

Coarctation of the aorta presenting as dilative cardiomyopathy: Exception or distinct mode of clinical presentation?

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While neonates with coarctation frequently present with congestive heart failure and critically reduced perfusion of the descending aorta following ductal closure, infants and older children with coarctation are usually oligosymptomatic and are diagnosed due to weak femoral pulses, heart murmur or arterial hypertension. We encountered several infants with coarctation, who presented with dilative cardiomyopathy (DCM) and reviewed our patients to determine, whether this presentation represents a rare exception or is relevant for the differential diagnosis of children with DCM.

101 infants were referred to our hospital for treatment of coarctation between 1/2001 and 12/2013. 24 of these patients were older than 4 weeks. Among them five children with severe coarctation presented at an age of 2.1-5.9 months under the clinical picture of DCM. These children accounted for 5% of those, who were diagnosed with coarctation in the first year of life and 21% of those, who presented in infancy. Echocardiography revealed a dilated left ventricle and markedly reduced function with shortening fractions between 9-19%. The aortic arch was normal in diameter with a circumscribed severe coarctation with some distance to the left subclavian artery. All patients underwent resection of the coarctation within 24 hours. Significant postoperative improvement of the left ventricle resulting in a normal shortening fraction occurred within a median interval of 1.7 months.

Coarctation of the aorta presenting as DCM in infancy has been rarely reported in the literature. In our study however this specific clinical presentation accounted for 21% of infants with discrete coarctation, who became symptomatic beyond 4 weeks of age. The stenosis was difficult to detect due to its distal location, the normal configuration of the aortic arch and the low isthmic gradient resulting from low cardiac output. Indirect hints are bicuspid aortic valve, thickened myocardium and reduced flow in the celiac artery. Following treatment of the coarctation left ventricular function appears to recover completely. Since these patients require urgent treatment differing completely from other forms of DCM, careful examination of the isthmic region should be included in the primary assessment of all infants presenting with DCM.