Cardiology in the Young

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Brief Report

Cite this article: Yamada Y and Kurosaki K (2024) Coronary artery compression by dilated pulmonary artery in an early infantile case of tetralogy of Fallot with absent pulmonary valve: ventricular fibrillation as the initial symptom of myocardial ischaemia. *Cardiology in the Young* **34**: 2033–2035. doi: 10.1017/S1047951124025940

Received: 30 December 2023
Revised: 3 June 2024
Accepted: 27 June 2024
First published online: 2 October 2024

Keywords:
Tetralogy of Fallot with absent pulmonary valve; ventricular fibrillation; coronary artery compression; dilated pulmonary artery

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Coronary artery compression by dilated pulmonary artery in an early infantile case of tetralogy of Fallot with absent pulmonary valve: ventricular fibrillation as the initial symptom of myocardial ischaemia

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Abstract

Coronary artery compression by a dilated pulmonary artery is a rare complication in patients with tetralogy of Fallot with absent pulmonary valve. We present a case in which this condition manifested at two months of age, with ventricular fibrillation as the initial symptom of myocardial ischaemia. It is important to recognise that this potentially fatal complication can occur in early infancy.

Introduction

Tetralogy of Fallot with absent pulmonary valve is a rare subtype within the tetralogy of Fallot spectrum. Clinically, respiratory distress due to airway compression by a dilated pulmonary artery is often a problem.[1] Reports of coronary artery compression by a dilated pulmonary artery in patients with tetralogy of Fallot with absent pulmonary valve are limited.[2–4] with no documented cases reported in the neonatal or early infancy period. We encountered a case of ventricular fibrillation as the initial symptom of myocardial ischaemia in early infancy at two months of age.

Case presentation

A prenatally diagnosed case of tetralogy of Fallot with absent pulmonary valve was delivered at 38 weeks and 3 days of gestation with a birth weight of 2,253 g. The pulmonary valve was absent as diagnosed in utero, and moderate pulmonary stenosis, moderate pulmonary regurgitation, and severe tricuspid regurgitation were observed. Respiratory distress was a concern, but the patient did not require mechanical ventilation and could be managed with non-invasive positive-pressure ventilation. Contrast-enhanced CT and echocardiography revealed proximity between the left coronary trunk and the pulmonary artery (Figure 1). No clinical signs or test results suggestive of myocardial ischaemia were observed. Since weaning from noninvasive positive-pressure ventilation was difficult, we deemed pulmonary artery plication necessary. However, given the patient's low body weight, we decided to postpone surgery until adequate weight gain was achieved. Respiration and circulation did not worsen, and the patient's weight exceeded 3,000 g at 59 days of age; as a result, surgery was scheduled for 64 days of age. At 61 days of age, while awaiting surgery, the patient experienced sudden deterioration in the clinical course. The patient suddenly developed ventricular fibrillation and cardiopulmonary resuscitation was performed. Spontaneous cardiac rhythm resumed after electrical defibrillation and adrenaline administration. Upon reviewing lead II on the bedside electrocardiogram monitor, intermittent ST-segment depression was noted several hours before the onset of ventricular fibrillation (Figure 2). In the 12-lead electrocardiogram, soon after resumption of spontaneous cardiac rhythm, there were no apparent signs of ischaemia. Ten hours after the resumption of spontaneous cardiac rhythm, the ST segments again depressed in lead II on the bedside ECG monitor. The subsequent 12-lead ECG revealed ST elevation in aVR and V1-2 and ST depression in II, III, aVF, and V5-6, which strongly suggested coronary ischaemia in the left coronary artery territory (Figure 2). Echocardiography revealed diffuse hypokinesis of the left ventricular wall. Blood tests revealed elevated levels of creatine kinase at 402 U/l (normal range: 62-287 U/l), CK-MB at 49 U/l (normal range: <12 U/l), and Troponin T at 0.941 ng/ml (normal range: <0.014 ng/ml). The policy was to increase the diastolic pressure in order to increase coronary blood flow. With the use of circulatory agonists, the blood pressure increased from 65/39 (47) mmHg to 92/56 (68) mmHg, and ST changes in the 12-lead electrocardiogram improved. Based on the clinical course of the above events, we diagnosed myocardial ischaemia

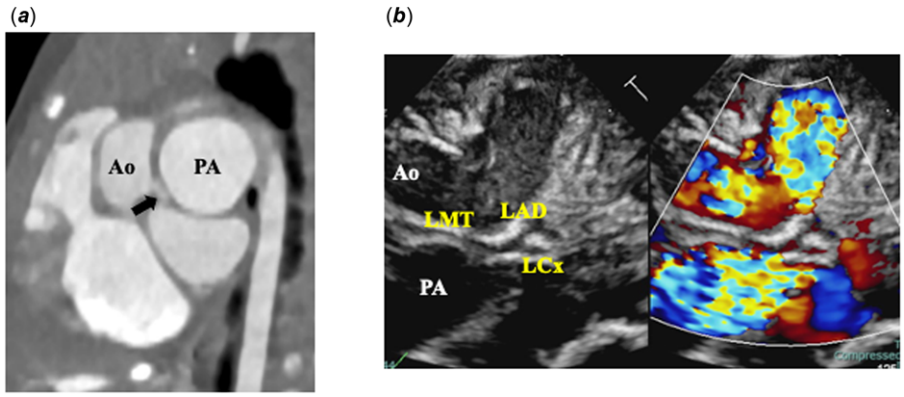

Figure 1. Contrast-enhanced CT (***A***) and echocardiography (***B***). The left main coronary trunk is in proximal to the dilated pulmonary artery. The black arrow in (***A***) indicates the left main coronary trunk. Ao: aorta, PA= pulmonary artery, LMT= left main coronary trunk, LAD= left anterior descending coronary artery, LCx, left circumflex coronary artery branch.

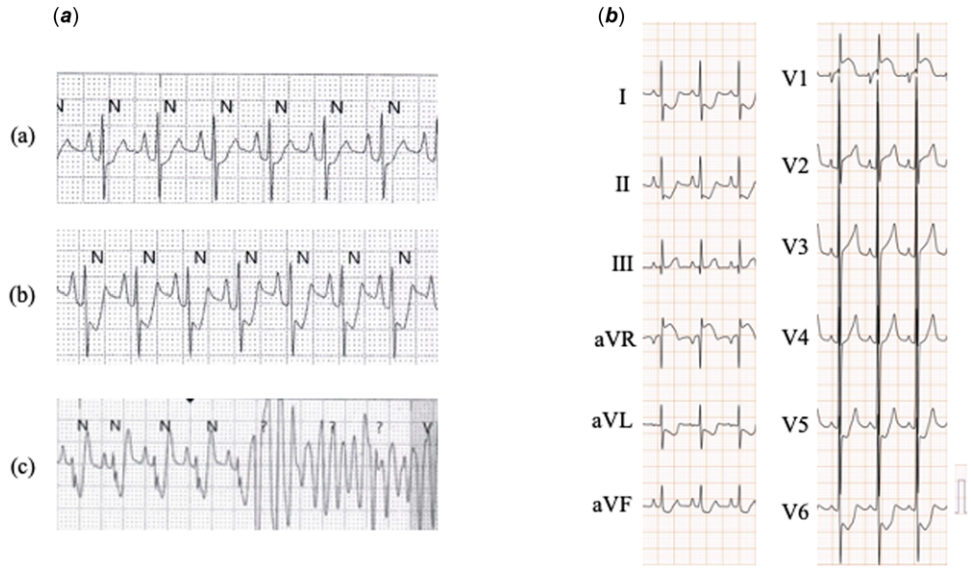

Figure 2. A lead II electrocardiogram on a bedside monitor (***A***). Two states: a state with a normal ST-segment (a) and a state with a depressed ST-segment (b), were intermittently repeated from several hours before the onset of ventricular fibrillation (c). A twelve-lead electrocardiogram during ST-segment depression on a bedside monitor (***B***). ST was elevated in aVR and V1-2, and ST was depressed in II, III, aVF, V5-6.

resulting from compression of the left coronary artery by a dilated pulmonary artery. Pulmonary artery plication, detachment of the main pulmonary artery, and systemic-to-pulmonary artery shunt were urgently performed. Subsequently, there was no recurrence of coronary artery ischaemia.

Discussion

In this patient, a rare complication of tetralogy of Fallot with absent pulmonary valve was observed, in which the left coronary artery was compressed by the dilated pulmonary artery. This uncommon complication manifests in early infancy, with ventricular fibrillation as the initial symptom of myocardial ischaemia. There have only been four reports of coronary artery compression by dilated pulmonary arteries in patients with tetralogy of Fallot with absent pulmonary valve.[2–4] The ages of onset in these reports were 6 months, 11 years, 16 years, and 17 years, respectively, and there were no cases of manifestation during the neonatal or early infancy periods. Diagnostic triggers were noted in two cases.[2,3] both of which were diagnosed by coronary angiography and contrast-enhanced CT. In those reports, cases in which ischaemic symptoms preceded the diagnosis, similar to this patient, were not identified.

There have been reports of left coronary artery compression by dilated pulmonary arteries in patients with idiopathic pulmonary arterial hypertension or CHD, such as atrial septal defect.[4,5] This pathophysiology has been widely recognised in recent years, especially in idiopathic pulmonary arterial hypertension, and its prevalence has been reported to be not necessarily low.[5] However, these reports described adult patients, and there have been no reports of cases in early infancy. A case report described a 22-year-old patient with persistent severe pulmonary hypertension after intermediate atrioventricular septal defect surgery, with compression of the left main coronary artery by a dilated pulmonary artery leading to the development of ventricular fibrillation. The authors suggested the possibility of the presence of similar pathophysiological cases among patients with pulmonary hypertension who experienced sudden death.[6] Tetralogy of Fallot with absent pulmonary valve is characterised by marked aneurysmal dilatation

https://doi.org/10.1017/S1047951124025940 Published online by Cambridge University Press

of the pulmonary arteries, and there may be cases of unrecognised coronary artery compression.

Our patient was unable to be weaned off noninvasive positive-pressure ventilation and was therefore considered a candidate for surgery, necessitating continued hospitalisation. This enabled prompt intervention when ventricular fibrillation occurred. In cases where respiratory impairment is mild, patients may be discharged home. However, it is essential to assess the risk of coronary artery compression by the dilated pulmonary artery before discharge in such cases.

In conclusion, in patients with tetralogy of Fallot with absent pulmonary valve, it is important to recognise the risk of developing fatal symptoms, including ventricular fibrillation due to coronary artery compression, which is caused by the dilated pulmonary artery, even in early infancy.

Acknowledgements. None.

Financial support. This research received no specific grant from any funding agency, commercial, or not-for-profit sectors.

Competing interests. The authors declare none.

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