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The joint hypermobility syndrome in a Cuban juvenile population

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ABSTRACT

In order to review the rise in joint hypermobility syndrome, identify the patterns associated with this diagnosis and correlate its most relevant symptoms, we did a descriptive transversal analytic study in a group of 280 young people of both genders between 15 and 17 years of age. A survey was carried out by the authors in order to get reach these objectives. The data were analyzed by means of descriptive statistics and processed with the Epidat3.1 software package. Results are shown in charts.
Results: The joint hypermobility syndrome (JHS) was diagnosed in 32 people (11.4% of the population studied) with mean age 15.7 years, predominance in the female sex and in the white skin group (P<.01). Fifty-three point one percent of those patients with hypermobility presented skin lesions associated to hereditary diseases of the connective tissue; among them the most relevant clinical sign was the presence of hematomas (P=.003). Symptoms of dysautonomia were associated to JHS (P≤.05) and correlated positively with the presence of hematomas. Moderate and severe chronic pain was also a feature of patients with the syndrome (P=.001) and was correlated in a positive manner to the hematomas.
Conclusions: Vascular affection as demonstrated by the formation of hematomas was the skin lesion more important among young people with JHS in this study. These lesions can be representative of the syndrome and a translate a larger damage at the connective tissue level.
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Síndrome de hipermovilidad articular en una población juvenil cubana

Palabras clave:
Síndrome de hipermovilidad articular
Población joven
Disautonomía
Dolor crónico
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RESUMEN

Para investigar la frecuencia de aparición del síndrome de hipermovilidad articular (SHA), identificar los cuadros asociados a este diagnóstico y correlacionar los síntomas más significativos, se realizó un estudio descriptivo-analítico en un grupo de 280 jóvenes de ambos sexos con edades comprendidas entre los 15 y los 17 años. Para la obtención de los objetivos se elaboró por parte de los autores un modelo de encuesta. Los datos se procesaron por estadística descriptiva utilizando el programa Epidat 3.1. Los resultados se muestran en tablas confeccionadas al efecto.
Resultados: El SHA se diagnosticó en 32 individuos (11,4% de los encuestados) con una media de edad de 15 años, predominio en el sexo femenino y en el grupo de población con color de piel blanca (p < 0,01). El 53,1% de los hipermóviles presentó lesiones cutáneas de las asociadas a las enfermedades hereditarias del tejido conjuntivo y, entre ellas, la presencia de los hematomas resultó significativa (p = 0,003). Los síntomas de disautonomía se asociaron al SHA (p ≤ 0,05) y se correlacionaron positivamente con los hematomas. El dolor crónico moderado y grave apareció en los individuos con el síndrome (p = 0,001) y también se correlacionó en forma positiva con los hematomas.
Conclusiones: La afectación vascular demostrada por la formación de hematomas fue el tipo de lesión cutánea más frecuente entre los hipermóviles jóvenes; esta lesión podría estar representando una forma más sintomática del síndrome y una mayor profundidad de daño a nivel de tejido conjuntivo.
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Introduction

Soft tissue rheumatism occupies one of the first places among the causes of morbidity due to non-transmissible chronic diseases in the juvenile population.[1] Among the wide group of situations that are currently mentioned as a probable origin of these types of problems, the hyperlaxitude or generalized joint hypermobility syndrome (JHS), stands out.[2]

Prevalence is estimated between 10% and 25% of the general population with variations according to gender, age and race.[3] The spectrum of clinical manifestations in JHS is wide and may vary from presenting single to multiple characteristics, with them being or not joint-related. The ones most frequently found are joint pain, arthritis, sprains, luxations, and skeletal deformities.[4] It has been associated to spontaneous pneumothorax, mitral valve prolapse,[5] anxiety,[6] and fibromyalgia.[7] However, in spite of the syndromes' symptoms pleomorphic behavior, it is impossible to predict its clinical behavior once it has been diagnosed, making it easy to overlook.

Its prevalence and associated symptoms are known in the Cuban pediatric population,[8] but its behavior in other age groups is unknown.

The present study had the objectives of investigating the frequency of JHS in a group of patients with ages ranging between 15 and 17 years of age, describing the most important symptoms associated to the diagnosis and identifying a relationship between them.

Patients and methods

A transversal, analytic-descriptive study was performed in 321 individuals of one gender or the other, with ages between 15 and 17 years, residing in the municipality of Güines. All of the patients were students at a technology center at their city for the period between January and June of 2005.

Inclusion criteria were:

1. Being registered at school.
2. No history of inflammatory or connective tissue disease.
3. No consumption of vitamin C, steroids, phenytoin, D-penicillamine or other drug or substance with an influence on connective tissue metabolism for the 6 month period prior to study inclusion.

Two hundred eighty individuals were included after signing an informed consent form. A survey designed by the authors was then applied to them to gather data such as: age, gender, skin color (grouped as white and non white, with the latter including patients of African or Asian descent or a mix of these 3 groups), data on chronic somatic pain (lasting ≥12 weeks) and its intensity (mild, moderate, and severe), skin signs (such as the formation of a skin fold, thinned skin lesions, and hematomas), bone and joint alterations detected upon physical examination and dysautonomous signs and symptoms, such as fainting, chest pain, orthostatic tachycardia and palpitations.

Criteria employed for the diagnosis of generalized joint hypermobility were those of JHS, but modified in the number of lax positive joints to 5 or more of the 9 mentioned by Beighton[9] for diagnosis at these ages.

Statistical analysis

The information obtained was put in a database and processed using Epidat 3.110 software. Data was analyzed as number and percentages. To evaluate the association to different factors, χ^2 with Yates correction was used, taking into account a 95% confidence interval (P≤05). To evaluate the correlation between hematomas and dysautonomy symptoms, Cramer's V correlation coefficient was used for nominal dychotomic variables. To evaluate the correlation with chronic pain, Spearman's rank coefficient was used for ordinal variables. Results are presented in tables.

Results

The total number of patients selected for this study was 280: eighty-nine male (31.7%) and 191 female (68.2%). By skin color, 221 (78.8%) were white and 59 (21.0%) non-white.

Table 1 shows the distribution of the 32 individuals (11.4%) who filled the JHS diagnostic criteria. Mean age was 15.7 years (SD, 0.71) with female (4:1) and white skin predominance (P≤.01). Some bone and joint alterations were found, their presence in hypermobility syndromes being statistically significant, such as lax foot (P<.05), knee valgum (P<.05), and somatic pain referred during survey application (P<.001) (Table 2).

53% of JHS patients had skin lesions (table 3), with spontaneous or minimal trauma induced hematomas being the most frequent lesion (p<0,003).

Table 4 presents symptoms of dysautomomous disorder seen in the studies patients and its relation to the JHS. The presence of chest pain (p<0,02), palpitations (p<0,05), intolerance to physical exercise (p<0,04) and orthostatic tachycardia (p<0,001) were statistically significant among hypermobile individuals.

Table 5 shows the correlation between the presence of skin lesions (hematomas) and dysautonomous syndrome, as well as between the presence of hematomas and chronic pain according to severity. Hematomas were seen to directly and significantly correlate with the presence of chest pain, palpitations and orthostatic tachycardia, as well as the mild, moderate and severe pain categories.

Discussion

The frequency of JHS among the subjects studied was 11.4%. This is within the universally calculated interval (range)[11] and is somewhat

Table 1
Distribution of the joint hypermobility syndrome according to age, gender, and skin color

Age, y	Male			Female		
	White	Non-white	Total	white	Non-white	Total
15	1	1	2	7	4	11
16	2	0	2	7	5	12
17	1	1	2	2	1	3
Total	4	2	6	16	10	26

Mean age, 15.75 (0.71). Skin color, white (P≤.01). Gender, 4:1

Table 2
Bone and joint alterations and its relation with the joint hypermobility syndrome

Bone and joint alterations	Non hypermobile		Hypermobile		P value
	No.	%	No.	%	
Scoliosis	47	19	9	28	.22
Khyphosis	10	4	1	3.1	.81
Hyperlordosis	12	4.9	1	3.1	.98
Knee varum	15	6	2	6.2	.72
Knee valgum	28	11.2	8	25	.05
Knee synovitis	0	0	1	3.1	.22
Recurvatum knee	31	12.5	6	18.8	.48
Ankle valgum	3	12	1	3.1	.94
Lax foot	46	18	11	34	.05
Flat metatarsum	28	11.3	1	0	.26
Somatic pain	59	23.8	32	100	.001

Source: survey.

Table 3
Skin alterations and its relation to the joint hypermobility syndrome

Lesion	Non hypermobile		Hypermobile		*P* value
	No.	%	No.	%	
Thinned skin lesion	71	28.6	6	18,8	.23
Hematoma	18	7.3	8	25	.003
Skin fold >4 cm	30	12	7	21	.12

Source: survey.

Table 4
Dysautonomy and hypermobility symptoms

Symptom	Non hypermobile		Hypermobile		*P* value
	No.	%	No.	%	
Exercise induced fainting	22	89	7	21.9	.04
Loss of consciousness	3	1.2	1	3.1	.94
Palpitations	14	5.6	5	15.6	.05
Thoracic pain	16	6.4	6	18.8	.02
Heat intolerance	11	4.4	3	9.4	.43
Orthostatic hypotension	1	4	0	0	.98
Orthostatic tachycardia	0	0	2	6.2	.001

Source: survey.

Table 5
Correlation between hematomas, dysautonomous syndrome, and chronic pain

Skin	Dysautonomous syndromes	R [a]	P value
Hematomas	Fainting	0.91	.0003
Hematomas	Thoracic pain	0.83	.001
Hematomas	Orthostatic tachycardia	0.44	.09
Hematomas	Palpitations	0.74	.0003
Hematomas	Chronic pain	0.58	.001
Hematomas	Chronic severe pain	0.59	.01
Hematomas	Chronic moderate pain	0.59	.01
Hematomas	Chronic mild pain	0.06	.45

[a] Value reached with the use of study correlations that can vary from -1 and 1.

less than that reported in other areas of the continent[12] but the same as that obtained in a study performed in a population of patients 11 to 14 years old done in our country.[8]

The prevalence of the syndrome was related to the ethnic origin of the patients, being larger in Asians and Africans than Europeans.[13-15] However, this larger prevalence in whites could be related to a greater representativity of this group among those studied and/or a marked admixture of the population, occasionally impossible to classify with ethnic criteria such as skin color or hair characteristics. This result could point out to a distinctive feature of JHS in our setting. The syndrome was also proven to be more frequent in females and, although described in equal proportions between genders in a pediatric population,[16] studies performed in older children point to a female predominance that rises until reaching the adult population.[17]

Some bone and joint alterations were associated to hypermobility as seen in other groups.[18] Somatic pain was present in all hypermobile patients. When compared with other studies in our setting, this symptom was seen more frequently with advancing age in patients with JHS, passing 60% at 13 years of age[8] to 100% in the present group (16 years). The result is the same to that seen by other researchers[19,20] and makes the syndrome the most frequent cause of chronic pain in youngsters this age.

On the other hand, in this series, 53.1% of patients had skin lesions described in association to connective tissue diseases (CTD). The most symptomatic cases of JHS were associated to the presence of these lesions.[21] Research has found low levels of tenascin-X (an extracellular matrix protein) in subjects with CTD and JHS. Cases presenting a greater deficiency in the expression of the B gene of tenascin X (TNX-B) have been related to the JHS.[22] These results suggest a bimodal behavior of the syndrome, which could be genetically determined and related with the presence or absence of skin lesions. However, other studies have not confirmed the relationship between skin elasticity and the formation of ulcers in the more symptomatic presentations of JHS.[23]

In the present serried, the formation of hematomas was significant; other researchers have also found a predominance of these lesions in the syndrome,[24] making this finding a possible diagnostic marker for JHS in the region.

Some dysautonomic syndromes were significantly present among the hypermobile patients in the study, as has been communicated by other authors.[25] The intensity of these symptoms is less than that referred in multiple sclerosis[26] and in some forms of epilepsy,[27] but the same as that described in fibromyalgia.[28] The origin of the dysautonomic syndrome in JHS is unknown. In this study it correlated positively with the presence of hematomas. Some authors recognize that the alterations in the vessel wall could be seen as a possible cause of hypertensive vascular dysfunction leading to dysautonomy.[29] As far as we know this possibility has not been explored in the syndrome, although our results suggest a very close relationship between hematoma formation and the appearance of dysautonomy symptoms in JHS.

Chronic pain also showed a relationship to hematomas in this study, which could point to the influence that the vascular lesion could have in the development of symptom intensity more than in its origin, because the relationship only existed in cases of moderate and severe chronic pain.

In summary, JHS was seen in 11.4% of young patients studied, with a bimodal pattern indicated by the presence of skin lesions. Vascular affection was demonstrated though the formation of hematomas and was the most statistically significant lesion seen among hypermobile patients and correlated with the appearance of dysautonomic symptoms and with a more intense chronic pain syndrome. In contrast with other authors, who did not find any relevance to the appearance of skin lesions,[30] we think that the formation of hematomas in patients with JHS corresponds to a more symptomatic form of the syndrome and could represent a marker which indicates a more profound damage at the level of connective tissue.

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