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Atrial standstill presenting as cerebral infarction in a 7-year-old girl

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Abstract

Atrial standstill is a rare arrhythmia defined by the absence of mechanical and electrical activity in the atria. Few cases of atrial standstill have been described in children, none of which have presented with cerebral infarction confirmed by imaging. We report a unique case of a 7-year-old girl presenting with expressive aphasia, central facial palsy and irregular pulse with cerebral infarction secondary to atrial standstill. This case illustrates that cardiogenic cerebral embolism in children can be caused by rare conditions like atrial standstill and should be considered in paediatric patients undergoing evaluation for stroke. There are no established treatment guidelines for atrial standstill. We recommend that treatment be directed towards any potential underlying cause. All patients with atrial standstill should receive long-term oral anticoagulation treatment and a permanent cardiac pacemaker implant to reduce the risk of further strokes or other cardiac events.

Keywords

Cerebral infarction, infarctus cerebri, pacemaker, paediatric

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Introduction

Atrial standstill is a rare arrhythmia defined by the absence of mechanical and electrical activity in the atria. It was first described in 1946 by Chavez et al.[1] The diagnostic criteria include: (a) abnormal or absent P-waves in surface and intracavity electrocardiograms (ECGs). If present, atrial activity is slow and erratic, with a prolonged, low-amplitude and low-frequency electrogram; (b) evidence of atrial asystole, that is, the absence of A-waves in jugular venous pulse and right atrial pressure tracings; (c) supraventricular QRS morphology; (d) immobility of the atria on fluoroscopy and (e) inability to stimulate the atria electrically.[2–5] Few cases of atrial standstill have been described in children.[6–9] We report a case of atrial standstill in a 7-year-old girl presenting with cerebral infarction with expressive aphasia and central facial palsy. Informed written consent for patient information and images to be published was provided by the patient's parents.

Case

A previously healthy 7-year-old girl was admitted to the hospital after waking up with acute onset expressive aphasia. The patient had no family history of cardiac or neurological disease and was not taking any medication. During a vacation, 3 years ago, the patient experienced an episode where she suddenly became confused and could not express herself coherently. She was assessed at a local hospital where neither cerebral imaging or ECG were performed. It was concluded that the patient had suffered a cerebral concussion despite no history of trauma. The patient had complete remission of all symptoms within a few hours.

Upon presentation at our hospital, the patient was awake, alert and fully orientated. Clinicians determined the presence of expressive aphasia and a mild central facial palsy on the right side, which regressed and disappeared within the same day. The patient did not complain of any cardiac symptoms, had normal heart sounds and a normal blood pressure of 106/51 mmHg. Her peripheral pulse was irregular with an average heart rate (HR) of 60 beats per minute (BPM).

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Cerebral magnetic resonance imaging (MRI) showed a recent infarction in the left frontal lobe and anterior insula cortex consistent with an occlusion of the frontal branch of the left middle cerebral artery. Furthermore, signs of earlier infarctions were seen in the parietal and temporal regions of the right hemisphere (Figure 1).

An initial 12-lead ECG showed a slow nodal escape rhythm with an HR of 54–83 BPM (Figure 2). A second ECG, done later the same day, showed atrial ectopic tachycardia (AET) with alternating 1:1 and 2:1 conduction with an HR of 180 BPM, probably originating from an ectopic focus in the left atria in the region of the pulmonary vein insertion. The ECG was repeated while intravenous adenosine was administered and showed an atrial rhythm with an HR of 210–220 BPM with positive P-waves in leads II and III, suggesting that the patient's arrhythmia was either a slow atrial flutter or AET. Transthoracic echocardiography (TTE) was normal apart from the discovery of a small patent foramen ovale (PFO). Transoesophageal echocardiography (TEE) confirmed the PFO and showed a normal left atrium with a normal auricle without any thrombus.

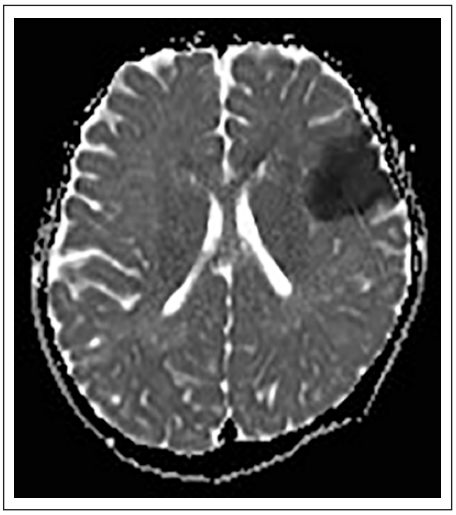

Figure 1. A cerebral MRI scan slice showing a fresh infarction in the left frontal lobe and anterior insula cortex as well as signs of old infarction in the parietal and temporal regions of the right hemisphere.

DC-cardioversion with a voltage of 20 J was attempted, resulting in asystole. Advanced cardiopulmonary resuscitation was begun with return of spontaneous circulation after 3 min. A temporary ventricular pacing (VVI) pacemaker (PM) was inserted and later replaced with a permanent epicardial VVI PM with a backup rate of 80 BPM. During the procedure, it was not possible to pace the right atrium, despite attempts from several positions and maximum output. Pacing on the left atrium was not attempted. No atrial contraction was observed on TEE during atrial or ventricular pacing (Figure 3).

It was concluded that the presenting cerebral insult originated from mural thrombus formation in the left atrium secondary to atrial standstill. Subsequently, the patient started oral anticoagulation. It was decided not to close the PFO.

Holter monitoring carried out 2 months after discharge showed a predominantly V-paced rhythm of approximately 80 BPM. Holter monitoring also revealed multiple episodes of narrow complex tachycardia with an irregular HR of 160–200 BPM, each lasting under 1 min. The patient was subsequently treated with metoprolol, 1 mg/kg once daily. Once on medication, Holter monitoring showed a predominant

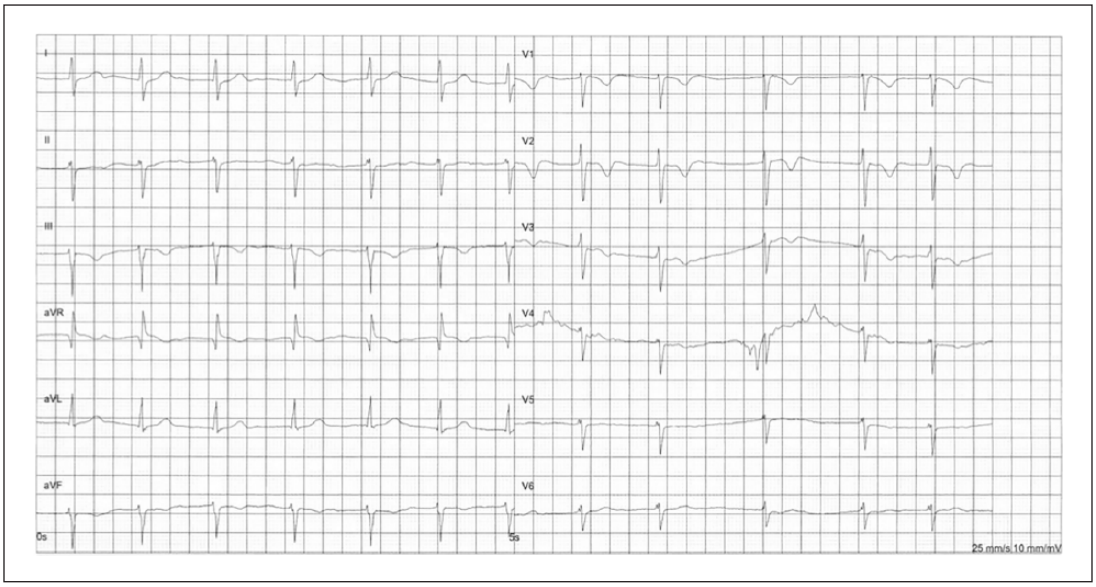

Figure 2. Twelve-lead surface ECG at the time of symptom presentation showing atrial standstill with a slow nodal escape rhythm with an HR of 54–83 BPM. There are no P-waves and the QRS complexes are of a supraventricular type.

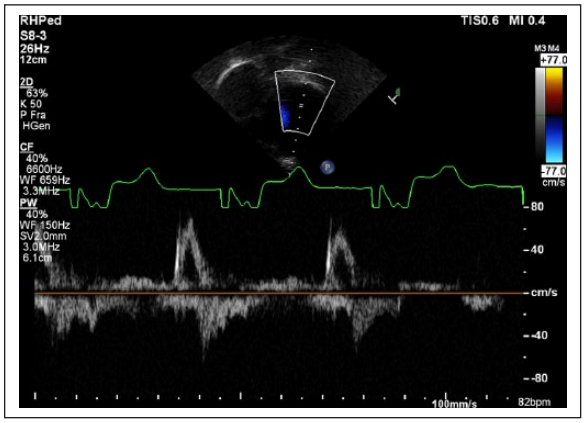

Figure 3. TTE showing lack of 'A'-wave in mitral valve pulse wave Doppler.

V-paced rhythm with an HR up to 112 BPM and an occasional nodal escape rhythm.

At 12 months follow-up, the patient was in good condition without any complaints of fatigue, palpitations or other neurological or cardiac symptoms and was physically active several times a week on the same level as her peers. Holter monitoring showed 99% V-pace with approximately 80 BPM without episodes of tachycardia. TTE showed a left ventricle with normal dimensions and normal systolic function. The PFO was not visible. The patient still received metoprolol and warfarin treatment.

Discussion and conclusion

We report a case of atrial standstill in a 7-year-old girl presenting as acute temporary expressive aphasia and central facial palsy due to MRI-verified cerebral infarction. Previous case reports have described atrial standstill in adults combined with syncope, acute ischemic stroke lesions, heart failure and sudden death.[3–5,10–12] To our knowledge, this is the first documented case of verified cerebral infarction as a result of atrial standstill in a paediatric patient.

Clinically, atrial standstill can be challenging to distinguish from sinus arrest, as both can have similar ECG findings with irregular slow escape rhythm, loss of P-waves and absent atrial fibrillatory waves.[10] Atrial standstill can be classified as partial (only involving one atrium) or total (involving both atria), as well as transient or persistent. Transient atrial standstill has previously been reported as a complication of myocardial infarction, hypoxia, myocarditis, hyperkalemia and digitalis or quinidine overdose.[3,4] Persistent atrial standstill is characterized by severe morphological changes with histologic fibrosis and cell death and has been reported in association with myocarditis, valvular dysfunction or Ebstein anomaly, neuromuscular dystrophy and amyloidosis.[3,4] Atrial standstill of idiopathic aetiology has also been described.[3,10,13] In familial atrial standstill, a cardiac sodium channel gene mutation has been identified.[8,9,11]

Nakazato et al.[12] examined adults with atrial standstill and described the condition as a progressive disease where conduction deteriorates progressively. A case report by Cappelli et al.[13] supports the theory that atrial standstill may be a progressive disease. Cappelli et al. described a 9-year-old girl, who received a VVI PM after being diagnosed with atrial standstill. After 13 years, electro-anatomical mapping showed a reduced extent of excitable atrial myocardium and an increased pacing threshold, suggesting progression of the disease.

In our case, atrial standstill was confirmed by the absence of P-waves in surface and intracavity ECGs, supraventricular type QRS morphology and the inability to stimulate the right atrium electrically. Since stimulation of the left atrium was not performed during electrophysiological investigation, we do not know whether the patient's atrial standstill was total or partial. The finding of previous cerebral infarction in the right hemisphere and the history of an earlier episode of confusion (possible transient ischemic attack) suggest that our patient's atrial standstill has been long standing. The presence of atrial arrhythmias, however, implies that the patient's atrial standstill may be of a transient nature, where previous infarcts could have been the result of atrial arrhythmias, such as supraventricular tachycardia, and not necessarily atrial standstill. It is still uncertain whether or not the patient's disease is progressive as only 1 year has passed since the time of her diagnosis. The underlying cause of the patient's atrial standstill remains unknown. Both parents were examined and had normal ECG and TTE findings. Genetic testing and myocardial biopsy were considered, but at our institution's Cardiogenetic and Electrophysiology Conference, it was decided that no further investigations would prove useful.

A multidisciplinary board of specialists concluded that the cerebral embolus in our patient most likely originated from the left atrium and was generated during atrial standstill and released during an episode of tachycardia. The PFO discovered was thought to be insignificant in the pathogenesis of the patient's thrombus. Malav et al.[5] reported a case of a 3-year-old child with an atrial septal defect (ASD) of the ostium secundum and atrial standstill that then led to congestive heart failure. The fact that the child had a secundum ASD and no clinical signs of stroke support the belief that the PFO observed in our patient was not the origin of the cerebral thrombus. It was decided not to close the PFO, as a paradoxical embolus was unlikely, and the procedure might complicate future electrophysiological examinations of the heart.

This case illustrates that cardiogenic cerebral embolism and cerebral infarction in children – most often secondary to thrombophilia, atrial flutter or left sided endocarditis – can also be caused by rare conditions like atrial standstill. Atrial standstill should be considered in patients with absent P-waves and a regular R–R interval in their surface ECGs. Since atrial standstill is rare, there are no established

treatment guidelines. Treatment should be directed towards any potential underlying cause. All patients with atrial standstill should receive long-term oral anticoagulation treatment and a permanent PM implant to reduce the risk of further strokes or other cardiovascular events.

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Ethics approval

Our institution does not require ethical approval for reporting individual cases or case series.

Informed consent

Written informed consent was obtained from a legally authorized representative(s) for anonymized patient information to be published in this article.

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