ANDES PEDIATRICA
REVISTA CHILENA DE PEDIATRÍA
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Andes pediatr. 2022;93(1):53-58
DOI: 10.32641/andespediatr.v93i1.3755

ORIGINAL ARTICLE

# Prevalence of joint hypermobility, postural orthostatic tachycardia syndrome (POTS), and orthostatic hypotension in school-children

## Prevalencia de la hipermovilidad articular, el síndrome de taquicardia postural (POTS) y la hipotensión ortostática en escolares

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Received: March 25, 2021; Approved: August 8, 2021

**What do we know about the subject matter of this study?**

Joint hypermobility (JH) is characterized by ligamentous laxity and musculoskeletal pain; postural orthostatic tachycardia syndrome (POTS) is the development of symptoms with upright positioning and increase heart rate that improve with recumbence. Children with both JH and POTS report dizziness, syncope, nausea, headache and palpitations.

**What does this study contribute to what is already known?**

This is the first study to assess the prevalence of POTS and orthostatic hypotension (OH), and the first study to assess the association between JD, OH and POTS in a community of school-children.

## Abstract

Children with joint hypermobility, postural orthostatic tachycardia syndrome, and orthostatic hypotension report autonomic symptoms such as dizziness, nausea, headaches, and palpitations. It is unclear if there is a pathophysiological link between connective tissue disorders and autonomic symptoms. There is no published data on the prevalence of disorder at the community level. **Primary objective:** To assess the prevalence of joint hypermobility, orthostatic hypotension, and postural orthostatic tachycardia syndrome in children. **Secondary objective:** To determine the relationship between joint hypermobility, orthostatic hypotension, and postural orthostatic tachycardia syndrome. **Patients and Method:** Participants aged 10 to 18 years were selected from public schools in three Colombian cities. The surveys included historical questions on the incidence of dizziness, nausea, headache, tremor, blurred vision, vertigo, anxiety, near syncope and syncope, sweating, palpitations triggered by standing in the two months prior to the investigation. Each of these signs and symptoms

**Keywords:**
Joint Hypermobility; Postural Orthostatic Tachycardia Syndrome; Postural Hypotension; Autonomic Symptoms; Beighton Scale

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Edited by:
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How to cite this article: Andes pediatr. 2022;93(1):53-58. DOI: 10.32641/andespediatr.v93i1.3755

was also assessed during the recumbency (10 minutes) and standing (2, 5 and 10 minutes) phases of the investigation. HR and BP measurements were obtained at the same intervals. Joint mobility was measured with a mechanical goniometer and assessed with the Beighton score. **Results:** Prevalence of joint hyperlaxity: 87 of 306 (28.4%). Prevalence of orthostatic hypotension: 5 of 306 (1.6%). Prevalence of postural orthostatic tachycardia syndrome: 6 of 306 (2.0%). Of 87 children with joint hyperlaxity, only 1 child had joint hyperlaxity at the same time as postural hypotension (1.2%) ($p = 0.6735$), and 1 child had joint hyperlaxity and postural orthostatic tachycardia syndrome simultaneously (1.2%) ($p = 0.5188$). **Conclusion:** Children with joint hyperlaxity did not have a higher prevalence of postural orthostatic tachycardia syndrome and orthostatic hypotension. It seems unlikely that connective tissue disorders are responsible for most cases of postural orthostatic tachycardia syndrome and orthostatic hypotension in the community. Of note, the pathophysiology of postural orthostatic tachycardia syndrome and orthostatic hypotension requires further investigation.

## Abbreviations

Blood pressure (BP), Ehlers-Danlos syndrome (EDS), Functional gastrointestinal disorders (FGIDS), Heart rate (HR), Joint hypermobility (JH), Orthostatic Hypotension (OH), Orthostatic intolerance (OI), Postural orthostatic tachycardia syndrome (POTS).

## Introduction

Joint hypermobility (JH) is a benign condition characterized by generalized ligamentous laxity and the presence of musculoskeletal pain without signs of systemic rheumatology disease[1]. It was clinically defined by the presence of a Beighton score of at least 4 out of 9[2]. Frequently, children report joint hypermobility as a sole symptom that does not interfere with daily life[3]. Few studies have assessed the prevalence data of JH in school-children using standardized and rigorous methods[4,5].

The development of somatic symptoms with the upright posture that improves with recumbence is defined as orthostatic intolerance (OI)[6]. Some of the children that develop symptoms with orthostasis meet diagnostic criteria for postural orthostatic tachycardia syndrome (POTS) and Orthostatic Hypotension (OH). The diagnostic criteria for POTS requires symptoms with upright positioning and increase of heart rate (HR) without a decrease in blood pressure (BP), while those children who also lower their BP are diagnosed as OH[3].

Children with both forms of joint hypermobility (h-EDS and JH) report similar autonomic symptoms to children with POTS and OH. Those include dizziness, syncope, nausea, headaches and palpitations[6]. Despite the common perception that symptoms of POTS are frequently found amongst children that consult to clinical practice, the prevalence of POTS and OH is not well known[7]. Moreover, no studies have assessed the prevalence of POTS and OH at the community level. It is also unclear if the presence of autonomic symptoms in JH is explained by a common pathophysiological link, or result from two common conditions that overlap due to chance.

The primary aim was to evaluate the prevalence of JH, POTS, and OH in a community of school-children. Our secondary aim was to determine the relationship between JH, POTS, and OH. We hypothesized that patients with JH are more likely to have POTS and OH than those without JH.

A previous study by our group that assessed the relation between functional gastrointestinal disorders (FGIDS), JH, POTS and OH found that children with FGIDs were not more likely to have POTS and OH. Therefore, we combined the data of children with and without FGIDs to study two important questions that have not yet been answered, what is the prevalence and whether there is an association between JH, POTS and OH at the school level?

## Patients and Method

### Study population

Participants were selected from public schools from three Colombian cities [Cali (southwest region), Palmira (southwest region) and Bucaramanga (northeastern region)]. Parents were sent informative letters explaining the study protocol, without revealing the aims and hypothesis of the study. Children between the ages of 10-18 years whose parents consented to their participation in the study and themselves assented to participate were included in the study. Physicians examined the children in school for signs of dehydration at baseline. Children with dehydration, history of organic gastrointestinal diseases, and/or eating disorders were excluded. Due to the low prevalence of connective tissue disorders such as Marfan Syndrome,

it wasn't taken into consideration when excluding patients for the probability of actually finding one within our sample.

**Study protocol**

Participants completed binary surveys (yes/no) regarding symptoms commonly associated with OI. Surveys included historical questions on the incidence of dizziness, nausea, headaches, tremors, blurred vision, vertigo, anxiety, near syncope and syncope, sweating, palpitations triggered by standing in the two months prior to the investigation. Each of these sign and symptoms was also assessed during the recumbence (10 minutes) and standing phase (2, 5, and 10 minutes) of the investigation. Baseline HR and BP measurements were obtained using an automatic blood pressure monitor (SCIAN LD-582) at the same intervals.

Joint mobility was measured using a mechanical goniometer (Goniómetro Profesional MSD), and was evaluated with the Beighton score (bilateral hypermobility of the fifth fingers, thumbs, knees, and elbows, and the ability to place palms flat on the floor with knees straight)[9].

**Environment**

Testing was done at each school in a quiet, temperature-controlled, isolated room absent of distractions. Members of the research team were readily available to help complete questionnaires during the study.

**Definitions**

Joint hypermobility was clinically defined by the presence of a Beighton score of at least 4 out of 9[1]. Orthostatic hypotension was defined as a drop in BP within 2 minutes of standing up of at least 20 mmHg systolic BP or at least 10 mmHg diastolic blood pressure[8]. Postural orthostatic tachycardia syndrome was defined as an increase of HR of at least 40 beats per minute during the time period of the upright position (10 minutes) without a decrease of systolic BP $\geq$ 20 mmHg[6], in adolescents 18 years or younger.

**Statistical analysis**

Data was calculated as percentages, means (standard deviation, [SD]; 95% confidence intervals, [95% CI]). Age, sex, and the frequency of diagnosis of POTS and/or OH were assessed and compared between the group of children with and without JH. The differences in characteristics between school-children with and without JH were compared with t-test for continuous variables and with a Fisher's exact test (2 $\times$ 2 contingency tables) for categorical variables. Univariate analysis and bivariate analysis were calculated. Odds ratios (OR) were calculated. Statistical significance was set at $P < 0.05$.

**Study approval**

The study was approved by the Institutional Review Board and Human Subjects Committee of Universidad del Valle of Cali, Colombia.

## Results

A total of 306 school-children (109 females, $P = 0.129$) from public schools from three cities were included in the study. The ethnicity of the population sample was 146 (47.7%) mixed-race, 128 (41.8%) black, 23 (7.6%) white and 9 (2.9%) indigenous; age was reported as 80 (26.1%) children between 10 and 12 years of age, and 226 (73.9%) adolescents between 13 and 18 years old. There were no children excluded due to dehydration.

**Aim 1. Prevalence**

Out of 306 participants, 87(28.4%) children met diagnostic criteria for JH, 6 (2.0%) were diagnosed with POTS and 5 (1.6%) with OH.

**Aim 2- Relation between JH, OH, and POTS**

Of the 87 children with JH, only 1 child had both JH and OH (1.2%) (P=0.6735) and only 1 child had both JH and POTS (1.2%) (P=0.5188). Overall, there was no significant difference in the prevalence of OH and POTS between those with JH and those without (table 1).

**Symptoms**

The three most common symptoms that were reported in the 2 months prior to the investigation and also developed during orthostatic testing were headache (11.0%), feeling hot (11.0%), and vertigo (10.1%) (table 2).

## Discussion

This is the first study to assess the prevalence of POTS and OH, and the first study to assess the association between JH, OH, and POTS in a community of school-children. Although symptomatic complaints that mimic autonomic disorders are common amongst children in clinical practice, until now, we were unable to establish the prevalence of OH and POTS in children at the community level.

Our primary aim was to evaluate the prevalence of JH, POTS, and OH in school-children. Similar to our previous school-based study that included 272 children and found that 28.3% had JH[1], in our current study, 28.4% of school-children had JH. This study confirms that JH is common outside of the clinical setting. Although symptoms of POTS are commonly reported in clinical prac-

**Table 1. Relation between joint hypermobility, orthostatic hypotension, and postural orthostatic tachycardia syndrome (POTS) in a community of school-children**

| | Beighton Score | | OR | IC95% | p |
|---|---|---|---|---|---|
| | Positive (%) 87 (28.4) | Negative (%) 219 (71.6) | | | |
| Orthostatic Intolerance | | | | | |
| No | 31 (35.6) | 82 (37.4) | 1.00 | | |
| Yes | 56 (64.4) | 137 (62.6) | 1.08 | 0.62-1.88 | 0.7672 |
| Orthostatic Hypotension | | | | | |
| No | 86 (98.9) | 215 (98.2) | 1.00 | | |
| Yes | 1 (1.2) | 4 (1.8) | 0.62 | 0.01-6.44 | 0.6735 |
| Postural orthostatic tachycardia syndrome | | | | | |
| No | 86 (98.9) | 214 (97.7) | 1.00 | | |
| Yes | 1 (1.2) | 5 (2.3) | 0.49 | 0.01-4.54 | 0.5188 |

**Table 2. Two-month history of symptoms compared to symptoms during testing**

| Symptoms | 2 month history (N = 306) | Children with a 2 month history of symptoms who also developed symptoms at 10 minutes of orthostasis | Children with a 2 month history of symptoms who were symptom free at 10 minutes of recumbence | Children with a 2 month history of symptoms that were consistent with testing |
|---|---|---|---|---|
| Dizziness | 190 (62.1%) | 41 (13.4%) | 14 (4.6%) | 23 (7.4%) |
| Nausea | 79 (25.8%) | 6 (2.0%) | 3 (1.0%) | 12 (3.8%) |
| Headache | 157 (51.3%) | 47 (15.4%) | 18 (5.9%) | 34 (11.0%) |
| Tremulousness | 50 (16.3%) | 5 (1.6%) | 1 (0.3%) | 6 (2.0%) |
| Blurry vision | 118 (38.6%) | 9 (2.9%) | 6 (2.0%) | 16 (5.1%) |
| Feeling hot | 96 (31.4%) | 20 (6.5) | 11 (3.6%) | 34 (11.0%) |
| Vertigo | 79 (25.8%) | 14 (4.6%) | 8 (2.6%) | 31 (10.1%) |
| Feeling anxiety | 76 (24.8%) | 18 (5.9%) | 4 (1.3%) | |
| Feeling faint | 59 (19.3%) | 10 (3.3%) | 5 (1.6%) | 26 (8.5%) |
| Sweatiness | 86 (28.1%) | 13 (4.2%) | 7 (2.3%) | 25 (8.1%) |
| Palpitations | 54 (17.6%) | 5 (1.6%) | 2 (0.7%) | 11 (3.7%) |
| Loss of consciousness | 6 (2.0%) | 0 (0.0%) | 0 (0.0%) | 0 (0.0%) |

tice, our findings suggest that the frequency of POTS is rare at the community level. The prevalence of OH and POTS in our sample was 1.6% and 2.0% respectively. Selection bias may explain the high prevalence of children with POTS consulting to tertiary care centers[9].

Our secondary aim was to determine the relationship between JH, POTS and OH. We found that those with JH were not significantly more likely to be diagnosed with OH, and/or POTS than those without JH. Again, selection bias may explain the frequent comorbidity of JH and autonomic dysfunction among patients seeking care in specialized clinics[9]. Based on our findings, in the community setting, JH may either not be related to autonomic dysfunction, or the diagnostic criteria for POTS and OH is too strict and may not capture some of the children that report signs and symptoms of autonomic dysfunction.

Joint hypermobility is a benign condition and comorbid autonomic symptoms should not be loosely attributed to this disorder. Although connective tissue disease has been linked to autonomic dysfunction due to vascular tissue laxity[10], our study suggests that, at

least at the community level, both POTS and OH are not related to JH.

Our study is not without limitations. The results of our study may not be generalizable to other settings. While we were able to establish the prevalence of JH and autonomic disorders in a sample of schoolchildren, we did not utilize a tilt-table-test despite, the gold standard for POTS diagnosis. However, based on surveys given to pediatric electrophysiologists, tilt-table-testing is of limited utility at least in patients with severe autonomic symptoms such as syncope[11] and therefore the absence of this test in our study may not be of relevance. Also, due to the subjective nature of most of the symptoms and the age of our patient sample, another limitation yet arises; nonetheless, subjective symptoms are common in almost every pathology and must be taken into consideration when defining patient's symptomatology.

Some of the strengths of this investigation include the school-based setting which allowed us to study children without the selection bias from those seeking medical attention for severe symptoms. Some of the additional strengths of our study include the use of the Beighton score and a goniometer to measure JH. The design of our labor-intensive and rigorous study included measuring BP and HR by physicians in a large number of children in a quiet, temperature-controlled environment.

In conclusion, while JH is common at the community level, POTS and OH are uncommon. Children with JH were not found to have a higher prevalence of POTS and OH. Thus, it is unlikely that connective tissue disorders are responsible for most cases of POTS and OH at least at the community level. The pathophysiology of POTS and OH warrants further investigation. Large multinational studies are needed to confirm our findings.

## Ethical Responsibilities

**Human Beings and animals protection:** Disclosure the authors state that the procedures were followed according to the Declaration of Helsinki and the World Medical Association regarding human experimentation developed for the medical community.

**Data confidentiality:** The authors state that they have followed the protocols of their Center and Local regulations on the publication of patient data.

**Rights to privacy and informed consent:** The authors have obtained the informed consent of the patients and/or subjects referred to in the article. This document is in the possession of the correspondence author.

## Conflicts of Interest

Authors declare no conflict of interest regarding the present study.

## Financial Disclosure

Authors state that no economic support has been associated with the present study.

## Acknowledgments

This article is part of a PhD Thesis at the University of Granada (Spain). Research line: Pathophysiology of medical-surgical diseases. We received no financial support for this investigation. There are no conflict of interest to disclose.

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