Neonatal isolated coronary artery dilatation: to treat (and how) or not to treat, that’s the question!

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Abstract

We describe the clinical presentation of a newborn, who received diagnosis of diffuse right coronary artery dilatation and a high origin of the same vessel, in the absence of intrauterine hypoxia or other identifiable causes of coronary artery ectasia. The relevance of the case is in the complex aetiology of congenital heart malformation. In addition, we also highlight the difficult therapeutic management of these patients, in lack of guidelines for newborn population.

Keywords

Coronary dilatation, cardiopathy, Kawasaki disease, Parvovirus B19, ASA, newborn.

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How to cite


Case report

P., born at 39 weeks of gestation from spontaneous vaginal delivery. Apgar score 1’ 8, 5’ 9, weight 2,920 g (10th-25th pc), length 49 cm (25th pc),


head circumference 34 cm (25th-50th pc). Maternal pregnancy investigation for congenital infections was completely negative (we found out IgG-positivity for Rubeo-virus, CMV, HSV in antenatal period). Obstetric morphologic ultrasound, performed at 21 weeks of gestation, was normal. Although the last obstetric ultrasound, performed at 35 weeks of gestation, highlighted fetal ascites, it was not confirmed at 37 weeks of gestation, when a dilated intestinal loop was discovered. Because of these findings, P. after birth underwent a complete malformation screening, which included cardiac, abdominal and cerebral ultrasound. The abdomen ultrasound on the 4th day of life did not confirm pathological findings emerged during pregnancy, and also fetal ascites was excluded.

However, pediatric cardiologists performed a cardiac ultrasound, and they found out an anomalous right coronary artery origin (so-called high origin) and also a dilatation of the same vessel throughout its course (proximal diameter 1.8 mm, z-score +3.13; median diameter 1.8 mm, z-score +3.8), without signs of aneurismatic lesions (Fig. 1). No other signs of congenital cardiopathy were detected. A complete cardiac evaluation (including cardiac ultrasound, ECG, blood pressure measurement and continuous pulse oximetry monitoring) was carried out for our newborn. During the following 20 days, ultrasound characteristics were unmodified, with a stable right coronary artery dilatation and perivessel hyperechoic signals in heart scans. We also measured artery blood pressure, daily diuresis, and all parameters were normal according to day of life. We did not find signs or symptoms of congestive heart failure. In addition, because of a high inflammatory index (CRP: 55, 10 times upper limit), without fever or other signs, suspecting an early onset neonatal sepsis, a 7-day double antimicrobial therapy was prescribed (ampicillin 50 mg/kg/die and gentamicin 4 mg/kg/die). A progressive normalization of CRP was observed. Blood culture was negative. To establish etiology of this condition, we investigated the main infectious causes of congenital heart defects. IgG for Parvovirus, VZV, Coxsackie, Enterovirus were positive (possible maternal origin); instead, IgM were negative. We also performed polymerase chain reaction (PCR) to detect Parvovirus B19 and CMV DNA on blood sample, and both of them were negative. In addition, our patient did not fulfil criteria for Kawasaki disease (absence of suggesting clinical, laboratory and instrumental data). Once excluded these causes, we concluded for a case of congenital coronary dilatation. We also faced the difficulty of therapeutic choice in a similar case. Considering the risk for aneurismatic evolution of the vessel dilatation, our cardiologists prescribed a prophylactic therapy with acetylsalicylic acid (ASA) at antithrombotic dosage (4 mg/kg/die), for at least 3 months, according to the latest guidelines for Kawasaki disease. We discharged P. on the 25th day of life, in overall good clinical conditions, with a weight of 3,240 g. Formulated milk was prescribed, because of a pathological loss of weight, and also antithrombotic therapy was confirmed (ASA 4 mg/kg/die). P. underwent a strict follow-up, at the beginning twice a month, then monthly, till now (10-month-old baby). To monitor therapy efficacy and also possible side effects, laboratory evaluation (including total blood cell count, coagulative indices, blood glucose and creatinine) was performed monthly. We also dosed once again antibodies IgG titre for Parvovirus B19, which showed a significant decay throughout this period. Cardiac ultrasounds performed during these months showed a stable situation of right coronary dilatation, without aneurismatic degeneration.

**Discussion**

Coronary ectasia has been defined as distension of a part of the artery (more than 150% of the diameter of an adjacent unaffected segment) which involves the whole vessel, in opposition to a classical aneurysmatic lesion. Our patient also presented a so-called high origin of the right coronary vessel [1, 2]. Normally, a coronary artery arises below the sinotubular junction (levels I or II) and, if the vessel arises on level III, it is

![Figure 1. Isolated right coronary artery dilatation and perivessel hyperechoic signals.](image-url)
considered high origin. Although an anomalous origin of coronary artery is a common and incidental finding, neonatal coronary dilatation is a rare condition, which provides Neonatologists with a wide differential diagnosis. Except for the ones associated with congenital heart malformations (bicuspid aortic valve and others) [3], a common cause of neonatal coronary ectasia is neonatal Kawasaki disease. This condition, though relatively rare in neonates, may present in this population in the absence of classical criteria. Because of a difficult early diagnosis, if untreated, the cardiac sequelae of this disease can be serious. Even if atypical or incomplete forms of Kawasaki disease are the most frequent in the newborn [4], we excluded this diagnosis for P., because he did not match diagnostic criteria, except for the high CRP and an early coronary enlargement [5]. Considering the prenatal transient ascites, infectious profile has been widely deepened [6]. Mother-foetal Parvovirus B19 infection has been associated with foetal anemia, hydrops fetalis, non-hydropic intrauterine foetal death and asymptomatic foetal infection [7]. Both maternal and newborn positivity for Parvovirus IgG, linked to a negative PCR for that virus on neonate’s blood, seemed to exclude a congenital infection, even if we cannot date mother’s one. This assumption is supported by the progressive decay of baby’s IgG titres, and could be confirmed at complete negative IgG titre until the first year of life. During the post-discharge follow-up, ultrasound evaluation showed a static imaging situation. In similar cases, the risk of developing aneurysm is strictly linked to the aetiology of the condition and antithrombotic therapy is still controversial. We have to highlight the extreme difficulty of this therapeutic choice, considering that ASA is off label in this range of age. We were aware that such a measure on one hand would expose the baby to possible side effects, on the other could result ineffective. In our case, despite a lack of information about aetio-pathogenetic factors, the increased risk of ischemic events, connected to anomalous rheology of blood in affected vessels, justified the therapeutic choice. In addition, also the duration of treatment was a subjective decision, because we extended the therapy over the classical 3 months. The last review on Kawasaki disease [8] represents the only document which approved pediatric antithrombotic therapy in these conditions.

**Conclusion**

This case has been really interesting because it points out some critical issues in neonatal clinical management. First of all, the negative consequence of a fragmented maternal infectious history. On the other hand, the importance of an extended screening for malformation in suspected newborn. Last but not least, the difficult therapeutic management of these borderline cases, when international diagnostic criteria are not completely fulfilled. Further studies are needed to evaluate the progression rate of the clinical findings and also to optimize drug therapy in this class of patients.

**Declaration of interest**

The Authors declare that there is no conflict of interest.

**References**