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Coarctation of The Aorta

Coarctation of the aorta is a constriction of a portion of the aorta which typically occurs just distal to the left subclavian artery. It classically presents with higher blood pressure in the upper extremities compared to the lower extremities, but may also present with asymptomatic hypertension of CHF. It is associated with Turner Syndrome. Diagnosis is typically made with echocardiography. Chest x-ray shows a classic 3 sign and rib notching. Treatment usually involves surgical repair.

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Pathophysiology

Constriction of a Portion of Aorta

Constrictor and A-orca

Coarctation of the aorta is a narrowing of the aorta. In 98% of patients, the narrowing is classically divided into infantile and adult forms. The infantile form is associated with a patent ductus arteriosus (PDA) and the coarctation occurs distal to the aortic arch, but proximal to the PDA. In the adult form, the coarctation occurs distal to the aortic arch and just across from or distal to the closed ductus arteriosus (called the ligamentum arteriosum).

Turner Syndrome

Turnip

Coarctation of the aorta is twice as common in males as in females. However, females with Turner Syndrome (45, XO) are frequently affected with coarctation of the aorta. Another important association is the increased risk of intracranial aneurysms (berry aneurysms).

Symptoms

Asymptomatic Hypertension

Thumbs-up Hiker-BP

Older infants and children are often asymptomatic and present with hypertension. They may also have murmurs caused by collateral blood flow or associated heart defects as well as chest pain or claudication. Adults typically present with hypertension, claudication and headaches.

BP Higher in Upper Extremities, Right Arm

BP-cuff on Upper Right Arm

The classic physical exam finding is higher systolic blood pressure in the upper extremities when compared to the lower extremities.

CHF

CHF Heart-balloon

In neonates with severe coarctation, heart failure may develop after the PDA closes because there is not enough time for the development of collateral blood flow or myocardial hypertrophy. Although heart failure rarely occurs beyond the neonatal period, adults may develop heart failure if severe hypertension is present.

Diagnosis

Echocardiogram

Echoing Cardiogram

Using an ultrasound, sound waves are used to visualize the structures of the heart and aorta. These can be performed in a both transthoracic and transesophageal fashion with the transesophageal echocardiogram (TEE) being the best choice to visualize the heart structures posteriorly.

Cardiac Catheterization

Heart Catheter-cat

Other tests used to diagnose the defect and measure the function of the heart include cardiac catheterization, MRI and CT.

3 Sign on Chest X-Ray

(3) Tree Chest X-Ray

The figure 3 sign on chest x-ray represents the contour abnormality of the aorta. It is formed by dilation of the aortic arch and left subclavian artery, indentation at the coarctation site and dilation of the descending aorta. It is important to note that dilation of the ascending aorta is common and may be visualized but the characteristic "3" sign is mostly due to dilation of the descending portion.

Rib Notching

Rib Notches

In order to bypass the coarctation and supply the descending aorta, the intercostal collateral vessels begin to dilate. The dilated vessels erode the inferior parts of the ribs, resulting in notching. Since collateral circulation takes time to develop, rib notching is unusual in patients less than 5 years old.

Treatment

Surgical Correction

Surgeon

For patients with recurrent coarctation (restenosis after a successful operation), balloon angioplasty with or without stent placement can be performed.

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