MARFAN SYNDROME: ECHOCARDIOGRAPHIC VALVULAR CHARACTERISTICS COMPARED TO HISTOLOGICAL FINDINGS

ACC Poster Contributions
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Background: Two major cardiovascular manifestations of Marfan syndrome are abnormalities of the aorta and the mitral valve (MV). Few studies have addressed the aortic (AV) and tricuspid (TV) valves in Marfan Syndrome patients (pts). The preoperative echo features of Marfan Syndrome pts undergoing AV replacement, MV replacement or repair, TV repair, the Bentall procedure, or ascending aortic replacement were compared with the valvular surgical histopathological findings.

Methods: From January 2004 to October 2009, preoperative echoes of 73 pts with Marfan Syndrome were retrospectively reviewed to identify the morphology and the hemodynamics of the cardiac valves, and to compare the findings with the histopathology, to summarize their echo characteristics and to evaluate the prevalence of the valve prolapse.

Results: Among 73 pts with Marfan Syndrome undergoing cardiac surgery, 66 pts had AV replacement or the Bentall procedure. Among these pts, 29 pts had available histology and all had valvular mucoid degeneration. Of 63 pts with echo determined moderate or severe AV regurgitation, only 4 pts (6.1%) had histologically thickened, floppy valves. Of 18 pts with echo determined MV involvement, 15 pts (20.5%) had MV prolapse, 11 pts had moderate to severe MV regurgitation of which 4 pts had MV repair and 7 pts had MV replacement; all 7 MV replacement pts had histologically thickened valve tissue and mucoid degeneration. Echo detected TV involvement in 8 pts (11.0%); all 8 pts had TV prolapse, 2 pts had severe TV regurgitation and underwent TV repair. Seven pts had both MV and TV involvement.

Conclusion: A survey of preoperative echoes of pts with Marfan Syndrome undergoing cardiac surgery showed that the MV has the highest incidence of cardiac valve involvement with findings that are readily detected, that when the TV is involved it is in combination with mitral valve lesions and that AV involvement had few echo manifestations. Preoperative echoes on pts with Marfan syndrome have distinctive and significant findings.