Cardiology in the Young

cambridge.org/cty

Cardiac resynchronisation therapy in a pace-dependent infant with tetralogy of Fallot

Ioana Sus[1,2], Carmen Suteu[1,2] and Dan Dobreanu[1,2]

[1]Emergency Institute for Cardiovascular Diseases and Transplantation, Tirgu Mures, Romania and [2]University of Medicine, Pharmacy, Science and Technology "G. E. Palade" of Tirgu Mures, Tirgu Mures, Romania

Brief Report

Cite this article: Sus I, Suteu C, and Dobreanu D (2022) Cardiac resynchronisation therapy in a pace-dependent infant with tetralogy of Fallot. *Cardiology in the Young* **32**: 849–852. doi: 10.1017/S1047951121004169

Received: 14 July 2021
Revised: 28 August 2021
Accepted: 18 September 2021
First published online: 13 October 2021

Keywords:
Cardiac resynchronisation therapy; tetralogy of Fallot; low cardiac output syndrome

Author for correspondence:
C. Suteu, Emergency Institute for Cardiovascular Diseases and Transplantation Tirgu Mures, 540136, Tirgu Mures, Romania. Tel/Fax: 0040 212 111 - 384 / 0040 265 216 368. E-mail: suteucarmen@yahoo.com

Abstract

We present the case of a 5-month-old infant with tetralogy of Fallot and congenital atrio-ventricular block that developed severe left ventricular dysfunction during apical left ventricular pacing, in which cardiac resynchronisation therapy was used as an emergency procedure due to persistent low cardiac output syndrome.

Case report

We present the case of a preterm girl (34 weeks of gestation, birthweight 1650 g) from a twin pregnancy obtained through in vitro fertilisation. At birth, the child had a heart rate of 90 bpm and was hypoxic with an oxygen saturation of 80%. The electrocardiogram revealed a Wenckebach atrio-ventricular block and the echocardiogram identified tetralogy of Fallot with hypoplastic pulmonary annulus but without anterograde pulmonary gradient shortly after birth. In the subsequent weeks, the pulmonary stenosis became important, with a maximum anterograde gradient of 108 mmHg. Four weeks after birth, the conduction disorder progressed to high-degree atrio-ventricular block with a ventricular rate of 50 bpm and, at a weight of 2380 g, a single chamber VVI pacemaker (Medtronic Sensia) was implanted through left thoracotomy, with a bipolar epicardial lead (Medtronic CapSure Epi) sutured on the apical-lateral wall of the left ventricle and the patient was discharged 1 week later, with a left ventricular ejection fraction of 50%. At the age of 4 months, the patient was admitted for severe hypoxic spells while on betablocker and a modified Blalock–Taussig shunt was performed using a 4-mm Goretex graft. One week later, the patient developed severe low-output syndrome and invasive mechanical ventilation together with inotropic treatment was necessary. On echocardiography, the shunt was permeable but the left ventricular systolic function was severely decreased with an ejection fraction of 25%, there was intraventricular (septal-to-posterior wall motion delay of 150 ms) and interventricular dyssynchrony (difference between pre-ejection times of the two ventricles 107 ms), severe mitral regurgitation (atrio-ventricular dyssynchrony) and left ventricular volume overload due to increased pulmonary return. The patient was started on adrenaline and milrinone infusion with persistent low output in the following 2 weeks, with a maximum creatin kinase-myocardial band of 287 U/l, lactate dehydrogenase 7033 U/l and aspartat aminotransferase 3809 U/l. In this context, given the impossibility to downscale the inotropic treatment, severe left ventricular dysfunction and intraventricular dyssynchrony, we opted for complete surgical correction of the congenital anomaly and upgrade to a triple-chamber pacemaker during the same procedure. The patient underwent surgery at the age of 5 months and 4000 g weight. The pulmonary annulus was enlarged with a transannular patch, the Blalock–Taussig shunt was interrupted and the ventricular septal defect was closed with a patch. Two additional bipolar epicardial leads were implanted, one on the right atrium and the other one on the right ventricle at the border between right ventricular outflow and inflow, and all three leads were connected to a triple chamber pacemaker (Biotronik Enitra 8 HF-T), placed subcutaneously above the right rectus abdominal muscle (Fig 1b). The pacemaker was programmed as DDD 110 bpm, with a sensed atrio-ventricular delay of 70 ms and a paced atrio-ventricular delay of 90 ms, interventricular (VV) delay 0 ms, optimised based on ECG and echocardiography (Fig 1a and c). Twenty-four hours after surgery, the patient had sustained ventricular tachycardia, resistant to overdrive pacing for which first lidocaine boluses, followed by mexiletine, and amiodarone were needed, with suppression of arrhythmia episodes after 3 days. All inotropic treatment (adrenalin, noradrenalin, milrinone) was stopped after 1 week, and the patient was haemodynamically stable, with a left ventricular ejection fraction of 45%, mild mitral regurgitation and a normal transmitral flow. During the following days, two important device-related events occurred. In the context of an infection with Klebsiella, the patient had sinus tachycardia interpreted as pacemaker-mediated tachycardia by the device, which correctly started the pacemaker-mediated tachycardia pacing protocol (Fig 2a), therefore we inactivated pacemaker-mediated tachycardia suppression function. Second, we observed that at a heart rate above 160 bpm, the pacemaker switched to right

© The Author(s), 2021. Published by Cambridge University Press.

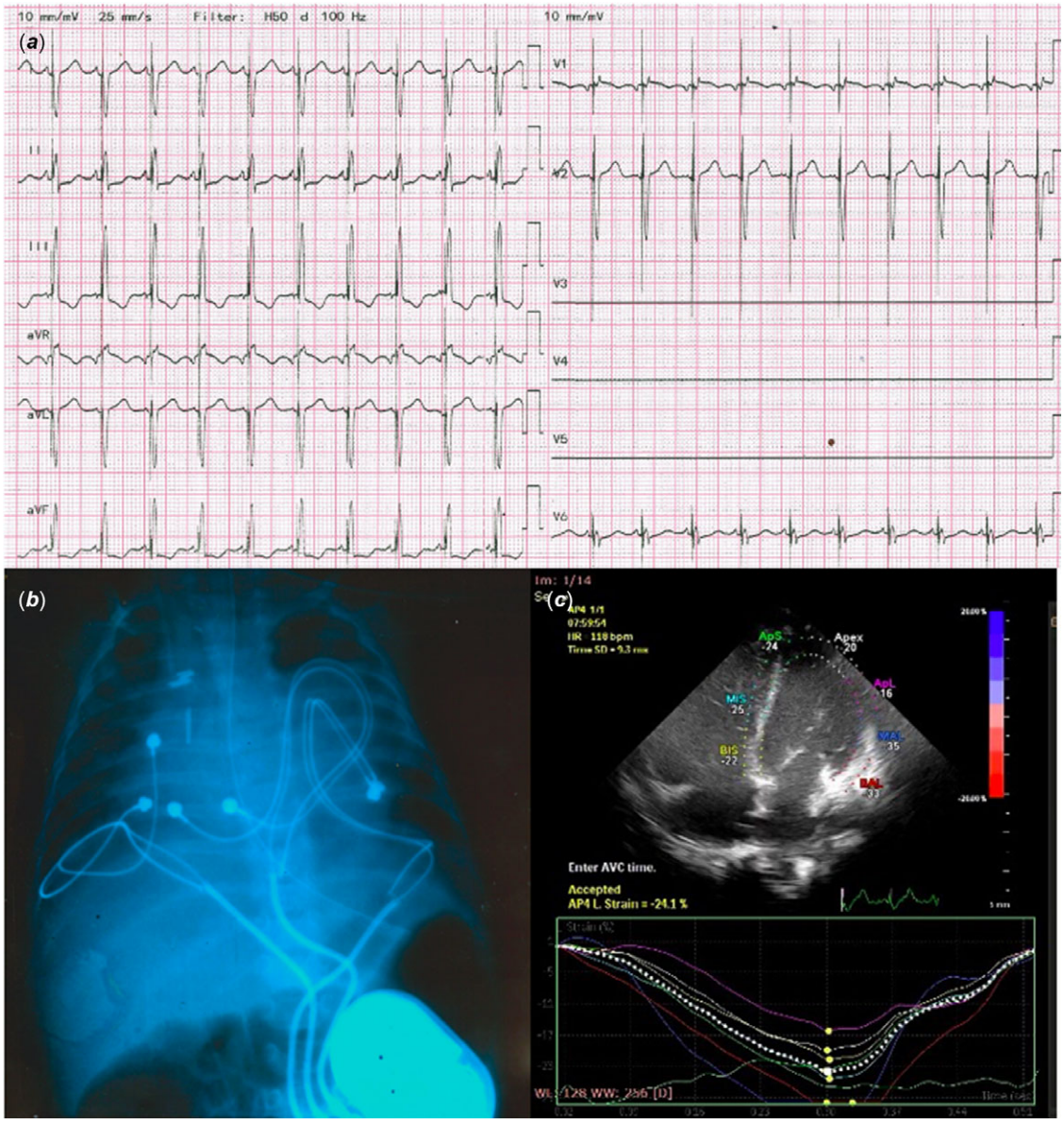


Figure 1. (***a***) Electrocardiogram after cardiac resynchronization therapy. (***b***) Thoracic x-ray after device implantation. (***c***) Apical four-chamber view with speckle tracking after resynchronization with optimal timing of contraction.

ventricular only pacing, losing the benefit of resynchronisation (Fig 2b). Unfortunately, this issue could not be solved through device programming, therefore, besides antibiotic treatment, we opted for propranolol and then ivabradine was associated in order to obtain sinus rates below 160 bpm. The evolution was favourable and the patient was evaluated 1 month, 6 months and 1 year after discharge, in good clinical status, with normal growth according to age and an improved left ventricular ejection fraction of 70% and mild mitral regurgitation.

Discussion

Cardiac resynchronisation therapy is now a widely used therapy in adult patients with decreased left ventricular ejection fraction and bundle branch block, recommended for heart failure therapy after 3 months of optimal medical treatment. The use of cardiac resynchronisation therapy in paediatric patients is a lot less standardised as there is a great anatomical heterogeneity. Three larger studies on cardiac resynchronisation therapy in paediatric patients[1–3] have shown that ventricular dyssynchrony due to single site pacing was the most frequent indication for cardiac resynchronisation therapy (44–77%), but in these cohorts the number of children with tetralogy of Fallot was low (<10%). At the same time, a decrease in LVEF was observed in less than 10% of children paced from the left ventricular apex or lateral wall.[4] Nevertheless, our patient developed biventricular dysfunction most probably both due to volume overload in uncorrected Fallot and left ventricular dyssynchrony due to pacing. In this context, as inotropic treatment could not be stopped, CRT at the time of total correction was considered the optimal solution. The greater challenge derived from cardiac resynchronisation therapy programming, as the software of triple chamber pacemakers is not adapted to paediatric heart rates. Therefore, even if sinus tachycardia at a rate of 160 bpm is not unusual in an infant, the pacemaker cannot pace using the left ventricular lead at that rate, so interventricular synchrony is lost. In this case, even if there was intermittently

https://doi.org/10.1017/S1047951121004169 Published online by Cambridge University Press

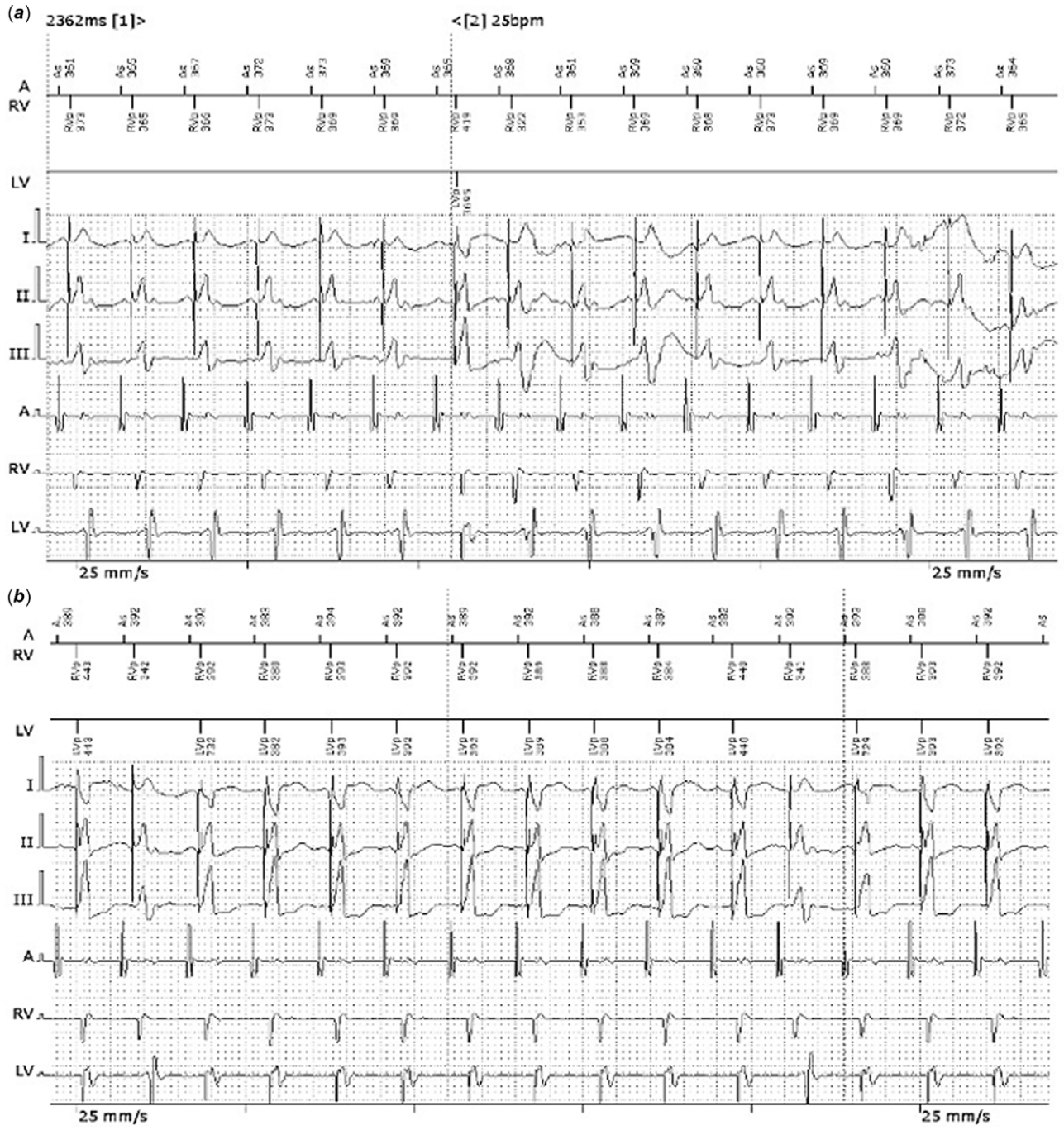


Figure 2. Device recorded intracardiac electrograms during (***a***) pacemaker mediated tachycardia supression protocol and (***b***) loss of left ventricular pacing at rates higher than 160 bpm

preserved atrio-ventricular conduction,[5] this was associated with bradycardia, so we aimed for permanent cardiac resynchronisation therapy pacing and we did not try to preserve intrinsic rhythm. In this context, balancing between physiologic tachycardia and the need to optimise cardiac resynchronisation therapy can be hard, and using ivbradine could be explored as an option. Further, cardiac resynchronisation therapy programming in children is a challenge not only due to physiology particularities but also due to technological limitations.

Acknowledgements. None.

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

Conflicts of interest. None.

References

1. Janoušek J, Gebauer RA, Abdul-Khaliq H, et al. Cardiac resynchronisation therapy in paediatric and congenital heart disease: differential effects in various anatomical and functional substrates. Heart 2009; 95: 1165–1171.

https://doi.org/10.1017/S1047951121004169 Published online by Cambridge University Press

2. Dubin AM, Janousek J, Rhee E, et al. Resynchronization therapy in pediatric and congenital heart disease patients: an international multicenter study. J Am Coll Cardiol 2005; 46: 2277–2283.
3. Cecchin F, Frangini PA, Brown DW, et al. Cardiac resynchronization therapy (and multisite pacing) in pediatrics and congenital heart disease: five years experience in a single institution. J Cardiovasc Electrophysiol 2009; 20: 58–65.
4. Janoušek J, van Geldorp IE, Krupičková S, et al. Permanent cardiac pacing in children: choosing the optimal pacing site: a multicenter study. Circulation 2013; 127: 613–623.
5. Bogdan S, Glikson M. Physiological pacing: a moving target? Eur Heart J 2015; 36: 141–142.

https://doi.org/10.1017/S1047951121004169 Published online by Cambridge University Press