Coarctation of the Aorta

:Objectives

1. Define the COA and its prevelance.
2. Recognise the pathophysiology of COA.
3. Identify the clinical manifestations of COA.
4. Recognise management of COA.

Coarctation of aorta is an obstruction in the descending aorta, located typically near the aortic attachment of the ductus arteriosus .It is basically a discrete localized constriction having a ridge or shelf like structure arising .from ductal end of the aorta

Coarctation of the Aorta

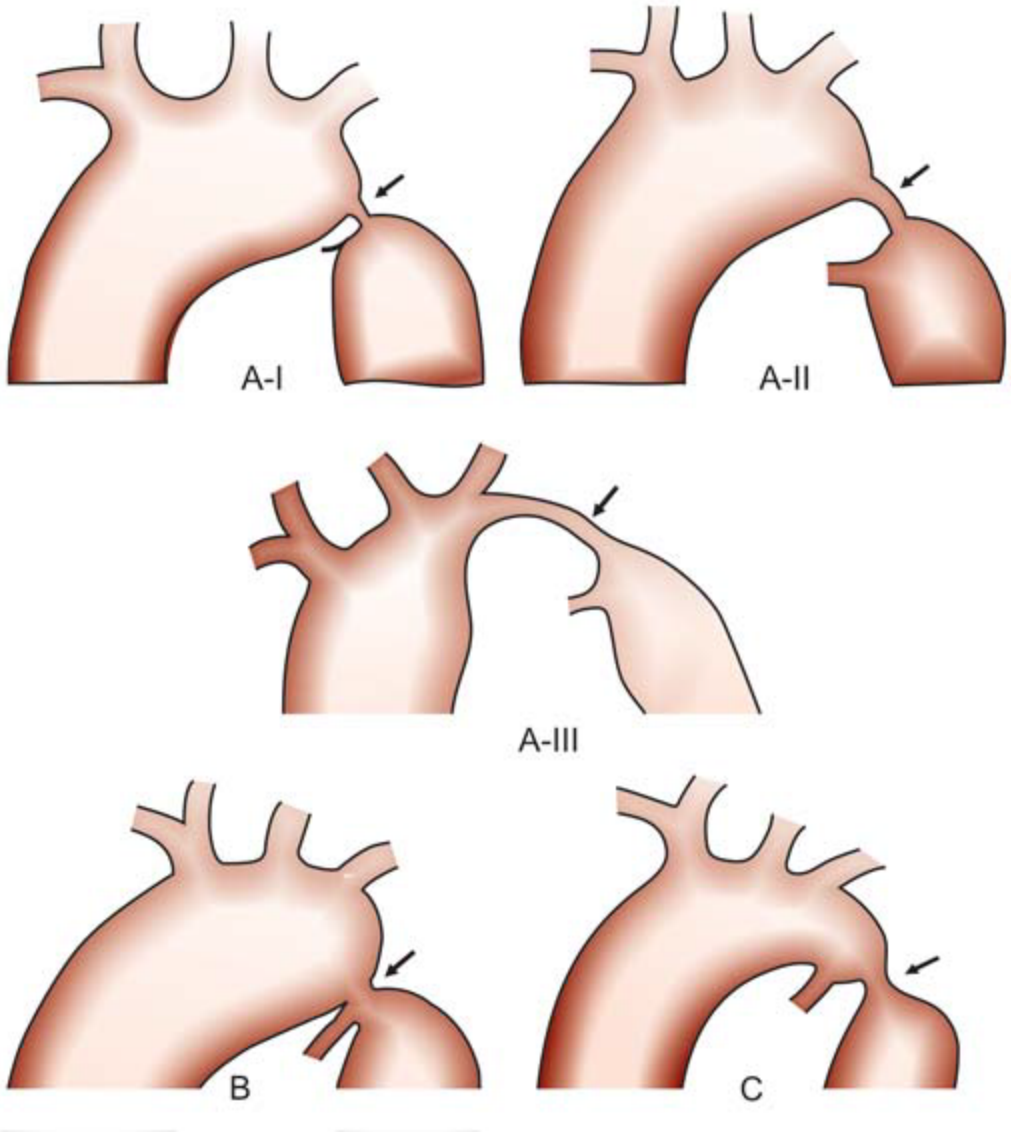
Prevalence

Coarctation of the aorta occurs in 8% to 10% of all cases of CHD. It is more common in males than in females (male-to-female ratio of 2 to 1). Among patients with Turner's .syndrome, 30% have COA

Pathology

The usual location of COA is juxtaductal, just distal. 1 to the left subclavian artery; less often it is proximal .to the origin of the left subclavian artery

The most common associated anomaly is bicuspid. 2 aortic valve, which occurs in more than 50% and up to 85% of all patients with COA. In *symptomatic infants* with COA, other associated cardiac defects such as aortic hypoplasia, abnormal aortic valve, VSD, and mitral valve anomalies are often present. The coarctation is almost always juxtaductal (i.e., located opposite the entry of the ductus arteriosus)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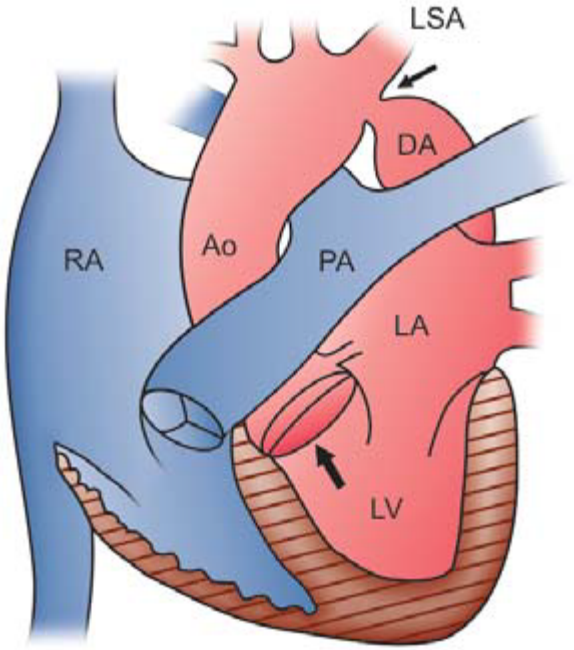
LSA
DA
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Symptomatic Infants
Clinical Manifestations
History

Poor feeding, dyspnea, or signs of acute circulatory shock may develop in the first 6 weeks of life. The newborn discharge examination may have been normal as a result of incomplete obliteration of the aortic end of the ductus, which would permit blood flow to the descending aorta. After ductal obliteration, the aortic lumen narrows with loss of the .space provided by the aortic end of the ductus

Physical Examination

Infants with COA are pale and experience varying degrees of. 1 respiratory distress. Oliguria or anuria, general circulatory shock, .and severe acidemia are common

Peripheral pulses may be weak and thready as a result of CHF.. 2 A blood pressure differential may become apparent only after improvement of cardiac function with administration of rapidly .acting inotropic agents

The S2 is single and loud; a loud S3 gallop is usually present.. 3 No heart murmur is present in 50% of sick infants. A nonspecific ejection systolic murmur is audible over the precordium. The .heart murmur may become louder after treatment

Electrocardiography

A normal or rightward QRS axis and RVH or right bundle branch block (RBBB) are present in most infants with COA rather than LVH; LVH is seen in older children

Radiography

Marked cardiomegaly and pulmonary edema or pulmonary venous congestion are usually present.

Natural History

About 20% to 30% of all patients with COA develop CHF by 3 month of .age

If COA is undetected and untreated in a symptomatic infant, early .death may result from CHF and renal shutdown

Management

Medical

In symptomatic neonates, PGE1 infusion should be started to. 1 promote ductal patency and establish flow to the descending aorta and .the kidneys

Intensive anticongestive measures with short-acting inotropic agents. 2 .(e.g.,dopamine, dobutamine), diuretics, and oxygen should be started

Metabolic disturbances (e.g., acidosis and hypoglycemia) should be recognized and treated promptly.

When the patient is stabilized, either surgical repair or balloon procedure should be performed because the improvement from anticongestive measures is usually temporary. 4

Asymptomatic Infants and Children

Clinical Manifestations

Most children are asymptomatic. Occasionally, a child complains of .weakness or pain (or both) in the legs after exercise

Physical Examination

.Patients grow and develop normally. 1

Arterial pulses in the leg are either absent or weak and delayed. There. 2 is hypertension in the arm or the leg systolic pressure is equal to or lower than the arm systolic pressure. In normal children, the oscillometric systolic pressure in the thigh or calf is 5 to 10 mm Hg higher .than that in the arm

With use of the auscultatory method, the leg systolic pressure may be as .much as 20 mm Hg higher than in the arm in normal children

A systolic thrill may be present in the suprasternal notch. The S2 splits. 3 normally, and the A2 is accentuated. An ejection click is frequently audible at the apex or at the base (or both), which may originate in the .associated bicuspid aortic valve or from systemic hypertension

A grade 2/6 to 3/6 systolic ejection murmur arising from the coarctation itself is usually best heard at the upper left sternal border, at the base, and in the left interscapular area posteriorly. If the coarctation is severe, the systolic murmur may be long and spill into diastole. Continuous murmurs may be prominent throughout the chest anteriorly, laterally, and posteriorly in patients with a well-developed arterial collateral system.

Electrocardiography

Leftward QRS axis and LVH are commonly found. The ECG is .normal in approximately 20% of patients

Radiography

.The heart size may be normal or slightly enlarged. 1

.Dilatation of the ascending aorta may be seen. 2

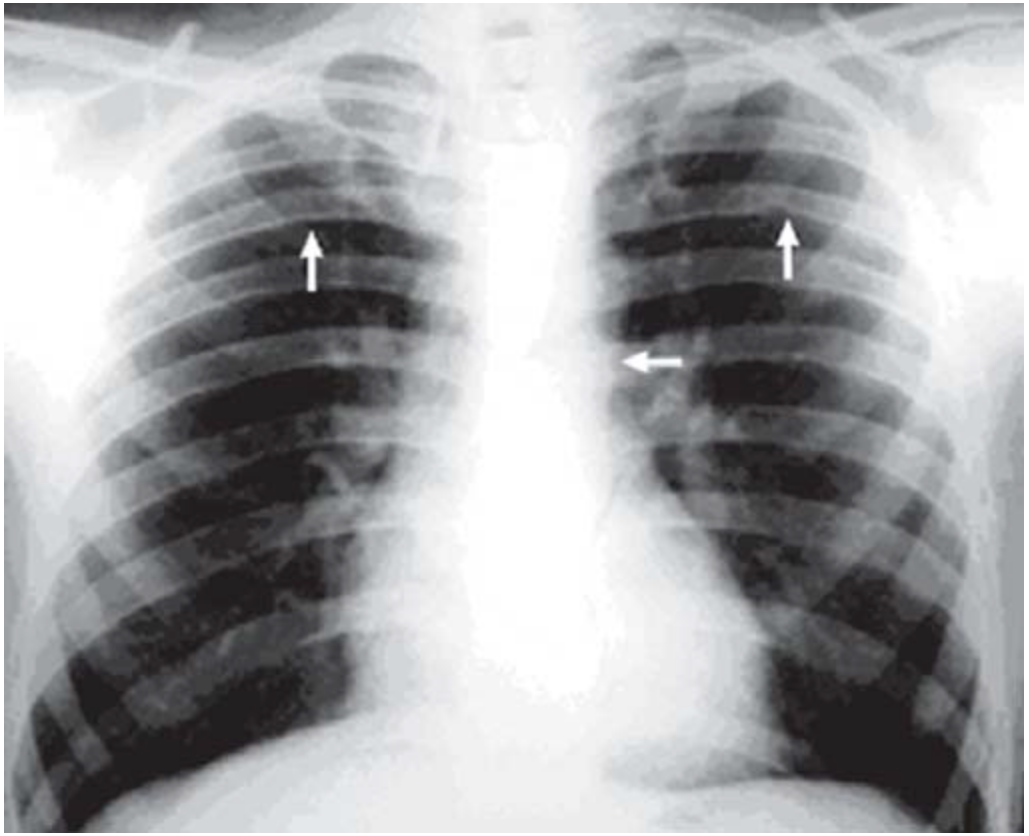
An E-shaped indentation on the barium-filled esophagus or. 3 .a "3 sign" on overpenetrated films suggests COA

Rib notching between the fourth and eighth ribs may be. 4 seen in older children but rarely in children younger than 5 .years of age

Echocardiography

The suprasternal notch 2D echo demonstrates a discrete shelf-. 1 like membrane in the posterolateral aspect of the descending aorta. Associated findings such as isthmus hypoplasia, poststenotic dilatation, and diminished pulsation in the descending aorta may be .present. Bicuspid aortic valve is frequently present

Doppler examination often demonstrates a pattern of diastolic. 2 runoff, especially in patients with robust collaterals or tight stenosis. Continuous wave Doppler flow profile distal to the coarctation is composed of two superimposed signals representing low-velocity flow in the proximal descending aorta and high-velocity flow across .the coarctation



Natural History

LV failure, rupture of the aorta, intracranial hemorrhage (i.e.,. 1 rupture of a berry aneurysm of the arterial circle of Willis), hypertensive encephalopathy, and hypertensive cardiovascular .disease are rare complications seen in adulthood

.A bicuspid aortic valve may cause stenosis or regurgitation with age. 2

Management

Medical

Children with mild COA should be watched regularly for. 1 hypertension in the arm and for increasing pressure differences between the arm and leg. Reduced BP readings in the lower extremities may be caused by femoral artery injuries resulting from .previous surgeries or interventional procedures

Balloon angioplasty for native unoperated coarctation is. 2 controversial, although most centers perform balloon dilatation for .recurrent coarctation

Some centers continue to use the balloon procedure for the native .COA, but other centers prefer a surgical approach

Surgical

Indications and Timing

Significant narrowing of the aorta with pressure gradient greater. 1
than 20 to 30 mm Hg is considered an indication for surgery in
.asymptomatic children

The preferred age for surgery varies from center to center; some. 2
centers prefer ages of 2 and 3 years, and others prefer delaying it
until 4 to 5 years of age. early surgery (i.e., before 1 year of age)
appears to increase the chance of recoarctation .risk of late
recurrence of coarctation is low if the surgery is performed after 2
.years of age

Procedures

Through a left thoracotomy incision, extended resection of the. 1
coarctation segment and end-to-end anastomosis is the procedure of
choice for discrete COA in children

Occasionally, subclavian artery aortoplasty or circular or patch. 2
.grafts may be performed