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Isolated supravalvular aortic stenosis: Classic case with hourglass configuration of ascending aorta

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Clinical Image Description

A one year old male child presented with feeding diffculties and failure to thrive. On examination body weight was 7 kg, blood pressure was 10/50 in right upper limb and 102/50 in left upper limb. Heart rate was 130/minute. Respiratory rate was 26/minute. On auscultation harsh grade IV/VI Ejection Systolic Murmur (ESM) was heard maximum over right first and second intercostal space, radiating to bilateral carotid arteries (right > left). Echocardiography of patient revealed features of Left Ventricular Outflow Tract (LVOT) obstruction. Patient was taken up for cath study which revealed characteristic hourglass deformity of ascending aorta on angiogram (Figure 1). Also, there was stenosis of left common carotid artery at its origin. Pull back pressure tracings showed increased left ventricular systolic pressure and raised aortic pressure just above the aortic valve level and pressure gradient of 60 mmHg at supravalvular level (Figure 2). The child had no abnormal facial features, no positive family history and a normal serum calcium levels. Based upon these findings, a diagnosis of sporadic Supravalvular Aortic Stenosis (SVAS) was made. Patient was subsequently referred to department of cardiothoracic vascular surgery and was offered corrective surgery. Patch aortoplasty was done. Patient recovered after the surgery.

SVAS is least common cause of LVOT obstruction, and is characterized by narrowing of ascending aorta in the region of sinotubular junction. It can occur in association with Williams syndrome (characterized by elfin facies, hypercalcemia and mental retardation) or as familial form (associated with autosomal dominant pattern of inheritance and positive family history) or sporadic form. Three recognized morphologic varieties have been described: the segmental variety, which results in characteristic hourglass configuration (most common type); hypoplastic type, in which there is tubular thickening and hypoplasia of ascending aorta and membranous type, consisting of a simple fibrous membrane. SVAS is elastic arteriopathy

and may involve other arteries as well. Echocardiography shows narrowed ascending aorta as visualized from suprasternal, subcostal and long axis parasternal view. Catheter angiography reveals characteristic "hourglass" appearance in segmental form and diffuse narrowing in hypoplastic form. Hemodynamic significance of stenosis can be inferred from trans-stenotic gradient. A gradient of more than 50 mm of Hg is present in significant obstruction. Surgical repair of SVAS is indicated when significant obstruction is present.

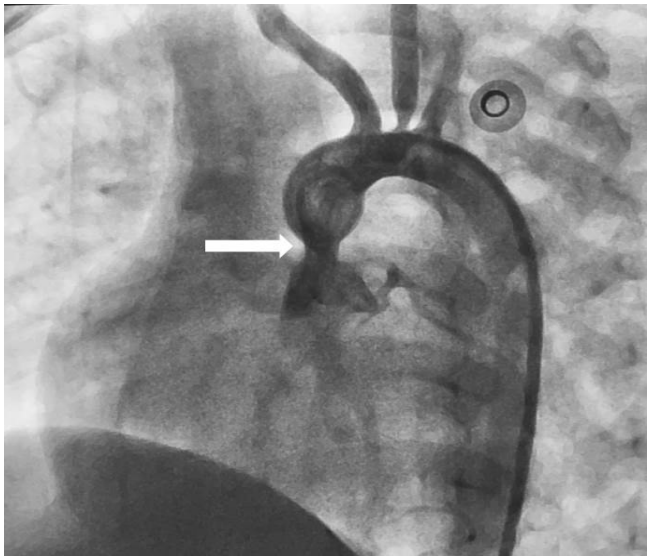

Figure 1: Angiogram showing classic hourglass appearance in ascending aorta at sinotubular junction (white arrow). Also note stenosis of left common carotid artery at its origin.

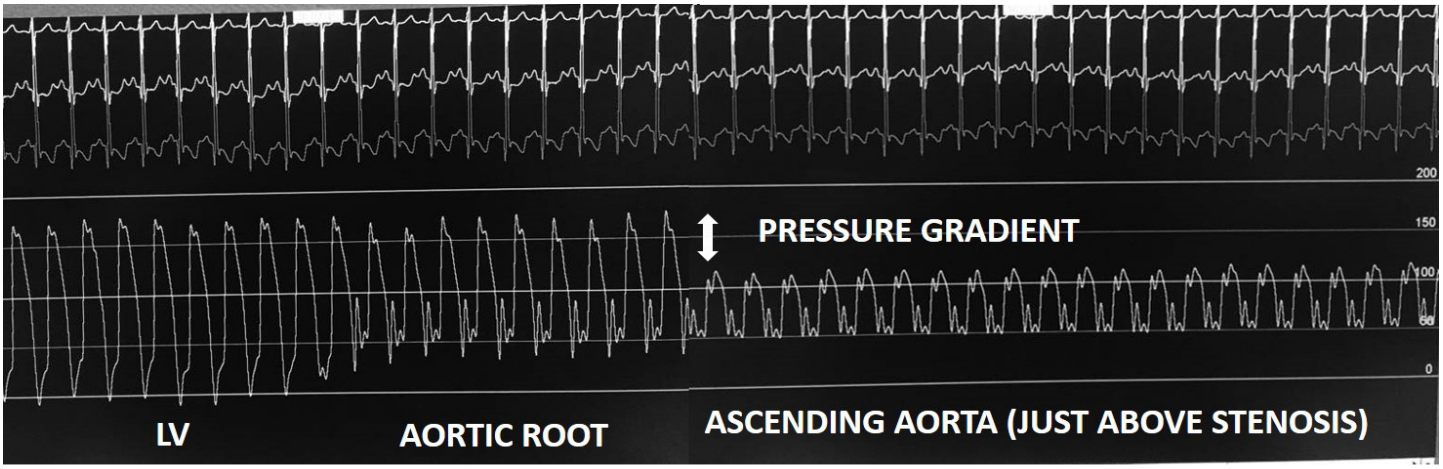

Figure 2: Pressure tracing showing no gradient by pullback between LV and aorta just above aortic valve level (LV pressure=170/35mmHg, Aorta pressure=170/50 mmHg) and a gradient of 60mm of Hg in ascending aorta, just superior to aortic valve (pressure=110/50 mmHg).

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