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Isolated left ventricular apical hypoplasia in a 9-year-old girl - A rare differential diagnosis for dilatated cardiomyopathy

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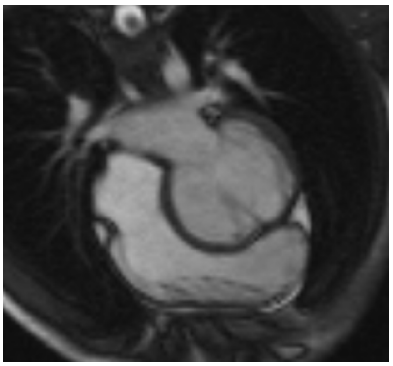

Introduction: Isolated left ventricular apical hypoplasia (LVAH) is a very rare primary cardiomyopathy with non-specific symptoms. This entity is probably contributed to an inadequate dilatation of chambers during partitioning.

Methods: In our female patient already prenatal ultrasound showed high echogenicity of the left ventricular myocardium. Postnatal echocardiograms showed a spherical dilated left ventricle with a normal function and no clinical symptoms. The diagnosis was congenital dilatative cardiomyopathy. During follow up echocardiographics enddiastolic diameter increased.
Therefore and due to the atypical left ventricular configuration, we performed a catheter study at the age of 9 years. There were no further pathological results apart from a mild elevated left-ventricular enddiastolic pressure (LVEDP) of 12 mmHg and the spheric configuration of the left ventricle. The histological analysis of myocardial biopsies showed no signs for an inflammation but a moderate interstitial myocardial fibrosis, consistent with dilatative cardiomyopathy.
Subsequent cardiac magnetic resonance imaging revealed the four typical morphological characteristics of LVAH consisting of a truncated and spherical left ventricular configuration with rightward bulging of the interventricular septum, a fatty replacement of the left ventricular apex, a complex papillary muscle network in the anterior apex, an elongation of the right ventricle wrapping around the deficient left ventricular apex and a left ventricular out-pouching (Figure). Due to the elevated LVEDP we started a treatment with Captopril.

Conclusion: To the best of our knowledge this is the first report of LVAH with abnormalities already observed during fetal echocardiography. Therefore this rare congenital heart defect should be considered as a differential diagnosis to dilatative cardiomyopathy in the newborn.