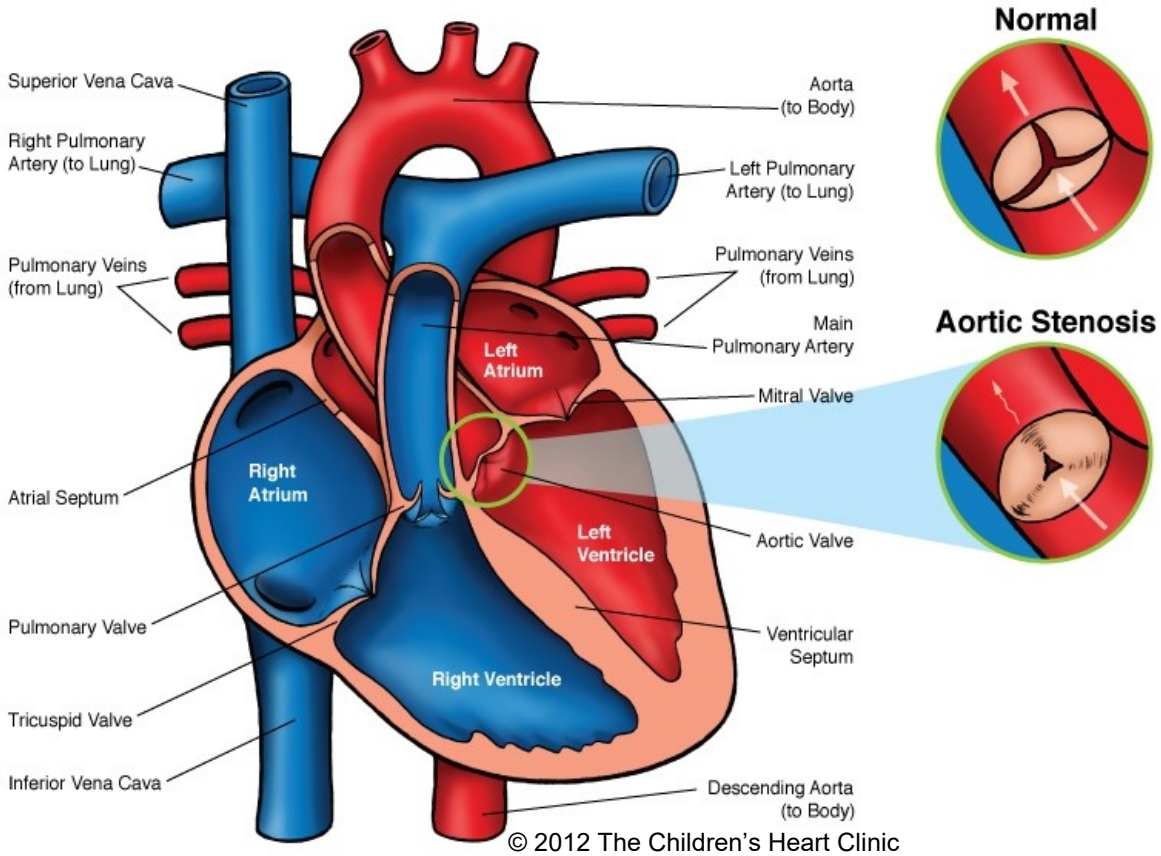
Aortic Stenosis - Valvar



Notes:

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Aortic Stenosis: Valvar

Aortic stenosis (AS) is severe narrowing or obstruction of the left ventricular outflow tract (LVOT) that occurs above, below, or at the level of the aortic valve. Valvar AS refers to narrowing at the valve and is the most common type (71%). A normal aortic valve has three leaflets or cusps (tricuspid). In 75% of children with valvar AS, the aortic valve has only two cusps (bicuspid) as a result a fused commissure (place where 2 leaflets are joined). This type is often not obstructive in childhood, but over time the valve becomes calcified and may lead to obstruction in adulthood. Unicuspid aortic valves and Dome aortic valves occur less commonly. These types of AS may result in *critical or severe AS*, requiring immediate intervention.

Physical Exam/Symptoms:

- In critical AS, neonates develop poor perfusion, pulmonary edema (fluid retention in the lungs) within days or weeks after birth as the ductus arteriosus closes. Clinical picture may resemble that of sepsis (severe infection).
- Most children are asymptomatic with mild to moderate AS.
- Fatigue, chest pain with exertion, or syncope (fainting) may occur in severe AS.
- Murmur: Harsh, grade II/VI systolic murmur heard best at the second left intercostal space, with transmission to the head and neck. An ejection click may be present. An early diastolic murmur of aortic regurgitation is sometimes present in patients with a bicuspid aortic valve.

Diagnostics:

- Chest X-ray: Normal heart size and pulmonary vasculature. The ascending aorta may appear dilated.
- EKG: Normal in mild cases. Left ventricular hypertrophy (enlargement) may be present in severe AS.
- Echocardiogram: Diagnostic.

Medical Management/Treatment:

- Annual echo and cardiology visits in asymptomatic children with mild to moderate stenosis, more frequent in severe AS.
- Prostaglandin E (PGE) infusion to keep ductus arteriosus patent prior to catheterization or surgical intervention in symptomatic neonates.
- Catheterization Lab Procedure: Balloon dilation in the cardiac catheterization lab is often the first step in the management of neonates with critical AS or symptomatic infants and children.
- Surgery: Indicated if balloon valvuloplasty results in severe aortic regurgitation (AR) or if the pressure gradient remains high (see Aortic Valve Replacement and/or Ross Procedure).
- Lifetime anticoagulation (Coumadin and Aspirin) is needed for people with mechanical aortic valve replacements to avoid clotting of the valve.
- Life-long cardiology follow up is needed.

Long-Term Outcomes:

- Stenosis may become severe over time, requiring long-term cardiology follow up and potential surgical intervention as an adult. 10-30% develop aortic regurgitation (leakage of the aortic valve) after balloon or surgical valvuloplasty.
- Subacute bacterial endocarditis occurs in 4% of patients with valvar AS.
- Mortality for children with valvar AS is 1-2%. Sick neonates with critical AS have a mortality rate approaching 10%.
- Developmental outcomes vary depending on the severity of valvar disease and other co-morbidities.