilian children.[22] Knee hyperextension had the highest positive predictive value (right knee 85.08% and left knee 83.96%). Little finger hyperextension had the highest negative predictive value. Screening for joint hypermobility may be considered using these four joints in children. There was no significant difference between right and left sided joint hypermobility. One sided scoring has been suggested based on similar findings by van der Giessen.[23] This was in contrast with previous studies conducted on adult population showing non-dominant side significantly more hypermobile than dominant side.[24] Age again appears to be an important factor in children showing no difference between dominant and non dominant side.

CONCLUSION

The frequency of BJHS is not very high in population group studied in Lahore, Pakistan; however, the presence of significant relation between musculo-skeletal complaints and joint hypermobility warrants the condition to be regarded as an important cause of arthralgias, recurrent sprains and joint dislocations in our population. BJHS needs to be considered while performing corrective joint or tendon surgery due to risk of over correction. Early recognition, especially in teen age girls also relieves the patient and parents from anxiety associated with recurrent symptoms.

REFERENCES

1. Remvig L, Jensen DV, Ward RC. Epidemiology of general joint hypermobility and basis for the proposed criteria for benign joint hypermobility syndrome: review of the literature. J Rheumatol 2007;34:804–9.
2. Murray KJ. Hypermobility disorders in children and adolescents. Best Pract Res Clin Rheumatol 2006;20:329–51.
3. Grahame R, Bird HA, Child A. The revised (Brighton 1998) criteria for the diagnosis of benign joint hypermobility syndrome (BJHS). J Rheumatol 2000;27:1777–9.
4. Qvindesland A, Jonsson H. Articular hypermobility in Icelandic 12 year olds. Rheumatology 1999;38:1014–6.
5. Birrell FN, Adebajo AO, Hazleman BL, Silman AJ. High prevalence of joint laxity in West Africans. Br J Rheumatol 1994;33:56–9.
6. Klemp P, William SM, Stansfield SA. Articular mobility in Maori and European New Zealanders. Rheumatology (Oxford) 2002;41:554–7.
7. Russek LN. Hypermobility syndrome. Phys Ther 1999;79:591–9.
8. Jansson A, Saartok T, Werner S, Renstrom P. General joint laxity in 1845 Swedish school children of different ages: age-and gender-specific distributions. Acta Paediatr 2004;93:1202–6.
9. Beynnon BD, Bernstein IM, Belisle A, Brattbakk B, Devanny P, Risinger R, *et al*. The effect of estradiol and progesterone on knee and ankle joint laxity. Am J Sports Med 2005;33:1298–304.
10. Gedalia A, Brewer EJ Jr. Joint hypermobility in pediatric practice: a review. J Rheumatol 1993;20:371–4.
11. Larsson LG, Baum J, Mudholkar GS. Hypermobility: features and differential incidence between the sexes. Arthritis Rheum 1987;30:1426–30.
12. el-Shahaly HA, el-Sherif AK. Is the benign joint hypermobility syndrome benign? Clin Rheumatol 1991;10:302–7.
13. Denko CW, Boja B. Growth hormone, insulin, and insulin-like growth factor-1 in hypermobility syndrome. J Rheumatol 2001;28:1666–9.
14. Adib N, Davies K, Grahame R, Woo P, Murray KJ. Joint hypermobility syndrome in childhood. A not so benign multisystem disorder? Rheumatology (Oxford) 2005;44:744–50.
15. Didia BC, Dapper DV, Boboye SB. Joint hypermobility syndrome among undergraduate students. East Afr Med J 2002;79:80–1.
16. Mogren IM, Pohjanen AI. Low back pain and pelvic pain during pregnancy: prevalence and risk factors. Spine 2005;30:983–91.
17. Ferrell WR, Tennant N, Sturrock RD, Ashton L, Creed G, Brydson G *et al*. Amelioration of symptoms by enhancement of

proprioception in patients with joint hypermobility syndrome. Arthritis Rheum 2004;50:3323–8.
18. Ofluoglu D, Gunduz OH, Ozaras N, Kayhan O. Early-onset hemochromatic arthropathy in a patient with idiopathic hypermobility syndrome. Rheumatol Int 2003;23:305–8.
19. Zapata AL, Moraes AJ, Leone C, Doria-Filho U, Silva CA. Pain and musculoskeletal pain syndromes in adolescents. J Adolesc Health 2006;38:769–71.
20. Manning J, Korda A, Benness C, Solomon M. The association of obstructive defecation, lower urinary tract dysfunction and the benign joint hypermobility syndrome: a case-control study. Int Urogynecol J Pelvic Floor Dysfunct 2003;14:128–32.
21. Skoumal M, Haberhauer G, Mayr H. Concomitant diseases in primary joint hypermobility syndrome. Med Klin (Munich) 2004;99:585–90.
22. El-Garf AK, Mahmoud GA, Mahgoub EH. Hypermobility among Egyptian children: prevalence and features. J Rheumatol 1998;25:1003–5.
23. van der Giessen LJ, Liekens D, Rutgers KJ, Hartman A, Mulder PG, Oranje AP. Validation of Beighton score and prevalence of connective tissue signs in 773 Dutch children. J Rheumatol 2001;28:2726–30.
24. Rikken-Bultman DG, Wellink L, van Dongen PW. Hypermobility in two Dutch school populations. Eur J Obstet Gynecol Reprod Biol 1997;73:189–92.

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