Right ventricle involvement in foetal critical aortic stenosis with heart failure - autopsy findings following prenatal intervention.

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Introduction: Foetal aortic stenosis came into focus since it started to be managed interventionally in utero. However, little is known about the developmental mechanisms underlying the spectrum of the disease. It can be stable throughout the foetal life, evolve to become the hypoplastic left heart or progress to the foetal heart failure. Some authors suggest the involvement of second heart field cells responsible for the formation of compact myocardium and fibrous framework of the cardiac musculature and postulate the secondary role of the aortic valve itself. The aim of the study was to assess the histological features of left and right ventricular wall in autopsy of hearts obtained from the foetuses after in utero aortic valvuloplasty and correlate the results with previously obtained echocardiographic data.

Methods: In years 2011-2014 37 foetal aortic valvuloplasties in 34 fetuses were performed at our institution. 3 fetuses died in utero, 3 in early neonatal period, 2 of them after premature delivery. The autopsy was performed in 3 cases. Samples of the right and left ventricular walls were taken and stained with hematoxylin and eosin, Van Gieson trichrome for connective tissue and orcein for elastic fibers. Foetal echocardiographic examinations of those patients before the foetal aortic valvuloplasty were reviewed.

Results: Left ventricular samples of all the cases presented the endocardial fibroelastosis, myocardial disarray and increased amount of interstitial connective tissue. In the right ventricular samples, myocardial disarray was present in all cases and increased amount of interstitial connective tissue in 2 of them. On echocardiography the foetuses presented signs of heart failure with severe impairment of left ventricular function. In all the cases there were signs of the right ventricular failure: shortened or monophasic inflow to the right ventricle and mild tricuspid regurgitation.

Conclusions: Both left and right ventricular myocardium presents microscopic structural abnormalities in foetal hearts with critical aortic stenosis and foetal heart failure. Post mortem findings are consistent with foetal echocardiographic features. The results support the role of formation of the compact myocardium in development of foetal critical aortic stenosis presenting with heart failure.