

## Successful re-vascularisation following renal angioplasty in a child with Takayasu arteritis

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### Introduction


Takayasu arteritis is the most common paediatric granulomatous large vessel vasculitis<sup>1</sup>. In children, it is insidious in onset with non-specific symptoms and later presents with complications, making it difficult to diagnose at an early stage<sup>2</sup>. We report a 14-year-old girl who presented with a hypertensive emergency and was diagnosed to have Takayasu arteritis. In view of severe renal artery stenosis and persistent hypertension requiring multiple antihypertensives, she underwent a successful left renal angioplasty.

### Case report

A 14-year-old girl presented with a history of easy fatiguability, intermittent headaches and gradually increasing breathlessness for one and a half years. The symptoms increased over the last two months for which she was admitted. On evaluation, she was found to have a hypertensive emergency with evidence of end organ damage. Four-limb blood pressure measurement revealed a significant difference with the mean upper limb pressures higher than the mean lower limb pressures by more than 10mm Hg. Two-dimensional echocardiography showed left ventricular hypertrophy with severe systolic dysfunction and an ejection fraction (EF) of 11%. A computed tomography (CT) angiography showed moderate to severe stenosis of the aorta with diffuse wall thickening and periaortic fat stranding. Right renal artery showed complete occlusion with the presence of a collateral supply. There was severe stenosis of the left renal artery. The blood parameters revealed anaemia (haemoglobin - 9 g%), thrombocytosis (platelet count 480,000 /cu mm) and elevated C-reactive protein (46.5 mg/L). The initial blood investigations were suggestive of an inflammatory aetiology.

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Coarctation of aorta (CoA) presents at a younger age with no signs of inflammation. Further, CoA is juxta-ductal and initial echocardiography excluded it. Hypertension and its complications are common manifestations of Takayasu arteritis (TA) and the significant difference in four limb blood pressure with lower limb pressure less than upper limb pressure suggested the possibility of TA. Imaging suggestive of involvement of infra diaphragmatic aorta and magnetic resonance findings of periaortic fat stranding, suggestive of inflammation, were all pointers to TA.

The child was diagnosed as TA and started on high dose steroid therapy. Injection methotrexate was added for long term maintenance. For the hypertensive emergency, she was started on nitroglycerin infusion, and gradually shifted to oral antihypertensives amlodipine, prazosin, clonidine and furosemide. In view of resistant hypertension despite multiple antihypertensives, and involvement of both renal arteries, a left renal angioplasty was planned. Left renal artery was dilated with a 4 mm renal angioplasty balloon (4 x 13 mm Mozec NC balloon; Figure 1). Post procedure, the child was continued on antihypertensives and immunosuppressive medications.

On 2-year follow up, child is now on reduced antihypertensives, off steroids and on continued methotrexate. A repeat CT angiogram showed patent left renal artery of normal calibre. There was diffuse wall thickening of descending thoracic aorta and upper abdominal aorta. The right renal artery is persistently occluded with presence of a collateral supply. A repeat echocardiography showed an EF of 48%.

### Discussion

Takayasu arteritis (TA) is a large vessel vasculitis causing granulomatous inflammation of the aorta and its branches<sup>1</sup>. The diagnosis of childhood TA is often difficult as it has an insidious onset with non-specific symptoms<sup>2</sup>. Our case also had symptoms for the last 2 years but they were nonspecific in the form of headaches and easy fatiguability. But while presenting to our hospital she had breathlessness, acute life-threatening hypertensive emergency and end organ damage.

As per Aeschlimann FA, *et al*<sup>3</sup>, arterial hypertension is the main presenting feature in children and is related to renal artery stenosis; 5-27% of children can have cardiovascular complications like cardiomyopathy<sup>4</sup>. Our child had hypertensive cardiomyopathy with an EF of 11% at presentation requiring emergency

management. The root cause was bilateral renal artery stenosis. In children, TA commonly involves aorta and infra-diaphragmatic renal and mesenteric arteries<sup>5</sup>. Blood parameters and imaging modalities help in identification of active inflammation. In our child, both acute inflammatory parameters were elevated as well as CT angiogram which was suggestive of inflammation along with renal artery stenosis.

In the index case treatment was approached in two prongs with initial management for control of hypertension and suppression of inflammation. This was followed by endovascular intervention in view of resistant renovascular hypertension requiring multiple antihypertensives. High dose steroid is the mainstay for

remission induction<sup>6</sup> which was given to our patient, followed by methotrexate, a steroid-sparing disease modifying anti-rheumatic drug.

Re-vascularization procedure in the form of angioplasty, with or without stenting, is done in steno-occlusive disease<sup>7</sup>. It reduces long-term complications and helps in managing the refractory hypertension in renal artery stenotic lesions. In a study by Hong S, *et al*<sup>8</sup>, a follow up imaging study of 9 patients was available showing lumen patency in 64% cases. Our case highlights that in patients of TA with persistent hypertension caused by renal artery stenosis, endovascular intervention and angioplasty should be considered.

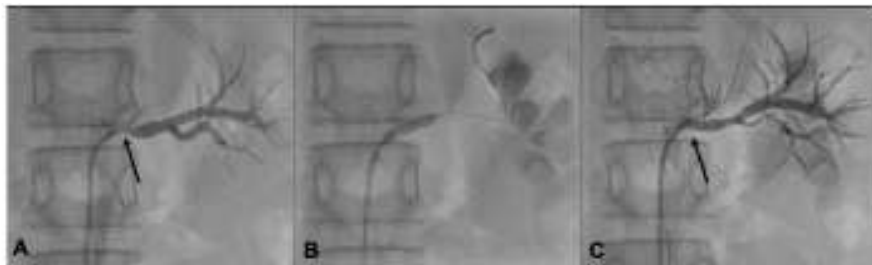


Figure 1: Selective left renal artery angiography shows severe short segment stenosis involving the origin and proximal portion. A. Balloon dilatation of the left renal artery. B. Post balloon dilatation selective left renal artery angiography reveals complete resolution of the stenosis. C.

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