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IMAGES in PAEDIATRIC CARDIOLOGY

Ponniah U, Overholt E. Severe reversible dilated cardiomyopathy associated with a large left ventricular thrombus in a young child with middle aortic syndrome. Images Paediatr Cardiol 2014;16(4):7-13.

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MeSH

Middle aortic syndrome, coarctation of the aorta, dilated Cardiomyopathy, left ventricular thrombus

Abstract

We report a case of a seven-year girl who presented with severe dilated cardiomyopathy (DCM) associated with a large thrombus in the left ventricle (LV). She had a long segment stenosis of the lower thoracic descending aorta, possibly due to non-specific aortitis and underwent successful stent angioplasty. The LV thrombus resolved after heparin without sequelae.

Background

Coarctation of the lower thoracic aorta or abdominal aorta, known as middle aortic syndrome (MAS) is a rare cause for aortic obstruction. Older children present with upper extremity hypertension, weak femoral pulses and a heart murmur. Presentation with LV dysfunction and severe DCM complicated by LV thrombus is unusual.

Case summary

A seven-year-old girl presented with abdominal pain and cough. She was afebrile, tachypneic, tachycardic, and had hepatomegaly (5cms), upper extremity hypertension (124/82 mm Hg), weak peripheral pulses and a gallop. Echocardiogram showed LV end diastolic dimension measuring 53 mm, diminished LV shortening fraction (SF) of 9%, a non-mobile LV thrombus measuring 4.2 x 1.1 centimeters. (Fig.1, Fig.2, Fig.3 and Fig.4) and a normal upper thoracic aorta.

Fig.1. Short axis view, 2 D echocardiogram showing a large LV thrombus in the LV apex (arrow)

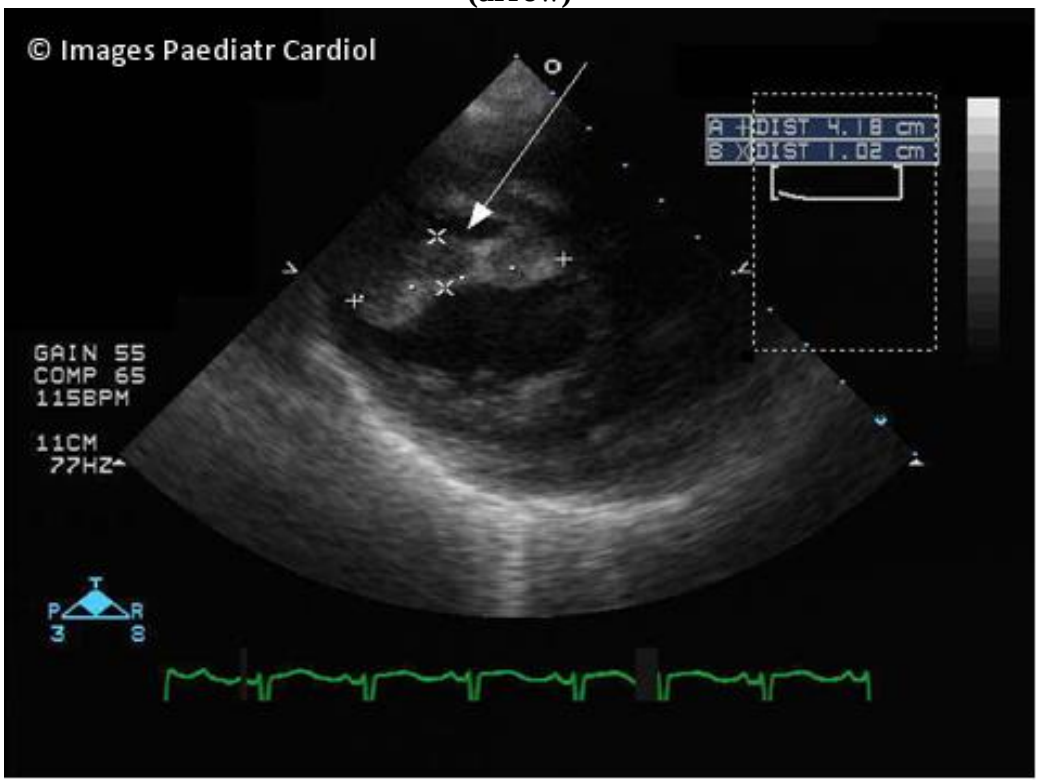


Fig.2. Four-chamber view, 2D echocardiogram showing a large non-mobile thrombus in the LV apex (arrow)

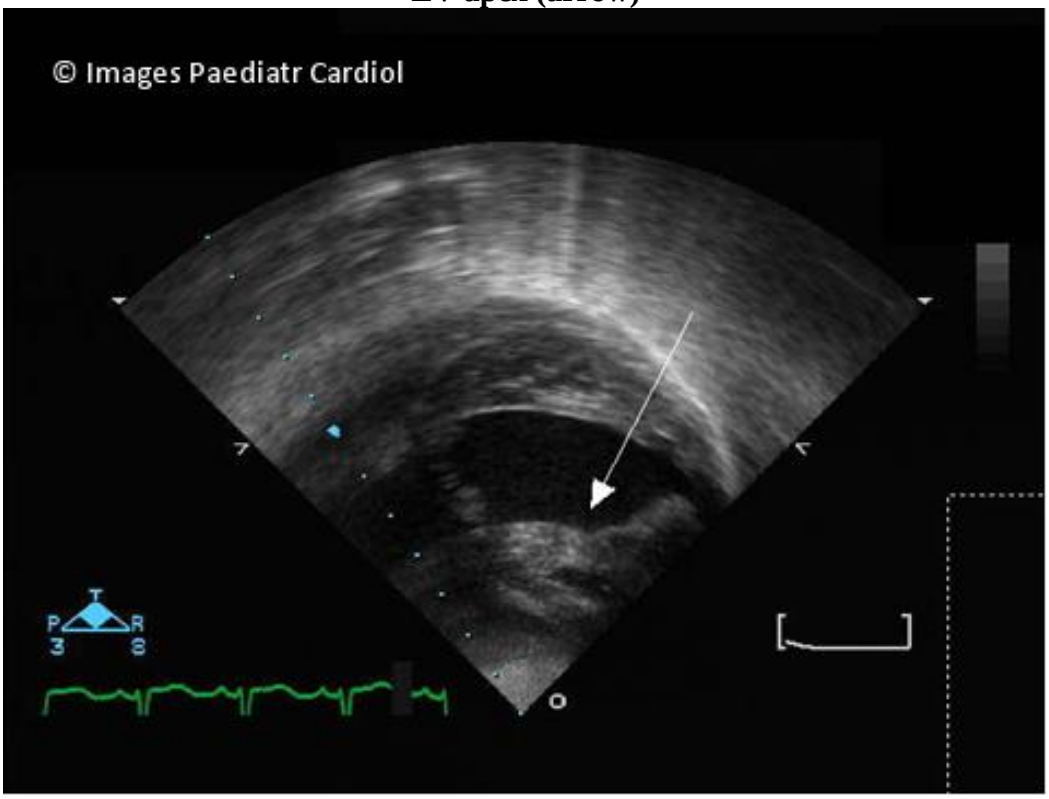


Fig. 3. Short axis view, 2 D echocardiogram showing a large LV thrombus in the LV apex (VIDEO)

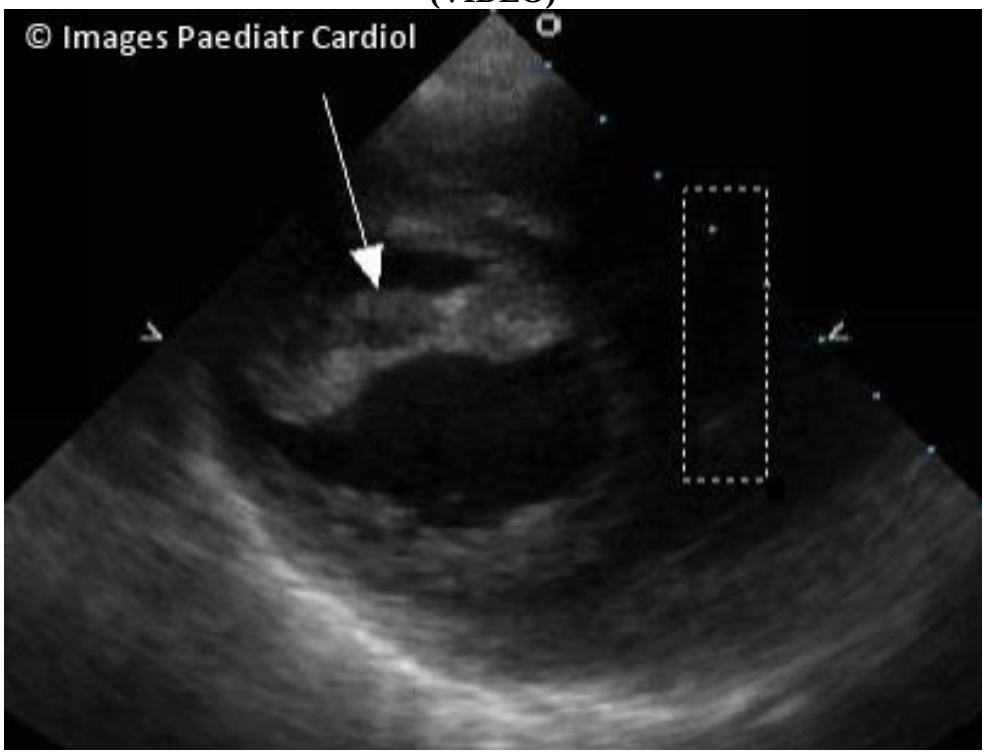
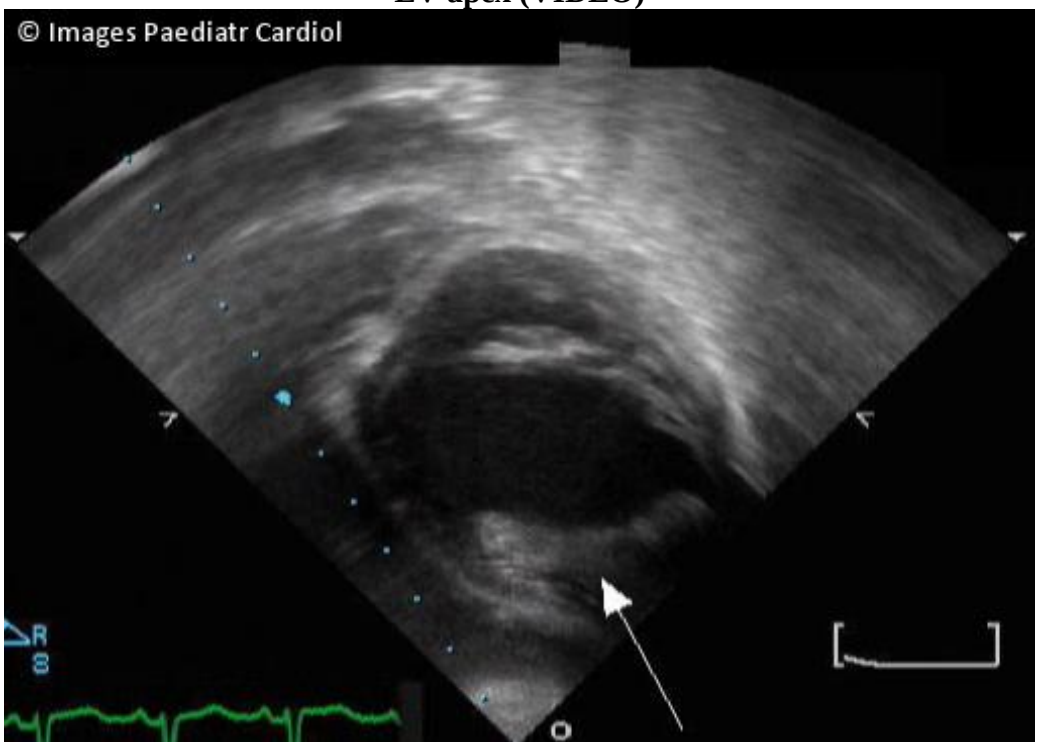


Fig. 4. Four-chamber view, 2D echocardiogram showing a large non-mobile thrombus in the LV apex (VIDEO)



Thrombophilia work up was negative and the ESR was 2 mm/hr. She was treated with inotropes and intravenous heparin followed by enoxaparin after 72 hours. She underwent cardiac catheterization, which showed severe long segment stenosis of the distal thoracic aorta with no collaterals (Fig. 5, Fig.6 and Fig.7), normal coronaries and no stenosis in the renal, celiac or

mesenteric vessels. She underwent successful stent angioplasty (Genesis XD 36mmx10mm) of the coarctation with no significant residual gradient (Fig.8 and Fig.9).

Fig.5. Aortography, AP view demonstrating a long segment stenosis (arrow) of the lower thoracic aorta with irregularity of the walls and mild post-stenotic dilation.

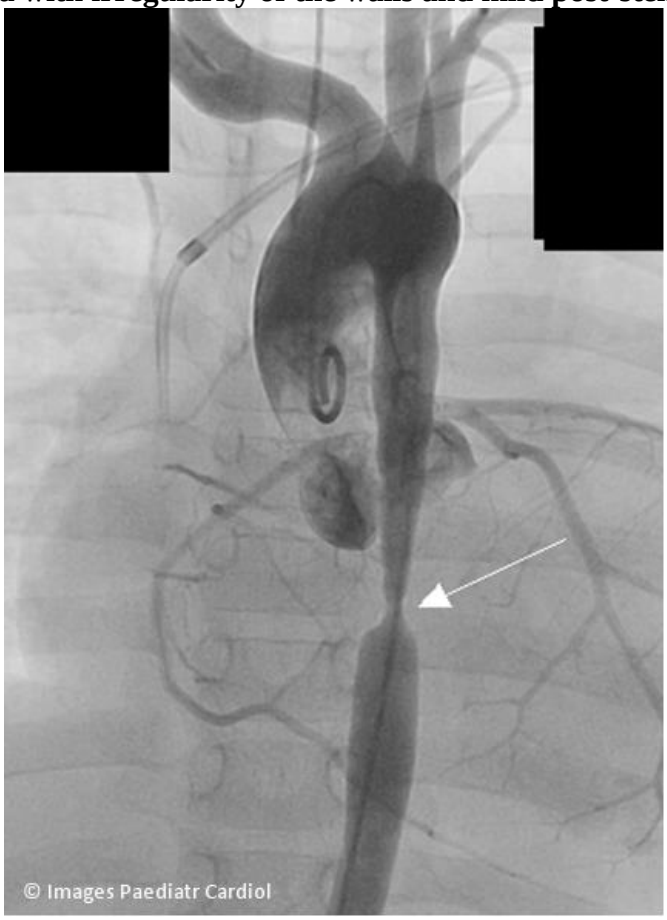


Fig.6. Aortography, AP view demonstrating a long segment stenosis (arrow) of the lower thoracic aorta with irregularity of the walls and mild post-stenotic dilation (VIDEO).

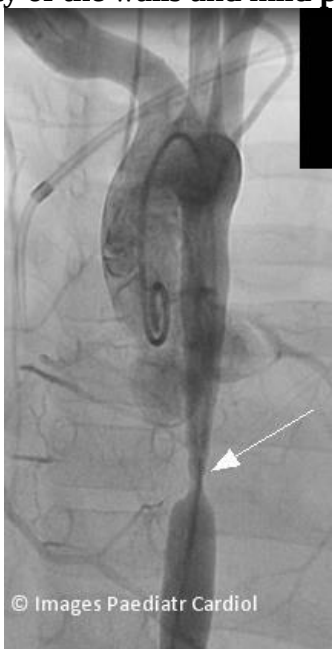


Fig.7. Aortography, lateral view demonstrating a long segment stenosis (arrow) of the lower thoracic aorta with irregularity of the walls and mild post-stenotic dilation (VIDEO).

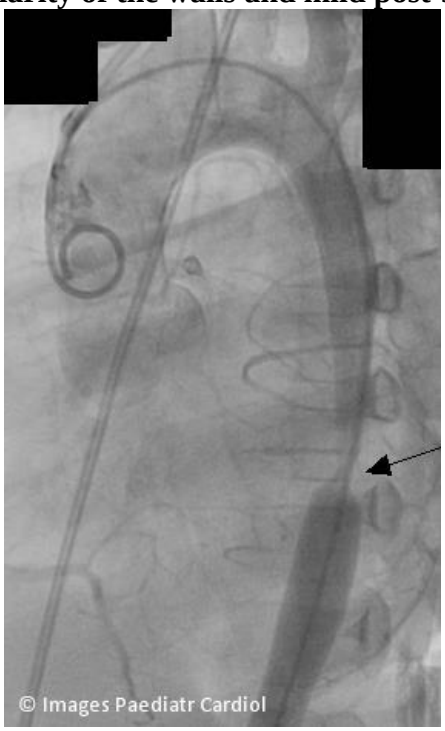


Fig.8. Aortography, post stent balloon angioplasty (arrows) with no residual coarctation.

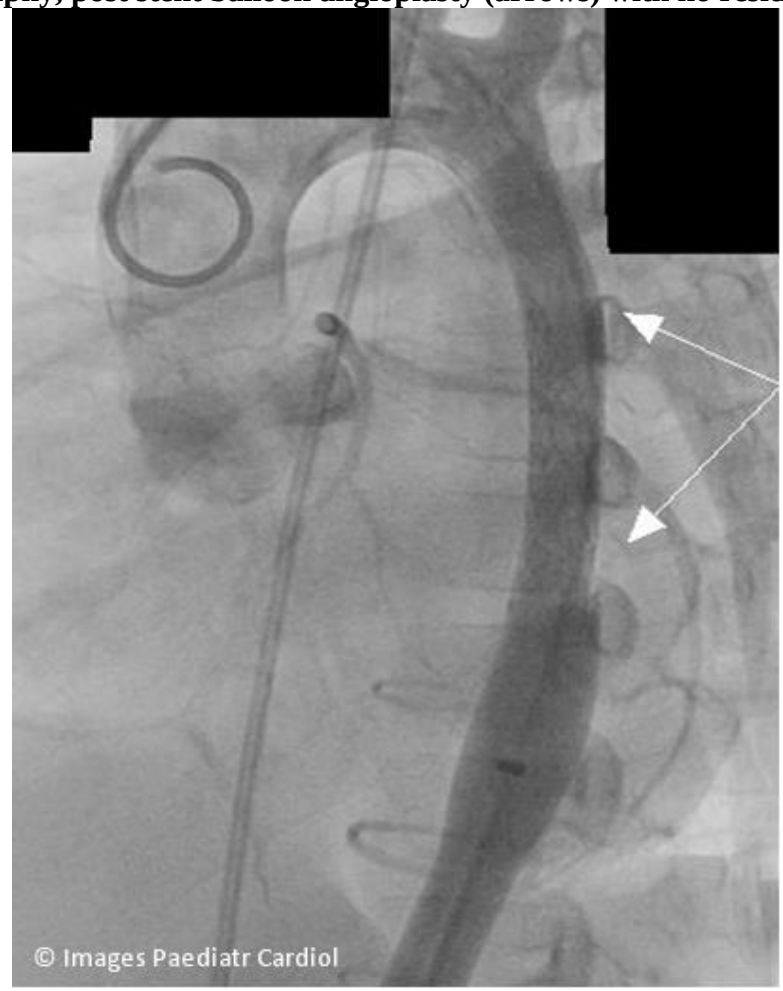
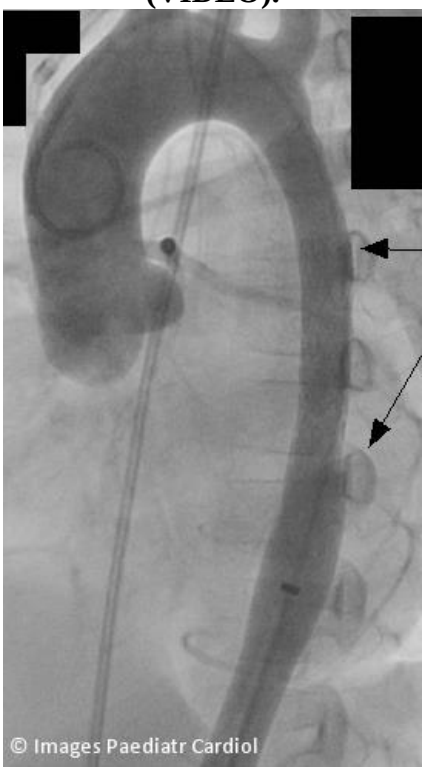


Fig.9. Aortography, post stent balloon angioplasty (arrows) with no residual coarctation (VIDEO).



She had no embolic complications and the LV thrombus resolved completely in six weeks. Seven years after two follow up angioplasty procedures, there is no residual coarctation, with normal upper extremity blood pressures, normal LV size and function (LVSF 34%).

Discussion

MAS first described by Sen [1] is a rare cause for aortic arch obstruction with a 0.5 to 2% incidence and commonly affects children and teenagers.[2] It occurs due to segmental narrowing of the distal thoracic aorta or abdominal aorta and is commonly acquired due to Takayasu's disease, aortitis syndrome, William syndrome, neurofibromatosis, and fibromuscular dysplasia.[2 3] Classic clinical findings include blood pressure difference between the arms and legs, weak or absent femoral pulses, bruit over the inter scapular area or abdomen.[1] Mild irregularity of the vessel wall seen in our patient suggests aortitis.

Diagnosis and treatment of MAS can be challenging. CTA or MRA are equally useful alternate imaging modalities for diagnosis. MAS may need surgery using bypass grafts particularly when renal and mesenteric vessels are involved.[2 3] Due to the young age and focal stenosis, endovascular stent intervention was accomplished successfully similar to other young patients.[3]

LV thrombus secondary to significant DCM can occur.[4] Since the LV thrombus was not mobile we elected to treat the patient conservatively with enoxaparin. Tissue thromboplastin inhibitor therapy has been used and surgical removal is recommended if there is imminent threat to systemic embolization with a large mobile mass.[5]

References

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