Journal of Advances in Medicine and Medical Research

22(8): 1-7, 2017; Article no.JAMMR.33561
Previously known as British Journal of Medicine and Medical Research
ISSN: 2231-0614, NLM ID: 101570965

The Benign Joint Hypermobile Syndrome among Sudanese's Children in Al-Khartoum State from November 2011-May 2012

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Authors' contributions

This work was carried out in collaboration between all authors. All authors read and approved the final manuscript.

Article Information

DOI: 10.9734/JAMMR/2017/33561
Editor(s):
(1) Nurhan Cucer, Erciyes University, Medical Biology Department, Turkey.
Reviewers:
(1) Terry Ellapen, University of KwaZulu-Natal, South Africa.
(2) Chia-Li Yu, National Taiwan University Hospital, Taiwan.
(3) Ayhan Goktepe, Selcuk University, Turkey.
Complete Peer review History: http://www.sciencedomain.org/review-history/19891

Original Research Article

Received 20th April 2017
Accepted 30th June 2017
Published 6th July 2017

ABSTRACT

Background: Joint hyper mobility (JH) defines as ligamentous laxity, it is due to a primary genetic disorder of connective tissue matrix proteins, Moreover, it is highly associations with the risk of soft tissue injury, chronic widespread pain, and early degenerative disease.
The aim at the study to assess the frequency of hyper mobility syndromes in Sudanese 'children, and the knowledge of mother toward to the disease, morbidity and importance of treatment else.
Methodology: This is, Hospital- based descriptive cross sectional study conducted at 4 hospital at Khartoum state in Sudan at(November-2011/may-2012), randomly selection 100 children their age between 3-13Y seen in the outpatient clinic or admitted to hospital namely above, receive the questioner about general information about on child and mother's ,also clinical measurement by Brighton criteria and Beighton score.
Results: Children had benign hyper mobile syndrome (BJHS) according to Beighton score 63%,

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while 56% of children according to Brighton criteria moreover, 45% of mother notice the BJHS while 9% of mother consider about the symptoms.
Conclusion: Hyper mobility syndromes are common in Sudanese children while the poor knowledge about this condition might lead to delay in diagnosis and application of treatment.

Keywords: Benign joint hyper mobile syndrome; Beighton score; Brighton criteria.

1. INTRODUCTION

Begin Joint hyper mobility syndrome (BJHS) defined as a heritable disorder [1], it was recognized as distinct pathology at 1967, it identified by a several names such as "hypermobility syndrome (HMS)," "joint hyper mobility syndrome," "hypermobility joint syndrome," and "benign hypermobile joint syndrome" [2], it presented by joint laxity [3] in passive movements and active movements [4], either in single joint or a multiple joints, [1,5].

It characterized by the occurrence of multiple musculoskeletal problems of with hypermobile individuals that do not have a systemic rheumatological disease [1].

It is a wide spread problem of with the word [6], increase among children [5], Its prevalence in the general population is between 10% and 15% [4], also it was common to women than in men [4,5], and more in Asian than African population [7].

The hormone status or physical training may increase the onset of joint hyper mobility include [7,8].

It was associated with involvement in other organs because of abnormal inherited collagen structure elsewhere [9]; such as association between joint hyper mobility syndrome and mitral valve prolapse, panic disorder [1,4], rectal, uterine prolapse [10], varicose veins, myopia, recurrent urinary tract infections ,asthma [1], and with juvenile episodic arthralgia [11].

Most of the children are asymptomatic [11], but the common symptomatic in children are joint inflammation [2], musculoskeletal pain commonly at night in lower-limb. [8], fatigue, often with reduction of exercise tolerance. [12], headaches, due to trapezius muscle spasm and poor sitting posture [13], easy bruising [13], clicking of joints [13], reduced core strength [13], fine motor control may be affected [13], and a small proportion may have problems with abdominal pain with or without bladder and bowel dysfunction [13].

BJHS rare associations with; postural orthostatic tachycardia syndrome (pots) [13], hernia: studies of children with hernia have shown that they show an increased prevalence of Beighton scored of 4 or more compared to the general population [14], and uterine or rectal prolapse [13] by the age of 50 common features are osteoarthritis changes in the knees, spine, and fingers with a resulting reduction in mobility [15], recurrent joint dislocation; patellar, elbow and shoulder dislocation and congenital hip dislocation [16], abnormal foot angle [17], scoliosis and postural deformity, skin abnormality [16], recurrent joint pain and effusion [18], and inflammatory poly arthritis :synovitis [2].

Moreover the small proportion of patients have one of the more serious conditions as Ehlers-Danlos syndrome, Marfan's syndrome or osteogenesis imperfect [9].

Generalize joint laxity affects up to 40% of young adolescents prevalence decreasing with age. it affected Girls more than boys; children of Asian ethnicity are more hypermobile than those of Caucasian ethnicity [19,20]. Mostly report on to the rheumatology literature [2], many children with high scores are not symptomatic [21-23].

The clinical assessment of hypermobility does not require to special equipment [11], commonly it assessed by the Beighton score which was devised in South Africa based [4,8], is a 9-point scoring system to quantify joint laxity and hypermobility. A higher score equates to higher joint laxity [24]. He excluded the other genetic diseases which associated with JH [2], and Brighton criteria, it contented both subjective and objective findings, also included the serious diseases with hypermobility, such as Ehlers-Danlos syndrome (EDS), Marfan syndrome (MF) and Osteogenesis Imperfecta (OI). Brighton criteria accepted diagnostic to incorporate the Beighton scoring system, in combination with the presence of persistent symptoms. It requires two major criteria or one major and two minor, in the absence of diagnosed Ehlers-Danlos or Marfan's syndromes. Two minor criteria are considered enough for diagnosis if it clearly affected [3].

The treatment process usually is length therefore it mostly depends on education of the patient and family, and facilitation of lifestyle and behavior modifications according to the plan [25].

Physical therapy session included modalities, activity modification, stretching and strengthening exercises in the affected joint, and osteopathic manipulative treatment [18].

Exercises in the Supervisor required improving joint stability and proprioception remains the mainstay of treatment [26].

2. METHODOLOGY

2.1 Study Area and Population

This is a Hospital- based descriptive cross sectional study, in one hundred children conducted at 4 hospitals in Khartoum state: Jafer Ibn Owf, Paediatric department of Military hospital, Saad Abo alilaa hospital, and Dentistry clinic in al-Neelain University in Khartoum state, from November 2011 to may 2012.

2.2 Inclusion and Exclusion Criteria

All Children, their age arrangement between 3_13 years from both sexes, seen in the outpatient clinic or admitted to hospital mentioned before.

2.3 Test and Measurements

We adopted Questionnaire and Clinical examination measurement. The questionnaire about general information about child and mother's, medical history of children and the awareness about the hypermobility, while the clinical examination measurement done by two scores: First Beighton Scoring System which is the old version of the Carter & Wilkinson scoring system is a simple validated system used to quantify joint laxity and hypermobility [2] secondly the Brighton criteria, used for diagnosis in the presence of two major criteria, one major and two minor criteria, or four minor criteria. The criteria are as follows [3]:

Major criteria [3]:

- Beighton score of 4 or more.
- Arthralgia for >3 months in four or more joints.

Minor criteria [3]:

- Beighton score of 1-3.
- Arthralgia for >3 months in one to three joints.
- Back pain for >3 months.
- Spondylosis/spondylolysis/spondylolisthesis.
- Dislocation/subluxation of more than one joint, or in one joint more than once.
- More than three soft tissue inflammatory conditions (eg, tenosynovitis, epicondylitis).
- Marfanoid habitus (tall, slim, span/height ratio >1.03, arachnodactyly, upper/lower segment ratio <0.89.
- Skin striae, thin skin, hyperextensible skin, papyraceous scars.
- Drooping eyelids, myopia or antimongoloid slant.
- Varicose veins, hernia or uterine/rectal prolapse.

Table 1. Beighton test

Joint	Finding	Pts
Left little finger	Passive dorsiflexion beyond 90° 1	Passive dorsiflexion < =90° 0
Right little finger	Passive dorsiflexion beyond 90°1	1 Passive dorsiflexion < =90° 0
Left thumb	Passive dorsiflexion to flexor aspect of forearm1	Cannot passively dorsiflex thumb to flexor aspect of forearm 0
Right thumb	Passive dorsiflexion to flexor aspect of forearm 1	Cannot passively dorsiflex thumb to flexor aspect of forearm 0
Left elbow	Hyperextend beyond 10°1	Extends < =10°0
Right elbow	hyperextend beyond 10o	1Extends < =10°0
Left knee	Hyperextend beyond 10°	1Extends < = 10°
Right knee	Hyperextend beyond 10°	1Extends < =10°
Trunk flexion with knees fully extended Palms and hands	can rest flat Palms and hands on the floor 1	cannot rest flat Palms and hands on the floor0

2.4 Data Analysis

Statistical analysis of the data was performed using SPSS soft ware for medical statistics. Descriptive analysis was also used to analyze the samples. To examine test significance,(t) test used. The significance level was set at ($p< 0.05$).

2.5 Ethical Considerations

The research conforms to the ethical principles of medical research developed by the World Medical Association Declaration of Helsinki. Ethical clearance was given by the Research Committee (El-Neelain University), as well as assent was obtained from children parent before filling the questionnaire.

3. RESULTS

100 cases, male =43, children less than 5 years =6 and 37 is more than 5y, female=57 children less than 5 years =16 and 41 is more than 5y, in age between (3_13) years) participated in this study. Children had (HMS) according to Beighton score 63%, while 56% of children according to Brighton criteria.

4. DISCUSSION

In the current study, 100 children were assessed by Beighton score and Brighton criteria to diagnosis the HJS the result show 63% of children were assessed by Beighton score had JH while the 56% of children were assessed by Brighton criteria had BJHS. Also arthralgia was common feature 58% followed by skin abnormality texture was 26.7%, joint dislocation and delay of wound healing were 19.6% and hernia was 7%. Moreover 45% of mother noticed the laxity of joint in their children while just 9% of mother, who consider about symptom.

Our result supported by clark and their collages, Their study aimed to investigated the prevalence of chronic widespread pain (CWP) and the extended neuro physiological features reported by a group of patients with JHS. Methods: Ninety patients with JHS which was diagnosed in accordance with the Brighton criteria, and 113 healthy volunteers with no musculoskeletal pain participated in the study. Results: CWP was reported by 86% of patients with JHS [27], as well as reported in one large survey in the United Kingdom, the combination of

Table 2. Demonstrated that number of children who affected according to their age and gender

	Gender of children		Children affected by HMS by Beighton score		Total
			Affected children	Not affected children	1.00
Male	Age	Less than 5	2	4	6
		Above than 5	24	13	37
	Total		26	17	43
Female	Ago	Less than 5	11	5	16
		Above than 5	26	15	41
	Total		37	20	57

Table 3. Demonstrated that percent of children had major and minor criteria

Criteria	Affected children	Unaffected children
Pain after play for long period	58.9%	40.9%
Pain after play for long period	41%	59%
Laxity joints	100%	11%
No laxity joints	0%	88%
Hernia	7%	11%
No hernia	92.7%	88%
Delayed wound heeling	19.6%	11%
No delayed wound heeling	80.3%	88%
Abnormal skin texture	26.7%	9%
Normal skin texture	73.2%	81%
Joint dislocation	19.6%	11.3%
No joint dislocation	80.3%	88%

Table 4. Demonstrated percent mother's awareness about (HMS)

Mother's awareness	Frequency
Number of mothers considered to HMS	45%
Number of mothers who did not considered HMS	55%
Total	100
Mothers think the hypermobility syndrome is a disease	9%
Mother don't think the hypermobility syndrome is a disease	91%
Total	100

joint hypermobility (JHM) and chronic widespread pain, which is typical of many patients with JHS, was found in 3 percent of a general population [28]. Also by jacop research which resulted the main clinical feature was joint laxity, which caused articular dislocations, subluxation and arthralgia, in the absent of evidence for any rheumatological disorder. Asymptomatic patients were rarely diagnosed while those with symptoms were commonly misdiagnosed [29].

Also the result of Remvig in 2007 showed, the major criterion of BJHS, is arthralgia it is major component of alleged hypermobility-related problems [30] Moreover Pain dominates the lives of many patients with hyper laxity syndromes, most commonly the Benign Joint Hypermobility Syndrome" was show in gerahame research in 2000 [31].

On another hand; hakim and gerahame in 2004 and gerahame R in 2008 were discussing the joint hypermobility syndrome (JHS) is very common in musculoskeletal disease clinics, but the diagnosis was often missed, while the actual prevalence of JHS was unknown [32,33]. There had been a lack of general population study or other studies of sufficient sample size to accurately estimate the prevalence of JHS [6].

5. CONCLUSION

According my results the Hypermobility syndromes is common in Sudanese, The arthralgia is common manifestations also we finding the lack of awareness among mother lead to delay in diagnosis and application of treatment.

6. RECOMMENDATIONS

We need to more study about JHS and increase the awareness among the community and physician to decreases the rate of complication.

CONSENT

As per international standard or university standard, patient's written consent has been collected and preserved by the authors.

ETHICAL APPROVAL

As per international standard or university standard, written approval of Ethics committee has been collected and preserved by the authors.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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Peer-review history:
The peer review history for this paper can be accessed here:
http://sciencedomain.org/review-history/19891