A 18-year-old  girl was referred to our hospital in November 2016 due to elevated arterial blood pressure, headache and palpitation since 1 year. Upon admission, the girl was in good general condition. The physical examination revealed tall narrow facies body built was below average. Thumb sign and wrist sign were positive with joint hypermobility. She had multiple nonpalpable purplish rash on abdomen and both thigh. Her B.P was 200/110mmHg on upper limb and 140/90mmHg on lower limbs. Her pulse was 84beat/min, regular, and all peripheral pulses were normal. Lower limb pulses were weak and there was no radiofemoral delay. The femoral pulses were present and equal. Her body weight and height were 36 kg (the 10th percentile) and 164 cm (the 25th percentile) respectively, arm span was 172 cm, lower segment 87 cm, high arched palate. She gave history of rash on abdomen and both thigh for 3 months. The neurological examination was unremarkable.

The family history was negative for essential hypertension and cardiovascular diseases.

On examination, there was visible apical impulse, apex beat was on 9cm from midline at 6th i.c.s and heaving, loud 1st heart sound, 2nd heart sound was normal. Abdominal bruit was present. Fundoscopic examination was normal.

Laboratory findings showed an elevated erythrocyte sedimentation rate (ESR) of 52 mm/h (normal value <20 mm/1 h) and serum C-reactive protein level of 1.13 mg/dl (normal value <0.5 mg/dl). The rest of the laboratory investigations, including serum creatinine, electrolytes, and urinalysis were normal. Chest X-Ray shows mild cardiomegaly, ECG normal, USG of whole abdomen showed bilateral polycystic ovaries. Color Doppler echo showed turbulent flow and spectral analysis suggested high peak velocity in the left and right renal artery and also in descending aorta at the origin of renal arteries. The gradient was 45mmHg with narrowing of descending aorta(Fig: 1), there was some collaterals arising from upper part of descending aorta. Aortic root was 28mm(Z Score >2STD) mild AR, mild left ventricular dysfunction with LVEF 48%, Mildly dilated LV, Hypoplastic right pulmonary artery.

CT Aorto pulmonary angiogram showed mild about 50% narrowing of mid abdominal aorta and about 3.5cm subtotal occlusion of both proximal renal artery.
Discussion:
The finding of hypertension and arterial bruits in young adults necessitates the examination of pulses and blood pressures in different limbs in order to detect asymmetry. Elevated ESR is a common finding; however, caution is advised, because up to 50% of patients may have active TA disease and a normal ESR.\textsuperscript{4,5,6}

On the basis of clinical manifestations and CT angiographic abnormalities, the diagnosis of TA and Marfan syndrome was made. At present, TA is diagnosed on the basis of the criteria proposed by European League Against Rheumatism Pediatric Rheumatology: Table-1

**Fig-1:** Color Doppler echo showing gradient in the descending aorta is 45mmHg

**Fig-2 and 3:** CT Aorto pulmonary angiogram showed about 50% narrowing of mid abdominal aorta and about 3.5cm subtotal occlusion of both proximal renal artery with severe ostial disease in Superior Mesenteric artery (SMA), mild infundibular narrowing, moderate narrowing in Right pulmonary artery (RPA), mildly dilated LV and small collateral from upper descending aorta to RPA.

<table>
<thead>
<tr>
<th>Criterion</th>
<th>Definition</th>
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<tbody>
<tr>
<td>Angiographic abnormality (Mandatory criterion)</td>
<td>Angiography (conventional, computed tomography or magnetic resonance imaging) of the aorta or its main branches and pulmonary arteries showing aneurysm/dilatation, narrowing, occlusion or thickened arterial wall not due to fibromuscular dysplasia or similar cause: changes usually focal or segmental.</td>
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<td>Pulse deficit or claudication</td>
<td>Lost/decreased/unequal peripheral artery pulse(s); Claudication: focal muscle pain induced by physical activity</td>
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<td>Blood pressure discrepancy</td>
<td>Discrepancy of four limb systolic blood pressure &gt;10 mmHg difference in any limb</td>
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<tr>
<td>Bruits</td>
<td>Audible murmurs or palpable thrills over large arteries</td>
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<tr>
<td>Hypertension</td>
<td>Systolic/diastolic blood pressure greater than 95\textsuperscript{th} percentile for height</td>
</tr>
<tr>
<td>Acute phase reactants</td>
<td>Erythrocyte sedimentation rate &gt;20 mm per first hour or C-reactive protein any value above normal (according to local laboratory)</td>
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</table>

TA is classified when the mandatory criterion is present plus any other criteria.
Takayasu arteritis (TA), also known as aortoarteritis and pulseless disease, is a rare condition. It is potentially life-threatening. It is a form of granulomatous arteritis, which affects large and medium-sized arteries, primarily the aorta and its large branches as well as proximal portions of pulmonary, coronary, and renal arteries. TA is predominantly a disease of young adults in the second and third decades of life, but it has also been reported in childhood and in adults older than 40 years. The youngest patient described was 6 months old, and the oldest one was 75 years. Females are more likely to be affected than males. However, hypertension is the most common sign in both groups.

The pathogenesis of arterial hypertension due to TA is complex, multifactorial, and not fully understood. At present, it is thought to be the result of three mechanisms: (a) mechanical, in which hypertension proximal to narrowed aorta (atypical coarctation) is due to high resistance to cardiac output imposed by narrowing; (b) neural, in which hypertension proximal to narrowed aorta results from aortic arch baroreceptors readjustment and this allows to ensure adequate blood supply to organs distal to narrowed aorta; and (c) hormonal, in which hypertension is caused by renal hypoperfusion due to stenotic lesions of one or both renal arteries or aorta alone.

Clinical manifestations of TA are nonspecific. The clinical course of the disease is divided into an early active inflammatory phase and late chronic phase. The active phase lasts for weeks to months and may have a remitting and relapsing course. It is characterized by systemic disease with symptoms of fever, general malaise, night sweats, loss of appetite, weight loss, headaches, dizziness, arthralgia, skin rashes, etc. In our patient, there is history of rash for three months.

In our patient, systolic arterial hypertension seemed to result from narrowing of abdominal aorta and bilateral renal artery stenosis.

Cardiac complications related to TA are due more to poorly controlled hypertension from aorto-renal arterial disease than to disease of the aorto-ostia of the coronary arteries. Aortic regurgitation that is secondary to aortic root dilation can occur in up to 20% of patients. Hypertension occurs in one third of patients and is usually caused by renal artery stenosis. Left ventricular dysfunction caused by myocarditis has been reported in up to 18% of cases.

Treatment of TA is based on the use of immunosuppressants such as prednisone and/or methotrexate to decrease or eliminate inflammatory activity. About 60% of patients with TA respond to glucocorticoids. However, as many as 40% relapse on tapering steroids. Alternative therapies such as azathioprine, cyclophosphamide, mycophenolate mofetil, and tacrolimus hydrate are also used in TA, especially for corticosteroid-resistant disease. Hypertension should be treated aggressively often with multidrug regimen, but pediatricians should be warned against ACE inhibitors until renal artery stenosis has been excluded.

In our patient, hypertension was treated using three medications namely: amlodipine (5 mg/day), Bisoprolol Fumerate 2.5 mg/day, Bisoprolol 2.5 mg + hydrochlorothiazide 6.25 mg/day. Initially the hypertension was not well controlled, so the option of stenting of the descending thoracic aorta and renal arteries had also been considered. But due to the fact that nonspecific markers of inflammation were elevated and this girl had never been treated before, she was qualified for continued medical treatment. The patient is currently under a long-term clinical surveillance by a cardiologist, rheumatologist and nephrologist.

Table-II
Clinical features of Takayasu arteritis related to ischemia

<table>
<thead>
<tr>
<th>The vessels involved</th>
<th>Clinical features</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Aortic branches</td>
<td>Malaise, decreased or absent pulse of upper extremities, dysfunction of upper extremities, headaches, dizziness, vision and orientation disturbances, syncope</td>
</tr>
<tr>
<td>2. Aortic arch</td>
<td>Congestive heart failure, aortic valve insufficiency, arterial hypertension</td>
</tr>
<tr>
<td>3. Coronary arteries</td>
<td>Ischemic heart disease, myocardial infarction</td>
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<tr>
<td>4. Pulmonary arteries</td>
<td>Chest pain, dyspnea, coughing, hemoptysis, congestive heart failure</td>
</tr>
<tr>
<td>5. Abdominal aorta or celiac trunk</td>
<td>Ischemia of the stomach and intestines, abdominal pain, nausea, vomiting</td>
</tr>
<tr>
<td>6. Renal arteries</td>
<td>Arterial hypertension, chronic renal failure</td>
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In the presence of symptomatic stenotic or occlusive lesions, endovascular revascularization procedures like bypass grafts, patch angioplasty, endarterectomy, percutaneous transluminal angioplasty, or stent placement should be taken into consideration.15

Conclusion:
Marfan syndrome is an autosomal dominant disorder of connective tissue involving the skeletal, ocular, and cardiovascular systems.5 In our case, the patient had concomitant Takayasu’s arteritis and Marfan syndrome, the former involving the bilateral renal arteries and abdominal aorta and causing obstruction, the latter causing aortic root dilatation with aortic regurgitation. To our knowledge, this is the first case in which a patient with concomitant Takayasu’s arteritis and Marfan syndrome.

References: