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## Case report

# Chorea: A rare manifestation of Takayasu's arteritis



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## ABSTRACT

The case of a girl with recurring chorea and a Takayasu's arteritis diagnosis is reported. This clinical manifestation has been reported in only one patient with this vasculitis in the pediatric group.

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## Coreia: uma manifestação rara da arterite de Takayasu

### RESUMO

Relatamos um caso de uma menina com coreia recorrente e diagnóstico de arterite de Takayasu. Esta manifestação clínica foi relatada em apenas um paciente com tal vasculite na faixa etária pediátrica.

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### Palavras-chave:

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## Introduction

Takayasu's arteritis (TA) is a chronic inflammatory disease with an unknown etiology, affecting the aorta and its major

branches, as well as the pulmonary artery.<sup>1</sup> It is a rare disease, especially in childhood, with pediatric patients corresponding to 20% of cases.<sup>2</sup> In the pediatric population, symptoms are insidious and systemic manifestations, such as fever, weight loss, and musculoskeletal symptoms predominate early in the

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disease, which contributes to a longer time elapsed before the diagnosis is made in younger patients.<sup>3,4</sup>

Hypertension is the main clinical finding of the disease in childhood and other findings include bruits, diminished or absent pulses, difference of blood pressure between limbs, heart failure, and claudication.<sup>4,5</sup> Central nervous system (CNS) manifestations are common in the pediatric population and might include headaches, dizziness, syncope, visual loss, seizures, and stroke.<sup>2,3,6</sup>

Many studies highlight an association of chorea with autoimmune rheumatic diseases, such as rheumatic fever, systemic lupus erythematosus, and antiphospholipid syndrome.<sup>7-10</sup> Reports on the association of chorea with Behcet's disease and Churg-Strauss syndrome have also been found.<sup>11,12</sup> However, only one case of chorea has been described in a child with Takayasu's arteritis.<sup>13</sup>

As this is a rare disease and only one case of chorea is reported in pediatric patients with this vasculitis, we describe the case of a child with TA manifesting chorea in the onset of the disease and over its course.

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## Case report

A 10-year-old girl started presenting with a daily pain in the hands and lower limbs since she was 4. The pain was more often felt late in the afternoon, worse in cold climate and when exercising and was improved with rest. Swelling and fever were not present. Two years after the onset of the condition, the painful symptoms were maintained and the patient started presenting involuntary movements on the left side of her body, which was worse with stress and improved during sleep. Chorea was diagnosed and its cause remained to be determined by the neurologist, who prescribed haloperidol (0.5 mg/day), with improvement of the condition after the medication was used for 1 week. Brain magnetic resonance imaging was normal and the EEG showed a posterior temporal and right occipital paroxysmal activity.

The patient remained asymptomatic for 2 years and then a daily headache and hypertension were found and thus she was referred to a cardiologist. A difference in blood pressure between the arms was present, blood pressure was not observable in both lower limbs, in addition to absent femoral, popliteal, pedal, and posterior tibial pulses. She underwent an angiography which showed severe coarctation of the descending aorta. The echocardiography showed a mild tricuspid regurgitation. Captopril and propranolol were prescribed and the patient was referred to the department of pediatric rheumatology with an initial diagnosis of rheumatic fever or TA. At the first visit to the pediatric rheumatology clinic, laboratory analyses showed slight elevation in inflammatory activity tests (erythrocyte sedimentation rate, and C-reactive protein) and absence of antinuclear antibody, anti-DNA, antibodies to extractable nuclear antigens, anticardiolipin, and antineutrophil cytoplasmic antibodies. TA was diagnosed and methotrexate 0.6 mg/kg/week, folic acid, amlodipine, and aspirin at antiaggregant dosage were introduced. She received 3 methylprednisolone IV pulses (30 mg/kg) and captopril, propranolol, and haloperidol were maintained. One month later, chorea recurred and then a penicillin G benzathine

prophylaxis (1.2 million units IMq3wk) was initiated. Six months later, an onset of right ankle, knee, and wrist arthritis, claudication, and headache was observed. At that time, a renal Doppler ultrasound showed >50% stenosis in the abdominal aorta above the kidneys and a <50% stenosis below the renal arteries emergence. Magnetic resonance angiogram showed infrarenal aorta and common iliac artery narrowing. Due to the major vascular impairment, cyclophosphamide was added into the pulse methylprednisolone therapy. One month later, haloperidol was withdrawn because of the absence of neurological symptoms (the treatment had been used for two and a half years). At the fourth cycle of cyclophosphamide, 3 months after haloperidol had been withdrawn, chorea reappeared for the third time and haloperidol was reintroduced (1 mg/day).

The patient currently maintains a visual blurring, ankle joint pain, long-distance claudication, and dyspnea on exertion. Blood pressure is controlled (by three antihypertensive drugs) and the patient had a slight improvement in lower-limb pulse amplitude. Following an 8-month treatment for chorea, the neurologist stopped the haloperidol and the patient has had no chorea for 1 year since the last recurrence. Inflammatory tests were normal. A new echocardiogram showed segmental coarctation of the low aortic arch with hemodynamic repercussion and mild concentric left ventricle hypertrophy. Following seven cyclophosphamide (1 g/m<sup>2</sup>) infusions, the patient maintained the complaints and a brain magnetic resonance angiogram showed an internal carotid artery stenosis. As a result, we decided to start infliximab (5 mg/kg). As the chorea was suggested to be associated with Takayasu's, we chose to stop the penicillin G benzathine prophylaxis. The patient is currently on methotrexate, folic acid, amlodipine, captopril, propranolol, aspirin, ranitidine, and infliximab. She has been clinically better on infliximab for 8 months, still presenting dyspnea on exertion, but the arthralgia and the visual blurring have improved.

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## Discussion

We have reported the case of a 10-year-old girl with recurring episodes of chorea associated with hypertension, diminished peripheral pulses, and an angiogram consistent with TA. Other rheumatic and nonrheumatic causes of chorea were ruled out.

The patient met the validated classification criteria for childhood TA,<sup>14</sup> which included angiographic abnormalities associated with hypertension, a difference in blood pressure between the limbs, diminished peripheral pulses, and claudication. The penicillin G benzathine prophylaxis was chosen at first because rheumatic fever is still the most common cause of chorea in childhood,<sup>15</sup> although the patient did not meet the criteria for the disease. The youngest had two more episodes of chorea in evolution, and one of the recurrences came off when the girl was in use of haloperidol, and the other when in secondary prophylaxis with penicillin benzathine, with questioning of the diagnosis of rheumatic fever.

Our patient presented left hemibody recurring chorea. This manifestation ceased only after a long immunosuppressive treatment. At that time, other causes of chorea were investigated. Brain magnetic resonance imaging and EEG had no

abnormal findings, which ruled out other neurological causes out. Thyrotoxicosis, also a cause for chorea, was ruled out. Negative antinuclear antibodies and antiphospholipid antibodies ruled out the diagnosis of systemic lupus erythematosus and the antiphospholipid syndrome as possible causes of chorea. The patient did not have clinical manifestations consistent with other primary vasculitides, such as Behcet's disease or polyarteritis nodosa. The most important differential diagnosis was rheumatic fever, but this assumption was unlikely because of the absence of other major criteria, such as carditis or arthritis early in the course, recurrence while on penicillin G benzathine prophylaxis and improvement from continuous immunosuppressive treatment. Constitutional symptoms and acute phase nonspecific signs may be common to both rheumatic fever and Takayasu's arteritis, however more unique findings, such as diminished peripheral pulses, hypertension and a difference in BP between the arms, in addition to imaging showing involvement of the aorta and its branches, led to the diagnosis of Takayasu's arteritis. In addition, the patient has been out of streptococcal infection prophylaxis for 18 months and no other chorea episodes occurred.

In the Brazilian multicenter study of 71 children and adolescents with Takayasu arteritis published this year, more than 70% of patients had neurological symptoms early in the disease.<sup>16</sup> These symptoms were represented by headache, convulsions, fainting and stroke. There was no description of chorea, except in the patient reported in this article.

The case of a child with TA presenting as a hemichorea has been recently published in a Spanish report.<sup>13</sup> Yet, no other reports were published.

Antibodies to basal ganglia, in addition to the presence of T lymphocytes, leading to an altered immune cell response, would possibly explain the occurrence of this rare manifestation. Furthermore, the involvement of intracranial vessels could lead to hypoperfusion of brain areas, especially the basal ganglia, and chorea emergence.

Through this case, we want to draw attention to a rare TA manifestation. The physician should be alert to atypical manifestations of the disease so that diagnosis and treatment are promptly accomplished.

### Conflicts of interest

The authors state no conflicts of interest.

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