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## Brief communication

# Prevalence of juvenile idiopathic arthritis in children aged 6 to 12 years in Embu das Artes, state of São Paulo, Brazil

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### ABSTRACT

The aim of the study was to study the prevalence of juvenile idiopathic arthritis (JIA) in school children in the city of Embu das Artes in São Paulo State. 2880 school children from seven public schools, aged between 6 and 12 years, were evaluated (clinical findings) by a pediatric rheumatologist. A board certified Pediatric Rheumatologist evaluated the subjects with suspected inflammatory arthropathy. Children with higher suspicion were referred to a specialized service. One hundred and forty-one children have presented abnormalities on examination of musculoskeletal system, with isolated pain on palpation the most common finding in the first evaluation (60.9%), with improvement in almost all cases in the second examination. Most of the abnormalities were related to recent injuries or congenital malformations. Six children have clinical findings suggestive of chronic arthropathy and were referred to a specialized pediatric rheumatology clinic. Of these, a 12 year-old girl fulfilled the criteria for JIA. The other diagnoses were aseptic necrosis of the hip ( $P = 1$ ) of and post-trauma synovitis ( $P = 4$ ). The prevalence of JIA in children aged between 6 and 12 years was 1/2.880 (or 0.34/1.000).

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## Prevalência da artrite idiopática juvenil em crianças com idades entre 6 e 12 anos na cidade de Embu das Artes, SP

### RESUMO

O objetivo do estudo foi determinar a prevalência da artrite idiopática juvenil (AIJ) em crianças escolares de na Cidade do Embu das Artes do Estado de São Paulo. 2880 escolares provenientes de 7 escolas públicas, com idades entre 6 e 12 anos, foram examinados por um especialista em Reumatologia Pediátrica. Nos casos de alteração do exame reumatológico os indivíduos foram examinados novamente por um outro especialista com título na área de atuação em Reumatologia Pediátrica. Os casos suspeitos de artropatia inflamatória foram encaminhados para um serviço especializado. Cento e quarenta e uma crianças apresentaram alterações no exame do aparelho locomotor, sendo a dor à palpação isolada o achado mais comum na primeira avaliação (60,9%), com melhora em praticamente 100% dos casos na segunda avaliação. A maioria das alterações foi relacionada a traumatismos recentes ou malformações congênicas. Seis crianças apresentaram exame suspeito de artropatia crônica e foram encaminhadas para avaliação em um centro de referência em

#### Palavras-chave:

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Dor

Exame físico

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Reumatologia Pediátrica. Destas, uma menina de 12 anos preencheu os critérios para AIJ. Os outros diagnósticos foram necrose asséptica da cabeça do fêmur (1 paciente) e sinovite pós-trauma (4 pacientes). A prevalência da artrite idiopática juvenil em crianças com idades entre 6 e 12 anos foi de 1/2.880 (ou 0,34/1.000).

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## Introduction

The diagnosis of juvenile idiopathic arthritis (JIA) is based on the criteria of the International League of Associations for Rheumatology (ILAR),<sup>1</sup> and requires the presence of arthritis lasting at least six weeks in the same joint in children aged less than 16 years, in addition to the exclusion of other causes of chronic arthritis. In many cases, aggressive arthritis and destruction of articular cartilage is observed, with decreased exercise capacity within a short period of time. However, this scenario can be minimized when the diagnosis is early and treatment is conducted according to universally accepted protocols.<sup>2,3</sup> However, many patients fail to seek specialized centers at the initial manifestation of the disease; even when submitted to appropriate treatment, they develop irreversible structural sequelae.<sup>4-6</sup>

The delayed diagnosis is due to of disease-specific factors, such as the fact that many patients with JIA are oligosymptomatic and, in the presence of pain, many parents associate it to a recent trauma. Another source of delay is the possibility of atypical clinical conditions, such as arthritis of small joints, in addition to the absence of specific laboratory tests for the diagnosis of JIA.<sup>6-8</sup> An additional factor that contributes significantly to the delay, both of diagnosis and treatment, is an ineffective referral system to specialists, who are typically found in large centers.

The development of strategies to optimize patient referral depends on the knowledge of epidemiological data for each disease. However, there are no Brazilian data on the prevalence of JIA. International data show a prevalence of 0.07 to 4.01/1,000 children,<sup>6,9-16</sup> based on studies with different methodologies, such as reviews of medical records, population registries, and field studies.<sup>17</sup>

In Brazil, there are no reliable data on the epidemiology of JIA at national or local level, as the disease is not of compulsory notification and there is no national or regional data registry. The objective of this study was to determine the prevalence of JIA in children in a city in the state of São Paulo, from a field study based on physical examination of children attending public schools.

## Methods

This was a cross-sectional, field study. Children and adolescents enrolled in the municipal and state schools in the city of Embu das Artes, a tourist resort located 27 km from the city of São Paulo, capital of the most populous state in Brazil. This city has 34 state and municipal public schools, attended by 98% of children and adolescents, with free education in all of them.

This study assessed children aged between 6 and 12 full years attending elementary school. According to the Education Secretariat of Embu das Artes, 17,289 children within this age group are enrolled in the city's schools. Seven schools were randomized, of which four were state and three were municipal.

After contacting the principals of each of the schools, letters were sent to all parents or guardians containing a brief explanation of the protocol and requesting them to sign an informed consent, which was signed by the parents of 2,880 children, who were then evaluated.

All children were assessed by a specialist in pediatric rheumatology (EY), with three years of clinical experience, in quiet rooms equipped with floor mats, always in the presence of a teacher or the principal. The rheumatologic examination consisted of inspection, palpation, and assessment of joint range of motion of the following joints: cervical and thoracic spine, temporomandibular joint, shoulders, elbows, wrists, small joints of the hands, sacroiliac joints, hips, knees, ankles, and small joints of the feet.

Arthritis was defined as the presence of joint swelling or the presence of two of the following alterations: pain, increased temperature, redness, and movement limitation. The examination included palpation of entheses to search for enthesitis.

All children who showed such alterations were evaluated within 30 days by a second board-certified physician with over 15 years of experience in the specialty (CAL).

Parents and principals were informed that children with arthritis in one or more joints, or any previously undiagnosed musculoskeletal abnormality, would be referred to the Outpatient Clinic of Rheumatology of the Department of Pediatrics to undergo imaging studies, laboratory tests, and regular monitoring.

The project, which was approved by the research ethics committee, has received research grants from the Fundação de Amparo à Pesquisa of the State of São Paulo (Process 05/50761-1).

## Results

Of the 4,210 children who took the letter to their parents, 68.4% brought the informed consent signed by their parents or guardians, totaling 2,880 children, who were included in the study (1,469 males and 1,411 females). The following age distribution was observed: 6 years, 12.1%; 7 years, 17.2%; 8 years, 24.1%; 9 years, 24.6%; 10 years, 18.3%; 11 years, 3.3%; and 12 years, 0.4%.

Upon physical examination of the musculoskeletal system, 187 alterations were detected in 141 children (4.9%)

(Table 1). Pain on palpation alone was the most common finding in the first assessment (60.9%), with improvement in almost 100% in the second evaluation.

Most of the alterations were related to recent trauma or congenital malformations. Six children had suspected chronic arthropathy on physical examination (joint limitation and/or increased volume), with no previous history of JIA. After verbal contact with parents, all were referred to the University's pediatric rheumatology outpatient clinic.

The definitive diagnosis of JIA was achieved only in one 12-year-old girl, born in Embu, without clinical follow-up since age 3 years. The mother reported that the patient presented with polyarthritis at 2 years of age, accompanied by intermittent fever, irritability, and inability to ambulate. The local medical service was sought and she underwent treatment with non-steroidal anti-inflammatory drugs for a few months. She was discharged, but since the age of 3 years she had intermittent pain in the ankles, knees, wrists, and elbows. In some situations, she presents difficulty to walk, and she does not practice sports activities due to her joint condition. On physical examination, performed at school and later at the pediatric rheumatology outpatient clinic, knee, ankle, wrist and elbow involvement were observed, with increased volume and temperature. The patient met the criteria for the diagnosis of JIA. She was enrolled and is being treated by the multidisciplinary team (rheumatologists, physical therapists, and psychologists).

The other diagnoses were aseptic necrosis of the femoral head (one patient without proper medical supervision) and post-traumatic synovitis (four patients).

With these data, the prevalence of JIA by active search through physical examination was determined to be 1/2,880 (or 0.34/1,000) in children aged 6 to 12 years in the city of Embu das Artes, Brazil.

## Discussion

In the present study, the prevalence of the main chronic arthropathy of childhood in a Brazilian population was determined, which was 0.34/1,000 children aged 6 to 12 years, from a field study carried out through the systematic examination of the musculoskeletal system of schoolchildren. The present data are consistent with the international literature, as the prevalence observed in several regional studies varies from 0.07 to 4.1/1,000 children.<sup>6,9-14,17</sup>

As emphasized by Manners and Bower,<sup>17</sup> the variability in prevalence observed in several epidemiological studies per-

formed in recent decades is influenced by the methodology used, difficulties in diagnosing the disease, variation in sample sizes, and time used by each author for data collection.

A community-oriented methodology was applied in the present study, focused on clinical examination of children with or without a previous diagnosis of arthritis, which has been used in two studies: that by Mielants et al. (Belgium, 1993)<sup>18</sup> and by Manners (Australia, 1999).<sup>6</sup>

In the Belgian study, a total of 2,990 children aged up to 18 years selected from a questionnaire on rheumatic manifestations were assessed; five of whom received a definitive diagnosis of chronic arthropathy, indicating a prevalence of 1.67/1,000 children. In the Australian study, 2,241 students were assessed at 12 years of age, and nine met the diagnostic criteria for chronic arthropathy, with a prevalence of 4.01/1,000 children. Of the nine cases identified by Manners,<sup>6</sup> seven had no prior diagnosis. Studies in the community tend to represent reality more faithfully than those based on data from clinical studies and hospitals, despite several possible limitations.

The determination of an age range for the study population, the methodology used in the present study and in the Australian study<sup>6</sup> may underestimate the true prevalence of JIA. A significant age group was excluded, and individuals aged 13 or older were not examined. This criticism, which is important, raises the question of the need for studies involving a broader age range. It is known that, in a considerable number of patients with arthritis, disease onset is between 13 and 16 years, especially the enthesitis-related arthritis and polyarticular arthritis with negative rheumatoid factor subtypes.

A second criticism of both studies is that patients with severe and disabling JIA might not be attending school, and therefore were not assessed. Certainly, studies that include home visits, focusing on bedridden children and those in wheelchairs would be useful for better data reliability. It is noteworthy that none of the schools visited had adequate facilities for disabled individuals, such as ramps, lifts, or even adapted classrooms and bathrooms, which certainly reflects the scenario observed in other schools in Brazil.

The present study, as well as those by Mielants et al.<sup>18</sup> and Manners,<sup>6</sup> identified children who met the criteria for chronic arthropathy who were not undergoing treatment, and therefore subject to the natural clinical course of the disease. The delay in starting the treatment is a reality observed in many pediatric rheumatology services.<sup>8</sup> A multicenter study performed in the state of São Paulo showed that 35% of patients experience delayed referral to centers specialized in pediatric rheumatology.<sup>19</sup>

Ehrmann-Feldman et al.<sup>8</sup> evaluated the medical history of 352 Canadian children with suspected JIA, and observed that the time between consultation with a nonspecialist clinician and a pediatric rheumatologist was 115 days on average. Joint destruction in JIA has an early onset, and can occur within the first six months of evolution, which limits the opportunity for treatment in the therapeutic window. Several factors are related to the delay in early diagnosis and early treatment: 1) relatively low prevalence, 2) absence of pathognomonic clinical signs or laboratory tests, 3) possibility of atypical presentations, such as involvement of small joints or asymptomatic picture,

**Table 1 – Alterations observed during the physical examination of the musculoskeletal system (n = 141 children).**

Abnormality	n	%
Pain at palpation	86	60.9
Movement limitation	36	25.5
Osteoarticular sequelae of trauma	26	18.4
Spinal column deviation	19	13.4
Congenital malformation	14	9.9
Increased articular volume	6	4.2
Total number of alterations	187	

and 4) problems associated with a low-efficiency health system and difficult access to pediatric rheumatology centers.

The present data show a prevalence of JIA of 0.34/1,000 children aged 6 to 12 years in a small city in the state of São Paulo, which cannot be extrapolated to Brazil. Epidemiological studies involving more cities from different regions are necessary to learn the prevalence of the disease in each one. The authors believe that the present study will encourage further investigations, which are necessary for the development of public health policies that can benefit thousands of children, who certainly have not been receiving early and appropriate treatment.

### Conflicts of interest

The authors declare no conflicts of interest.

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