

# Atypical coarctation of aorta

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## Abstract

*Atypical coarctation of aorta (CoA) is characterized by localized or extended narrowing of the ascending aorta, of the descending thoracic aorta at the level of the diaphragm, or of the abdominal aorta. Symptoms are attributed to upper body arterial hypertension and may include headache, abdominal angina, leg fatigue at exercise, cold feet and intermittent claudication. A pressure difference between arms and legs may exist as it is the case for the isthmic CoA. A systolic vascular murmur can be heard over the region of stenosis. This disease is often caused by an arteritis (Takayasu arteritis or aortitis) or a fibromuscular dysplasia. Some forms may be congenital. It can be associated with neurofibromatosis such as von Recklinghausen disease or Williams syndrome. This rare condition affects 0.5% to 2% of individuals with CoA. The symptomatic treatment consists of antihypertensive medication. The causal treatment is either percutaneous transluminal angioplasty with or without endovascular stent placement or surgery.*

## Keywords

Coarctation, aortic isthmus, atypical, ectopic, Takayasu arteritis, neurofibromatosis (Recklinghausen disease), Williams syndrome, rubella syndrome, tuberous sclerosis, Alagille syndrome.

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## Disease name and synonyms

- Atypical coarctation of the aorta
- Coarctation of the abdominal aorta
- Middle aortic syndrome
- Mid-aortic dysplastic syndrome

## Definition

In coarctation of aorta (CoA), stenosis is characteristically located at the junction of the distal aortic arch and the descending aorta, immediately beyond the origin of the left subclavian artery near the insertion of the *ligamentum arteriosum*. In rare circumstances, coarctation is located far from the aortic isthmus and is called "atypical coarctation".

Atypical CoA is characterized by a localized or extended narrowing of the ascending aorta, of the descending thoracic aorta at the level of the diaphragm or of the abdominal aorta. Coarctation of the descending thoracic aorta or of the abdominal aorta is also called "middle aortic syndrome" or "mid-aortic dysplastic syndrome". Major arterial branches and visceral arteries (renal, superior mesenteric or hepatic arteries) may be involved.

## Frequency

Atypical CoA is a rare condition, affecting 0.5% to 2% of individuals with CoA.

**Clinical presentation****Atypical CoA in various syndromes**

In Takayasu arteritis, thoracic and abdominal aorta is commonly involved. Granulomatous vasculitis can cause intima proliferation and consecutively aneurysm or stenosis of various degrees or even the occlusion of the aorta and its branches. High incidence of concomitant renal artery stenosis is reported, while celiac or superior mesenteric artery involvement is less often observed.

Williams syndrome is associated with supravalvular aortic stenosis, localized or extended narrowing of the ascending aorta beyond the superior margin of the sinuses of Valsalva. Supravalvular aortic stenosis becomes more apparent and progresses with age. Occasionally, Williams syndrome is associated with coarctation of the abdominal aorta and renal artery stenosis. Arterial hypertension may develop, secondary to progressive renal artery stenosis or to stiffness and thickness of the arterial walls.

In [neurofibromatosis type 1](#) (von Recklinghausen disease), atypical CoA is reported in the descending thoracic and abdominal aorta. The renal arteries are rarely involved. Stenosis may be attributed to the proliferation of Schwann cells within the vessels wall.

In all cases, the aortic narrowing may cause a pressure gradient across the stenosis and a consecutive pre-stenotic arterial hypertension and post-stenotic hypotension. Arterial flow in the visceral arteries distal to stenosis is reduced. Impaired renal perfusion results in renovascular hypertension. Collateral arteries may originate between the pre- and post-stenotic region.

**Main symptoms**

Depending on the etiology and degree of stenosis, atypical CoA may present in childhood but is often diagnosed in adolescents or adults.

Symptoms are attributed to upper body arterial hypertension and may include headache, abdominal angina, leg fatigue at exercise, cold feet, and intermittent claudication. A pressure difference between arms and legs may exist as it is the case for the isthmus CoA. A systolic vascular murmur can be heard over the region of stenosis.

**Etiology**

Atypical CoA is often caused by an arteritis (Takayasu arteritis or aortitis) or fibromuscular dysplasia. Some forms may be congenital. The disease can be associated with neurofibromatosis such as Von Recklinghausen disease and Williams syndrome. Occasionally atypical coarctation or hypoplasia of the

abdominal aorta is associated with rubella syndrome, tuberous sclerosis or Alagille syndrome.

**Diagnostic methods****ECG**

ECG may show left ventricular hypertrophy.

**Imaging**

Aortic narrowing can be shown and also quantified by magnetic resonance imaging, computed tomography and aorto-/arteriography. Echocardiography or abdominal sonography is generally of limited help due to restricted accessibility.

**Treatment**

The treatment of atypical CoA mainly depends on its etiology.

The symptomatic treatment consists of antihypertensive medication. The treatment of underlying arteritis, *i.e.* in Takayasu arteritis, consists of corticosteroids, cyclophosphamide or methotrexate.

The causal treatment is either percutaneous transluminal angioplasty with or without endovascular stent placement or surgery. Surgery comprises reconstruction of the aorta with resection of the stenotic segment, interposition of grafts or aorto-aortic bypasses or enlargement with patches.

The indication for the repair or treatment of possible associated intracardiac (resection of the supravalvular aortic stenosis in Williams syndrome) or vascular defects (revascularisation of the visceral vascular tree) needs to be evaluated.

Systematic follow-up for the detection of late post-operative or post-interventional complications is strongly recommended.

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