Nandita Sharma et al., J Clin Exp Cardiolog 2016, 7:6(Suppl)
http://dx.doi.org/10.4172/2155-9880.C1.036

Congenital aneurysm in the mitral-aortic intervalvular fossa in a 7-year old

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The mitral–aortic intervalvular fibrosa (MAIVF) 1 is a region of the heart that connects the anterior mitral leaflet to the posterior aortic root and is in communication with the left ventricular (LV) outflow tract, this area contains fibrous, relatively avascular tissue and is the weakest area of the aortic ring. As a consequence of its poor vascularization, this structure is susceptible to injury during aortic or mitral valve replacement or destruction from bacterial infective endocarditis leading to development of an aneurysm or pseudoaneurysm of intervalvular fibrosa (P-MAIVF). This condition is rare in pediatric age group and is usually preceded by aortic valve surgery, active or prior infective endocarditis, and congenital heart disease.

Case Report: A 7-year old girl previously healthy was referred for evaluation of murmur and abnormal electrocardiogram which was suggestive of biventricular hypertrophy. Transthoracic echocardiogram (TTE) revealed a pulsatile aneurysmal structure (1.1/0.8 cm) posterior to the aortic root and extending to the mitral valve in the intervalvular mitral-aortic fibrosa. There was turbulent flow on color Doppler, filling and expanding the structure during systole and emptying during diastole. (Fig 1). There was no mitral or aortic regurgitation. A cardiac MRI was performed which confirmed the findings of echocardiogram. Her case was discussed with the pediatric cardio-thoracic surgeons and since she was asymptomatic, and her aneurysm size was deemed small, not compressing on adjacent structures or causing regurgitation, it was decided to manage the aneurysm conservatively. She has been followed up for 4 years now with mild increase in her aneurysm to 1.7/1.4 cm.

Discussion: We present a congenital mitral-aortic intervalvular fossa aneurysm in a previously healthy 7 year old without any preceding event which reflects a truly congenital nature of the aneurysm. Clinical manifestation may vary from being asymptomatic to shortness of breath, congestive heart failure, cerebral vascular accidents or other embolic events, chest pain, active endocarditis infection.

Enlargement of the aneurysm may lead to aortic or mitral regurgitation secondary to failure of leaflet coaptation, compression of adjacent structures including the left atrium, coronary arteries, conduction system and pulmonary artery. Close proximity to the left atrium and aorta may result in fistulous communications with these structures. Rupture into the pericardium resulting in cardiac tamponade is the most dreaded complication, though such an event appears to be very rare. There is a possibility of coexistent aneurysms reinforcing it being a congenital defect.

Echocardiography usually establishes the diagnosis. Visualization of the echo-free space with systolic expansion and diastolic collapse of the aneurysmal structure is an important diagnostic feature. However, this phenomenon can be less prominent in a patient with a fistulous communication because the pressure variations within the aneurysm are blunted. Diagnosis is more accurate with a transesophageal echocardiogram than with a transthoracic echocardiogram. Real time 3D echocardiography can also be complimentary to 2D. Computed tomography and MRI both provide excellent multiplanar and 3D images of the aneurysm, its relationship with the coronary arteries, aortic root, and cardiac chambers, which are invaluable to the operating surgeon.

The usual recommended treatment is surgery due to potential fatal complications. While there have been some case reports of conservative management and monitoring, there is insufficient data to predict natural course of congenital subvalvular aneurysms. Surgery is technically challenging and associated with high risk of morbidity or mortality The high-risk factors which increase predisposition to rupture include active endocarditis, size of P-MAIVF >3 cm, bicuspid aortic valve, regurgitation, presence of fistula to cardiac chamber or aorta, thrombus in P-MAIVF, compression of adjacent structures(coronary or pulmonary artery). These high risk features would likely contribute to the progression of the pseudoaneurysm and the development of complications. There have been suggestions that patients with thick fibrous and calcific walls might be at lower risk for rupture.