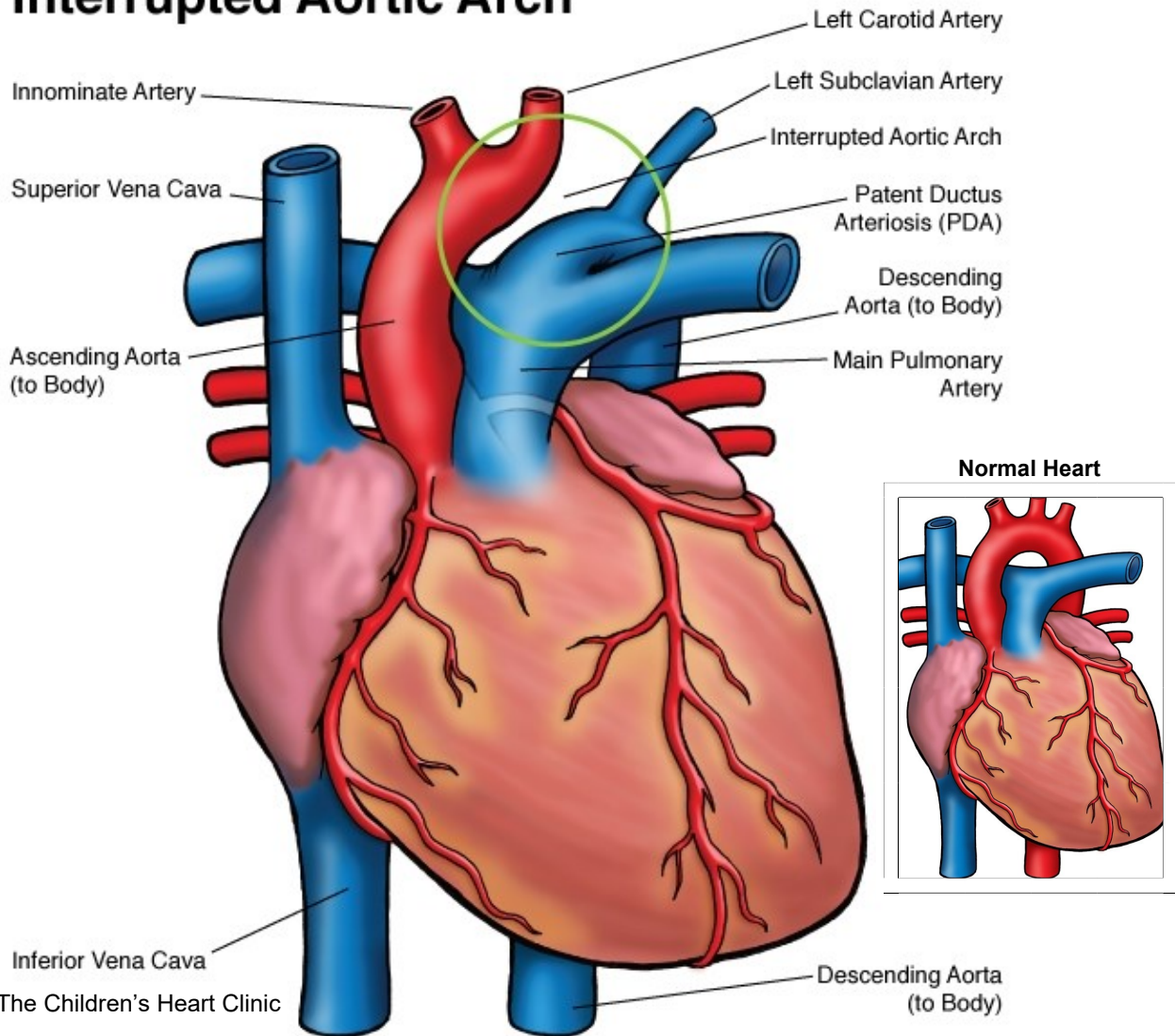


Interrupted Aortic Arch



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Notes:

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Interrupted Aortic Arch (IAA)

Interrupted aortic arch (IAA) is an extreme type of coarctation of the aorta in which a portion of the aorta arch is atretic (closed) or a segment is absent. IAA occurs with patent ductus arteriosus (PDA) and ventricular septal defects (VSD) more than 90% of the time. Bicuspid aortic valve, mitral valve deformities, truncus arteriosus, or subaortic stenosis may also be associated with IAA. DiGeorge syndrome occurs in more than 15% of patients. IAA is rare, accounting for 1% of all congenital heart defects.

Types:

- Type A: Occurs in 30% of cases of IAA. The interruption is distal to the left subclavian artery.
- Type B: Occurs in 43% of cases of IAA. The interruption is located between the left subclavian and left carotid arteries. Often, the right subclavian artery is aberrant, meaning it arises abnormally from the descending aorta. 50% of children with Type B IAA have DiGeorge syndrome.
- Type C: Occurs in 17% of children with IAA. The interruption is located between the innominate and left carotid arteries.

Physical Exam/Symptoms:

- Within the first days of life, infants develop respiratory distress, poor pulses and perfusion, cyanosis (blue color). In rare cases, the ductus arteriosus stay open longer, leading to later presentation.
- Cardiogenic shock.

Diagnostics:

- Chest X-ray: Cardiomegaly (enlarged heart), increased pulmonary vascular markings and pulmonary congestion/edema are present.
- EKG: May show right ventricular hypertrophy (enlargement) in some cases.
- Echocardiogram: Diagnostic.

Medical Management/Treatment:

- Prostaglandin E (PGE) therapy should be started as soon as possible after birth or diagnosis to keep the ductus arteriosus patent.
- Genetic screening for DiGeorge syndrome is done shortly after birth.
- Intubation and inotropic support as needed preoperatively.
- Infants will need surgical repair shortly after birth/diagnosis (see Repair of Interrupted Aortic Arch).
- Bacterial endocarditis prophylaxis prior to any dental procedure ONLY for children with valve replacements, prosthetic arch repairs, or unrepaired cyanotic heart disease.
- Life-long cardiology follow up is needed.

Long-Term Outcomes:

- Hypertension may persist postoperatively, requiring medications for management.
- In general, growth and development are normal in the absence of other congenital heart disease or co-morbidities such as DiGeorge Syndrome.