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# Takayasu's arteritis a rare cause of hypertension and cardiac hypertrophy in Bangladeshi children- a case report and literature review.

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**Abstract**

Takayasu's arteritis or pulseless disease is a chornic large vessel vasculities the cause of which is largely unknown. Takayasu's arterities is seldom reported, because it is a rare disease affecting about only 3 (three) people of every million indivisuals throughout the world, most commonly among Asian female of childbearing age. It rarely causes happertension in children.

We present a 11 years old girl without typical symptomatic manifestation and with a unique presentation of headache & swelling of the body for one year. She also complaints of examinational dyspnoea for same duration and abdominal pain for six months. Her clinical examination revealed very feeble left upper limb pulse with no radio-femoral delay, different blood pressure of both arms and limbs, systolic bruits audible in left 2nd intercostal space and grade-1 apical pansystolic murmur. Her coronary angiogram report showed diffuse narrowing of a long segment of abdominal aorta, severe stenosis of proximal left subclavian artery and stenosed right renal artery. In this article we will describe a Takayasu's arteritis (TA), a rare cause of childhood hypertension.

**Keyword :** Takayasu's arteritis (TA);Vasculitis; Hypertension.

## Introduction

Takayasu's arteritis or Pulseless disease is a chronic vasculitis of large vessels. It is rare, but most commonly seen in Japan, South East Asia, India, and Maxico. In 1905 Takayasu, professor of ophthalmology at Kanazawa University Japan presented the case of a 21 year old women with characteristic fundal arteriovenous anastomoses due to narrowing of large and medium size vessels claudication, hypertension, dizziness, headache and visual problems can occur.Using a stethoscope, one may also hear "bruits"(harsh,"whoosing" sounds made by the flow of blood through abnormally narrowed vessels.) Smaller vessels are spared.

Takayasu's arteritis (TA) is a type of vasculitis. Also known as "Aortic arch syndrome and pulseless disease" causes is unknown but studies that it may occur after certain type of infection.It also may be an autoimmune causes of blood vessel inflammation. In takayasu's arteritis due to chronic inflammation damages the aorta, the large artery that carries blood from heart to the rest of the body and the main branches due to inflammation of vessels leads to wall thickening, fibrosis, stenosis, thrombosis, end organ ischaemia and aneurysm formation. As a result weak or absnt pulses occurs in peripheral part of extremities. Although it has been reported worldwide, it is more common in young Asian women. Females are about 8-9 times more likely to get it than males. People usually get the disease between 15 to 30 years of age. It is rarely reported in children[1].We here by reported a rare cases of hypertension and cardiac hypertrophy in a secondary to takayasu arteritis (TA).

## Case Report

A 11 years old girl presented with history of difficulty of respiration on exertion, headache and swelling of the whole body for last one year, abdominal pain and low grade irregular fever for last 6 months. There was no history of oliguria, hematuria, rash, joint pain, jaundice, cough or contact with tuberculosis. Physical examination revealed a conscious, oriented, anemic, febrile (temperature was 100°F), mildly edematous girl with puffy faces. Pulse was 104 beat per minute which was regular. Right radial artery pulse was normal in volume but in left radial, left & right femoral arterial pulse volume was feeble, there was no radio-radial & radio-femoral delay, blood pressure was 180/100 mm Hg in right hand and 120/90 mm Hg in both lower limbs and

left hand, common carotid pulsations were equal on both side and absent carotid bruit but bruit audible over right renal angle. Examination of cardiovascular system revealed jugular venous pulsation was raised, visible cardiac impulse present, apex beat was thrusting in character, which is shifted to left 6th inter-costal space lateral to mid clavicular line and grade-1 apical pan systolic murmur radiating to the axilla, systolic bruits audible in left 2nd intercostal space. Respiratory system and other systemic examination were normal. The optic fundi were normal.

**Laboratory investigation were as follows**

Hemoglobin was 8.2gm/L,total leucocyte count - 19000/cmm, Neutrophils-66%, Lymphocyte-25%, Monocyte-07%, Eosinophils-02%, Basophil-00%, erythrocyte sedimentation rate (ESR)-100 mm in first hour, C-reactive protein (CRP) was increased.Blood glucose,serum electrolyte,lipid profile were normal, Mantoux test, VDRL, AntiHCV, Anti HIV, TPHA, HBsAg, ANA Screening ,Antids-DNA, cANCA, p-ANCA were negative, SerumC3, and C4, liver function test & renal function test were normal. Urine analysis showed spot urine protein creatinine ratio-1, Ultrasonography of Kidney, Ureter & Urinary bladder (KUB)-normal findings of both kidney, Tc-99m DTPA showed-Rt. kidney-50%, Lt. Kidney-50%,

For evaluation of cardiovascular system, ECG showed left ventricular hypertrophy(LVH) with strain, Chest–X-Ray Posterior-Anterior view showed cardiomegaly.

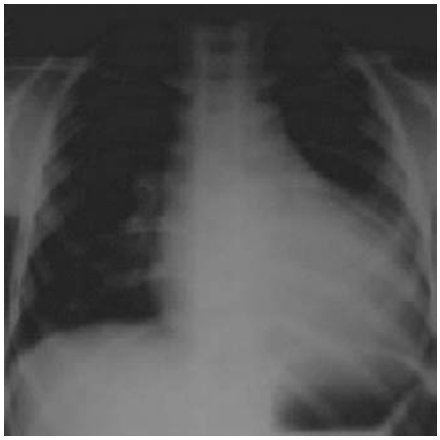
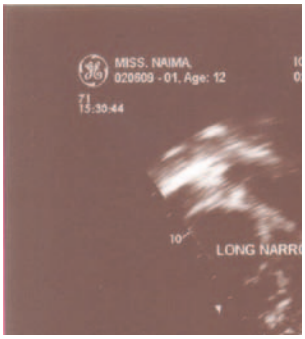


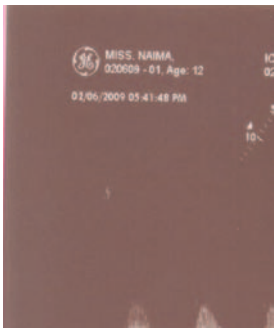
*FIG: 1 Chest X-Ray (Posterior-Anterior) View*

Echocardiography showed concentric hypertrophy of left ventricle, thick myxomatous mitral valve, annulus is 24.0 mm with mild mitral regurgitation, long segment narrowing in abdominal aorta at the level of diaphragm, left ventricular ejection fraction (EF)-40%, which is suggestive of large artery vasculitis-Takayasu arteritis (TA).

FIG.:2



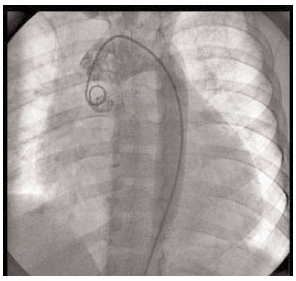

Echocardiography-2 (a) Narrowing of abdominal aorta




Echocardiography-2 (b) Supra sternal view showed arch of aorta

Coronary Angiogram report showed: diffuse narrowing of a long segment of abdominal aorta, severe stenosis of proximal left subclavian artery and right renal artery is stenosed at stoma and for about 1.5cm.

Fig-3 CAG +PCI



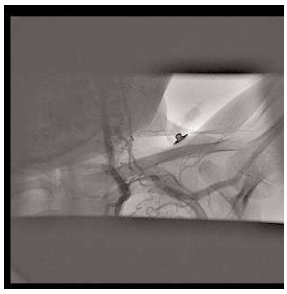
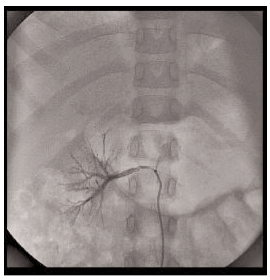


Fig.:3-a)Narrowing of Abdominal Aorta Fig.:3-b)Left Subclavian artery stenosis



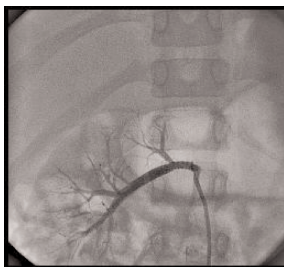


Fig.:3-c)Right renal artery stenosis Fig.:3-d)After PCI to Right renal artery

PTA (Percutanous transluminal angioplasty) to right renal artery and right subclavian artery was done with bare metal stent (4.0X 19 mm) in same setting and prednisolone started orally and then discharged with antihypertensive, antiplatelet drugs and advised to come for follow-up after 3 weeks. During follow-up patient was seen clinically improved, erythrocyte sedimentation rate (ESR) fall to18 mm in 1st hour and her blood pressure was controlled. It was decided to continue steroid for next 4-6 months and observe for any symptoms and plan surgical correction if symptom persist.

## Discussion

Takayasu arteritis (TA) the commonest cause of renovascular hypertension in India[2].The etiology of takayasu arteritis is unknown,but an associatiation with tuberculosis has been reported[3]. Pathogenesis of takayasu arteritis starts in genetically susceptible indivisual, with perhaps a specific humoral milieu, followed by exposure to a yet unidentified antigen, leading to immune response that targets the large vessels[4]. Takayasu arteritis affects patient in 2nd and 3rd decade of life. Females are affected more commonly than males 2.5:1[5]. Absent pulses along with hypertension & vascular bruits could be the commonest mode of presentation.[6] The index case was also a female child in the second decade of life and presented similarly.

Takayasu arteritis is classified according to the site of involvement[6]. Type-I-Aortic arch, Type-II-Descending thoracic aorta,Type-III- Descending thoracic aorta and abdominal aorta, Type-IV-Abdominal aorta only, Type-V-Aortic arch, descending thoracic aorta & abdominal aorta. In a study by Hata et.al. Japanese patients showed a higher frequency of involvement of the ascending aorta, aortic arch & its branches than did the Indian in whom the frequency of involvement of the abdominal aorta &/ or renal artery was higher[7-8]. In the present case the proximal descending aorta & renal arteries were involved.

Therapeutic modalities include steroids, immunosuppressive agents and antihypertensive drug therapy.20%-100% success rate of steroids has been reported in different studies[8]. Cyclophosphamide and methotrexates are often needed to control intense inflammatory response.In addition balloon dilatation or stenting is often necessary[9]. Angioplasty of renal artery results reduce the hypertension.More than 50% cases achieve remission,but 28% of cases never achieve remission.The 5 years mortality is as high as 35%.There is no basis for genetic counseling.The best outcome appears to be associated with early diagnosis & institution of medical & surgical therapy.

The aim of this case reporting was to sensitize all practicing physicians in the field of medicine as well as pediatrics diagnosis of takayasu's arteritis should not get delayed or missed if any child present with hypertension.

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