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Brief Report

Cardiac features in the presymptomatic period in a neonate with anomalous left coronary artery arising from the pulmonary trunk

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Abstract We describe a neonate presenting with a cholestatic jaundice due to Alagille syndrome. On echocardiography as part of the diagnostic work-up, we noted a slight dilation of the normally functioning left ventricle at the initial examination in the third week of life. Over the next 2 weeks, serial echocardiograms showed slowly progressive dilation and dysfunction of the left ventricle, together with a persistent mild elevation of levels of cardiac Troponin-I in the serum. These findings, unrelated to the underlying Alagille syndrome, prompted cardiac catheterisation, which confirmed that the main stem of the left coronary artery was originating from the pulmonary trunk. Surgery was successfully performed still in the presymptomatic period. The association of Alagille syndrome with anomalous left coronary artery arising from the pulmonary trunk is most unusual. We emphasise the cardiac findings prior to detection of the anomalous origin of the coronary artery.

Keywords: Alagille syndrome; newborn; Troponin-I

ANOMALOUS ORIGIN OF THE LEFT CORONARY artery from the pulmonary trunk is a rare congenital anomaly, accounting for no more than 0.5% of all congenital cardiac malformations. Children with this anomaly usually remain asymptomatic, without suspicious physical signs, until they become symptomatic within the first months of life with signs of congestive heart failure due to ischemic left ventricular dysfunction.[1]

Alagille syndrome is characterized by paucity of intralobular bile ducts, cholestasis, cardiovascular anomalies, typically peripheral pulmonary stenosis, vertebral anomalies such as the butterfly arrangement, ocular anomalies with a characteristic triangular facies, renal disease, and optional involvement of other organs. The association of anomalous origin of the left coronary artery from the pulmonary trunk in this syndrome, as far as we are aware, has previously been reported only once.[2] We now report a second case, in particular because the sequence of diagnosis gave the unique opportunity to observe the course of anomalous origin of the left coronary artery from the pulmonary trunk in the period prior to development of symptoms.

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Case report

A male newborn was referred at the age of 16 days because of progressing jaundice, beginning a few days after birth. After an uneventful twin pregnancy, he was born spontaneously at term, and was small for gestational age with a birth weight of 2325 g, the weight of his twin sister at birth being 3450 g. In the days immediately prior to his admission, the boy was drinking poorly. The baby presented with clinical signs of dehydratation, with a weight still below that measured at birth. He showed a greenish icteric skin colour, an enlarged liver, and a grade II systolic murmur in the back, with no signs of cardiopulmonary decompensation. Except for a triangular facial form, with frontal bossing, there were no other dysmorphic signs. Laboratory tests revealed a direct hyperbilirubinemia, elevated liver enzymes, renal involvement with elevated creatinin, and a metabolic acidosis. The clotting studies were normal. Ultrasound of the abdomen showed an

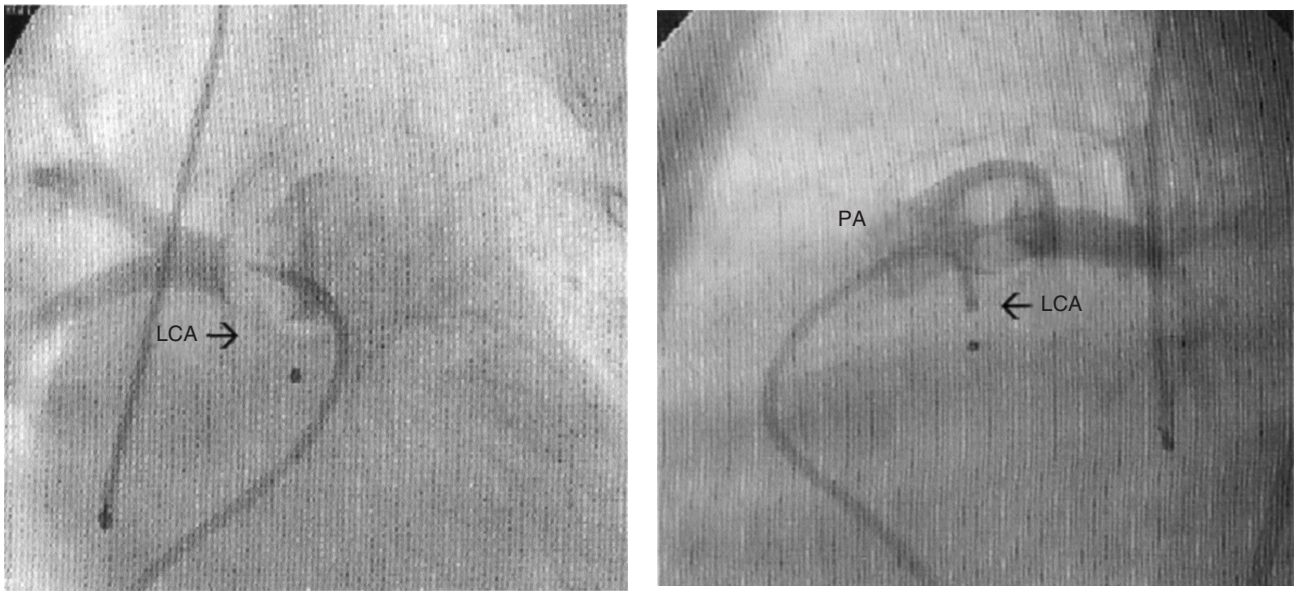

Figure 1.
Biplane angiography in the pulmonary trunk, with a balloon inflated in the distal part of the trunk (PA), shows that the left coronary artery (LCA) originates from the inferior aspect of the trunk.

enlarged liver with a hyperechogenic texture, a small gallbladder, and non-dilated biliary vessels. Ocular examination showed bilateral posterior embryotoxon. The molecular genetic analysis confirmed a mutation typical for Alagille syndrome in the JAG1 gene.

Echocardiography revealed bilateral stenosis of the branches of the pulmonary trunk, with gradients of 28 mmHg on each side. The first echo examination showed a slightly dilated left ventricle, with an enddiastolic diameter of 22 mm, with normal function, the ejection fraction being 68%. At that time, an elevated level of cardiac Troponin-I was found in the serum, at 2.6 mg/l. Repeated echocardiograms over the next 10 days showed progressive dilation of the left ventricle to 24 mm enddiastolic diameter, and deterioration of left ventricular function, with the ejection fraction decreasing to 42%. There was also the development of mild mitral regurgitation. Repeated measures of Troponin-I in the serum showed an ongoing mild troponinemia of between 1.4 and 2.8 mg/l.

Due to this course, which seemed unrelated to the Alagille syndrome, we suspected anomalous origin of the left coronary artery from the pulmonary trunk. After an electrocardiogram showed typical alterations,[3] with deepened wide Q waves in leads I and aVL, the baby was taken to the catheterization laboratory, where a blocked angiogram in the pulmonary trunk (Fig. 1) confirmed the diagnosis. Operation, with direct reimplantation of the left coronary artery in the aortic root, was performed. There were no serious perioperative complications. After open-heart surgery, the levels of Troponin-I normalized within 9 days. By 10 weeks after the operation, echocardiography showed normalization of cardiac function, ejection fraction returning to 64%, and left ventricular enddiastolic diameter reducing to 20 mm, in the absence of mitral regurgitation.

Discussion

The most common cardiovascular anomalies in patients with Alagille syndrome are stenosis of the branches of the pulmonary trunks and tetralogy of Fallot. Any other malformation can occur, but they are reported to be very rare. We found only one patient previously reported[2] with anomalous origin of the left coronary artery from the pulmonary trunk in association with Alagille syndrome.

Besides the very unusual association of these two diseases, the interesting observation in our patient, from the stance of the cardiologist, was the possibility to observe the presymptomatic course of anomalous origin of the left coronary artery from the pulmonary trunk, thanks to the early echocardiography performed for routine work-up in Alagille syndrome. Except for the typical murmur of stenosis of the branches of the pulmonary trunk, our patient did not show cardiac symptoms at that point. Together with the typical echocardiographic signs of a increasing left ventricular dilation, and progressive impairment of left ventricular function, we have been able to demonstrate a mild but persistent troponineamia in the early presymptomatic course. Together with the other electrocardiographic and echocardiographic signs that have

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already been established as helping in discrimination between anomalous origin of the left coronary artery from the pulmonary trunk and dilated cardiomyopathy,[3,4] a persistent elevation in levels of Troponin might also be of possible help as a clue to early diagnosis in similar cases. We are not aware of any other report giving details of the presymptomatic evolution of cardiac features in this disease. Thanks to modern day surgical techniques,[5,6] the long-term prognosis of these patients is good. Even so, all signs that might lead to earlier diagnosis and treatment would be of value for these children.

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