



**Description:** Incomplete fusion of the endocardial cushions.

Consists of a low ASD that is continuous with a high VSD and clefts of the mitral and tricuspid valves, which create a large central AV valve that allows blood to flow between all four chambers of the heart. The directions and pathways of flow are determined by pulmonary and systemic resistance, left and right ventricular pressures, and the compliance of each chamber, although flow is generally from left to right. It is the most common cardiac defect in children with Down syndrome.

**Pathophysiology:** The alterations in hemodynamics depend on the severity of the defect and the child's pulmonary vascular resistance. Immediately after birth, while the newborn's pulmonary vascular resistance is high, there is minimum shunting of blood through the defect. When this resistance falls, left-to-right shunting occurs, and pulmonary blood flow increases. The resultant pulmonary vascular engorgement predisposes the child to development of HF.

**Clinical manifestations:** Patients usually have moderate to severe HF. There is a loud systolic murmur. There may be mild cyanosis that increases with crying. Patients are at high risk for developing pulmonary vascular obstructive disease.

**Surgical treatment:**