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Acute Myocardial Infarction in a Neonate Caused by a Coronary Thrombosis: a Considerable Diagnostic and Therapeutic Challenge



Infarto agudo de miocardio en un neonato causado por trombosis coronaria: un gran reto diagnóstico y terapéutico

To the Editor,

Although neonatal acute myocardial infarction secondary to coronary thrombosis is extremely rare, it has high mortality (about 90%)¹ and its early diagnosis requires a high level of clinical suspicion. Of the few cases described in the literature, most are of unknown origin.² Reported risk factors include prothrombotic conditions, myocarditis, prematurity, neonatal asphyxia, Kawasaki disease, and placement of an umbilical venous catheter.^{1–4} Various therapeutic strategies are available, such as surgical thrombectomy,⁵ systemic or local thrombolytic treatment (although controversial in neonates, there have been some successful results),^{2,4} and use of an extracorporeal membrane oxygenator as hemodynamic support.^{2–6}

The present scientific letter concerns a male neonate, with no relevant obstetric history except for maternal gastroenteritis at 12 days before birth, who was born at 37 + 5 weeks gestation via emergency cesarean section due to pathological cardiotocography (DIP II). His Apgar score was 6/9. At birth, he required positive pressure ventilation for 3 min due to poor respiratory effort. He was admitted to the neonatal intensive care unit under clinical management and continuous monitoring. There, a marked progressive clinical worsening was seen, with a poor general status and mixed acidosis that required intubation, mechanical ventilation, and refractory intensive inotropic support. For the latter, he was administered stress doses of hydrocortisone. Echocardiography was performed to rule out cardiogenic shock as a differential diagnosis, revealing severe ventricular dysfunction with ejection fraction < 15%, with minimal motion of the lateral wall but with preserved right ventricular function. Both coronary arteries were adequately visualized, but no visualization was possible of left coronary artery

flow. In addition, a 12-lead electrocardiogram showed deep Q waves in DI, aVL, and V₅–V₆ (Figure 1).

With the diagnosis of cardiogenic shock due to acute left ventricular dysfunction, treatment was begun with prostaglandins to maintain systemic cardiac output through the arterial conduit. The differential diagnosis included myocarditis (due to the maternal history of gastroenteritis), coronary thrombosis, and an anomalous origin of the left coronary artery in the pulmonary artery (although it was unlikely because symptoms do not usually begin until pulmonary resistance decreases). Treatment was started with intravenous gamma-globulins and acyclovir, and it was decided to perform cardiac catheterization. Due to the patient's considerable hemodynamic instability, catheterization was delayed until the fourth day of life, but revealed a filling defect of 1 m in diameter in the left coronary artery, compatible with a thrombosis or dissection of the left main coronary artery (Figure 2, Video of the supplementary material), confirming the diagnosis of acute myocardial infarction due to thrombosis of the left coronary artery. Initial treatment was to consist of intravenous heparin and systemic r-TPA (recombinant tissue plasminogen activator) or acetylsalicylic acid but, due to the shock-induced coagulopathy, the considerable risk of brain bleeding, and the late diagnosis, treatment was begun with intravenous heparin alone for 7 days, at a dosage of 1.5 mg/kg body weight every 12 hours. The first percutaneous treatment considered was placement of an intracoronary stent, but this approach was rejected due to the risk-benefit of stent implantation in a neonate. Another possibility was intracoronary administration of r-TPA, but recovery of myocardial viability was considered unlikely due to the delayed diagnosis. In the following days, reopening of the coronary artery was seen on ultrasound, but with minimal recovery of ventricular function (30% ejection fraction). Serological and polymerase chain reaction studies of the blood for the presence of virus were negative, as were thrombophilic studies. The patient remained in the neonatology unit for 94 days due to his considerable hemodynamic lability in response to minimally invasive procedures and intercurrent processes. He was eventually discharged with diuretic

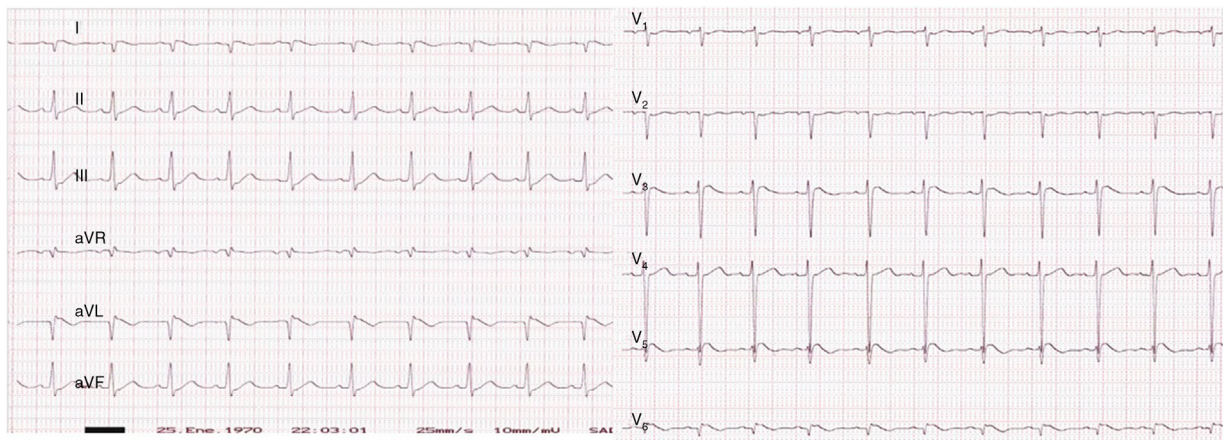


Figure 1. Electrocardiogram showing deep Q waves in DI, aVL, and V₅–V₆.



Figure 2. Catheterization of the patient with visualization of a filling defect of the left coronary artery.

therapy, digoxin, carvedilol, acetylsalicylic acid, and nasogastric feeding. During his clinical course, he was admitted several times to the pediatric intensive care unit for cardiac decompensation in the presence of catarrhal symptoms. He was included on the transplant list at 12 months of life and received a transplant 5 months later, with good outcome. Histopathology of the explanted heart showed a dilated left ventricle, with extensive areas of fibroelastosis, and almost half of the external myocardium in the free wall had been replaced by adipose tissue and fibrosis. No coronary obstruction was detected.

Acute myocardial infarction in the neonatal period is exceptional and represents a considerable diagnostic and therapeutic challenge for clinicians and cardiologists. Early diagnosis via electrocardiography, echocardiography, and catheterization is important for adequate treatment and to promote coronary reperfusion because these patients often lack adequate collateral circulation. Prompt treatment could avoid the irreversible myocardial necrosis^{3,6} that culminates in heart transplantation,

as in our patient. Various therapeutic options have been described, but there is no universal consensus because few such patients have been described and because newborns are highly susceptible to immaturity-related complications.² The most recent articles advocate local thrombolytic treatment with low-dose intracoronary r-TPA, together with the administration of intravenous heparin, with good results and without complications.^{2,4–6}

SUPPLEMENTARY MATERIAL



Supplementary material associated with this article can be found in the online version available at [doi:10.1016/j.rec.2015.06.012](https://doi.org/10.1016/j.rec.2015.06.012).

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Familial Paralysis of the Atrium Due to a Mutation in SCN5A



Parálisis auricular familiar debida a una mutación en SCN5A

To the Editor,

We present a family with a history of atrial arrhythmias requiring a pacemaker (PM) due to sick sinus syndrome (SSS) and paralysis of the atrium. Genetic study revealed the presence of the SCN5A^{p.Arg219His} variant already associated with familial paralysis of the atrium (FPA), conduction disorders, and dilated cardiomyopathy. To the best of our knowledge, this is the first report of FPA in a Spanish family.

The index case (III.3) was a 30-year-old man with no relevant medical history who was admitted for dizziness; on arrival at hospital, the electrocardiogram (ECG) revealed absence of atrial

activity and idioventricular rhythm (Figure 1A). The echocardiogram showed a severely dilated left atrium (60 mL/m²), while the other chambers were of normal size. The left ventricular ejection fraction (LVEF) in the magnetic resonance imaging study was 48% and there was no late enhancement. A dual-chamber PM was implanted. The patient improved without any complications and left ventricular ejection fraction completely recovered after 9 months of ramipril therapy.

The patient's father (II.3) required a PM at the age of 31 years, also for paralysis of the atrium. In the most recent ECG, sinus rhythm alternated with VVI pacing (Figure 1B). A paternal aunt (II.2) required a PM aged 62 years and his son (III.1) had a stroke aged 38 years and required a PM for SSS. A first cousin (III.3) had atrial fibrillation and atrial flutter from the age of 29 years, with several cardioversions, and ablation of the cavotricuspid isthmus, after which he was in sinus rhythm with propafenone (Figure 1C).