Cerebral Infarction in a Patient with Congenital Complete Heart Block

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Summary: Stokes-Adams attacks are fairly common in children with congenital complete heart block, but the occurrence of cerebral infarction is quite unusual. We present the case of a 13-year-old boy with congenital heart block and an embolic stroke involving the cerebral artery. Echocardiography revealed no valvular regurgitation, hypokinetic segments, mural thrombus, or myxoma. Electrocardiographic monitoring demonstrated good response of ventricular rate to exercise and no episodes of atrial or ventricular dysrhythmia. It is assumed that embolism occurred due to bradycardia.

Key words: congenital complete heart block, cerebral infarction

Case Report

A 13-year-old boy with congenital heart block was admitted to our hospital because of the abrupt onset of left hemiparesis and confusion. While he was asleep, his father noted his eyes rolled back and he was difficult to arouse. His speech was unintelligible and his left arm and leg were weak.

Bradycardia had been detected in the patient before birth. Electrocardiography demonstrated complete heart block with a ventricular rate of 60 beats/min and an atrial rate of 150 beats/min at birth. He grew up at a normal rate and was without symptoms such as syncope, dizziness, or exercise intolerance until this episode. He was followed by our clinic three times a year. The last electrocardiogram (ECG) before the stroke showed complete heart block with a ventricular rate of

54 beats/min and an atrial rate of 80 beats/min. The QRS axis was normal with a duration of 0.08 s. Twenty-four h Holter monitoring obtained 3 months before the stroke demonstrated that the ventricular rate varied from 39 beats/min to 120 beats/min with no episodes of ventricular dysrhythmia or asystole. Neither atrial fibrillation nor sinus dysrhythmia were detected. Treadmill exercise testing demonstrated an increase in ventricular rate to 120 beats/min (Fig. 1). No pacemaker had been placed because it was felt to be unnecessary in the absence of symptoms.

On admission, the patient weighed 45 kg and measured 158 cm in height. Pulse rate was 54 beats/min, his blood pressure was 140/80 mmHg, and respiratory rate was 20/min. Examination of the heart revealed no thrills or heaves, and the first heart sound varied in intensity. A grade 1/6 vibratory systolic ejection murmur was heard along the upper left sternal border. There was no hepatosplenomegaly. Left hemiparesis was noted, and Babinski's sign was present on the left side.

Magnetic resonance imaging of the brain demonstrated a high intensity area in the left temporal region consistent with infarction (Fig. 2). Cardiomegaly was not detected on chest radiography. Complete blood count, blood gas analysis, urinalysis, coagulation studies, serum cholesterol, and triglyceride concentrations were normal. The plasma levels of protein C and protein S were normal. Antinuclear antibodies and anticardiolipin antibodies were not present. A urine sodium nitroprusside test to diagnose homocystinuria was negative, and plasma amino acid concentrations were normal. The serum lactate and pyruvate concentrations were normal, excluding mitochondrial encephalopathy with lactic acidosis and stroke-like episodes (MELAS). Echocardiography demonstrated no valvular regurgitation, hypokinetic segments,

FIG. 1 Heart rate response to treadmill exercise testing using the Bruce protocol. The ventricular rate increased up to 120 beats/min.

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Received: August 27, 1997

Accepted with revision: November 6, 1997

FIG. 2 Magnetic resonance imaging (A) and magnetic resonance angiography (B). Note the high intensity area in the right paraventricular region, which is consistent with a recent infarction. Magnetic resonance angiography shows obstruction of the right middle cerebral artery.

mural thrombus, or myxoma. Atrial septal defect or ventricular septal defect were not detected; left ventricular function was normal. Furthermore, a patent foramen ovale with right-to-left shunt, which can be responsible for paradoxical embolism, could not be diagnosed by transthoracic color Doppler or saline contrast echocardiography during Valsalva maneuver. Two months after the stroke, a permanent DDD pacemaker set at a rate of 70 beats/min was implanted to prevent a second stroke. His left hemiparesis has improved but not completely resolved.

Discussion

Stokes-Adams attacks have been reported in children with congenital complete heart block.^{1,2} However, cerebrovascular infarction in this group of patients has never been reported, except in patients after intravenous pacemaker implantation.³ We describe a 13-year-old boy with congenital heart block and an embolic stroke involving the right internal cerebral artery.

The precipitating factors of the infarction remained unclear. Stroke during childhood has been reported in patients with homocystinuria,⁴ MELAS,⁵ and antiphospholipid syndrome.⁶ However, these conditions were excluded in our patient. A paradoxical embolism due to right-to-left shunt through a patent foramen ovale can be responsible for stroke and ischemic cerebral events;⁷ however, a patent foramen ovale was not found by color Doppler or saline contrast echocardiography with the Valsalva maneuver. Although 24-

h ECG monitoring showed that the ventricular rate did not decrease to < 35 beats/min and there was no evidence of atrial fibrillation; in view of the close relation of cerebral infarction and lowest heart rate at night, we assumed that the embolism occurred due to bradycardia secondary to complete heart block. We hypothesize that an embolus developed because of bradycardia. We recommend the regular examination of thrombus.

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