Coarctation of the Aorta (CoA) in Children — Symptoms and Treatment

Coarctation of the aorta is characterized by the constriction of the aorta usually opposite to the patent ductus arteriosus insertion site. Patients might present with signs and symptoms suggestive of congestive heart failure early in life or with hypertension later in life. Echocardiography and magnetic resonance imaging or computed tomography are the mainstay diagnostic modalities for the disease. Medical therapy for congestive heart failure mainly consists of diuretics and inotropic drugs. Beta-blockers might be used for the treatment of hypertension in patients with coarctation of the aorta but a surgical intervention to relieve the aortic obstruction is usually preferred.

Overview

Coarctation of the aorta (CoA) is characterized by the abnormal constriction of the aorta due to localized medial layer thickening. This constriction might appear as a shelf-like structure or a membranous curtain-like structure. The involved region might be discrete or a long segment. A discrete short segment of CoA is the most common form of the disease.
Epidemiology of Coarctation of the Aorta

CoA is one of the common forms of congenital heart diseases with an estimated incidence of 6 to 8% among patients with congenital heart disease. CoA is more common in European and North America countries compared to Asia.

The condition is more common in boys. The incidence of CoA in symptomatic infants is not different between the two sexes. The most common age of presentation is 5 years.
Young children usually present with congestive heart failure while adults are more likely to present to the clinic because of persistent hypertension.

The condition might be isolated or more commonly associated with other congenital heart defects such as a bicuspid aortic valve, and ventricular septal defects. The condition can cause symptoms of impaired peripheral circulation in young infants and can be associated with significant mortality due to the increased risk of congestive heart failure.

Patients with CoA are at an increased risk of developing subacute bacterial endocarditis, hypertension, and a slightly decreased survival curve compared to healthy individuals. It is estimated that up to 90% of the patients with CoA who do not undergo surgical treatment of the pathology will end up dead by the age of 50 years.

The patient’s age at presentation, the size of the involved segment, and the presence of other preexisting heart defects are the most important predictors of mortality and morbidity in patients with CoA.

**Classification of Coarctation of the Aorta**

The coarctation of aorta can be classified into three main types;

**Preductal coarctation**

This type of aortic narrowing occurs proximal to the patent ductus arteriosus. As the blood flow to the aorta distal to the coarctation is dependent on the ductus arteriosus, hence, severe narrowing of this type can be life-threatening.

The main reason for this type of coarctation is the intracardiac anomaly during the fetal life, which eventually decreases the blood flow through the left heart side, making the hypoplastic development of the aorta. 5% of the infants with Turner’s syndrome also suffer from this type of coarctation.

**Ductal coarctation**

In this type of coarctation, the aorta narrows at the insertion of the ductus arteriosus. This type of the coarctation basically arises during the closing of ductus arteriosus.

**Postductal coarctation**

In this type of coarctation, the aorta narrows distally to the point of insertion of the ductus arteriosus. In this type of coarctation, although ductus arteriosus is open, the blood flow to the lower parts of the body is impaired. This type is more prevalent in the adults.

The other associated conditions with this type of coarctation are; hypertension in the upper limb, notching of the ribs (due to collateral circulation), and weak pulses in the lower limb.

The main reason for this coarctation is the extension of a muscular artery, during the fetal life, into an elastic artery, which leads to the fibrosis and coarctation of the ductus arteriosus upon the birth, narrowing the aortic lumen. Aortic narrowing can present in the forms of either aortic coarctation or the aortic stenosis.
The exact etiology of CoA is unknown in most cases but certain risk factors have been linked to the condition. Turner syndrome and Takayasu arteritis are two possible diseases that have been associated with an increased risk of CoA.

Patients with CoA have a significantly increased afterload pressure on the left ventricle. This is associated with increased stress on the left ventricular wall, ventricular hypertrophy and eventually can put the patient at an increased risk of developing congestive heart failure.

Patients with CoA are at an increased risk of developing persistent hypertension. The exact mechanism of hypertension in this group of patients is poorly understood. It is, however, noted that patients with CoA usually have an exaggerated activation of the renin-angiotensin system which might explain why they have persistent hypertension.

Patients with CoA might eventually develop a slight improvement in their symptoms due to the formation of arterial collaterals that pass the obstructed region of the aorta.

Clinical Presentation of Coarctation of the Aorta

Patients with other congenital heart defects are more likely to present earlier to the clinic. Symptoms of the early presentation of CoA include difficulty during feeding, tachypnea and the development of congestive heart failure. Patients who have severe CoA might present with acute congestive heart failure especially at the time of ductus arteriosus closure.

Patients presenting with CoA later in life usually are diagnosed incidentally while they are being investigated for persistent hypertension. Patients at this stage can complain of headaches, chest pain, and fatigue. Due to the association between CoA and intracerebral aneurysms, the presentation of CoA in some patients can be that of life-threatening intracranial hemorrhage.

Patients who present late to the clinic might have a pressure difference between the arms and legs of more than 20 mm Hg, a sign that is highly suggestive of CoA.

Symptoms

In all mild cases, the symptoms may be absent. However, when present, the symptoms include:
The heart is unable to pump the blood completely to the systemic circulation and pulmonary circulation is also disturbed resulting in difficult breathing and shortness of breath.

When the blood supply to the brain is reduced, the patient can experience episodes of fainting or near fainting.

Heart is unable to properly pump the blood which results in the reduced oxygenation of body tissue. The reduced oxygenation is the main reason for the fatigue, tiredness, and weakness experienced by the patient.

Infants have trouble breathing and have difficulty in feeding

During the narrowing of aorta in COA, the normal blood flow is disturbed

The heart is pumping against the high pressure that causes it to pump poorly. Infants appear pale and may become very fussy or very weak. The symptoms can quickly progress to shock and require admission to an intensive care unit.

There are basically 2 signs of the coarctation of aorta which should be checked upon physical examination:

1. **High blood pressure (BP) in the arms while low blood pressure in the legs simultaneously.** The blood pressure is higher in the blood vessels present before the obstruction which in this case are in the arms and is lower in the area beyond the point of obstruction. This difference in the blood pressure between the arms and the legs is referred as a gradient.
2. **The quality of the pulses in the arms and the legs will also be different, with bounding pulses in the upper extremities and weak or totally absent pulses in the lower extremities.**

If both of these signs are identified by the clinician, then these patients are referred to a cardiologist for their further evaluation.

**Diagnostic Workup for Coarctation of the Aorta**
Patients with CoA might present with cardiogenic shock. In that case, care must be taken to exclude sepsis as the cause of shock. Blood, urine and cerebrospinal fluid cultures are indicated in patients presenting with shock.

Patients might have elevated levels of creatinine and blood urea nitrogen due to impaired kidney perfusion especially if they are started on angiotensin converting enzyme inhibitors.

An electrocardiogram might reveal left ventricular hypertrophy. Patients with early onset of CoA semiology usually have signs suggestive of congestive heart failure on their chest.
x-rays such as pulmonary edema, cardiomegaly, and increased pulmonary vascular markings. Patients who present late might have cardiomegaly and rib notching due to collateral vessels formation.

Echocardiography is very helpful in the evaluation of congenital heart diseases. It can help in the exclusion of other preexisting cardiac abnormalities. Doppler echocardiography is useful in the measurement of the gradient pressure at the site of the coarctation to determine the degree of obstruction.

Other imaging modalities such as magnetic resonance imaging and computed tomography scanning of the thoracic and abdominal aorta are very helpful in the identification of the aortic pathology and the determination of the size and site of the coarctation.

In few patients, cardiac catheterization might be needed to determine the degree of the aortic obstruction, the exact anatomical location of the obstruction, and the size of the obstruction. Recent imaging modalities such as echocardiography and magnetic resonance imaging have made cardiac catheterization a second-line diagnostic modality for the evaluation of CoA.

After surgical correction, a biopsy might be examined for histologic characterization of the lesion. When the histological examination is performed, one might see a thickened medial layer of the aortic wall that is usually opposite to the insertion of the patent ductus arteriosus.

<table>
<thead>
<tr>
<th>Chest x-ray</th>
<th>With imaging the resorption of the lower part of the ribs can be seen. The classic “figure 3 sign” seen on X-ray results from the post-stenotic dilation of the aorta.</th>
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<tbody>
<tr>
<td>Echocardiogram</td>
<td><strong>It is the definitive test for coarctation.</strong> It can provide the images of the heart and aorta and helps in the identification of the exact area of the obstruction.</td>
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<tr>
<td>MRI</td>
<td>An MRI (magnetic resonance imaging) is an imaging study which is done inside a magnet that allows very finely detailed images of the heart, collateral blood vessels, and the aorta.</td>
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**Differential Diagnosis of Coarctation of the Aorta**

The successful diagnosis of the coarctation of aorta depends on the history of the patient, presentation of the symptoms by the patients and vigilant detection of signs by the clinician. The further confirmation can be done with the help of lab and imaging investigations. For the accurate diagnosis of patients with coarctation of the aorta, the main symptoms and signs should be considered, which have been mentioned above. The other clinical conditions which can confuse the diagnosis of coarctation of aorta due to similar presentation have been named below:

<table>
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<th>Condition</th>
<th>Symptoms/Signs</th>
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| Endocardial fibroelastosis | History: Excessive sweating, breathlessness, wheezing, failure to thrive.  
Physical Examination: Tachypnea during feeding and grunting respiration with intercostal retractions. Expiratory wheezes or rales in lung bases. |
| Dilated cardiomyopathy | Symptoms: Dyspnea on exertion, shortness of breath, Orthopnea, edema, fatigue  
Signs on physical ex: Tachypnea, Tachycardia and hypertension/hypotension. |
| Pediatric Valvar Aortic stenosis | Symptoms vary based on the age at the presentation and the severity of the obstruction.  
Signs on Physical ex: Neonates: reduced/absent pulses, tachycardia, Tachypnea  
Childhood: systolic ejection murmur present at the left middle and right upper sternal border, 4th heart sound present |
### Treatment of Coarctation of the Aorta

The choice of treatment depends on a number of factors which include:

- The age of the patient
- The risks and benefits of the procedures
- The anatomy of the aortic obstruction

#### Surgical Treatment

Surgical repair is the most time-tested and traditional treatment for the most patients of coarctation of the aorta, particularly for the *infants* and the *younger children*. The procedure of surgical treatment is done through an incision on the left side of the chest cavity and the heart-lung machine is not required for it.

The hospital stay is usually from 4 to 7 days following the surgery with the first few days stay in an intensive care unit to monitor the vitals vigilantly.

High blood pressure is commonly observed in these patients after surgery and it is managed with intravenous (I/V into the bloodstream) medicines given in the hospital, and then medications taken by mouth (oral) that usually continue after their departure from the hospital.

The risks of this surgical procedure are very low, and the observed outcomes are excellent.

#### Catheterization Treatment

The cardiac catheterization is basically a test done by using the *catheters* (small tubes) placed into the peripheral (in any arm or leg) blood vessels and then maneuvered into the large blood vessels and the heart. In the recent past 25 years, a number of such procedures using the specialized catheters have been developed for the repairing the heart.

The coarctation of the aorta can be opened up from the inside using the balloon dilation method or by placing a stent inside. The catheterization procedure usually takes several hours and is performed under anesthesia.

The patients are strictly observed overnight in the hospital and can otherwise resume their normal activities within a few days of the procedure. In case stents are placed, *aspirin* is recommended for a number of months to prevent the formation of blood clots.

Other precautions to prevent the risk of infection are also needed. *Antibiotics* are also given before general dental work and some other usual medical procedures (this is termed as the bacterial endocarditis prophylaxis).

Following the successful treatment, the patients with the coarctation of the aorta are able to lead a normal lifestyle and most likely have a near-normal lifespan. However, limited
exercise restrictions can be indicated for the patients with continued aortic obstruction, high blood pressure, or the dilation of the aorta. These patients need lifelong regular cardiology evaluations with the imaging of their heart so that they can be monitored for later problems.

The recoarctation is more prevalent when the repair is done during the infancy. Balloon angioplasty is the usual treatment option for the recoarctation, although repeated surgery can be indicated for some patients.

Complications of Coarctations of the Aorta

High blood pressure

It is usually common in these patients with coarctation as they get older, even after the very successful repair through either surgery or the catheterization, and they need antihypertensive medications for a long time. This problem is more likely when the repair is done in the older children or the adults.

High blood pressure can lead to the thickening of the heart muscle and can bring about changes in the blood vessels.

The elevated blood pressure increases the risk of stroke, coronary artery disease, kidney damage and other health problems. It is very important to maintain the normal blood pressure for future health.

The repaired area of the aorta (either by surgery or catheterization) may become narrowed again (which is called recoarctation) with the return of the same blood pressure difference between the arms and legs (major sign for diagnosis of the coarctation of the aorta) on physical examination and further evidence of aortic obstruction by the echo-cardiogram or MRI.

Aneurysms (the weakness of the walls of the aorta) can also occur at the site of the repair after both the surgery and the catheterization, although the significance of this is still unclear to the clinicians and surgeons.

The bicuspid aortic valve, which is often associated with the coarctation of the aorta, may develop some sort of narrowing (termed as stenosis) or leaking (termed as regurgitation) over the time and need further treatment.

References


Adult Congenital Heart Association: http://www.achaheart.org

American Heart Association Website: http://www.heart.org

Boston Children’s Hospital Website: http://childrenshospital.org

medscape.com
