Cardiology in the Young

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Accessory-pathway-mediated dilated cardiomyopathy in an infant

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Case Report

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Abstract

We report the case of a 10-month-old infant presenting with dilated cardiomyopathy caused by septal dyssynchrony linked to a right septal accessory pathway-mediated pre-excitation. No atrioventricular reciprocating tachycardia was reported, but this pre-excitation resulted in left septal dyssynchrony. After the introduction of flecainide therapy, the antegrade pre-excitation disappeared on ECG, leading to normalisation of left ventricular function.

Brief report

We report the case of a female infant who presented at two months of age at the paediatric cardiology consultation due to a cardiac murmur. The infant was asymptomatic, exhibiting good growth in both weight and height. Her electrocardiogram (ECG) showed a regular sinus rhythm at 126 beats per minute, a PR interval of 63 ms, and a QRS duration of 108 ms, with a pattern of left ventricular conduction delay corresponding to ventricular pre-excitation. The delta wave morphology suggested an anteroseptal accessory pathway Figure 1(a). Transthoracic echocardiography (TTE) revealed a patent ductus arteriosus with a high velocity exceeding 4 m/s, a maximum velocity in the pulmonary veins of 0.8 m/s, left atrial dilation (left atrial/aortic root ratio: 1.5), and left ventricular (LV) dilation (z-score +3). Despite the septal dyssynchrony observed on M-mode imaging, the left ventricular systolic function remained preserved, with an ejection fraction (LVEF) between 58 and 64% (Figure 2(a)).

The percutaneous closure of the patent ductus arteriosus was successfully performed at 4 months of age without complications. One month after cardiac catheterisation, transthoracic echocardiography (TTE) showed persistent left ventricular (LV) dilation with a preserved LV ejection fraction (LVEF) of 62%. At 10 months of age—six months post-closure—TTE revealed worsening LV dilation (z-score + 4) and a decreased LVEF of 35% (Figure 2(b); Video S1). Additionally, the infant exhibited slowed weight gain since 6 months of age, with a recorded weight of 7.8 kg. ECG continued to show ventricular pre-excitation.

A comprehensive cardiomyopathy evaluation ruled out infectious, immunological, metabolic, and genetic aetiologies. Cardiac MRI estimated an LVEF of 43%, revealing significant septal dyssynchrony but no late gadolinium enhancement. Pharmacological management was initiated with an angiotensin-converting enzyme inhibitor and an aldosterone antagonist.

Given that accessory pathway-mediated dyssynchrony and LV dysfunction were the most likely diagnoses, antiarrhythmic therapy with oral flecainide was introduced at a dose of 3 mg/kg/day in three daily doses (60 mg/m^2/day). The treatment was well tolerated, and within 24 hours, ECG demonstrated the disappearance of ventricular pre-excitation, an increased PR interval (143 ms), and narrowing of the QRS complexes (69 ms) (Figure 1(b)).

Regular Holter monitoring was performed, and the flecainide dosage was gradually increased to 6 mg/kg/day in two daily doses due to the persistence of intermittent pre-excitation, which disappeared during sinus tachycardia but remained absent of atrioventricular reentrant tachycardia (AVRT). Six months following flecainide therapy initiation, TTE parameters showed normalisation, with an LV end-diastolic dimension z-score of 1.8, an improved LVEF of 72%, and resolution of septal dyssynchrony (Video S2). Concurrently, the infant exhibited weight gain.

Comment

This case highlights an infant under 12 months of age presenting with LV dysfunction caused by septal dyssynchrony secondary to a right anteroseptal accessory pathway, without any episodes of AVRT. Flecainide therapy successfully blocked antegrade conduction through the accessory pathway, resolving the septal dyssynchrony responsible for the cardiomyopathy.

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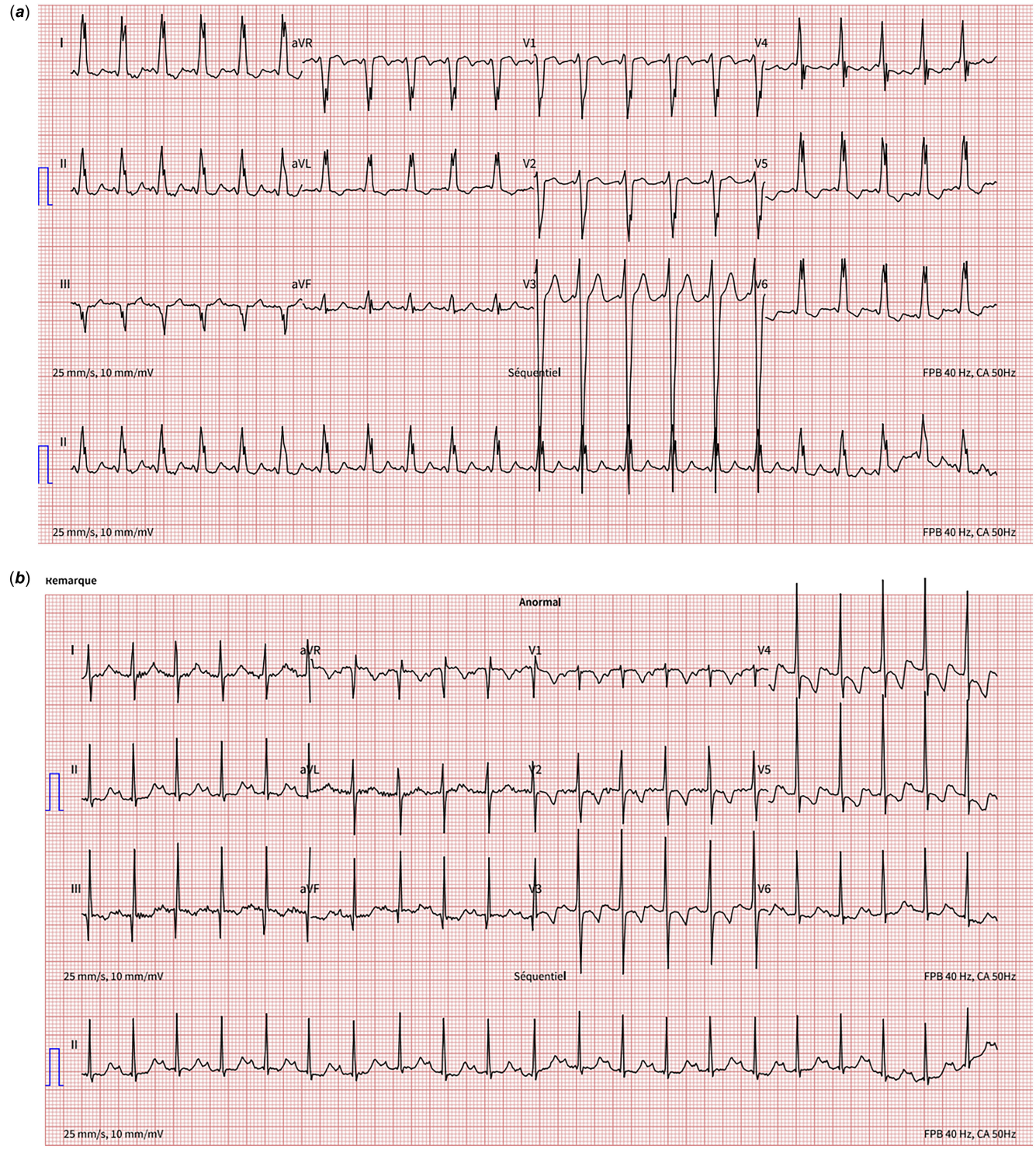


Figure 1. (***a***) Electrocardiogram prior to oral flecainide administration showing a pattern of left ventricular conduction delay consistent with ventricular pre-excitation. (***b***) Normalisation of the electrocardiogram following oral administration of flecainide, marked by the disappearance of ventricular pre-excitation, prolongation of the PR interval, and narrowing of the QRS complexes.

This condition is rare and less recognised compared to arrhythmia-induced cardiomyopathies associated with AVRT; however, a few cases have been reported in the literature. Ventricular dyssynchrony, particularly intraventricular, is known to contribute to ventricular remodelling and heart failure.[1–4] A distinguishing aspect of this case was the coexistence of a patent ductus arteriosus with left-to-right shunting, which led to LV dilation and likely masked the initial impact of the accessory pathway.

Following multidisciplinary discussion, pharmacological therapy with oral flecainide was chosen over percutaneous ablation, particularly due to the patient's young age and the potential for spontaneous regression of the accessory pathway within the first years of life. Additionally, catheter ablation of an

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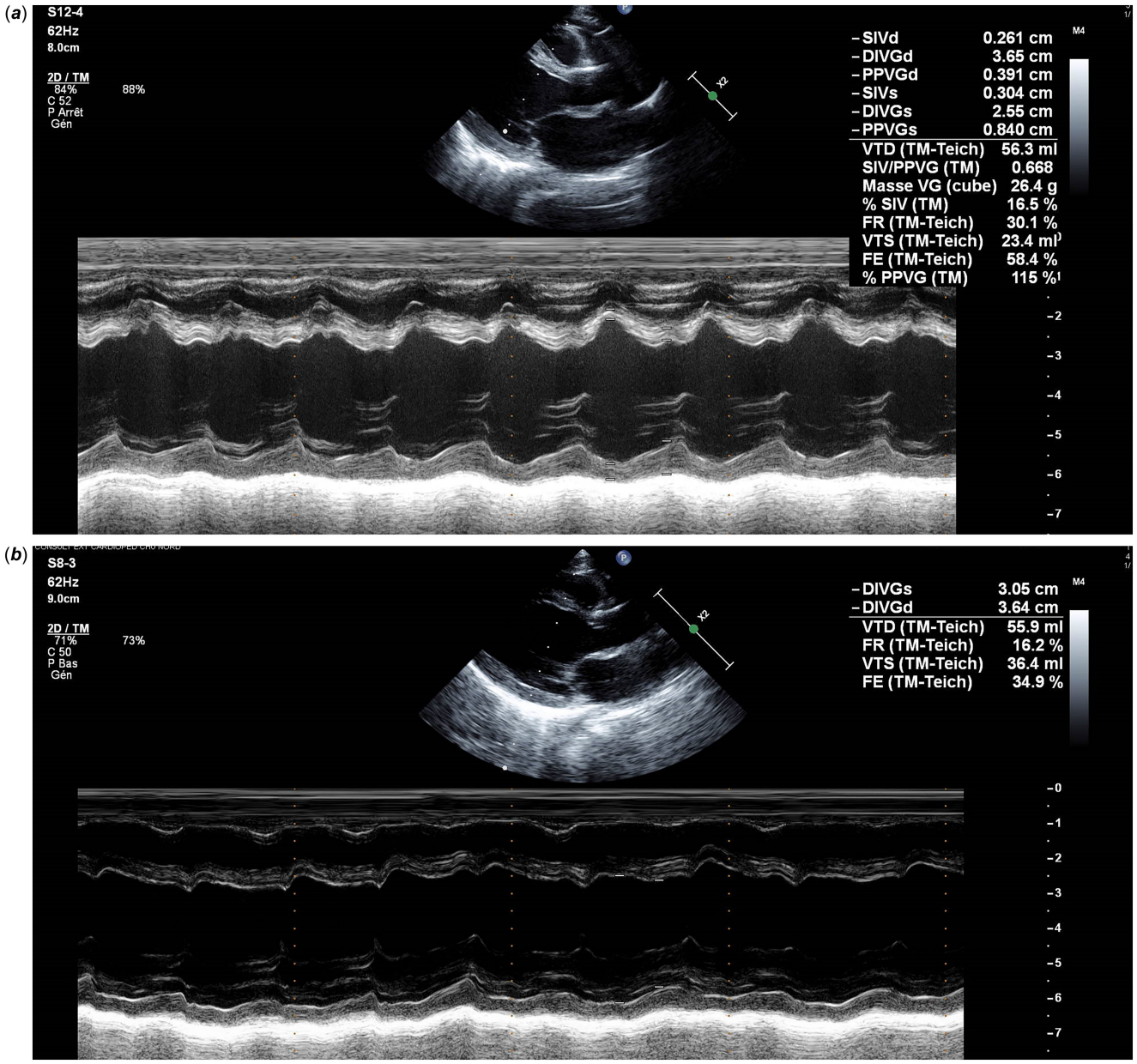


Figure 2. (***a***) Echocardiography, parasternal long-axis view, M-mode at the time of percutaneous patent ductus arteriosus closure, showing left ventricular dilation with preserved left ventricular ejection fraction. (***b***) Echocardiography, parasternal long-axis view, M-mode, at six months after patent ductus arteriosus closure, prior to initiation of flecainide therapy, showing left ventricular dilation with impaired left ventricular systolic function.

anteroseptal accessory pathway carries a risk of iatrogenic atrioventricular (AV) block.

Yi Zhang et al. recently reported their experience with right accessory pathway ablation in 10 infants (mean age: 6.7 months, average weight: 8.1 kg) with LV dysfunction, with no notable complications.[5] The recovery time for LV function depended on initial LVEF severity; patients with mild LV dysfunction ($40 \leq$ LVEF $< 50\%$) showed rapid recovery within one week, similar to our patient, whose improvement occurred later with oral flecainide therapy requiring dosage adjustments based on Holter monitoring due to intermittent pre-excitation.

The use of flecainide for this indication was first successfully reported in two infants without congenital heart defects in 2019[6] and later in 2024 in an infant with Ebstein's anomaly.[7] Flecainide is generally not a first-line therapy in patients with LV dysfunction due to potential proarrhythmic effects.[8] However, given the unique mechanism of accessory-pathway-mediated LV dysfunction in this case, flecainide was deemed the most appropriate choice.

For our patient, the therapeutic plan involves monitoring for spontaneous regression of the accessory pathway in the coming years. If it persists, catheter ablation may be considered when the risk of AV block is lower. Several studies have demonstrated the efficacy and clinical safety of oral flecainide in infants for AVRT management.[9,10] The choice between pharmacological therapy and percutaneous ablation in infants with accessory pathway-related dilated cardiomyopathy depends on each centre's expertise, as this condition remains rare.[2]

We believe that using flecainide—which was very well tolerated—as a first-line therapy to block the accessory pathway and restore good left ventricular function was the appropriate therapeutic

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choice. This approach allowed us to avoid the potential risk of atrioventricular block associated with a percutaneous procedure in this 10-month-old infant, especially considering the possibility of spontaneous regression of the accessory pathway.

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Competing interests. None.

References

1. Udink ten Cate FEA, Kruessell MA, Wagner K, et al. Dilated cardiomyopathy in children with ventricular preexcitation: the location of the accessory pathway is predictive of this association. J Electrocardiol 2010; 43: 146–154.
2. Miyazaki A, Uemura H. Perspective of preexcitation induced cardiomyopathy; early septal contraction, and subsequent rebound stretch. J Cardiol 2022; 79: 30–35.
3. Emmel M, Balaji S, Sreeram N. Ventricular preexcitation associated with dilated cardiomyopathy: a causal relationship? Cardiol Young 2004; 14: 594–599.
4. Couchonnal LF, Chicos AB, Knight BP. An unusual level of block in a patient with an unusual accessory pathway. Cardiovasc Electrophysiol 2012; 23: 334–335.
5. Zhang Y, Jiang H, Cui J, Li MT, Zhou HM, Li XM. Ablation of ventricular preexcitation to cure preexcitation-induced dilated cardiomyopathy in infants: diagnosis and outcome. Circ Arrhythm Electrophysiol 2023; 16: e011569.
6. Sekine M, Masutani S, Imamura T, et al. Improvement in dyssynchrony with pharmacological ablation of right-sided accessory pathway-induced cardiomyopathy in infants. Int Heart J 2019; 60: 1201–1205.
7. Uthayakumaran G, Asakai H, Gnanappa G. Pharmacological resynchronisation with flecainide in an infant with Ebstein anomaly and Wolff-Parkinson-White pattern: a case report. Eur Heart J Case Rep 2024; 8: ytae442.
8. Cardiac Arrhythmia Suppression Trial (CAST) Investigators. Preliminary report: effect of encainide and flecainide on mortality in a randomized trial of arrhythmia suppression after myocardial infarction. N Engl J Med 1989; 321: 406–412.
9. Cunningham T, Uzun O, Morris R, et al. The safety and effectiveness of flecainide in children in the current era. Pediatr Cardiol 2017; 38: 1633–1638.
10. Kohli V. Oral flecainide is effective in management of refractory tachycardia in infants. Indian Heart J 2013; 65: 168–171.

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