

## Hemichorea as a Presentation of Acute Rheumatic Fever

Wan-Shan Lin, MD; Wen-Jen Su<sup>1</sup>, MD; FACC, Kuang-Lin Lin<sup>1</sup>, MD;  
Jing-Long Huang<sup>1</sup>, MD; Huei-Shyong Wang<sup>1</sup>, MD

There has been a decline in the incidence of acute rheumatic fever in recent decades in developed countries and in Taiwan. Sydenham's chorea, a major manifestation of rheumatic fever, was the most common cause of chorea in children in the past. But the incidence of Sydenham's chorea has declined in recent years in concert with the decline in rheumatic fever. Sydenham's chorea is usually bilateral and female predominant. Hemichorea is rare. We report on a 10-year-old boy who presented with progressive right side involuntary movements, an apical systolic murmur, prolonged PR interval, and elevated antistreptolysin O titer, who was diagnosed with acute rheumatic fever. (*Chang Gung Med J 2006;29:612-6*)

**Key words:** hemichorea, rheumatic fever, Sydenham's chorea.

Although a resurgence of acute rheumatic fever was noted in the mid to late 1980s, there has been a decline in the incidence and severity of the disease in recent decades in developed countries and in Taiwan.<sup>(1-4)</sup> Currently, younger doctors may have never seen a patient with rheumatic fever and have become unfamiliar with the disease. Sydenham's chorea was reported in 10-15% of patients with rheumatic fever but the incidence was relatively low, around 1-8%, in South and East Asia.<sup>(3,5-12)</sup> Sydenham's chorea is usually bilateral and female predominant.<sup>(7,11,13)</sup> Hemichorea is rare, in concert with the declined incidence of rheumatic fever. We herein describe our experience with a boy who presented with hemichorea and finally was diagnosed with acute rheumatic fever.

### CASE REPORT

A 10-year-old boy presented with a 3-week history of progressive involuntary movement of his right limbs. The case had been treated as focal epileptic seizures and anticonvulsants had been

given at another hospital. On examination, he twisted his right hand and leg and had a clumsy gait and frequent jerking movements. During the week prior to presentation, he had noticed a deterioration in his handwriting skills and had been having increasing difficulties with speaking, walking, and daily activities such as manipulating chopsticks and drinking from a glass. The involuntary movements disappeared during sleep and became prominent when the boy felt anxious. Lability of mood was also noted during this period of time. He had a mild cough 2 weeks prior to the onset of the involuntary movements and had visited primary physicians, but no studies were done. The medical history and family history were unremarkable.

Review of systems revealed palpitations, tachycardia and claudication. No fever, sore throat or arthralgia was noted. The initial physical examination revealed a slightly increased heart rate about 110 per minute and a grade 3/6 high-pitched, blowing systolic heart murmur at the apex. No skin rash, subcutaneous nodules or joint swelling was found. His neurological examination was remarkable for right

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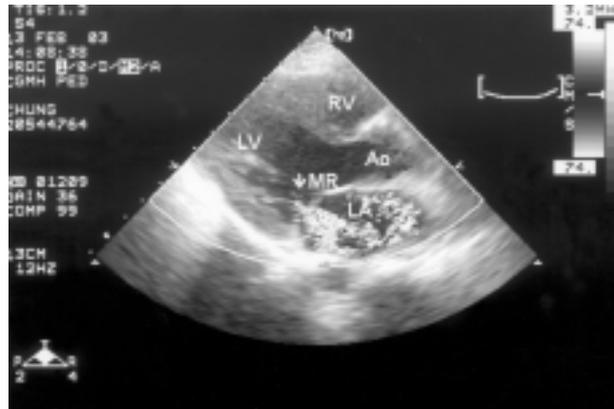
From the Department of Pediatrics, Chang Gung Memorial Hospital, Keelung, <sup>1</sup>Department of Pediatrics, Chang Gung Children's Hospital, Taipei.

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Correspondence to: Dr. Kuang-Lin Lin, MD, Department of Pediatrics, Chang Gung Children's Hospital, 5, Fushing Street, Gueishan Shiang, Taoyuan, Taiwan 333, R.O.C. Tel.: 886-3-3281200 ext. 8200; Fax: 886-3-3288957; E-mail: lincgh@cgmh.org.tw

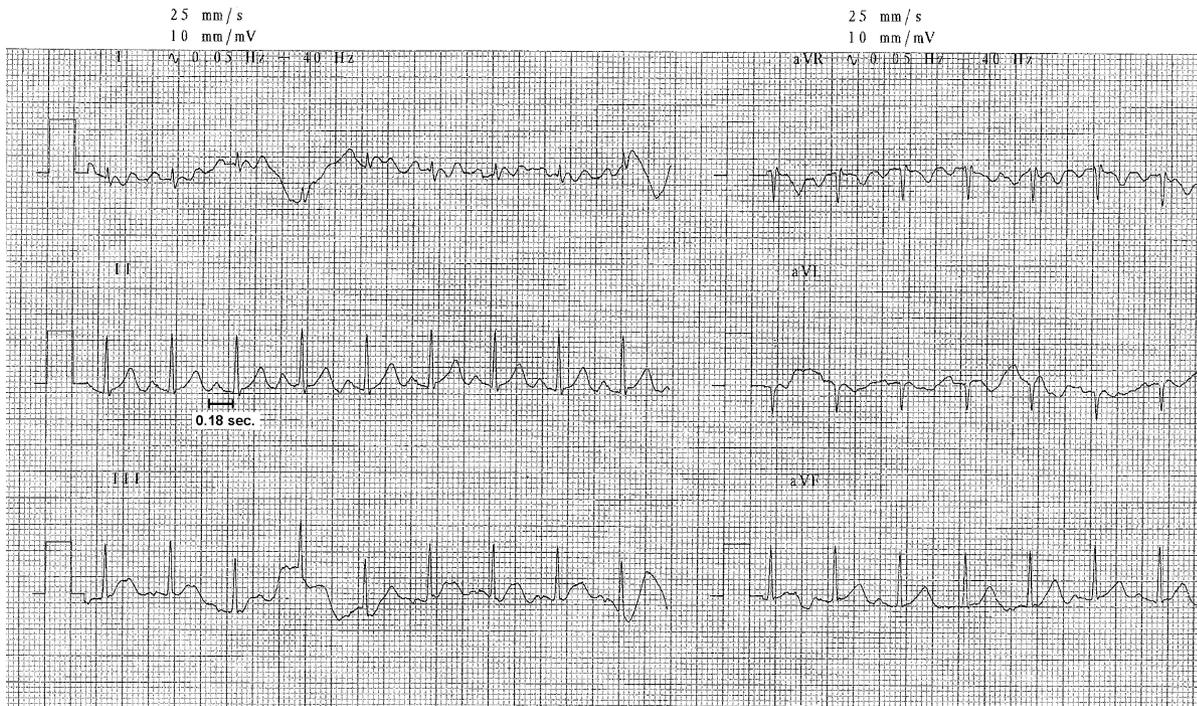
hemichorea and slurred speech. In addition to involuntary, jerking movements of his right upper and lower extremities (more prominent in the arm), he had frequent facial grimaces and choreic movements of his shoulders and neck. Deep tendon reflexes were slightly decreased, but the muscle power was normal.

Due to the chorea and significant heart murmur, acute rheumatic fever was suspected, although hemichorea is not a common presentation of acute rheumatic fever. Laboratory studies revealed an elevated antistreptolysin O titer of 803 IU/ml and an increased erythrocyte sedimentation rate of 30 mm/hr. A chest roentgenogram showed cardiomegaly with a cardio-thoracic ratio of 56%. Echocardiography demonstrated moderate mitral regurgitation (Fig. 1) and dilatation of the left atrium and left ventricle with mild ventricular compromise. An electrocardiogram revealed a prolonged PR interval (Fig. 2). An electroencephalogram showed mild diffuse cortical dysfunction over the bilateral hemispheres. A diagnosis of acute rheumatic fever with Sydenham's chorea was made based on the presence of carditis, chorea, elevated acute phase reactant,



**Fig. 1** Color flow Doppler echocardiography in the precordial approach. Note direction of jet and mosaic pattern. Parasternal long axis view shows moderate mitral regurgitation. Ao, aorta; LA, left atrium; LV, left ventricle; MR, mitral regurgitation; RV, right ventricle.

prolonged PR interval and presumptive evidence of prior streptococcal infection. Haloperidol 0.03 mg/kg/day was given and the chorea improved 3 days after therapy. Treatment of the carditis included prednisolone 2 mg/kg/day for 2 weeks followed by a



**Fig. 2** Electrocardiogram at rest demonstrating a prolonged PR interval of 0.18 seconds. The upper limit of the PR interval for the 10-year-old child with the same heart rate is 0.15 seconds.<sup>(8)</sup>

tapering of the dose and aspirin 100 mg/kg/day (after prednisolone tapering) for 1 month followed by gradual withdrawal. Digoxin 4 mcg/kg/day and furosemide 1.5 mg/kg/day were used for mild heart failure. Two months after therapy, the PR interval and heart size were normal. He was given benzathine penicillin prophylaxis, and still had mitral regurgitation 2 years later.

## DISCUSSION

There is still no single symptom, sign, or laboratory test that is pathognomonic or diagnostic of acute rheumatic fever. The guidelines for the diagnosis of acute rheumatic fever are based on the 1992 updated Jones criteria.<sup>(1)</sup> Chorea consists of rapid, involuntary movements observed on physical examination which do not have a cause which can be demonstrated by laboratory tests. It is worsened by stress and anxiety and subsides during sleep. Sydenham's chorea, first described in 1686 by Thomas Sydenham, is generally used to signify a diagnosis of a form of rheumatic fever. It is a cardinal feature of rheumatic fever and is sufficient alone to make the diagnosis.<sup>(1)</sup>

Sydenham's chorea is a disease of childhood or early adolescence. Most reported patients were 5 to 15 years old. Sydenham's chorea was reported in about 10-15% of patients with rheumatic fever but the incidence was relatively low, around 1-8%, in Africa, the Pacific, South and East Asia and the Arabian Peninsula.<sup>(3,5-12)</sup> In most patients, Sydenham's chorea is bilateral, and hemichorea has been reported in only 15-20% of patients.<sup>(7,9)</sup> It occurs considerably more often in girls than boys in the teen years, occurring at a ratio of approximately 2:1.<sup>(5,7)</sup> Because the onset of chorea is usually 1 to 6 months after the provocative streptococcal infection, other rheumatic manifestations may not be found at presentation.<sup>(7,14)</sup> In our patient, the hemichorea was treated as focal seizures, and tachycardia was not recognized as an early sign of heart failure. The diagnosis of acute rheumatic fever was not confirmed until 2 weeks after the initial presentation. Although hemichorea in rheumatic fever is an unusual presentation, we emphasize the importance of the differential diagnosis of focal seizures and hemichorea.

Sydenham's chorea is a neuro-psychiatric disorder consisting of neurologic and psychiatric signs.

The neurologic signs include choreic movement, hyporeflexia and hypotonia and the psychiatric signs are emotional lability, nightmares, obsessive-compulsive symptoms, and separation anxiety disorder.<sup>(8,9,15)</sup> One hypothesis is that when genetically vulnerable children are exposed to group A beta-hemolytic Streptococcus infection, antibodies mistakenly attack cells in the basal ganglia and cause inflammation.<sup>(9)</sup>

Sydenham's chorea is the most common cause of acquired chorea in children.<sup>(14)</sup> There are other diseases that cause chorea in childhood. The differential diagnosis is mainly between Sydenham's chorea, lupus-associated chorea and drug-induced chorea.<sup>(12)</sup> Sometimes, chorea induced by another disease is misdiagnosed as Sydenham's chorea. The common causes of chorea include atypical seizures, brain tumor, cerebrovascular accident, collagen vascular disorders such as systemic lupus erythematosus and Behcet's disease, drug intoxication, endocrine disorders such as hyperthyroidism and hypoglycemia, hereditary disorders such as Huntington's chorea and Wilson's disease, pregnancy and viral encephalitis.<sup>(13,14,16)</sup>

The causes of hemichorea are the same as those of bilateral chorea. Hemichorea is defined as an involuntary, hyperkinetic disorder involving unilateral parts of the body, which may be due to selective involvement of the contralateral caudate nucleus, putamen, globus pallidus and subthalamic nucleus.<sup>(17-19)</sup> One recent study showed that blood flow was significantly decreased in the basal ganglia contralateral to the chorea and increased in the thalamus, reflecting the loss of pallidal inhibitory input from the pallidum to the thalamus.<sup>(18)</sup>

For a child with chorea, the initial examination includes chest roentgenography, echocardiography, electrocardiography, antistreptococcal antibody studies, ESR and CRP. If the initial examination does not support a diagnosis of Sydenham's chorea, further approaches include evaluation of thyroid function, antinuclear antibody titer, antiphospholipid antibodies, copper metabolism, and neuroimaging studies. Other diagnostic tests depend on the degree of probability developed from the history and findings on physical examination. We conclude that hemichorea, though uncommon, may occur in rheumatic fever and requires medical attention.

## REFERENCE

1. Special Writing Group of the Committee on Rheumatic Fever, Endocarditis, and Kawasaki Disease of the Council on Cardiovascular Disease in the Young of the American Heart Association. Guidelines for the diagnosis of rheumatic fever: Jones criteria, 1992 update. *JAMA* 1992;268:2069-73. Correction. *JAMA* 1993;269:476.
2. Bisno AL. The resurgence of acute rheumatic fever in the United States. *Annu Rev Med* 1990;41:319-29.
3. Lue HC. Rheumatic fever and rheumatic heart disease in Taiwan. *Formos J Med* 1998;2:261-73.
4. Veasy LG, Wiedmeier SE, Orsmond GS, Ruttenberg HD, Boucek MM, Roth SJ, Tait VF, Thompson JA, Daly JA, Kaplan EL, Hill HR. Resurgence of acute rheumatic fever in the intermountain area of the United States. *N Engl J Med* 1987;316:421-7.
5. Carapetis JR, Currie BJ. Rheumatic chorea in northern Australia: a clinical and epidemiological study. *Arch Dis Child* 1999;80:353-8.
6. Lue HC, Chen CL, Wei H, Okuni M, Mabilangan LM, Dharmasakti D, Hanafiah A. The natural history of rheumatic fever and rheumatic heart disease in the orient. *Jpn Heart J* 1979;20:237-52.
7. Nausieda PA, Grossman BJ, Koller WC, Weiner WJ, Klawans HL. Sydenham chorea: An update. *Neurology* 1980;30:331-4.
8. Park MK. *Pediatric cardiology for practitioners*. 4th ed. St. Louis: Mosby Inc., 2002:41 and 306.
9. Swedo SE. Sydenham's chorea: A model for childhood autoimmune neuropsychiatric disorders. *JAMA* 1994;272:1788-91.
10. Griffiths SP, Gersony WM. Acute rheumatic fever in New York City (1969-1988): A comparative study of two decades. *J Pediatr* 1990;116:882-7.
11. Eshel G, Lahat E, Azizi E, Gross B, Aladjem M. Chorea as a manifestation of rheumatic fever - A 30-year survey (1960-1990). *Eur J Pediatr* 1993;152:645-6.
12. Bitar FF, Hayek P, Obeid M, Gharzeddine W, Mikati M, Dbaibo GS. Rheumatic fever in children: a 15-year experience in a developing country. *Pediatr Cardiol* 2000;21:119-22.
13. Klawans HL, Brandabur MM. Chorea in childhood. *Pediatr Ann* 1993;22:41-50.
14. Fenichel GM. *Clinical pediatric neurology: a signs and symptoms approach*. 4th ed. Philadelphia: W.B. Saunders Co., 2001:281-6.
15. Swedo SE, Leonard HL, Schapiro MB, Casey BJ, Mannheim GB, Lenane MC, Rettew DC. Sydenham's chorea: physical and psychological symptoms of St Vitus dance. *Pediatrics* 1993;91:706-13.
16. Higgins DS. Chorea and its disorders. *Neurol Clin* 2001;19:707-22.
17. Chung SJ, Im JH, Lee MC, Kim JS. Hemichorea after stroke: clinical-radiological correlation. *J Neurol* 2004;251:725-9.
18. Kim JS, Lee KS, Lee KH, Kim YI, Kim BS, Chung YA, Chung SK. Evidence of thalamic disinhibition in patients with hemichorea: semiquantitative analysis using SPECT. *J Neurol Neurosurg Psychiatry* 2002;72:329-33.
19. Emery ES, Vieco PT. Sydenham Chorea: magnetic resonance imaging reveals permanent basal ganglia injury. *Neurology* 1997;48:531-3.

## 以單側舞蹈症表現之急性風濕熱

林萬山 蘇文鈺<sup>1</sup> 林光麟<sup>1</sup> 黃璟隆<sup>1</sup> 王輝雄<sup>1</sup>

急性風濕熱在最近一、二十年在已開發國家及台灣變得少見，多數年輕醫師不曾見過急性風濕熱病患並對其診斷陌生。Sydenham's chorea 為急性風濕熱主要表現之一，在以往亦為孩童舞蹈症最常見之原因，近年來其發生率隨急性風濕熱之減少而大幅減少。Sydenham's chorea 多為雙側表現，以單側舞蹈症 (hemichorea) 表現之情形為罕見。本報告提出一位十歲男童病例，因右側肢體漸進性不自主運動而求診，外院醫學中心診斷為部分性癲癇發作 (focal epileptic seizure)，在本院最後確定診斷為急性風濕熱。我們認為以單側舞蹈症表現之急性風濕熱現今並不常見，其診斷有賴醫師之高度警覺。(長庚醫誌 2006;29:612-6)

**關鍵字：**單側舞蹈症，急性風濕熱，Sydenham's chorea。

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長庚紀念醫院 基隆院區 小兒科；<sup>1</sup>長庚兒童醫院 台北院區 兒童內科部

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通訊作者：林光麟醫師，長庚兒童醫院 兒童內科部。桃園縣333龜山鄉復興街5號。Tel.: (03)3281200轉8200; Fax: (03)3288957; E-mail: lincgh@cgmh.org.tw