Complete AV canal defect

:Objectives

1. Define AV canal defect (atrioventricular defect) and pathology
2. Describe clinical manifestations and physical signs of AV canal defect.
3. Recognise investigations for patients with AV canal defect
4. Describe the management of AV canal defect.

Complete Endocardial Cushion Defect

Prevalence

Complete ECD (also known as AV canal defect, complete AV canal defect, or AV communis) occurs in 2% of all CHDs. Of children with Down syndrome, about 40% have .CHDs, and 50% of the defects are ECD

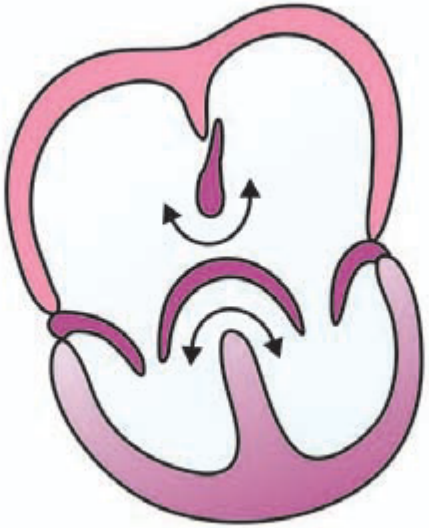
Pathology

Abnormalities seen in complete ECD affect the structures normally derived from the endocardial cushion tissue. Ostium primum ASD, VSD in the inlet ventricular septum, and clefts in the anterior mitral valve and the septal leaflet of the tricuspid valve (forming the common AV valve) are all present in the complete form of ECD .The combination of these defects may result in interatrial and interventricular shunts, LV-to-RA shunt, and AV valve regurgitation. Although rare, the entire atrial septum may be absent (common atrium). When two AV valve orifices are present without an interventricular shunt, the defect is called *partial ECD* or *ostium primum*

Interatrial shunt

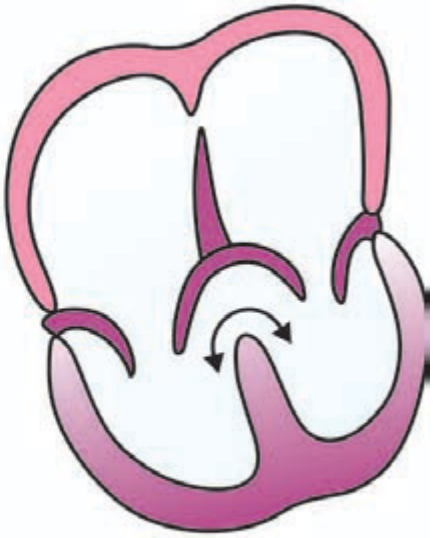
Bridging leaflets attached to ventricular septum

Interatrial and interventricular shunt



Floating bridging leaflets

Interventricular shunt



Bridging leaflets attached to atrial septum

Whereas in complete ECD, a single valve orifice connects the atrial and ventricular chambers, in the partial form, there are separate mitral and tricuspid orifices. The common AV valve .usually has five leaflets

Clinical Manifestation

History

Failure to thrive, repeated respiratory infections, and signs of CHF are .common

Physical Examination

Infants with ECD are usually undernourished and have tachycardia. 1 and tachypnea (signs of CHF). This defect is common in infants with .Down syndrome

Hyperactive precordium with a systolic thrill at the lower left sternal. 2 .border is common

The S1 is accentuated. The S2 narrowly splits, and the P2 increases in. 3 intensity. A grade 3 to 4 of 6 pansystolic murmur is usually audible along the lower left sternal border. The systolic murmur may transmit well to the left axilla and be heard well at the apex when mitral .regurgitation (MR) is significant

A mid-diastolic rumble may be present at the lower left sternal border or at the apex as a result of relative .stenosis of the tricuspid or mitral valve

Signs of CHF (e.g., hepatomegaly, gallop rhythm) may. 4 .be present

Electrocardiography

Superior" QRS axis with the QRS axis between −40. "1 and −150 degrees is characteristic of the defect

Most of the patients have a prolonged PR interval. 2 .(first-degree AV block)

RVH or RBBB is present in all cases, and many. 3 .patients have LVH, too

Radiography

Cardiomegaly is always present and involves all four .cardiac chambers

Pulmonary vascular markings are increased and the main PA segment is prominent

Natural History

Patients with complete ECD develop heart failure 1. 1 to 2 months after birth, and recurrent pneumonia is .common

Without surgical intervention, most patients die by. 2 .the age of 2 to 3 years

In the latter half of the first year of life, survivors. 3 begin to develop pulmonary vascular obstructive disease. These survivors usually die in late childhood .or as young adults

Management

Medical

In small infants with CHF, anticongestive management. 1 with diuretics and ACE inhibitors should be started. Digoxin may also be used

.Nutrition should be optimized.2

Surgical

Indications

The presence of complete ECD indicates the need for surgery because an important hemodynamic derangement is usually present. Most of these infants have CHF that is unresponsive to .medical therapy, and some have elevated PVR

Timing

Although timing varies among institutions and with the hemodynamics of the defect, most centers perform the repair at 2 to 4 months of age. Early surgical repair is especially important for infants with Down syndrome with complete ECD because of their known tendency to develop early pulmonary vascular obstructive .disease