

# Neonatal Cardiac Assessment and Congenital Heart Disease

compiled by Violet Stephens based on the works of [Patricia Hartley](#)

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## Objectives

1. Identify methods of cardiac assessment
2. Identify hereditary diseases in which congenital heart disease is a frequent finding
3. Discuss cardiac development and fetal circulation
4. Describe cyanotic cardiac lesions
5. Describe the methods of treatment

### Introduction

Each year approximately one percent of all babies born in the United States are diagnosed with congenital heart disease. As many as one-third of these babies will be critically ill and require care by cardiologists in the first days to weeks of life. Depending on the type of heart problem, initial signs and symptoms may include tachypnea, cyanosis and/or a heart murmur. With severe forms of CHD there may be marked cyanosis, respiratory distress and rapid progression to advanced states of shock. Prompt effective care of neonates with CHD can reduce secondary organ damage, improve short and long-term outcomes and reduce mortality.

Cardiovascular assessment of the newborn requires great skill with the techniques of inspection, palpation, and auscultation. Inspection of the general activity of the neonate, breathing patterns, presence or absence of cyanosis, and activity of the precordium are all important. Palpation of pulses, peripheral perfusion, and thrills is also imperative. Auscultation, however, is the main focus of the exam. This is when the examiner assesses heart rate, rhythm, regularity, and heart sounds (especially murmurs). The dynamic properties of the newborn heart make this assessment more difficult than the cardiac assessment of an adult.

The cardiovascular exam constantly changes over the first few hours, days and weeks of life as the neonate changes from fetal circulation with the placental circuitry to the newborn lung circuitry. Because changes in ductal flow, decreasing pulmonary vascular resistance, and increasing systemic vascular resistance occur over the first few hours and days of life, cardiovascular assessments should be done shortly after birth, at six to twelve hours of age, and again at one to three days of life in addition to regular intervals after discharge.

## Fetal Circulation

Knowledge of the normal route of fetal blood flow is essential for understanding the circulatory changes that occur at delivery. Fetal circulation is anatomically and physiologically different from adult circulation in several important ways. In the fetus, oxygenation of the blood, removal of carbon dioxide and wastes occurs in the placenta, which is a low-resistance circulatory pathway. Because placental oxygenation is not as efficient as pulmonary oxygenation, the fetus' arterial oxygen tension (PaO<sub>2</sub>) is approximately 20 to 30 torr.

Since fetal hemoglobin binds more tightly to oxygen and the fetal oxyhemoglobin dissociation curve is located to the left of the adult curve, this oxygen tension corresponds to an arterial oxygen saturation of 60 to 70 percent. Fetal circulation involves three unique anatomic features that are not present in the adult other than the placenta and the umbilical vein and arteries. The ductus venosus permits the majority of blood from the placenta to bypass the liver and enter the inferior vena cava. When this blood enters the right atrium, most of it is diverted toward the atrial septum. The foramen ovale is the opening in the interatrial septum that permits a portion of blood to flow from the right atrium directly to the left atrium. This blood then enters the left ventricle and aorta, to perfuse the head and upper extremities of the fetus.

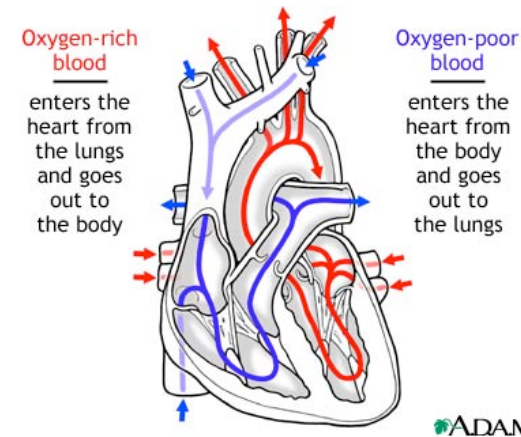
Venous return from the head and upper extremities passes to the heart through the superior vena cava. Most of this blood flows through the right atrium into the right ventricle and enters the pulmonary artery. Since pulmonary vascular resistance is very high and systemic vascular resistance is low most of the blood in the main pulmonary artery flows

through the ductus arteriosus and into the descending aorta to perfuse the trunk and lower extremities.

The patent ductus arteriosus (PDA) is a tubular communication between the pulmonary artery and the descending aorta that allows blood to flow between the pulmonary artery to the aorta, bypassing the fetal lungs. Only about eight percent of fetal cardiac output enters the lungs; 92 percent is diverted through the ductus arteriosus into the descending aorta. Fetal circulation can be described as two parallel circuits rather than the serial circuit present in extrauterine life.

## Neonatal Circulation

The clamping of the umbilical cord and the subsequent removal of the placenta causes immediate circulatory changes in the neonate. With the first breath and occlusion of the umbilical cord, systemic resistance is elevated, which reduces blood flow through the ductus arteriosus. Cord occlusion causes a prompt rise in blood pressure and a corresponding stimulation of the aortic baroreceptors and the sympathetic nervous system. The onset of respirations and lung expansion causes a decrease in pulmonary vascular resistance secondary to the direct effect of oxygen and



carbon dioxide on the blood vessels. Resistance decreases as arterial oxygen increases and arterial carbon dioxide decreases.

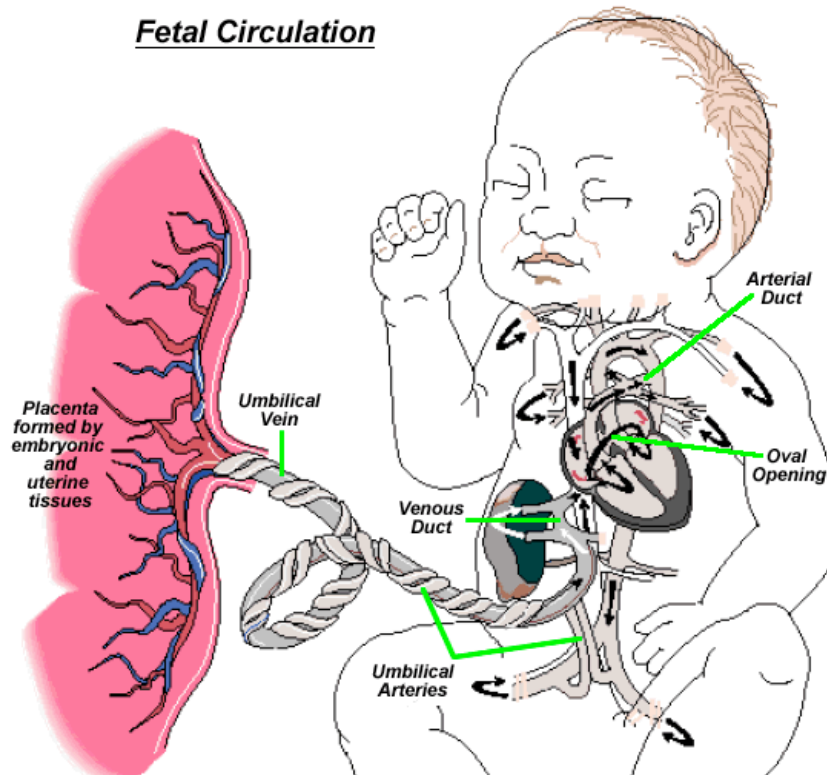
The major portion of the right ventricular output flows through the lungs and increases the pulmonary venous return to the left atrium. The increased amount of blood in the lungs and heat causes increased pressure in the left atrium. The increased pressure in the left atrium combined with the increased systemic resistance functionally closes the foramen ovale. In most individuals, the foramen ovale becomes sealed by the deposit of fibrin and cell products during the first months of life. This process is referred to as anatomic closure of the foramen ovale.

In approximately 25 percent of the population, however, the foramen ovale is not anatomically sealed, so it remains probe-patent beyond adolescence. This means that a catheter can be passed from the right to the left atrium during cardiac catheterization, or that probe can be passed through the foramen ovale during cardiovascular surgery.

Until the foramen ovale is anatomically sealed, anything that produces a significant increase in right atrial pressure can reopen the foramen ovale, making it patent. Due to the structure of the opening, the shunt through the patent foramen ovale is primarily from the right to the left atrium. However, if both atria become much enlarged, the foramen ovale may become stretched open, permitting bi-directional shunting of blood at the atrial level.

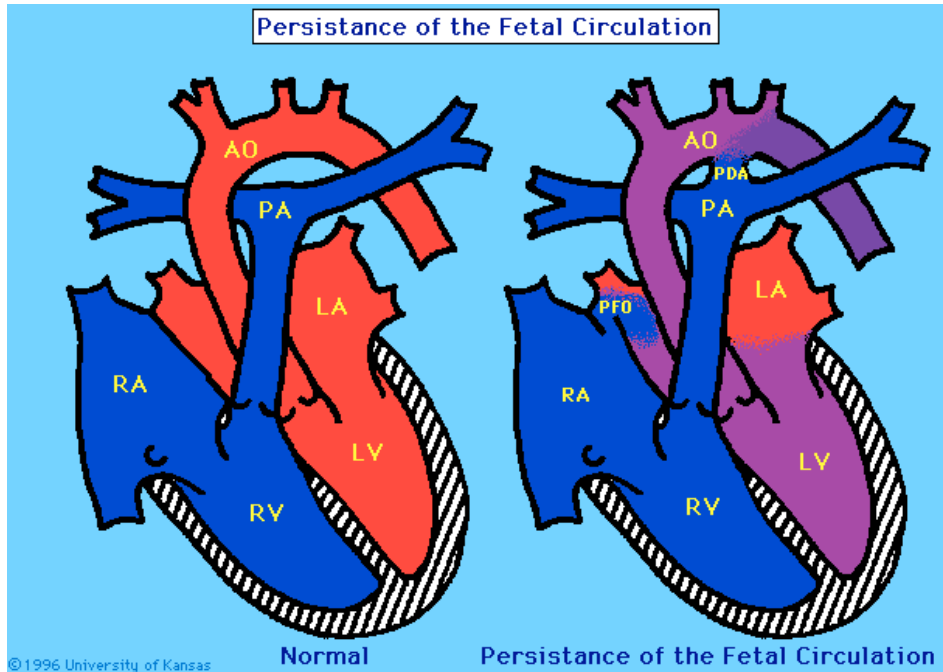
The three major fetal shunts, the ductus venosus, the foramen ovale and the patent ductus arteriosus are normally eliminated within the first days of life. Following closure of these shunts, postnatal circulation is established. Systemic venous blood enters the right atrium from the superior and inferior vena cavae. This poorly oxygenated blood enters the right ventricle, and then passes through the pulmonary artery and into the pulmonary

## Fetal Circulation



circulation, where it becomes oxygenated. The pulmonary venous blood then returns to the left atrium through the pulmonary veins. This blood passes through the left heart and into the aorta to supply the systematic circulation.

When the lungs expand and become air filled, the fetal lung fluid is primarily absorbed into the pulmonary capillaries. Since the lungs provide more efficient oxygenation of the blood than does the placenta, the neonate's arterial oxygen tension rises. This rise is thought to be the most potent stimulus to constriction of the ductus arteriosus. The rise in the oxygen tension of the blood bathing the ductus may also contribute to ductal constriction. Acidosis and a fall in endogenous prostaglandin levels also promote ductal closure.



If the infant has a congenital heart defect producing increased pulmonary blood flow, such as a patent ductus arteriosus or a ventricular septal defect, pulmonary vascular resistance will not fall normally after birth. Such neonates demonstrate a delayed and less marked drop in pulmonary vascular resistance during the first four to twelve weeks of life (this drop may occur over a shorter period in the premature neonate). Because of this delay, a fall in pulmonary vascular blood flow often fails to become apparent until the child is four to twelve weeks of age. These symptoms may develop earlier if the infant is premature. (Normal cardiac circulation on right)

## Cardiac Assessment

Review of the maternal, fetal, and neonatal history is helpful in cardiac evaluation of the newborn.

History that may be associated with congenital heart defects may include: Maternal infections, especially viral and protozoal infections, early in pregnancy, maternal use of tobacco, alcohol, or drugs.

## Maternal drugs known to cause congenital heart defects:

Diphenylhydantem	Fetal hydantoin syndrome, PS, AS
Trimethadoin	VSD, TET
Thalidomide	TET, Truncus
Lithium	Epstein's anomaly
Alcohol	Fetal alcohol syndrome, VSD, ASD

## Neonatal history of:

- Cyanosis
- Tachypnea without pulmonary disease
- Sweating
- Poor feeding
- Edema

## Physical Assessment

Evaluate the newborn's **activity**: sleeping or awake, alert or lethargic, anxious or relaxed. Check **respiratory effort**, including the presence of signs of respiratory distress such as nasal flaring, expiratory grunting, stridor, retractions, or paradoxical respirations. Note skin **color** in a well lit room. (Assess mucous membranes in dark skinned neonates).

### Normal:

Acrocyanosis – peripheral cyanosis or bluish discoloration of hands and feet not involving the mucous membranes it often resolves by 48 hours or with stabilization of the infant.

Circumoral cyanosis – bluish discoloration around the mouth which is associated with nipple or breast feeding and should resolve following the feeding

### Abnormal:

Central cyanosis – bluish discoloration of the tongue and mucous membranes caused by desaturation of arterial blood indicating cardiac and/or respiratory dysfunction.

Cyanosis may be visible with 3 to 5 gm/dL of reduced hemoglobin.

Infants with polycythemia (Hgb > 20 gm) may appear cyanotic even when adequately oxygenated.

Infants with anemia (Hgb < 10 gm) may not appear cyanotic even when adequately hypoxicemic.

Pallor may indicate vasoconstriction.

Physiologic jaundice may be prolonged.

If cyanosis is present, one must differentiate between peripheral and central cyanosis and whether it improves with crying, does not change or becomes worse with crying.

Check for presence of sweating.

Assess for precordial bulging or precordial activity without bulging.

Check for pectus excavatum, which may cause a pulmonary systolic ejection murmur or large cardiac silhouette on an anteroposterior chest radiograph because of the decreased anteroposterior chest diameter.

Check skin perfusion – normal capillary refill time is  $\leq 3$  seconds. Signs of shock can be observed with abnormal skin perfusion when capillary refill is  $> 3$  seconds, prolonged in lower body compared with upper body and mottling associated with other symptoms.

## Palpation

During palpation the nurse should do the following:

Note any hyperactivity. There are two classes of heart disease in which the pericardium appears quite active. Cases of volume overload present in CHD with large left-to-right shunts, such as PDA or VSD. Cases of severe valvular insufficiency, such as aortic or mitral insufficiency.

Check for thrill. A thrill is a fine vibration felt by the hand and corresponds to the sound of a murmur. Thrills are best detected with the palm of the hand, rather than the fingertips, although the fingertips are needed to feel a thrill in the suprasternal notch or over the carotid arteries.

Determine the point of maximal impulse (PMI). This will aid in determining whether the right or left ventricle is dominant. If the right ventricle is dominant the impulse is maximal at the lower left sternal border. If the left ventricle is dominant, the impulse is at the apex.

Count the peripheral pulse rate, noting any irregularities or inequalities of rate or volume.

Evaluate the carotid, brachial, femoral and pedal pulses to detect differences between sides and upper and lower extremities. If pulses are unequal, obtain four extremity blood pressures. A marked difference may be caused by coarctation of the aorta. Cuff size is critical. A cuff that is too narrow gives falsely high readings and too large a cuff may yield low readings.

Assess for bounding pulses.

Palpate the abdomen to determine the size, consistency, and location of the liver and spleen.

## Auscultation

Expert auscultation of the neonatal heart requires much practice over time. The neonatal heart should be auscultated with the infant inactive and quiet. When auscultating, a pediatric or neonatal stethoscope with a diaphragm and bell is very helpful. The pediatric stethoscope has a smaller chest piece than the adult model, and a stethoscope with an even smaller chest piece is used for examining premature infants. The tubing is usually longer to reach inside an isolette. They both have two types of chest pieces.

The open bell conducts sound with practically no distortion, but it makes all sounds loud and may be difficult to maintain an airtight seal. Since low-frequency sounds are hard to hear, the bell is well suited for them. If properly sized, the diaphragm maintains its own seal and is useful for high-pitched sounds. The closed diaphragm has a larger diameter than the bell. It is important to note that the bell piece functions as a diaphragm chest piece when applied too tightly to the skin. The skin acts as a diaphragm, and low-frequency sounds are not as easy to discern. The binaurals should fit comfortably. The ear tubes must be inclined anteriorly to conform to the direction of the normal ear canal. If the chest piece is too large, proper positioning may be difficult to achieve resulting in a harsh noise by intermittent contact of skin with the diaphragm. The harsh noise sounds like a pericardial friction rub.

At a minimum, the four traditional auscultatory areas should be examined. These are the aortic area (second intercostals space, right sternal angle), pulmonic area (second intercostals space, left sternal angle), tricuspid area (fourth intercostals space, left sternal angle), and mitral area (fourth intercostals space, left midclavicular line). A more thorough examination is recommended. There are six anterior areas and three posterior areas for auscultation.

## Anterior

1. **Left Ventricular Area** – centered around the apex of the heart. It extends laterally to the anterior axillary line. The following heart sounds are best heard in this area:

- Mitral insufficiency murmur
- Summation gallops
- Subaortic stenosis murmur
- Aortic insufficiency murmur
- Aortic ejection click in aortic stenosis
- Click and late systolic murmur in mitral valve prolapse
- Mitral stenosis murmur

2. **Right Ventricular Area** – encompasses the lower part of the sternum and the third and fourth intercostals spaces on both sides of the sternum. The following are heart sounds best heard in the right ventricular area:

- Summation gallop
- Pulmonary insufficiency murmur
- Ventricular septal defect murmur
- Aortic insufficiency murmur
- Tricuspid stenosis murmur

3. **Left Atrial Area** – murmurs associated with the left atrium are best heard at the apex:

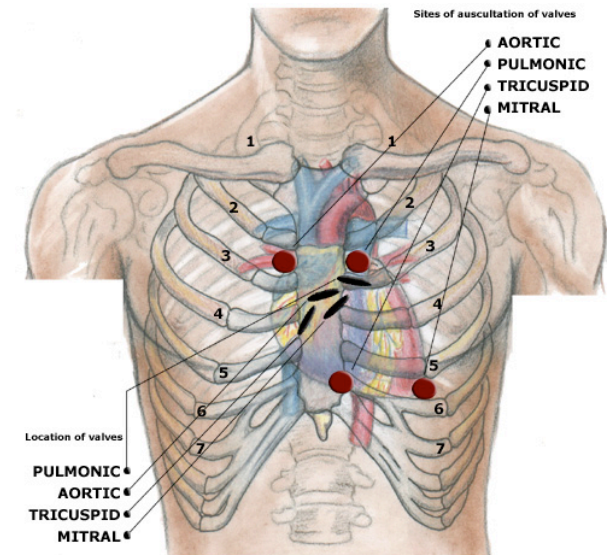
- Mitral stenosis
- Mitral insufficiency

4. **Right Atrial Area** – extends 1–2 cm to the right of the sternum in the fourth and fifth intercostals spaces.

- The murmur of tricuspid insufficiency is best heard here.

5. **Aortic Area** – corresponds to the region of the aortic root and part of the ascending aorta. It begins at the third left intercostals space and extends across the manubrium to the first, second, and third right inter-spaces. The aortic area includes the suprasternal notch and the head of the right clavicle. The following heart sounds are best heard in the aortic area:

- Aortic stenosis murmur
- Aortic stenosis murmur
- Aortic insufficiency murmur
- Sounds caused by increased aortic flow or dilation of the ascending aorta
- Sounds produced by abnormalities of the carotid and subclavian arteries





- The aortic component of the second heart sound

**6. Pulmonary Area** – encompasses the second and third left interspaces close to the sternum. The following sounds are best heard over the pulmonary area:

- Pulmonary stenosis murmur
- Pulmonary insufficiency murmur
- Murmurs caused by increased flow of the pulmonary artery
- Pulmonary ejection click
- The pulmonary component of the second heart sound
- Murmur of patent ductus arteriosus

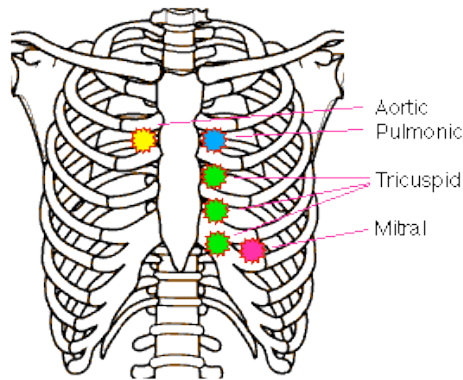
## Posterior

1. **Left Atrial Area** – overlies the fifth, sixth, seventh and eighth posterior interspaces.

- mitral insufficiency murmur

2. **Aortic Area** – overlies the fourth to eighth thoracic vertebral bodies to the left of the midline. The following murmurs are heard in the aortic area:

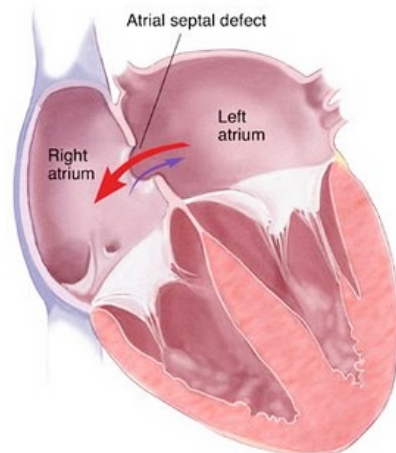
- Aortic stenosis murmur
- Aortic insufficiency
- Coarctation of the aorta



3. **Pulmonic area** – overlies the fourth and fifth thoracic vertebrae and the corresponding interspaces to the left and right of the spine. The murmurs heard there are:

- Pulmonary stenosis murmur
- Pulmonary insufficiency murmur
- Atrial septal defect

Diagram of atrial septal defect



## Identification of Heart Sounds

There are four individual heart sounds: S<sub>1</sub>, S<sub>2</sub>, S<sub>3</sub>, and S<sub>4</sub>. S<sub>3</sub> and S<sub>4</sub> are rarely heard in the newborn. S<sub>1</sub> is the sound resulting from closure of the mitral and tricuspid valves after atrial systole. It is best heard at the apex or lower left sternal border. S<sub>1</sub> is the beginning of ventricular systole. Splitting of S<sub>1</sub> is infrequently noted in

newborns. Wide splitting of S<sub>1</sub> is heard in a newborn with right bundle branch block or Epstein's anomaly.

S<sub>2</sub> is the sound created by closure of the aortic and pulmonary valves, which marks the end of systole and the beginning of ventricular diastole. It is best heard in the upper left sternal border or pulmonic area. Evaluation of the splitting of S<sub>2</sub> is important diagnostically. The timing of the closure of the aortic and pulmonary valves is determined by the volume of blood ejected from the aorta and pulmonary artery and the resistance against which the ventricles must pump.

In the immediate newborn period, there may be no appreciable splitting of S<sub>2</sub>. Because the right and left ventricles pump similar quantities of blood and the pulmonary pressure is close to the aortic pressure, these valves close almost simultaneously. Thus, S<sub>2</sub> is heard as a single sound. As the pulmonary vascular resistance falls, the pulmonary resistance decreases and becomes lower than the aortic pressure, causing a splitting of S<sub>2</sub> as the valve leaflets on the left side of the heart (aortic valve) close before those on the right (pulmonary valve).

By 72 hours of life, S<sub>2</sub> should be split. The absence of split S<sub>2</sub> or a widely split S<sub>2</sub> usually indicates an abnormality. A fixed, widely split S<sub>2</sub> occurs in conditions that prolong right ventricular ejection time or shorten left ventricular ejection time. A narrowly split S<sub>2</sub> occurs in conditions in which there is early closure of the pulmonary valve (pulmonary hypertension) or a delay in aortic closure. A single S<sub>2</sub> is significant because it could represent the presence of only one semi lunar valve (aortic or pulmonary atresia, truncus arteriosus).

The relative intensity of the aortic and pulmonary components of S<sub>2</sub> must be assessed. In the pulmonary area, the aortic component is usually louder than the pulmonary component. Increased intensity of the pulmonary component, compared with the aortic component, occurs with pulmonary hypertension. Conditions that cause decreased diastolic pressure of the pulmonary artery (critical pulmonary stenosis, tetralogy of Fallot (TOF), tricuspid atresia) may cause decreased intensity of the pulmonary component. Since S<sub>3</sub> and S<sub>4</sub> are rarely heard in the neonatal period, their presence denotes a pathologic process. Likewise, a gallop rhythm, the result of a loud S<sub>3</sub> and S<sub>4</sub>, and tachycardia are abnormal.

After evaluation of the individual heart sounds, the systolic and diastolic sounds are evaluated. The ejection sound or click occurs after S<sub>1</sub> and may sound like splitting of S<sub>1</sub>. The ejection click is best heard at the upper left or right sternal border or base. The pulmonary click can best be heard at the second or third left intercostals space and is louder with expiration. The aortic click best heard at the second right intercostals space does not change in intensity with change in respiration.

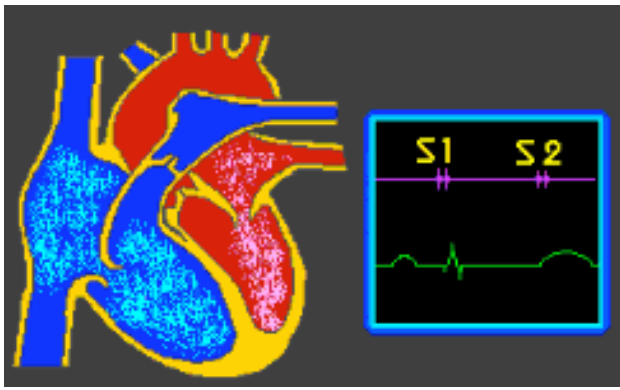
## Murmurs

Cardiac murmurs should be evaluated as to intensity (grades 1 to 6), timing (systolic or diastolic), location, transmission, and quality (musical, vibratory, or blowing):

- Grade 1: barely audible
- Grade 2: soft but easily audible
- Grade 3: moderately loud; no thrill
- Grade 4: loud; thrill present
- Grade 5: loud; audible with stethoscope barely on chest
- Grade 6: loud; audible with stethoscope not touching the chest

The murmur grade is recorded as 1/6 and so on.

The next step in evaluating a murmur is its classification in relation to S<sub>1</sub> and S<sub>2</sub>. The three types of murmurs are systolic, diastolic, and continuous. An infant with no



murmur may still have a significant cardiac disease.

### Systolic Murmurs

Most heart murmurs are systolic, occurring between S<sub>1</sub> and S<sub>2</sub>. Systolic murmurs are either ejection or regurgitation murmurs. They are a normal finding during the routine

physical exam of a healthy infant. Studies have shown that as many as 90% of healthy children have a benign murmur at some time.

Ejection murmurs are caused by flow of blood through stenotic or deformed valves or increased flow through normal valves. Regurgitant systolic murmurs begin with S<sub>1</sub>, with no interval between S<sub>1</sub> and the beginning of the murmur. Regurgitation murmurs generally continue throughout systole. Regurgitation systolic murmurs are caused by flow of blood from a chamber at a higher pressure throughout systole than the receiving chamber. Regurgitation systolic murmurs are associated with only three conditions: 1) ventricular septal defects (VSDs), 2) mitral regurgitation, and 3) tricuspid regurgitation.

### Diastolic Murmurs

Diastolic murmurs are classified according to their timing in relation to heart sounds as early diastolic, mid-diastolic, or pre-systolic. They are usually pathologic. They result from aortic regurgitation and pulmonary insufficiency. With aortic regurgitation the murmur is high pitched and blowing. It begins with the second heart sound and is loudest in early diastole. It may be missed because it is often very soft or may be mistaken for breath sounds because of its high pitch. Bounding pulses are present.

The murmur of pulmonary insufficiency is a distinctive diastolic murmur. It is low-pitched, early in onset and of short duration. It ends well before the first heart sound. It occurs with postoperative TOF, pulmonary hypertension, postoperative pulmonary valvotomy for pulmonary stenosis, or other deformity of the pulmonary valve.

Mid-diastolic murmur results from abnormal ventricular filling. The murmur results from turbulent flow through the tricuspid or mitral valve due to stenosis. They are associated with mitral stenosis or large left-to-right shunt VSD or PDA, producing relative mitral stenosis secondary to increased flow across the normal-sized mitral valve. It is seen in atrial septal defect (ASD), total or partial anomalous pulmonary venous return (TAPVR, PAPVR), endocardial cushion defects, or abnormal stenosis of the tricuspid valve.

### Continuous Murmurs

Most continuous murmurs are not audible throughout the cardiac cycle. They begin in systole and extend into diastole. They are a pathologic finding. They can be produced in three ways: rapid blood flow, high-to-low pressure shunting, and localized arterial

obstruction.

The most significant is the PDA high-to-low shunting. The patency of the ductus is normal in the first 24 hours of life, but a few weeks later a patent ductus is abnormal. It is more common in girls (sex ratio of 3:2), tends to affect siblings, and may be a complication of maternal rubella. It is six times more common in infants born at high altitudes and more common in premature infants. If the ductus is large there may be vigorous pericardial activity, a systolic thrill and bounding pulses, there may be symptoms of congestive heart failure (CHF).

### Congenital Heart Defects

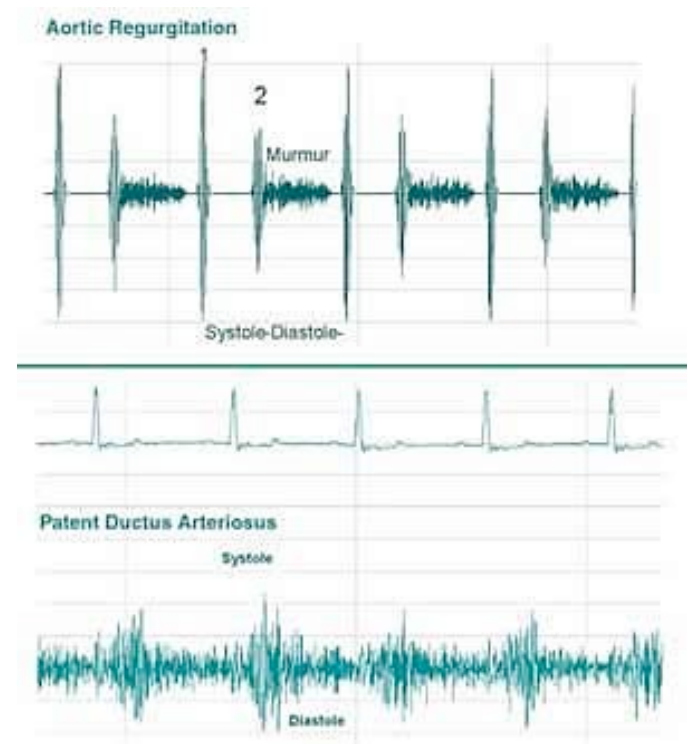
Cardiac development occurs during the first seven weeks of gestation. Causes are classified as chromosomal (ten to twelve percent), genetic (one to two percent), maternal or environmental (one to two percent), or multifactorial (85 percent). The vast majority are considered to be of multifactorial origin. These defects are probably the result of an interaction effect of the other causes. The following diagram of a normal heart is supplied for reference when reading the descriptions of abnormalities.

### Acyanotic Heart Defects

Acyanotic heart defects are those that produce a left-to-right shunt. Typically these defects do not produce cyanosis because there is sufficient oxygenated blood in the circulation. The left-to-right or right-to-left shunts produce increased pulmonary blood flow and increased workload on the heart.

### Patent Ductus Arteriosus (PDA)

PDA is the failure of the ductus to close in response to increased arterial oxygen concentrations after the initiation of pulmonary function. The persistence of the ductus arteriosus is beyond 24 hours. A systolic murmur is heard. Bounding peripheral pulses help to differentiate a PDA from a Ventricular Septal Defect (VSD). The precordium is usually active. Some infants will have widened pulse pressures. Infants weighing <1,000 grams are likely to have reduced systolic



and diastolic pressures. The volume overload of blood in the left atrium and left ventricle lead to increased pulmonary venous engorgement. In addition to the systolic murmur and bounding pulses symptoms of CHF are tachypnea, dyspnea, hoarse cry, frequent lower respiratory tract infections and coughing, and poor weight gain.

**Timing of PDA treatment is controversial with three broad approaches to timing:**

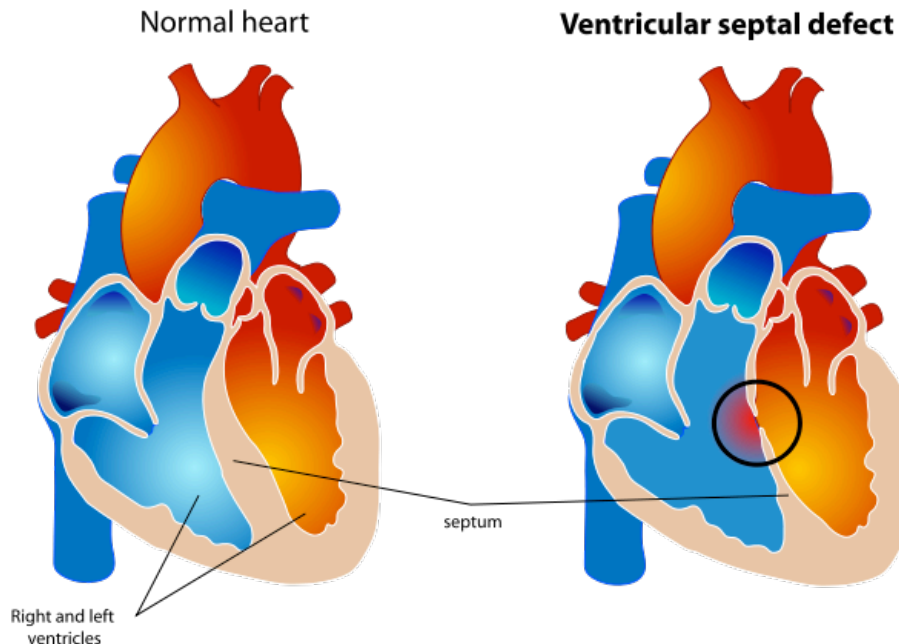
1. Treating when the PDA becomes clinically symptomatic
2. Targeted presymptomatic treatment
3. Prophylactic treatment

*None of these approaches has shown clear benefits in short and long-range outcomes. It is important to note that the prophylactic approach to PDA treatment is the only strategy that has been shown to have benefits of any sort. Medical management includes prophylactic antibiotics against bacterial endocarditis and prostaglandin inhibitors. Prostaglandins prevent the ductus from closing. Definitive treatment is surgical ligation.*

## Ventricular Septal Defect (VSD)

VSD can occur anywhere in the ventricular septum. The size of the defect and the degree of pulmonary vascular resistance are more important to severity than location. X-ray is normal. There is usually a loud harsh pansystolic murmur. Symptoms depend on severity, and range from asymptomatic to poor exertional tolerance, recurrent pulmonary infections and symptoms of CHF. With severe VSD, there may be pulmonary hypertension and cyanosis.

Management of VSD includes monitoring for CHF and treatment with diuretics and digitalis. Unless there is pulmonary hypertension there is no activity restriction. Prophylaxis against bacterial endocarditis may be implemented prior to surgical closure of the VSD.



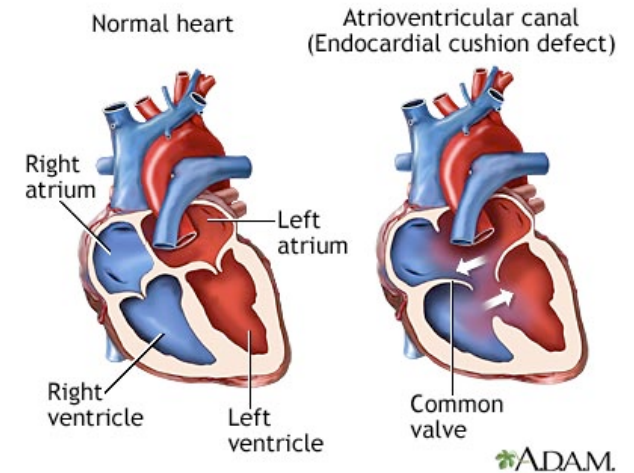
## Atrial Septal Defect (ASD)

ASD is a communication between the right and left atria. There is increased blood flow to the right ventricle through the pulmonary valve. This creates the typical ejection murmur, usually grade II/VI. The infant is usually asymptomatic unless the murmur is present. X-ray may show enlargement and increase in pulmonary vascularity. Untreated ASD can lead to CHF, pulmonary hypertension, and atrial arrhythmias.

Spontaneous closure of ASDs occurs in the first five years of age in up to 40 percent of children, medical management includes prevention and treatment of CHF. Activity is not restricted. Surgical correction is accomplished by a simple patch or with direct closure.

## Endocardial Cushion Defects

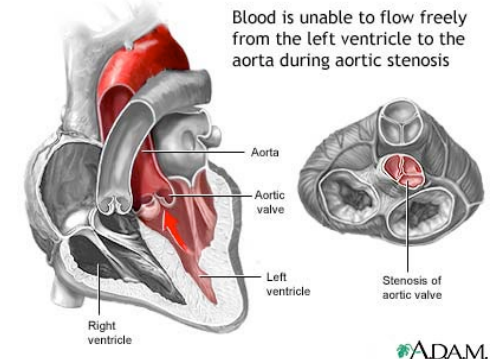
Endocardial cushion defects are lesions that produce abnormalities of the atrial septum, ventricular septum, and AV valves. Symptoms result from increased pulmonary blood flow caused by the abnormal connection between both ventricles and the atria. The infant may present with respiratory distress, signs of CHF, tachycardia, and a murmur. The mitral regurgitation may be heard as grade III holosystolic murmur that transmits to the back. Chest x-ray reveals generalized cardiomegaly and increased pulmonary vascularity. The infant has recurrent respiratory infections and failure to thrive.



Management is aimed at preventing or treating CHF and bacterial endocarditis. Surgical closure of ASD and VSD with reconstruction of the AV valves is required. In some cases, pulmonary artery banding may be performed as a palliative procedure if there is not significant mitral regurgitation. Surgery is indicated when CHF is unresponsive to medical therapy, recurrent pneumonia, failure to thrive, or a large shunt with development of pulmonary hypertension and increasing pulmonary vascular resistance.

## Aortic Stenosis

Aortic stenosis is one of a group of defects that produce obstruction to ventricular outflow. There is narrowing or thickening of the aortic valvular region. Symptoms depend on severity.





Mild stenosis can be asymptomatic. More severe stenosis can cause activity intolerance, chest pain, and CHF. There may be narrow pulse pressure and a higher systolic pressure in the right arm. Cardiomegaly is present with CHF.

Management of aortic stenosis includes prevention and treatment of CHF with fluid restriction, diuretics, and digitalis. Some activity restrictions may be required to prevent increased demand on the heart in moderate to severe cases. Balloon valvuloplasty may be performed during cardiac catheterization to improve circulation. In critical cases, maintenance of the patency of the ductus arteriosus with prostaglandin E<sub>1</sub> to prevent hypoxia may be needed. The type and timing of surgical correction depends on the exact location and severity of the defect.

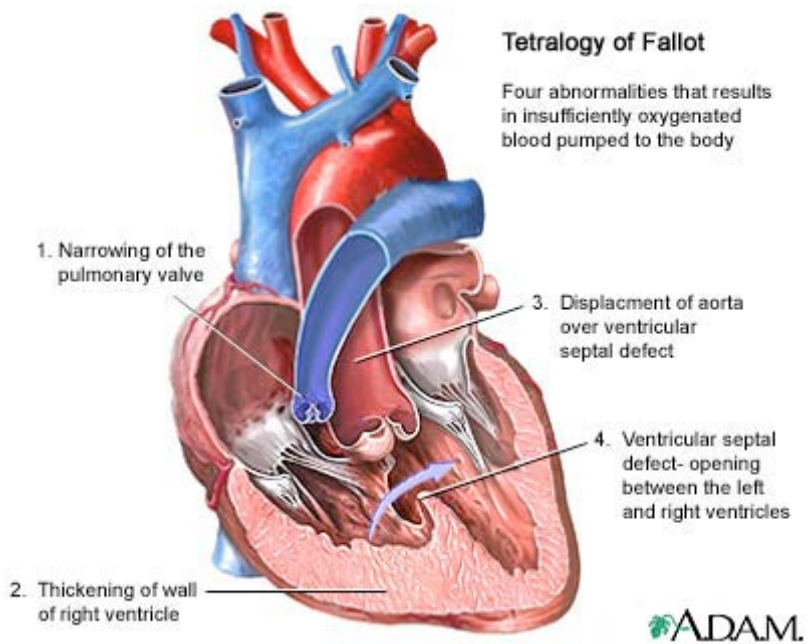
## Cyanotic Heart Defects

Cyanotic heart defects are those defects with right-to-left shunt with either reduced or increased pulmonary blood flow.

## Tetralogy of Fallot (TOF)

TOF is composed of the following abnormalities:

- Ventricular septal defect
- Pulmonic outflow obstruction
- Aorta overriding right and left ventricles
- Right ventricular hypertrophy



In cases of TOF blood is shunted right to left through the VSD and the overriding aorta. The murmur of TOF is loudest along the left sternal border between the third and fourth interspaces. A thrill will be present if the murmur is at least a grade III. There is usually a single second heart sound. Cyanosis depends on the varying

degrees of pulmonary outflow obstruction. The chest x-ray demonstrates enlargement of the right ventricle, small main pulmonary artery, and a normal left atrium and left ventricle. It has been called “boot-shaped” heart. The EKG would show right ventricular hypertrophy.

Cardinal signs include cyanosis, hypoxia, and dyspnea. Severe decompensation or “tet” spells are common in infants or children but can occur in neonates. Children instinctively assume a squatting position, which traps venous blood in the legs and decreases systemic venous return to the heart. Chronic arterial desaturation stimulates erythropoiesis, causing polycythemia that may lead to increased blood viscosity, microcytic anemia, and cerebrovascular accident.

Definitive therapy for TOF is surgical repair. Medical management includes prevention and treatment of hypoxemia, polycythemia, infection and microcytic hypochromic anemia. It is important to avoid dehydration to prevent increased risk of cerebral infarcts because of hemoconcentrations. Surgical management may be either palliative or corrective with palliative procedures undertaken to improve pulmonary blood flow by creating a pathway between systemic and pulmonary circulation.

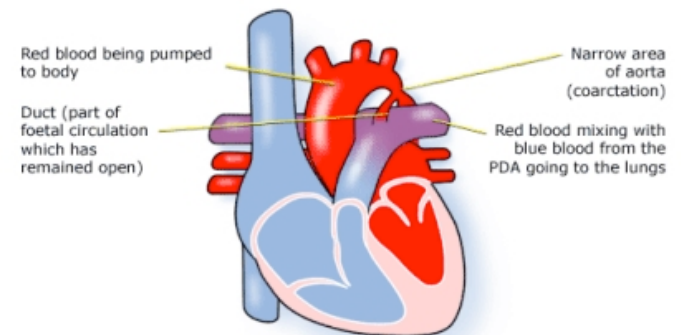
## Coarctation of the Aorta

The coarctation of the aorta obstructs flow from the proximal portion of the aorta to its distal portion. If it is proximal to the insertion of the ductus arteriosus, the lower half of the body will be supplied by the right ventricle through the ductus, and should be cyanotic. The upper half of the body will be supplied by the left ventricle, and should be totally oxygenated. Collateral circulation will not be stimulated during fetal life. After birth the circuitry persists. On chest x-ray cardiomegaly is evident. The EKG will consistently show right ventricular hypertrophy. Hypertension in the upper extremities and a lower pressure in the lower extremities can be expected. Femoral pulses will be present but weaker.

Normally no murmur is present. The second sound will be closely split. If the coarctation is distal to the insertion of the ductus arteriosus, collateral circulation will be established during fetal life to permit perfusion to the lower half of the body. This x-ray shows an indentation of the aorta that resembles the number 3. The infant will present with CHF, absent, weak, or delayed pulses in the lower extremities, and bounding pulses in the upper extremities. There may be a systolic thrill felt at the suprasternal notch.

Surgery is performed at three to five years of age if signs and symptoms can be medically controlled. Surgery is performed earlier if medical management is not

Coarctation of the aorta





successful in providing adequate oxygenation, preventing CHF and avoiding sub acute bacterial endocarditis. Prostaglandin E1 may be needed to maintain ductal patency if the constricted segment is at the level of the ductus arteriosus.

## Pulmonary Atresia

Pulmonary atresia results in the absence of communication between the right ventricle and the pulmonary artery. The right ventricle is usually hypoplastic, with thick ventricular walls. The presence of a PDA, ASD, or patent foramen ovale to allow mixing of blood is crucial for survival. Cyanosis and tachypnea present without other signs of obvious respiratory distress.

A soft systolic murmur is heard at the upper left sternal border. Heart size may be normal or enlarged. There are decreased pulmonary vascular markings.

Prostaglandins are used to maintain ductal patency until balloon atrial septostomy can be performed to promote mixing of systemic and pulmonary venous blood in the atria. Surgical correction is performed by creating a systemic-pulmonary artery shunt between the left subclavian artery and the left pulmonary artery.

## Pulmonary Stenosis

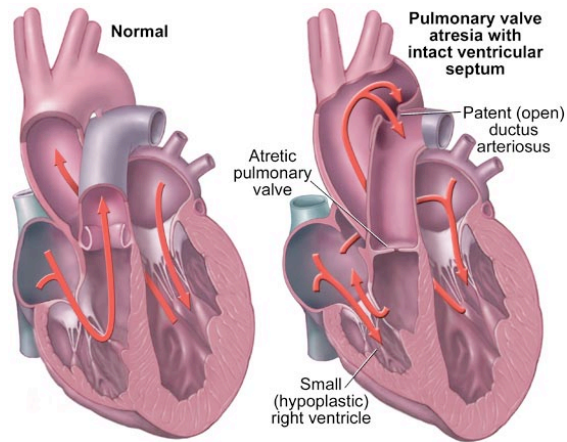
Pulmonary Stenosis is when narrowing of the pulmonary valve causes the right ventricle to pump harder to get blood past the blockage. Cyanosis depends on the severity of the stenosis. Valvuloplasty may be done during cardiac catheterization to stretch the valve. Moderate stenosis may cause easy tiring. Severe or critical pulmonary stenosis will cause CHF. There is a pulmonary systolic ejection click at upper left sternal border and widely split S2 or systolic ejection murmur (grade 2 to 5/6), at the upper left sternal border and transmits across the back.

Surgical correction is performed in children when the right ventricular pressure measures 80 to 100 mm Hg and balloon valvuloplasty is not successful. Infants with critical pulmonary stenosis and CHF require prostaglandin infusion to maintain ductal patency until surgery is performed.

## Truncus Arteriosus

This results from inadequate division of the common great vessel into a separate aorta and pulmonary artery. Cyanosis may be present depending on the amount of pulmonary blood flow. A systolic click and harsh VSD murmur may be present. On x-ray the heart size is increased. CHF, bounding arterial pulses, and widened pulse pressures are present. If truncus arteriosus is not detected in the newborn period the infant will feed poorly, fail to thrive, have frequent respiratory infections, and worsening CHF.

Treatment involves control of CHF and prophylaxis with antimicrobial agents.



Pulmonary artery banding is performed as a palliative measure in small infants with increase pulmonary blood flow and CHF that does not respond to medical management. The definitive surgical correction is performed during infancy.

## Transposition of the Great Arteries (TGA)

TGA is the result of inappropriate septation and migration of truncus. The aorta receives unoxygenated blood and returns it to the systemic circuit. There are two separate parallel circulations. Marked cyanosis is present as well as signs of CHF.

There will be loud harsh systolic murmur. Hypoglycemia, hypocalcemia, and metabolic acidosis are frequently present. On x-ray, the heart is enlarged and has a narrow base. It is described as egg-shaped.

TGA is a cardiac emergency. Immediate management includes correction of acidosis, hypoglycemia, and hypocalcemia. Oxygen and prostaglandins are administered. Cardiac catheterization with balloon atrial septostomy is done.

Prognosis for TGA without surgical intervention is poor. Definitive surgical correction is done by switching the right and left sided structures at the ventricular level, the artery level, or the atrial level.

## Tricuspid Atresia

In this condition there is no tricuspid valve, so no blood can flow from the right atrium to the right ventricle. The right ventricle is small and survival depends on an ASD or VSD.

Most of the poorly oxygenated blood goes from the left ventricle into the aorta and on to the body. The rest of the blood flows through the VSD to the small right ventricle to the pulmonary artery and back to the lungs. The infant is cyanotic.

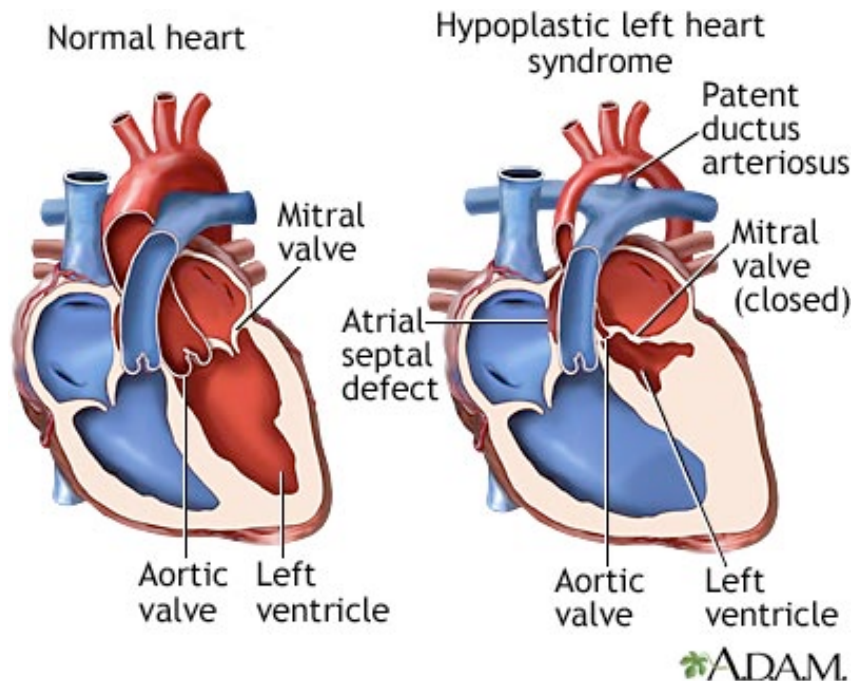
## Total Anomalous Pulmonary Venous Return

The pulmonary veins drain into the right atrium (rather than the left atrium). There is no direct connection between the pulmonary veins and the left atrium. Cyanosis is present, as is respiratory distress. Feeding is associated with increased cyanosis, the infant tires easily and has progressive growth failure.

Increased cyanosis associated with feeding is secondary to the compression of the common pulmonary vein by the filled esophagus. A grade II to III systolic ejection murmur is heard at the left sternal border. A precordial bulge and hyperactive right ventricular impulse may be seen. The PMI is at the xiphoid process or lower left sternal border.

The pulmonic sound may be pronounced. A quadruple or quintuple gallop rhythm is heard. X-ray findings include mild to moderate cardiomegaly and increased pulmonary markings. The characteristic "snowman sign" occurs because of anatomic appearance of left superior vena cava, the left innominate vein, and the right superior vena cava.

Treatment is focused at preventing CHF and hypoxemia. Diuretics may be administered to decrease pulmonary edema and balloon atrial septostomy performed to enlarge the interatrial communication that will promote better mixing of blood. Surgery is delayed until infancy if medical management is successful.



## Hypoplastic Left Heart Syndrome (HLHS)

HLHS consists of a group of defects including a small aorta, aortic and mitral valve stenosis, and a small left atrium and ventricle. The infant presents with progressive cyanosis, pallor, and mottling. Tachycardia, tachypnea, dyspnea, and pulmonary rales are present. The second heart sound is loud and single. Poor peripheral pulses and vasoconstriction of the extremities is noted on the exam. On x-ray, mild to moderate heart enlargement and pulmonary venous congestion is seen.

Management is aimed at prevention of hypoxemia and correction of metabolic acidosis. Prostaglandins are administered to maintain ductal patency. Balloon atrial septostomy is done to decompress the left atrium. Surgical correction of HLHS is experimental and has high mortality. Transplantation is a more common option for these infants who typically have 100 percent mortality.

## Drug Treatment of Congenital Heart Defects

### Diuretics

Diuretics are used in the treatment of CHF to decrease fluid overload and fluid retention:

- Furosemide (Lasix) – 1 mg/kg/dose IV
- Spirolactone (Aldactone) – 1.5 – 3.0 mg/kg/day PO
- Chlorothiazide (Diuril) – 20 – 40 mg/kg/day PO

### Inotropic Agents

These are used to increase myocardial performance by increasing the strength of contraction of the heart muscle. There is often a concomitant increase in heart rate during administration of these agents. It is also used to increase renal perfusion and increase heart rate, increase venous return to the heart, and decrease pulmonary vascular resistance. They have a short half-life and must be infused continuously. The major complications are tachyarrhythmias and tissue necrosis following extravasation.

- Dopamine – 2 - 20 µg/kg/min IV continuous infusion
- Dobutamine – 2-10 µg/kg/min IV continuous infusion
- Isoproterenol – 0.05-0.5 mg/kg/min
- Amrinone – 0.75 – 3 mg loading dose over three minutes, maintenance infusion of 5 µg/kg/min

### Prostaglandin

Prostaglandin is indicated to maintain patency of the ductus arteriosus to provide adequate systemic or pulmonary blood flow in infants with specific heart defects. It directly relaxes smooth muscles in arteriolar and venous walls; increases cardiac output if the decrease is secondary to myocardial dysfunction.

Prior to initiation of therapy, other causes of hypoxia should be excluded. Prostaglandin E1 may precipitate respiratory depression or systemic hypotension in neonates with RDS, pulmonary disease, sepsis, or intracerebral hemorrhage. It has a rapid onset of action. It produces vasodilatation, smooth muscle relaxation of ductus arteriosus, and pulmonary and systemic circulations. There is increased arterial saturation by 25 to 100 percent. It is important to monitor B/P. Vasopressors may be required. Apnea, flush, fever, seizure-like activity, and decreased heart rate are common side effects.

- PGE1 – 0.05-0.1 mg/kg/min IV continuous infusion

### Prostaglandin Synthetase Inhibitors

This is indicated for the pharmacologic closure of the patent ductus arteriosus (PDA). During gestation, the patency of the ductus is maintained by the production of prostaglandins. It promotes ductal closure by inhibition of prostaglandins in the wall of the ductus.

Failure of the ductus to close postnatally often complicates recovery from respiratory distress syndrome (RDS) in premature infants. Despite initial improvement in the RDS with subsequent decrease in pulmonary vascular resistance, the infant's condition worsens due to a large left-to-right shunt through the ductus. This often results in increased supplemental oxygen requirements, ventilator dependence, and CHF.

Indomethacin is a nonsteroidal anti-inflammatory drug that inhibits prostaglandin production by blocking the action of cyclooxygenase on arachidonic acid, thus accelerating ductal closure. Its onset is 12 to 24 hours. It is essential to monitor renal function, bilirubin, electrolytes, glucose, platelets, and bleeding. Repeat dosing in premature infants may be required. Failure to close the ductus after three courses of Indocin may require surgical closure.

- Indomethacin (Indocin) – 0.2 mg/kg IV q 24 hr