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ISOLATED LEFT VENTRICULAR APICAL HYPOPLASIA IN A 9-YEAR-OLD GIRL - A RARE DIFFERENTIAL DIAGNOSIS FOR DILATATED CARDIOMYOPATHY

INTRODUCTION

Isolated left ventricular apical hypoplasia (LVAH) is a very rare primary cardiomyopathy with non-specific symptoms and up to now just appeared as incidental findings. This entity is probably contributed to an inadequate dilatation of chambers during partitioning. Patients faced with LVAH should be monitored regularly due to life-threatening consequences. Abnormalities during early fetal echocardiographics have not been reported yet.

CASE REPORT

In our female patient already prenatal ultrasound showed high echogenicity of the left ventricular myocardium (17th week of pregnancy). Therefore, the newborn was presented postnatal in our clinic for further diagnostic workup. The echocardiograms showed a spherical dilated left ventricle with a normal function. No clinical symptoms were observed. The diagnosis was congenital dilatative cardiomyopathy. During follow up echocardiographics at the age of 8 years, enddiastolic diameter increased (z-score 3.09;Figure 1).

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 borderline elevated left-ventricular enddiastolic pressure (LVEDP) of 12 mmHg and the spheric configuration of the left ventricle (Figure 2). The histological analysis of myocardial biopsies showed no signs for an inflammation but a moderate interstitial myocardial fibrosis, consistent with dilatative cardiomyopathy.

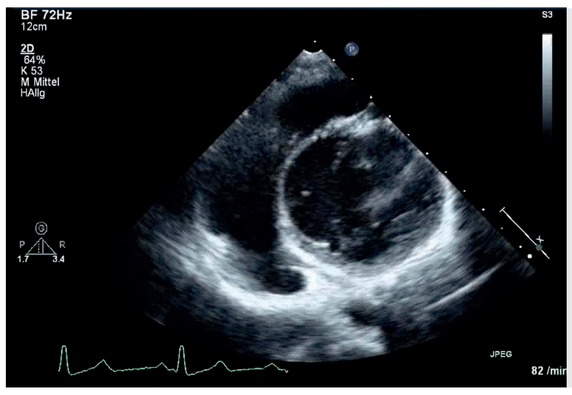

Figure 1
Echocardiogram showing a spherical dilated left ventricle with a normal function.

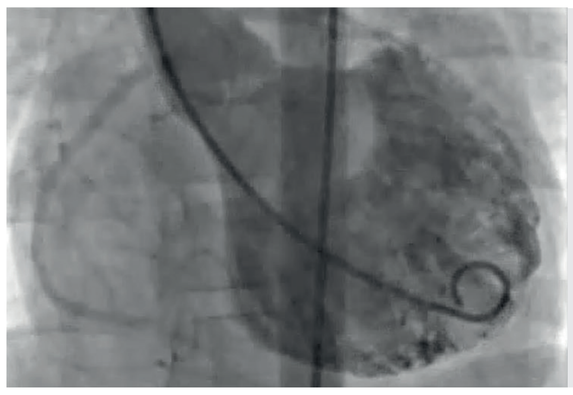

Figure 2
Catheter study at the age of 9 years showing a spheric configuration of the left ventricle (30°RAO view) and a borderline elevated left-ventricular enddiastolic pressure (LVEDP) of 12 mmHg.

Subsequent cardiac magnetic resonance imaging revealed the four typical morphological characteristics of LVAH

- 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 outpouching (Figure 3).

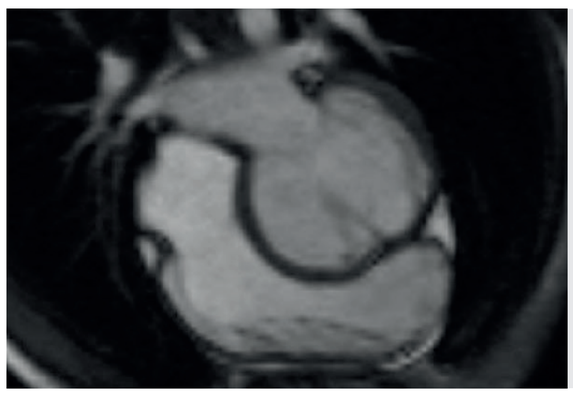

Figure 3
Cardiac MRI presenting the typical morphological characteristics of left ventricular apical hypoplasia.

Due to the borderline 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 early fetal echocardiography. Therefore this rare congenital heart defect should be considered as a differential diagnosis to dilatative cardiomyopathy in the newborn.

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