

# **McCance: Pathophysiology, 6th Edition**

## **Chapter 31: Alterations of Cardiovascular Function in Children**

### **Key Points – Print**

#### **SUMMARY REVIEW**

##### Development of the Cardiovascular System

1. The heart arises from the mesenchyme and begins as an enlarged blood vessel with a large lumen and a muscular wall. By approximately the eighth week of gestation, all structures of the fetal heart and vascular system are present.
2. The endocardial cushions are instrumental in closing the atrial septum, dividing the AV canals into the right and left AV orifices, and closing the septum.
3. In the fetus the pulmonary and systemic circulatory systems are connected by the foramen ovale, an opening between the atria; by the ductus arteriosus, a fetal vessel that joins the PA to the aorta; and by the ductus venosus, a fetal vessel that connects the inferior vena cava to the umbilical vein.
4. Fetal circulation is different from postnatal circulation because of the presence of fetal shunts and altered metabolic needs of the various organs.
5. Fetal blood flow depends on resistance for its distribution through the body. Resistance in the pulmonary circulation is higher than resistance in the systemic circulation, so myocardial thickness is about the same in the right heart and the left heart.
6. After birth, systemic resistance increases and pulmonary resistance decreases.
7. Pulmonary vascular resistance drops suddenly at birth because the lungs expand and the pulmonary vessels dilate. It continues to decrease gradually during the first 6 to 8 weeks after birth. Decreased resistance causes the right myocardium to thin out.
8. Systemic vascular resistance increases markedly at birth because severance of the umbilical cord removes the low-resistance placenta from the systemic circulation. Increased systemic resistance causes the left myocardium to become dominant and thicken over time.
9. Changes in resistance cause the fetal connections between the pulmonary and systemic circulatory systems to disappear. The foramen ovale closes functionally at birth and anatomically several months later; the ductus arteriosus closes functionally 15 to 18 hours after birth and anatomically within 10 to 21 days; and the ductus venosus closes within 1 week after birth.
10. At birth a series of circulatory changes occur that affect blood flow, vascular resistance, and oxygen tension. The most important change is the shift of gas exchange from the placenta to the lungs.
11. After birth, significant postnatal changes occur, including thinning of the right ventricular myocardium as the pulmonary vascular resistance drops. As the systemic vascular resistance

increases, the left ventricular myocardium becomes thicker and more dominant as it is in the adult heart.

### Congenital Heart Defects

1. Most congenital cardiovascular defects have begun to develop by the eighth week of gestation, and most have many causes, both environmental and genetic.
2. Environmental risk factors associated with the incidence of congenital heart defects typically are maternal conditions. Among these are viral infections, diabetes, drug intake, alcohol intake, metabolic disorders, and advanced maternal age.
3. Genetic factors associated with congenital heart defects include but are not limited to Down syndrome, trisomy 13, trisomy 18, cri du chat syndrome, and Turner syndrome. It now appears, however, that most genetic mechanisms of causation are multifactorial.
4. Classification of congenital heart defects is based on whether they (a) cause blood flow to the lungs to increase or decrease, (b) obstruct ventricular blood flow patterns, or (c) cause mixing of unoxygenated and oxygenated blood.
5. Symptoms of HF are usually the result of congenital heart defects that increase blood volume and pressure in the pulmonary circulation. Clinical manifestations are almost the same as the manifestations of CHF in adults. A unique manifestation in children is FTT.
6. Cyanosis, a bluish discoloration of the skin, indicates that the tissues are not receiving normally adequate oxygenated blood. Cyanosis can be caused by defects that (a) reduce pulmonary blood flow; (b) overload the pulmonary circulation, causing pulmonary hypertension, pulmonary edema, and respiratory difficulty; and (c) cause large amounts of unoxygenated blood to shunt from the pulmonary to the systemic circulation.
7. Congenital defects that maintain or create direct communication between the pulmonary and systemic circulatory systems cause blood to shunt from one system to another, mixing oxygenated and unoxygenated blood and increasing blood volume and pressure on the receiving side of the shunt.
8. The direction of shunting through an abnormal communication depends on differences in pressure and resistance between the two systems. Flow is always from an area of high pressure to an area of low pressure. The resistance to flow determines the volume of the shunting.
9. Acyanotic congenital defects that increase pulmonary blood flow consist of abnormal openings (PDA, ASD, VSD, AVC defect, or truncus arteriosus) that permit blood to shunt from left (systemic circulation) to right (pulmonary circulation). Cyanosis does not occur because the left-to-right shunt does not interfere with the flow of oxygenated blood through the systemic circulation.
10. If the abnormal communication between the left and right circuits is large, volume and pressure overload in the pulmonary circulation leads to CHF.
11. In truncus arteriosus the main trunk fails to divide longitudinally into the aorta and PA. All blood from both ventricles enters the truncus, so that mixed blood is delivered by both circulatory systems, causing varying degrees of cyanosis and HF.

12. In heart defects that decrease pulmonary blood flow (TOF, tricuspid atresia), myocardial hypertrophy cannot compensate for restricted right ventricular outflow. Flow to the lungs decreases, and cyanosis is caused by mixing of systemic and pulmonary venous return.
13. Obstruction of ventricular outflow commonly is caused by PS, AS, COA, interrupted aortic arch, or hypoplastic left heart syndrome.
14. Despite obstruction, ventricular outflow remains normal because of compensatory ventricular hypertrophy stimulated by increased afterload and, in postductal COA, development of collateral circulation around the coarctation.
15. Signs of HF can occur with pulmonary overcirculation or myocardial failure.
16. Complex congenital defects that depend on mixing of the pulmonary and systemic circulations for survival during the postnatal period include complete transposition of the great arteries and total anomalous pulmonary venous connection. This mixing results in desaturated systemic blood flow and cyanosis.
17. In complete transposition of the great vessels, the circulatory systems are not connected serially or through a shunt, so that oxygenated blood remains permanently in the pulmonary circulation and unoxygenated blood remains in the systemic circulation. Survival depends on patency of the ductus arteriosus; after that, surgical intervention is mandatory.
18. Total anomalous pulmonary venous connection is caused by the persistence of the fetal common PA and the lack of pulmonary venous return to the LA. All blood from the pulmonary and systemic circulations enters the RA. Mixed blood enters the LA through an ASD; it then flows into the systemic circulation and causes cyanosis.
19. Treatment for all hemodynamically severe congenital defects is surgical or interventional correction of the anomaly and management of cyanosis and HF.

#### Acquired Cardiovascular Disorders

1. The most common acquired cardiovascular disorders of childhood are Kawasaki disease, rheumatic heart disease, and hypertension.
2. Kawasaki disease is an acute systemic vasculitis that also may result in the development of coronary artery aneurysms and thrombosis.
3. Essential hypertension in children is the same as that in adults, except that it is more likely to be diagnosed in an early, asymptomatic stage. Most cases of hypertension in young children have an underlying cause.
4. Obesity in childhood is an epidemic in the United States and other countries.
5. Obese children are at risk for acquiring numerous other serious and potentially life-threatening illnesses, such as asthma, sleep apnea, hypertension, type 2 diabetes mellitus, and cardiovascular disease.