

# **INTRODUCTION TO CONGENITAL HEART DISEASE**

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# Fetal Anatomy & Physiology

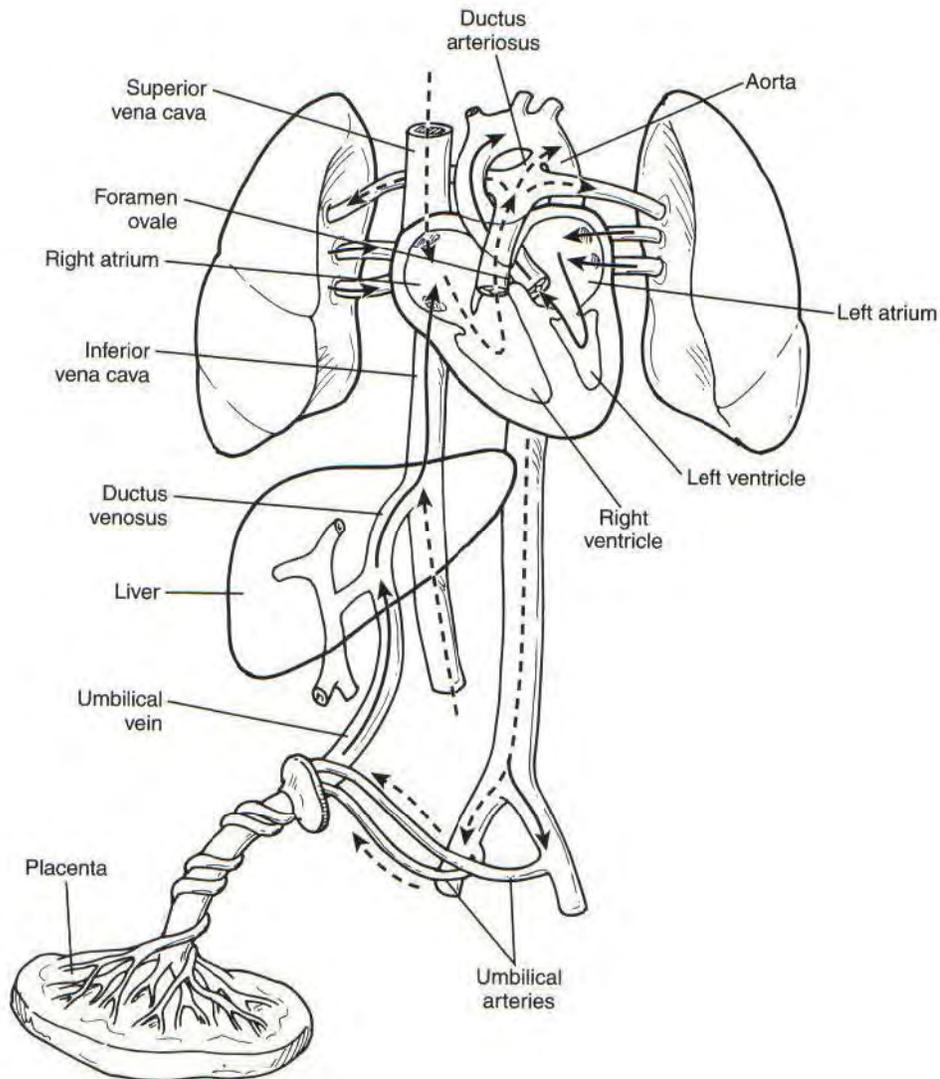


Figure 1: Fetal circulation (*The Pediatric Anesthesia Handbook, 2<sup>nd</sup> Ed*  
Bell C, Kain ZN, Hughes C. Reprinted with permission)

## Anatomy

Two umbilical arteries originate from the internal iliac arteries and deliver fetal blood to the placenta where it is oxygenated. One umbilical vein carries oxygenated blood from the placenta to the fetus. The majority of umbilical vein blood bypasses the liver via the ductus venosus and empties into the inferior vena cava (IVC) where it mixes with less oxygenated blood from the lower half of the body. The IVC blood entering the right atrium is directed by the Eustachian valve across foramen ovale into the left atrium. The left ventricle pumps this blood to the heart and upper body through the great vessels of the aortic arch. Deoxygenated superior vena cava (SVC) blood enters the right atrium and

primarily crosses the tricuspid valve into the right ventricle. Only a small amount of SVC blood enters the left atrium via the foramen ovale. High pulmonary vascular resistance (PVR) forces the right ventricular output to enter the systemic circulation via the ductus arteriosus. The ductus arteriosus originates from the pulmonary artery (PA) and inserts into the aorta in the region of the distal arch and descending thoracic aorta.

Fetal circulation is characterized by:

- high PVR secondary to fluid filled lungs and a hypoxic environment
- low systemic vascular resistance (SVR) secondary to the large surface area of the low resistance utero-placental bed
- the most oxygenated blood from the umbilical vein perfuses the brain and heart preferentially by shunting across the liver via the ductus venosus and shunting across the right heart via the foramen ovale
- lesser oxygenated blood perfuses the lower body by shunting across the ductus arteriosus and into the descending thoracic aorta

### **Physiology**

Umbilical vein PaO<sub>2</sub> is 30-35 mmHg. Therefore, oxygen transport must be achieved in a relatively hypoxic environment. Approximately 80% of fetal hemoglobin is Hgb F compared to an adult who has over 90% Hgb A. The PaO<sub>2</sub> at which Hgb is 50% saturated is called the P50. Hgb F (P50 19mmHg) is left shifted in comparison to Hgb A (P50 26mmHg). Hgb F has a greater affinity for oxygen than Hgb A which improves oxygen uptake at the placenta. Fetal pH (normal values 7.25-7.35) is lower than in adults. The lower fetal pH improves oxygen unloading at the tissue level, because Hgb F binds oxygen more avidly than Hgb A. Fetal hemoglobin is high compared to adult levels, which raises oxygen carrying capacity.

Oxygen content of blood (CaO<sub>2</sub>) is defined by the following equation:

$$\text{CaO}_2 \text{ (ml O}_2\text{/dl blood)} = (\text{SaO}_2 \times \text{Hgb} \times 1.34) + (\text{PaO}_2 \times 0.003)$$

SaO<sub>2</sub> is expressed as a fraction

Hgb in grams/dl

1.34 ml O<sub>2</sub>/Hgb (gram/dl). Amount of O<sub>2</sub> in mls carried per gram/dl of Hgb

PaO<sub>2</sub> in mmHg

0.003 ml O<sub>2</sub>/mmHg. Amount of dissolved O<sub>2</sub> in mls per mmHg partial pressure O<sub>2</sub>

At normal levels of PaO<sub>2</sub>, the amount of oxygen dissolved in blood is negligible since it is multiplied by 0.003 ml O<sub>2</sub>/mmHg. Therefore the CaO<sub>2</sub> is calculated by determining the amount of oxygen carried bound to hemoglobin. For example, an adult with a SaO<sub>2</sub> of 98% and a Hgb of 14 grams/dl would have the following oxygen content:

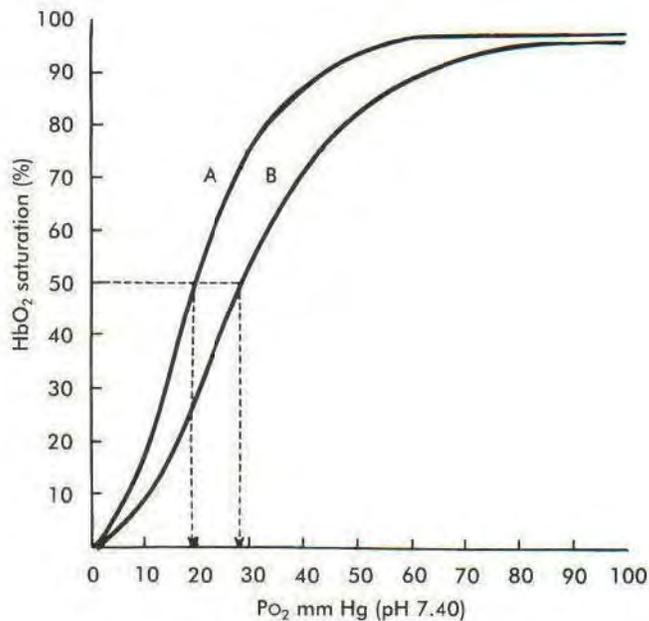
$$\text{CaO}_2 = 0.98 \times 14 \times 1.34 = 18.4 \text{ ml O}_2\text{/dl of blood}$$

The fetus maintains CaO<sub>2</sub> through two mechanisms. Firstly, Hgb F is left shifted which means that at any given PaO<sub>2</sub> fetal Hgb is more saturated than adult Hgb. Umbilical vein

PaO<sub>2</sub> is between 30-35 mmHg. Fetal Hgb is approximately 70%-80% saturated at this PaO<sub>2</sub> in comparison to adult Hgb which would have a SaO<sub>2</sub> of 50-60%. This significant difference occurs because a PaO<sub>2</sub> of 35mmHg falls in the steepest part of the oxygen-hemoglobin dissociation curve. Secondly, hemoglobin levels in utero are elevated which raises CaO<sub>2</sub>. Using the equation for CaO<sub>2</sub> for a fetus with a Hgb of 18 grams/dl and a SaO<sub>2</sub> of 75% in the umbilical vein yields the following:

$$\text{CaO}_2 = 0.75 \times 18 \times 1.34 = 18.1 \text{ ml O}_2/\text{ dl blood}$$

The effect of the left shifted Hgb F and polycythemia produce an oxygen carrying capacity in the fetus that is nearly equal to adults despite an in utero SaO<sub>2</sub> of 75%.



A: Fetal Hgb (P50 19 mmHg)

B: Adult Hgb (P50 26 mmHg)

Figure 2: Oxygen-hemoglobin dissociation curve (*Fetal and Neonatal Physiology, 3rd Ed Polin RA, Fox WW, Abman SH. Reprinted with permission*)

### Transitional Circulation

The events precipitating the transition from fetal to adult circulation are clamping of the umbilical cord and inflation of the lungs. Cord clamping removes the low resistance placenta from the circulation and raises SVR. Lung inflation and increased PaO<sub>2</sub> lowers PVR dramatically, causing increased pulmonary blood flow and increased blood return to the left atrium. The left atrial pressure rises above right atrial pressure closing the flap of tissue covering the foramen ovale. A true patent foramen ovale (PFO) is rare in adulthood but a probe patent PFO persists in approximately 25% of adults.

The ductus arteriosus remains patent in utero due to the effects of hypoxia, mild acidosis and placental prostaglandins. Removal of these factors after delivery initiates the process

of ductal closure. Functional closure occurs because left atrial pressure rises above right atrial pressure. Anatomic closure of the ductus arteriosus takes weeks to occur. The ductal remnant is known as the ligamentum arteriosum. A patent ductus arteriosus (PDA) often occurs in premature infants with lung disease. Indomethacin, via its anti-prostaglandin action can be used to induce closure of the PDA. During the period before anatomic closure of the ductus arteriosus and foramen ovale, certain physiologic stresses can cause the newborn to revert to fetal circulation. This is characterized by increased pulmonary vascular reactivity, raised PVR and right-to-left shunting at the PFO and PDA. The clinical result is cyanosis. Hypothermia, hypercarbia, acidosis, hypoxia and sepsis can all cause a reversion to fetal circulation.

There are many types of congenital heart disease (CHD) presenting in the newborn period in which either pulmonary or systemic blood flow is dependent on shunting through the ductus arteriosus. Prenatally, the fetus is stable because of ductal flow but postnatally closure of the ductus arteriosus would be fatal. The discovery of prostaglandin E1 (PGE1 or Alprostadil™) has revolutionized the care of these newborns allowing stabilization of their condition while a thorough assessment is undertaken and the appropriate clinical decisions made. PGE1 can be started immediately after delivery when prenatal studies suggest CHD with a need for ductal patency. It can also be started after functional closure of the ductus arteriosus and is usually successful in restoring ductal patency. Higher doses (greater than 0.03 mcg/kg/min) can result in apnea requiring some neonates to be intubated. When PGE1 is infused for weeks a generalized edematous state can result. Surgeons will often note that the tissues lack the normal tensile strength and tone.

## **Neonatal Myocardium**

General immaturity and a decreased number of myofibrils result in both reduced contractility (inotropy) and relaxation (lusitropy). Afterload increases are poorly tolerated because of the reduced inotropic state. Preload reductions are poorly tolerated because of the reduced lusitropic state. Calcium is critical for both ventricular contraction and relaxation. The neonatal T tubule system and sarcoplasmic reticulum is immature resulting in poor release and re-uptake of intracellular calcium. Consequently, the neonate relies much more on a normal serum level of ionized calcium. Contrary to some traditional teaching the neonate does respond favorably to volume loading through the Frank-Starling mechanism. However, it does so within the restrictions posed by the immaturity of its contractile and relaxation properties. Therefore rapid volume loading can “over stretch” and distend the ventricles. Given these limitations, the neonate’s cardiac output is more reliant on heart rate than that of an adult. The right ventricle and left ventricle are equal in a neonate because the right ventricle has hypertrophied in response to the high PVR in utero. Thus, the ventricles are inter-dependent and failure of one can rapidly lead to bi-ventricular failure. Ventricular remodeling occurs as PVR falls and SVR rises. The left ventricle hypertrophies and forms the apex of the heart while the right ventricular hypertrophy regresses. Parasympathetic innervation is well developed but sympathetic innervation is poor and potent stimuli may result in a profound vagal response and bradycardia.

# Congestive Heart Failure

The usual determinants of cardiac function (preload, afterload, systolic contractility, diastolic relaxation) apply in the pediatric heart as they do in adults. Congestive heart failure (CHF) in adults is caused by left ventricular dysfunction or valvular disease. Left ventricular dysfunction can be either systolic, diastolic or both. Aortic or mitral valve disease can also lead to CHF. Regardless of the cause, eventually elevated pulmonary venous pressures lead to pulmonary hypertension and interstitial edema. The same set of circumstances can apply in infants and children but more commonly CHF is by pulmonary over circulation rather than ventricular dysfunction or valvular disease.

## Left-to-right shunts

Pulmonary over circulation from a large left-to-right shunt results in CHF. In this case, CHF results from excessive forward circulation and not elevated left atrial and pulmonary venous pressures. Cardiac function is normal and often even hyperdynamic. Preload is increased as a large component of left ventricular output bypasses the systemic circulation, entering the lungs and rapidly returning to the left side of the heart. Symptoms of CHF in infancy are tachypnea, wheezing, tachycardia, diaphoresis and failure to thrive with poor weight gain. Classically tachypnea and diaphoresis occur with feeding. Initial management consists of diuretics (furosemide). The Qp:Qs is the ratio of pulmonary to systemic blood flow. In left-to-right shunts the Qp:Qs can only be determined by hemodynamic catheterization.

Qp:Qs	Shunt	Clinical status
<1	right-to-left	cyanosis
1-2	minimal left-to-right	asymptomatic
2-3	moderate left-to-right	mild symptoms of CHF
>3	large left-to-right	severe symptoms of CHF

## Mixing lesions

The other scenario in which there is pulmonary over circulation and possible CHF is mixing lesions. Mixing lesions occur when a functional single ventricle ejects the mixed systemic and pulmonary venous return. These patients are cyanotic and may be dependent on a PDA to ensure pulmonary or systemic blood flow. The Qp:Qs ratio of pulmonary to systemic blood flow can be determined roughly from the systemic oxygen saturation by the following simple equation:

$$Qp:Qs = \frac{SaO_2 - SvO_2}{SpvO_2 - SpaO_2}$$

SaO<sub>2</sub>: Aortic O<sub>2</sub> saturation  
 SvO<sub>2</sub>: SVC O<sub>2</sub> saturation  
 SpvO<sub>2</sub>: Pulmonary vein O<sub>2</sub> saturation  
 SpaO<sub>2</sub>: Pulmonary artery O<sub>2</sub> saturation

The explanation of how this equation is derived is included in the section on Hemodynamic Calculations. The following four assumptions are made:

1. Fractional inspired oxygen ( $F_{iO_2}$ ) is 0.21 and pulmonary venous blood is fully saturated.
2. Oxygen consumption ( $VO_2$ ) is normal, with a  $SvO_2$  of 25-30% less than  $SaO_2$ .
3. The patient is not severely anemic. (This lowers SVC  $O_2$  saturation)
4. Complete mixing with aortic and pulmonary artery oxygen saturations being equal.

In most cases, the assumptions are valid allowing a rapid bedside determination of  $Q_p:Q_s$  based only on the  $SpO_2$ . Consider a patient with tricuspid atresia. All systemic venous blood shunts from right-to-left at the atrial level. The patient is on PGE1 to maintain a PDA. The single ventricle (left ventricle morphology) ejects mixed blood into the aorta. The pulmonary circulation is supplied via the PDA. The measured  $SpO_2$  is 80%. Based on the above assumptions, oxygen saturation in the SVC is 55%, the PA 80% and the pulmonary vein 100%. The  $Q_p:Q_s$  is 1.25. There is cyanosis but normal ventricular work load. Now consider a patient with hypoplastic left heart. All pulmonary venous blood shunts from left-to-right at the atrial level. The patient is on PGE1. The single ventricle (right morphology) ejects mixed blood into the pulmonary artery. The systemic circulation is supplied via the PDA. The measured  $SpO_2$  is 90%. Based on the assumptions, the  $O_2$  saturation in the SVC is 65%, the PA 90% and the pulmonary vein 100%. The  $Q_p:Q_s$  is 2.5. There is almost normal oxygen saturation but greatly increased work on the single ventricle.

When the systemic oxygen saturation rises, the increased cardiac output is represented by the higher  $Q_p:Q_s$ . The crucial point to understand is that the  $Q_p:Q_s$  reflects the amount of increased myocardial work. Imagine a 3 kg newborn with a mixing lesion. The cardiac output required to meet systemic demand would be approximately 300 ml/minute (100 ml/kg/minute). When the  $SaO_2$  is between 75-80%, the  $Q_p:Q_s$  is close to 1 or unity and the cardiac output will be 300 ml/minute. Now consider the same patient with a  $SpO_2$  of 90% and a  $Q_p:Q_s$  of 2.5. The systemic demand remains 300 ml/minute but the cardiac output must be 2.5 times the systemic demand to compensate for the fact that pulmonary blood flow is 2.5 times systemic blood flow. The cardiac output needed is 750 ml/minute which is 2.5 times the systemic demand.

The balanced  $Q_p:Q_s$  of 1 in a mixing lesion is the goal because in the short term, cyanosis is better tolerated than a high  $Q_p:Q_s$  with pulmonary over circulation, ventricular volume overload, increased myocardial work and possible CHF. The following graph illustrates the relationship between systemic oxygen saturation and  $Q_p:Q_s$ . Once the  $SaO_2$  is over the 80%, the curve flattens and further increases in oxygen saturation come at the price of dramatically increased ventricular work load. (Figure 3)

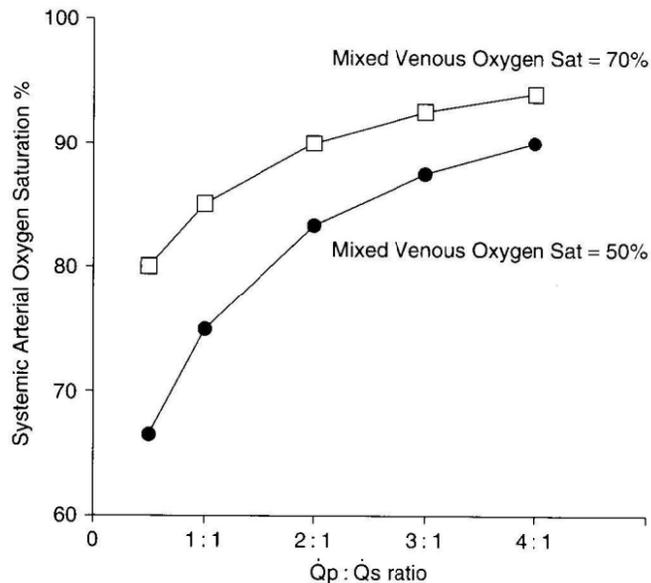


Figure 3: Relationship between systemic oxygen saturation and Qp:Qs in mixing lesions  
*(Congenital Diseases of the Heart: Clinical-Physiological Considerations 2<sup>nd</sup> Ed. Rudolph AM. Reprinted with permission)*

## Hemodynamic Calculations

Cardiac catheterization remains the gold standard for evaluation of hemodynamics. Pulmonary blood flow is referred to as Qp and systemic blood flow as Qs. Qp:Qs is the ratio of pulmonary to systemic blood flow. A normal heart without any shunting has a Qp:Qs of 1 or unity. Calculating Qp:Qs relies on the Fick principle and the oxygen content equation. The patient should be breathing room air for the most accurate data.

Fick equation: O<sub>2</sub> consumption (VO<sub>2</sub>) equals cardiac output (Q) multiplied by the arterio-venous O<sub>2</sub> difference  $VO_2 = Q \times (CaO_2 - CvO_2)$

VO<sub>2</sub> is O<sub>2</sub> consumption in ml/min                      Q is cardiac output in L/min

CaO<sub>2</sub> is O<sub>2</sub> content of arterial blood in ml O<sub>2</sub>/L

CvO<sub>2</sub> is O<sub>2</sub> content of venous blood in ml O<sub>2</sub>/L

Rearranging the equation yields:  $Q = VO_2 / (CaO_2 - CvO_2)$

Equation for CaO<sub>2</sub> or CvO<sub>2</sub> is:  $SaO_2 \times Hgb \times 1.34 + (PaO_2 \times 0.003)$

PaO<sub>2</sub> x 0.003 is disregarded because it is so small and the equation becomes:

$SaO_2 \times Hgb \times 1.34 \times 10$  (multiplying by 10 converts ml O<sub>2</sub>/dl to ml O<sub>2</sub>/L)

Accurate measurement of VO<sub>2</sub> has proven difficult so an assumed number is used based on tables that take into account the variables of the patient's age, weight, heart rate etc. Knowing VO<sub>2</sub> and being able to measure the O<sub>2</sub> saturation at different locations allows the calculation of Qp and Qs independently and therefore the ratio of Qp:Qs. Qp requires

measuring the O<sub>2</sub> saturation in the pulmonary vein and pulmonary artery. Q<sub>s</sub> requires measuring the O<sub>2</sub> saturation in the aorta and the superior vena cava.

$$Q_p = VO_2 / (C_{pv}O_2 - C_{pa}O_2) \quad Q_s = VO_2 / (CaO_2 - CvO_2)$$

C <sub>pv</sub> O <sub>2</sub> = O <sub>2</sub> content in the pulmonary vein	(SpvO <sub>2</sub> x Hgb x 1.34 x 10)
C <sub>pa</sub> O <sub>2</sub> = O <sub>2</sub> content in the pulmonary artery	(SpaO <sub>2</sub> x Hgb x 1.34 x 10)
CaO <sub>2</sub> = O <sub>2</sub> content in the aorta	(SaO <sub>2</sub> x Hgb x 1.34 x 10)
CvO <sub>2</sub> = O <sub>2</sub> content in the SVC	(SvO <sub>2</sub> x Hgb x 1.34 x 10)

With this data, an exact Q<sub>p</sub> and Q<sub>s</sub> are calculated and the ratio of Q<sub>p</sub>:Q<sub>s</sub> is determined. A much quicker method of determining Q<sub>p</sub>:Q<sub>s</sub> uses only the O<sub>2</sub> saturation because the other variables in the equation are constants and cancel each other out

$$Q_p:Q_s = \frac{VO_2}{C_{pv}O_2 - C_{pa}O_2} \quad \text{divided by} \quad \frac{VO_2}{CaO_2 - CvO_2}$$

$$Q_p:Q_s = \frac{CaO_2 - CvO_2}{C_{pv}O_2 - C_{pa}O_2}$$

$$Q_p:Q_s = \frac{Hgb \times 1.34 \times 10 (SaO_2 - SvO_2)}{Hgb \times 1.34 \times 10 (SpvO_2 - SpaO_2)}$$

$$Q_p:Q_s = \frac{SaO_2 - SvO_2}{SpvO_2 - SpaO_2}$$

This method does not determine cardiac output but is more accurate in calculating Q<sub>p</sub>:Q<sub>s</sub> because VO<sub>2</sub> is omitted from the equation. Using VO<sub>2</sub> introduces more error into the Q<sub>p</sub>:Q<sub>s</sub> because VO<sub>2</sub> is derived from tables and not measured.

The other key measurement in the cardiac catheterization lab is resistance.

SVR = $\frac{MAP - CVP}{\text{Systemic cardiac index}}$	PVR = $\frac{\text{Mean PAP} - \text{PA wedge}}{\text{Pulmonary cardiac index}}$
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With shunting, the systemic cardiac output is not equal to the pulmonary blood flow and both must be calculated separately using the Fick equation. In adults the denominator is cardiac output and the final number is multiplied by 80 to yield metric units of dynes-sec-cm<sup>-5</sup>. In pediatrics the 80 is omitted and the denominator is cardiac index. Resistance is quoted in Wood units (mmHg/L/min/m<sup>2</sup>). The normal PVR is 2-3 Wood units and SVR is 10-15 Wood units. Neonatal values are slightly higher for PVR and slightly lower for SVR.

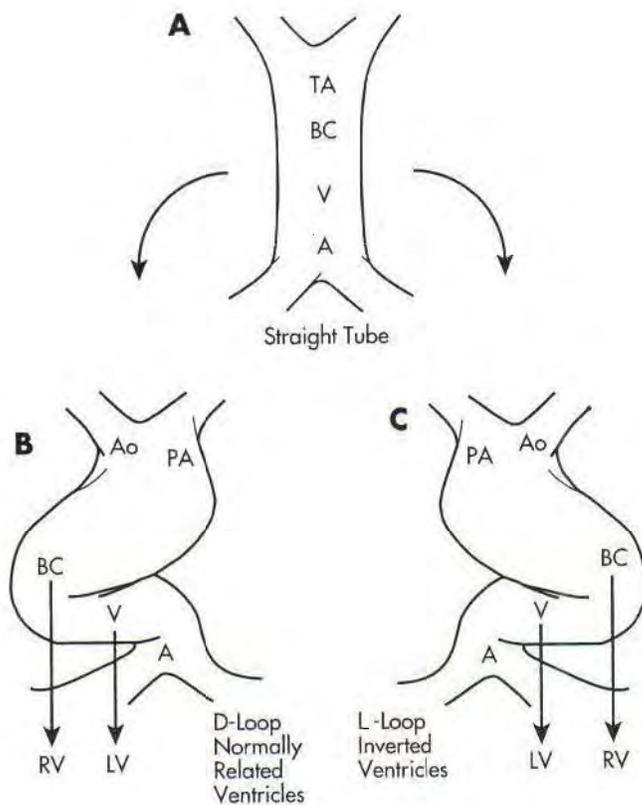
## Echocardiography

The initial investigation for CHD is trans-thoracic echocardiography (TTE) and in many instances no further testing is needed. Trans-esophageal echocardiography (TEE) is routinely used intraoperatively. Conscious sedation used for TEE in adults is usually inadequate for children and consequently anesthesiologists will be asked to provide either general anesthesia or deep sedation when a TEE is required outside the setting of the operating room. Echocardiography will reveal the anatomic lesion and allow assessment of cardiac function and measurement pressure gradients. Gradients are calculated by measuring the velocity of blood flowing between two points. The velocity in meters/sec is converted to a pressure gradient using the modified Bernoulli equation which states the gradient in mmHg is equal to  $4 \times \text{velocity}^2$ .

In aortic stenosis, the blood accelerates across the stenotic valve and if the velocity of that blood is measured at 4 meters/sec the gradient becomes  $4 \times 4^2 = 64$  mmHg. A common practice is to measure the velocity of the tricuspid regurgitation (TR) jet. (Most people have a physiologic amount of TR that can be measured). The velocity reflects the difference in pressure between the right ventricle (driving force) and right atrium (resisting force). If the velocity of the TR jet is 3 meters/sec, the Bernoulli equation calculates the gradient to be  $4 \times 3^2 = 36$  mmHg. Right ventricular systolic pressure is therefore 36 mmHg plus the right atrial pressure. Right atrial pressure can be qualitatively estimated as low, normal or high or CVP is measured from a central line. In the preceding example a TR jet of 3 meters/sec equates to a pressure gradient of 36 mmHg and if the measured CVP is 15 mmHg, the estimated right ventricular systolic pressure is 51 mmHg. The right ventricular systolic pressure is equal to PA systolic pressure if there is no right ventricular outflow tract obstruction or pulmonary stenosis. In this way a simple noninvasive test has been used to assess the presence and degree of pulmonary hypertension. Another example is a coarctation gradient which is estimated by measuring the velocity acceleration across the aortic narrowing. A velocity of 3 meters/sec is a gradient of  $4 \times 3^2 = 36$  mmHg.

## Cardiac Embryology

The heart begins as straight tube loosely divided into segments.



A – primitive atrium V – primitive ventricle BC – bulbus cordis TA – truncus arteriosus

Figure 4: Embryologic development of the heart (*Critical Heart Disease in Infants and Children* Nichols DG, Cameron DE, Greeley WG, Lappe LG, Ungerleider RM, Wetzel RC. Reprinted with permission)

The sinus horns flow into the primitive atrium and develop into the systemic venous return to the heart. Beyond the truncus arteriosus is the aortic sac which develops into the aortic arches. The bulbus cordis initially encompasses the primitive right ventricle, an area called the conus cordis and the truncus arteriosus. The truncus arteriosus divides into the origins of the aorta and pulmonary artery. The conus cordis develops into the right (RVOT) and left ventricular outflow tracts (LVOT). The truncus arteriosus and the conus cordis together are called the cono-truncal region and abnormalities in this region are linked to certain genetic syndromes. The remainder of the bulbus cordis becomes the right ventricle.

The primitive cardiac tube loops and folds on itself. Normal looping is rightward which is called D-looping (D for dextro or right). This results in the morphologic right ventricle being on the right and the morphologic left ventricle being on the left. Abnormal leftward or L-looping (L for levo or left) causes the ventricles to be inverted with the right atrium connected to the morphologic left ventricle on the right side of the heart and the left atrium connected to the morphologic right ventricle on the left side of the heart.

Organogenesis is complete by eight weeks. The guiding principle of cardiac formation is that normal organogenesis leads to the establishment of normal blood flow which then produces normal growth and development. A lesion that disturbs normal blood flow impairs the development of the downstream structures. For example mitral valve atresia causes the distal structures of the left ventricle, aortic valve and ascending aorta to be severely underdeveloped and leads to hypoplastic left heart syndrome.

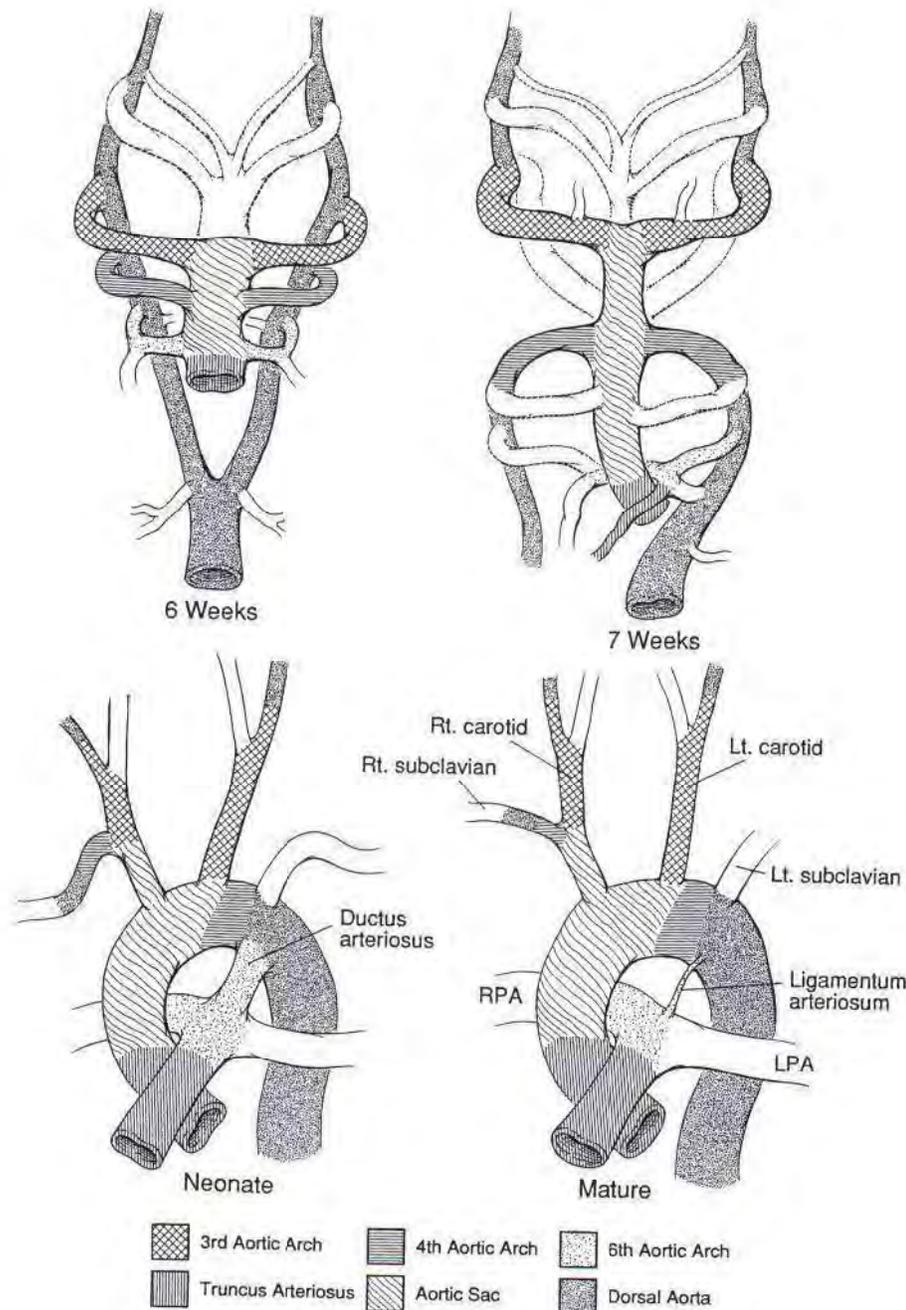


Figure 5: Embryologic development of the aortic arch and pulmonary artery  
*(Critical Heart Disease in Infants and Children. Nichols DG, Cameron DE, Greeley WG, Lappe LG, Ungerleider, RM Wetzel RC. Reprinted with permission)*

## Nomenclature

The name of a cardiac lesion usually describes its anatomic features (ex. atrial septal defect, tricuspid atresia). Abnormal connections are similarly contained in the name (ex. double outlet right ventricle). Transposition strictly refers to the condition where the right ventricle is connected to the aorta and the left ventricle is connected to the pulmonary artery. This is also described as ventriculo-arterial discordance.

There is a three letter system of nomenclature. Familiarity with the system allows one to understand the language of the pediatric cardiologists. The system was devised by Richard Van Praagh and now bears his name. It describes the heart as a series of segmental connections. The three major segmental connections or relationships are:

1. Viscero-atrial
2. Atrio-ventricular
3. Ventriculo-arterial

Viscero-atrial:

This is the relationship of the abdominal viscera to the atria. The normal arrangement is for the liver to be on the right, the stomach and spleen on the left and the right sided IVC flows into the morphologic right atrium. The morphologic right atrium and left atria are distinguished by the differences in their atrial appendages. The term situs refers to position and solitus means normal. A normal anatomic relationship between the abdominal organs and the atria is situs solitus (S) or “normal sidedness”. Situs inversus (I) is the mirror image arrangement (left sided liver, right sided stomach and spleen) and situs ambiguous (A) is the condition referred to as heterotaxy in which abdominal organ position is very abnormal and variable. Thus the first letter in the Van Praagh system is either S, I or A.

Atrio-ventricular:

This is the relationship of the atria to the ventricles. As previously explained, the relationship is either D-loop (D) or L-loop (L). In D-looping, the right atrium connects to the right ventricle and the left atrium connects to the left ventricle. With L-looped ventricles, there is atrio-ventricular discordance. The right atrium is connected to the left ventricle and the left atrium is connected to the right ventricle. A point to remember is that in L-looped ventricles, the valves associate with their normal ventricular attachment. For example, the right atrium connects to the left ventricle across the mitral valve and the left atrium connects to the right ventricle across the tricuspid valve. The second letter in the Van Praagh system is either D or L.

Great arteries:

This is the relationship of the great arteries to each other. The normal relationship has the aorta posterior and to the right of the pulmonary artery which is situs solitus (S) of the great vessels. The other common abnormal positions for the aorta in relation to the pulmonary artery is anterior and to the right (D) or to the left (L). The third letter in the Van Praagh system is S, D or L.

The Van Praagh system for a normal heart would be situs solitus of the viscero-atrial relationship, D-looped ventricles and situs solitus of the great arteries which is S, D, S.

## **Anesthetic Drugs in Congenital Heart Disease**

There is a dearth of good data on anesthetic effects in CHD. What follows is brief summary with references to relevant papers. How do anesthetics alter the balance between SVR and PVR, thereby changing the nature and direction of shunt flow? How do anesthetics affect the key determinants of cardiac output which are preload, afterload, heart rate, contractility and shunt flow?

### **Propofol**

Propofol is commonly used for the induction. For procedural sedation it is frequently administered via infusion to a spontaneously breathing child with a natural airway. This popular technique avoids airway instrumentation. The rapid clearance of propofol enables a prompt, smooth awakening which is usually free from nausea, vomiting and emergence agitation. Children can tolerate the respiratory depressant effects of propofol far better than adults. Infusion rates in excess of 200 mcg/kg/min may be required. In healthy children without CHD, propofol causes a decrease in blood pressure of 30% due to decreases in SVR (15%) and heart rate (10-20%).<sup>1</sup> Data in children with CHD demonstrates propofol's primary effect is a drop in blood pressure through a decrease in SVR.<sup>2</sup> Systemic cardiac output increased without a change in heart rate or PVR. In children without shunt there was a small decrease in PaO<sub>2</sub> presumably due to decreased respiratory drive but no increase in PVR. Importantly, in children with shunts the decrease in SVR was consequential. Left-to-right shunting decreased and right-to-left shunting increased. The effects are dose dependent with usual infusions rates between 100-200 mcg/kg/min in most studies. While most children tolerate propofol well, it should be used with caution as the sole agent in the setting of right-to-left shunts and particularly in those for whom a decrease in systemic afterload is dangerous (aortic stenosis, hypertrophic cardiomyopathy, severe ventricular dysfunction).

### **Ketamine**

Ketamine combines anesthesia, analgesia, cardiovascular stability and lack of respiratory depression with maintenance of airway reflexes. Drawbacks to ketamine are prolonged action, emergence phenomena and a dissociative anesthetic state that may result in patient movement. Anesthetic doses of ketamine (50-75 mcg/kg/min) are much greater than analgesic doses (5-10 mcg/kg/min). In children with CHD, ketamine (50-75 mcg/kg/min) resulted in the maintenance of the relationship between SVR and PVR. Systemic blood pressure increased through an increase in cardiac output with little change in heart rate.<sup>3</sup> Increased inotropy may be beneficial for children with significant ventricular dysfunction. A common question is whether ketamine's sympathomimetic action raises PVR in children with pulmonary hypertension (PHTN). Evidence suggests that in the setting of normocarbia with supplemental oxygen administered, PVR is not increased.<sup>4</sup> Combined with 0.5 minimal alveolar concentration (MAC) sevoflurane in spontaneously breathing children with severe PHTN, ketamine did not raise PVR and overall hemodynamics were stable.<sup>5</sup>

## **Etomidate**

Etomidate is little studied in children with or without CHD. Bolus dosing of 0.3 mg/kg was well tolerated with maintenance of systemic blood pressure and preservation of the balance between SVR and PVR.<sup>6</sup> Transient adrenal suppression does occur even with bolus dosing and could be consequential in the sickest children. Other drawbacks of etomidate are pain on injection and emesis.

## **Volatile anesthetics**

Halothane is no longer available in the USA and will not be addressed here other than to state that at equivalent MAC levels to isoflurane and sevoflurane, there is greater myocardial depression and suppression of the baroreceptor mediated increase in heart rate. The Qp:Qs ratio is unchanged with halothane, sevoflurane or isoflurane if ventilation is controlled even with a high FiO<sub>2</sub>.<sup>7</sup> Cardiac output with isoflurane was maintained even at 1.5 MAC as the decrease in SVR and increase in heart rate offset reduced inotropy. Sevoflurane was similar but an overall decrease in cardiac output resulted from the lack of a compensatory increase in heart rate combined with reduced inotropy. Children with significant ventricular dysfunction may not hemodynamically tolerate MAC levels of volatile anesthesia.

## **Fentanyl/Midazolam**

Combining a synthetic opioid with a short acting benzodiazepine has long been favored by pediatric cardiac anesthesiologists. When the combination is the only anesthetic or sedative, its duration of action usually limits its use to sick children who will remain intubated at the conclusion of the procedure. The ratio of Qp:Qs is unchanged as long as ventilation is controlled.<sup>7</sup> Fentanyl at doses of 25 mcg/kg maintains SVR, PVR and systemic blood pressure.<sup>8</sup> The caveat here is that significant bradycardia is avoided. Therefore the general tone of the sympathetic nervous system greatly influences the blood pressure and patients can easily become hypotensive due to changes in the loading conditions of the heart while inotropy is preserved. Particularly sympatholytic is the combination of synthetic opioids with volatile anesthetics, midazolam or propofol.

## **Dexmedetomidine**

Dexmedetomidine is a selective  $\alpha$ -2 agonist with an emerging role pediatric anesthesia.<sup>9</sup> It is an analgesic and sedative with minimal respiratory depression. The sedation of dexmedetomidine has been likened to a natural sleep state. Decreased sympathetic outflow is usually marked by relative bradycardia and stable blood pressure. However, side effects detrimental to children with CHD are hypertension (peripheral  $\alpha$ -2 agonist effect), bradycardia and hypotension. The pros and cons of dexmedetomidine for CHD surgery has recently been debated.<sup>10,11</sup> Dexmedetomidine may be poorly tolerated in heart rate dependent neonates and infants. Its lack of respiratory depression is an attractive feature for ICU sedation in children at risk for obstructive sleep apnea such as Down syndrome.

## Atrial Septal Defect

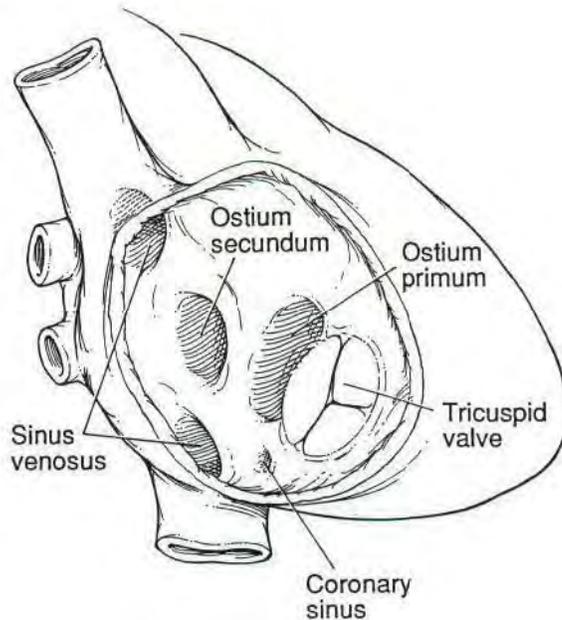


Figure 6: Types of atrial septal defects (*Critical Heart Disease in Infants and Children*. Nichols DG, Cameron DE, Greeley WG, Lappe LG, Ungerleider RM, Wetzel RC. Reprinted with permission)

As an isolated condition an atrial septal defect (ASD) is usually asymptomatic, with the lesion discovered incidentally or the murmur of a widely split P2 (right ventricular volume overload causes the pulmonary valve to close after the aortic valve) may be heard. An untreated ASD (usual  $Q_p:Q_s > 2$ ) causes right sided volume overload (right atrial and right ventricular dilation) with increased pulmonary blood flow eventually leading to pulmonary hypertension, symptoms of CHF and increased PVR. The time frame for the development of significant pulmonary hypertension with an isolated ASD is usually many decades. Repair is generally done between 3-5 years of age, either by percutaneous device closure in the cardiac catheterization laboratory or surgery. Secundum ASDs are the only type that can close spontaneously and smaller lesions of this type have a high rate of closure over time.

Isolated ASDs, even if large are generally well tolerated, and anesthetic induction and maintenance are usually uneventful. Lesions with left-to-right shunting are best managed by optimizing the balance between SVR and PVR with the goal of decreasing the  $Q_p:Q_s$ . Since blood flow is left-to-right across the defect, the  $Q_p:Q_s$  is increased by factors that increase SVR (raise the “driving force” of the shunt) or decrease PVR (lower the “resisting force” of the shunt). The most important variable raising SVR is systemic hypertension and the most critical variables for PVR are arterial carbon dioxide ( $PaCO_2$ ) and the  $FiO_2$ . Hyperventilation with  $FiO_2$  1.0 can dramatically decrease PVR with a correspondingly large increase in pulmonary blood flow in the setting of a left-to-right shunt. These comments are generic to all left-to-right shunt lesions but actually less relevant to an ASD. The reason is that the driving and resisting forces governing shunt flow in an ASD are left atrial and right atrial pressure respectively. These atrial pressures

tend to fluctuate within a relatively narrow range despite large changes in SVR or PVR which primarily affect the ventricles. For ASD repair, either surgical or percutaneous, TEE is used during the procedure intraoperatively to assess the repair and confirm there is no residual ASD.

Sinus venosus defects (2-5% of all ASD) are rare and often associated with right sided partial anomalous pulmonary venous return (PAPVR) to the right atrium. Therefore the total volume of the left-to-right shunt is greater than a lone ASD. Repair of sinus venosus defects is surgical and involves closing the ASD with a patch placed in such a way that the anomalous pulmonary venous return is baffled across the defect into the left atrium. If there is PAPVR it is important to confirm the patency of pulmonary venous return with TEE after separation from cardiopulmonary bypass (CPB).

Primum defects involve failure of the septum primum to merge with the endocardial cushions. There is a high association of a cleft in the anterior leaflet of the mitral valve with resulting mitral regurgitation. The volume overload from mitral regurgitation increases left atrial pressure and increases the degree of left-to-right shunting. Primum ASDs (5-7% of all ASDs) are endocardial cushion defects and therefore are very often part of an atrio-ventricular canal lesion. Repair of primum ASD is surgical, at which time the anterior mitral leaflet cleft is also repaired. Repair of a cleft mitral valve is usually not technically difficult but needs to be confirmed with TEE after separation from CPB. Damage to the conduction system may occur with primum ASD repair and heart block may ensue. Heart block unfortunately could be permanent but may also be transient due to myocardial edema in the region of the conduction system.

The ostium secundum is an opening that forms in the septum primum and should eventually be covered by the septum secundum. Secundum ASDs (85% of all ASDs) that have an adequate rim of surrounding septal tissue may be closed using a device placed percutaneously in the cardiac catheterization laboratory. Very large defects or those without a sufficient amount of circumferential tissue are closed surgically. Device closure of an ASD is now a common procedure associated with minimal serious complications and remarkably, discharge from hospital the next day. Hemodynamic instability during the procedure is rare. General anesthesia is required because TEE is used to guide placement of the device.

## Ventricular Septal Defect

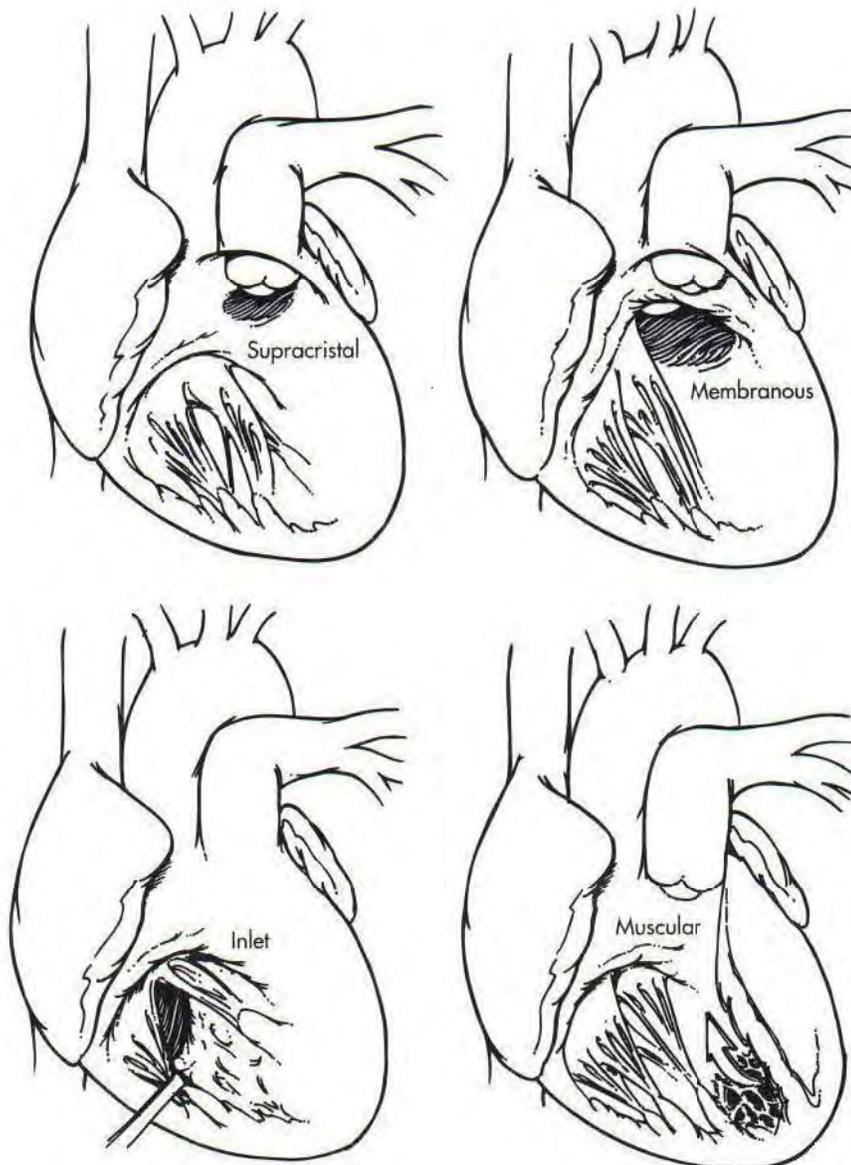


Figure 7: Types of ventricular septal defects (*Critical Heart Disease in Infants and Children. Nichols DG, Cameron DE, Greeley WG, Lappe LG, Ungerleider RM, Wetzel RC. Reprinted with permission*)

Types of ventricular septal defects (VSD):

1. Muscular (single or multiple)
2. Perimembranous
3. Inlet (beneath the septal leaflet of the tricuspid valve)
4. Supracristal
5. Malalignment (the infundibular septum dividing the outflow tracts is malaligned in addition to having a septal defect)

In the absence of other complicating lesions, the pathophysiology of a VSD is pulmonary over circulation due to the left-to-right shunt. Large VSDs eventually cause pulmonary hypertension followed by increases in PVR. Histological changes in the pulmonary vasculature ensue with reduced ability to respond to pulmonary vasodilators. The shunt direction then reverses and this condition is referred to as Eisenmenger's Syndrome. In developed countries, an undiagnosed VSD progressing to Eisenmenger's Syndrome is extremely rare. A VSD usually causes little right ventricular volume overload as blood shunted from left-to-right is ejected in systole and goes directly out the right ventricular outflow tract (RVOT) into the pulmonary circulation. However, there is left ventricular volume overload because of the amount of blood that rapidly recirculates through the lungs. For instance, a Qp:Qs of 3:1 means that the left heart must pump three times the normal volume of blood in order to meet the usual systemic demands.

A VSD is termed restrictive if their small size limits pulmonary over circulation. Small muscular or perimembranous VSDs have a high rate of closure over time. The flow across a large unrestrictive VSD is determined by the balance between SVR and PVR. As PVR falls in the first months of life the flow across the VSD can increase greatly. The Qp:Qs may be extremely high (over 3:1). An isolated large VSD is managed medically with diuretics in the first few months of life and then repaired surgically later in infancy. Without surgery, standard diuretic therapy will eventually fail and the patient will suffer progressive CHF, characterized by poor feeding, reduced weight gain and increased incidence of lower respiratory infections. The progression to Eisenmenger's Syndrome is measured in years for a VSD rather than the decades expected for an ASD. An ASD results in simply increased pulmonary blood flow. However, a VSD causes increased blood flow at an increased blood velocity. Recall that the "driving force" for left-to-right flow across a VSD is left ventricular systolic pressure which is the same as systemic systolic blood pressure. The high velocity pulmonary blood flow results in greater shear forces that cause accelerated damage to the vasculature. Anesthetic principles for VSD management are simply those for a left-to-right shunt and are covered in the section on ASD. One important caveat is that the degree of shunting with a VSD is generally much larger than with an ASD. Consequently, management of CHF may require aggressive diuresis to the point that the infant is intravascularly volume depleted and may become hemodynamically compromised with the induction of anesthesia and initiation of positive pressure ventilation. A clue to the intensity of diuresis preoperatively is the serum bicarbonate. (Furosemide causes a metabolic alkalosis). Standard treatment of VSD remains surgical but muscular defects may be closed percutaneously in the cardiac catheterization laboratory.

Inlet VSDs are most often associated with AV canal defects. Malalignment VSDs occur in context of other cardiac lesions. In addition to the VSD, there is malalignment of the infundibular septum which divides the ventricular outflow tracts. Anterior malalignment leads to RVOT obstruction and posterior malalignment leads to left ventricular outflow tract (LVOT) obstruction. The respective outflow tract stenosis must be addressed in addition to closing the VSD to prevent obstruction to blood flow. The classic example of a posterior malalignment VSD is Tetralogy of Fallot (TOF).

Isolated muscular or perimembranous VSDs are approached surgically via the right atrium. Avoiding a right ventriculotomy greatly decreases the chances of right ventricular

dysfunction and arrhythmia, both postoperatively and in the long term. The conduction system runs along the ventricular septum and is at risk during perimembranous VSD repair. Patients should be observed for conduction disturbances and TEE is needed to rule out residual VSD after separation from CPB.

Closure of a VSD with a percutaneous device is an exciting new therapy. In the United States, the Food & Drug Administration (FDA) has restricted use of these devices to muscular VSDs. Given that most muscular VSDs close spontaneously, device closure is not a common procedure. Anesthetic induction and maintenance principles are those for any significant left-to-right shunt. Specific to the procedure there can be arrhythmia, transient heart block, hemodynamic disturbance secondary to right ventricular obstruction from the deployment catheter, right ventricular perforation and device embolization.

The FDA has withheld approval of perimembranous VSD closure devices because of a significant incidence of complete heart block. The worrisome feature is that heart block can occur well after successful and uncomplicated device deployment. Presumably an area of inflammation and subsequent fibrosis develops around the perimembranous VSD closure device which is in close proximity to the conduction system. Device closure of a perimembranous VSD is a much more technically demanding procedure often marked by hemodynamic instability. Similar to a muscular VSD, the anesthetic principles are those of a significant left-to-right shunt. All the procedural complications of a muscular VSD device closure apply but in addition the subvalvular structures of the tricuspid and mitral valves can be damaged during deployment. The tricuspid valve is at risk of functional stenosis due to the deployment catheter. Additionally, the left ventricular aspect of a perimembranous VSD is immediately below the aortic valve. Deployment of the device may impinge on the aortic root and cause sudden aortic insufficiency.

## **Patent Ductus Arteriosus**

A patent ductus arteriosus (PDA) is a common lesion, especially in premature infants with respiratory disease (meconium aspiration, respiratory distress syndrome). Other conditions which result in hypoxia, hypercarbia, acidosis or primary pulmonary hypertension of the newborn can result in a PDA. The physiology governing closure of the PDA is discussed in the section on Transitional Circulation.

A PDA is a left-to-right shunt causing pulmonary over circulation and congestive heart failure. There is volume overload of the left side of the heart because of the amount of blood recirculated through the lungs. Left-to-right shunting is abundant because flow through the PDA is continuous through systole and diastole. Diastolic run off into the pulmonary circulation lowers diastolic BP compromising distal perfusion, particularly mesenteric/renal and lowering coronary perfusion to the left ventricle.

Initial management in neonates is medical, using indomethacin to induce ductal closure through its anti-prostaglandin activity. Surgical closure is via left thoracotomy, taking care to preserve the left recurrent laryngeal nerve which loops around the underside of the aortic arch in close proximity to the PDA. It is recommended to have an indicator of distal perfusion during PDA ligation. This can be a lower extremity arterial line or more commonly a pulse oximeter on the foot. When a trial clip is placed on the PDA, loss of

distal perfusion indicates that the clip was actually placed on the aorta. Although this may seem implausible, a large PDA can be difficult to distinguish from the descending aorta. Ligation of a PDA is very common operation in severely premature infants. Those rare patients presenting later in childhood require a thorough assessment to assess the severity of pulmonary hypertension. Infants and older children are candidates for percutaneous coil occlusion of a PDA.

## **Complete Atrio-Ventricular Canal**

Complete atrio-ventricular canal (CAVC) is an endocardial cushion defect with free communication between all four chambers of the heart. There is a primum ASD, an inlet VSD and a common atrio-ventricular (AV) valve. The common AV valve may be regurgitant due to abnormal valve tissue. Transitional AV Canal refers to a lesion in which the VSD component is relatively minor because it is filled with sub-valvular tissue from the common AV valve. The majority of children with CAVC have Down syndrome.

Free communication between all four cardiac chambers results in a very large left-to-right shunt. Shunting is minimized by the high PVR of the newborn period but as PVR falls, the rising Qp:Qs will lead to CHF and failure to thrive. Surgery is undertaken at this point (usually less than four months of age) before the high Qp:Qs leads to pulmonary vascular changes which can develop early in CAVC. For unknown reasons this is especially true in infants with Down syndrome. Close attention to the anesthetic principles of left-to-right shunt physiology is needed. Similar to a large VSD, aggressive diuresis to prevent CHF may result in intravascular volume contraction and marked hypotension on induction of anesthesia.

Surgery is technically challenging and involves:

- closure of the primum ASD
- closure of the inlet VSD
- creation of a competent mitral and tricuspid valve from the common AV valve

TEE is done after separation from CPB to rule out:

- residual ASD
- residual VSD
- AV valve regurgitation

Significant problems with residual septal defects or AV valve regurgitation require returning to CPB to attempt the repair again. The conduction system can be damaged in the repair leading to heart block. High pulmonary blood flow prior to repair can result in increased postoperative pulmonary vascular reactivity. This is more likely if the child presented late with significantly elevated Qp:Qs. Late presenting children may require cardiac catheterization to fully assess hemodynamics and calculate the PVR prior to surgery.

## Coarctation

Coarctation of the aorta is a common congenital heart defect. The ductus inserts into the aorta at the junction of the distal arch and descending thoracic aorta in the region of the left subclavian artery. Coarctation occurs because of excessive growth of tissue from the ductus arteriosus into the aorta. A frequent association is a bicuspid aortic valve.

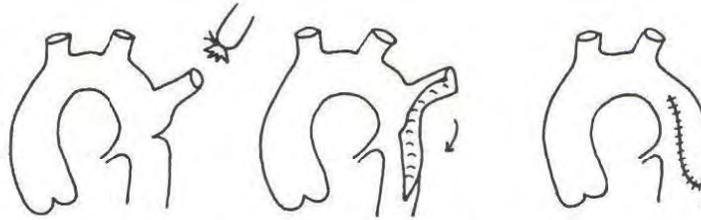
Neonates with critical coarctation present with circulatory collapse, shock and acidosis due to poor distal perfusion and mesenteric ischemia. Resuscitation requires volume, bicarbonate and PGE1 to restore ductal patency. Distal systemic perfusion is now ductal dependent. The neonate will be maintained on PGE1 until surgery. Surgery is scheduled once the neonate is hemodynamically stable and the metabolic derangements of the shock state have resolved.

The surgical approach is via left thoracotomy. Various operations have been tried to try and reduce the incidence of recoarctation. Most commonly the coarctation segment is resected and the aorta repaired with an oblique end-to-end anastomosis. The left subclavian artery may be variously included or sacrificed. The diagram on the following page illustrates various coarctation repair techniques. During infancy, the left subclavian artery can be safely sacrificed because of collateral perfusion to the arm will rapidly develop. However, the peripheral pulses will likely always have a reduced volume. This should be kept in mind when considering blood pressure measurement or arterial placement in a patient with a prior coarctation repair.

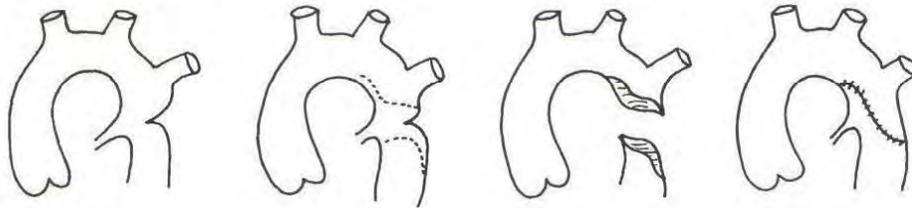
Blood pressure must be measured in the right arm during the period of aortic cross clamp because the clamp will be proximal to the left subclavian artery. The blood pressure in the right arm will reflect cerebral perfusion. The pliable cardiovascular system of newborns and infants copes well with the hemodynamic stress of such a proximal aortic cross clamp. If necessary, the rise in blood pressure during aortic clamping is easily treated by deepening anesthetic depth. A blood pressure difference between upper and lower extremities may persist for hours after surgical repair. Post operative hypertension is frequently a problem and requires potent vasodilators to reduce blood pressure and protect the aortic suture line. The period of aortic clamp is usually brief (less than 20 minutes) and distal organ dysfunction (spinal cord, gut, renal) is rare. Neonates are often left intubated and sedated in the immediate postoperative period to aid in the tight blood pressure control required after coarctation repair.

Coarctation presents in childhood with upper extremity hypertension, decreased femoral pulses, left ventricular hypertrophy on ECG or in advanced cases with CHF. The initial treatment is usually with balloon dilation in the cardiac catheterization laboratory. Balloon dilation is usually accompanied by stenting. Balloon dilation is also the preferred method for treating re-coarctation after surgical repair. Those presenting in childhood will often have hypertension which persists after surgery since the main risk factor for long term hypertension is the duration of uncorrected coarctation.

Left Subclavian Flap Angioplasty



End-to-End repair with resection of coarctation segment



Left Subclavian Translocation Angioplasty

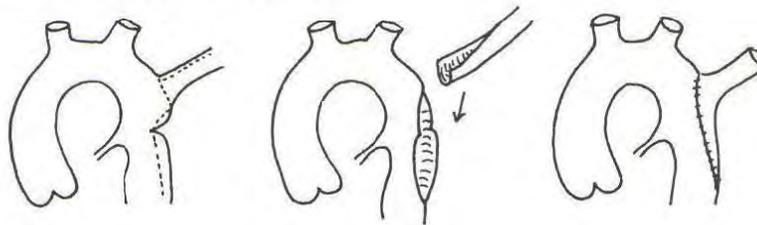
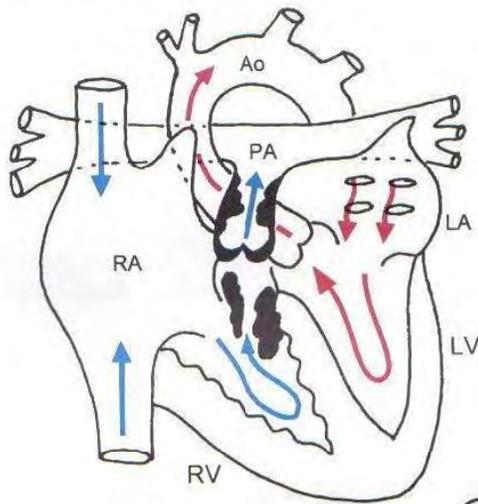


Figure 8: Surgical repair of coarctation (*Pediatric Heart Surgery, A Ready Reference for Professionals, 3<sup>rd</sup> Ed. May, LE. Reprinted with permission*)

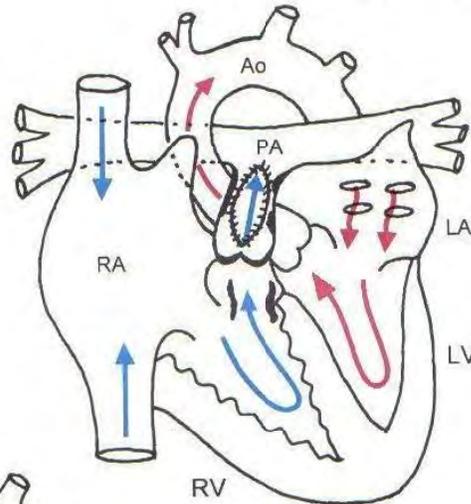
## Pulmonary Stenosis

Pulmonary stenosis (PS) is a common congenital heart defect. Usually it is part of other complex lesions. Isolated PS presents with a murmur. The degree of symptoms depends of the severity of the obstruction. Severe disease leads to right ventricular hypertrophy. If right sided pressures are high and a PFO/ASD is present, there may be a right-to-left shunting and cyanosis. The obstruction is normally valvar but can be can be subvalvar or supra-valvar. Initial treatment is usually balloon dilation of the lesion in the cardiac catheterization laboratory. Right ventricular pressures are measured before and after the balloon dilation. The goal is usually to reduce to the right ventricular pressures to less than 50% systemic. Operative treatment is reserved for severe disease or situations in which balloon dilation is unsuccessful. Ideally the repair preserves the integrity of pulmonary valve but if a trans-annular patch is required, free pulmonary insufficiency results. (See Tetralogy of Fallot section for discussion on long term consequences of free pulmonary insufficiency)

Pulmonary Stenosis -  
Valvar, Supravalvar, and  
Infundibular



Repair with Pulmonary  
Valve Annulus Intact



Repair with Transannular Patch

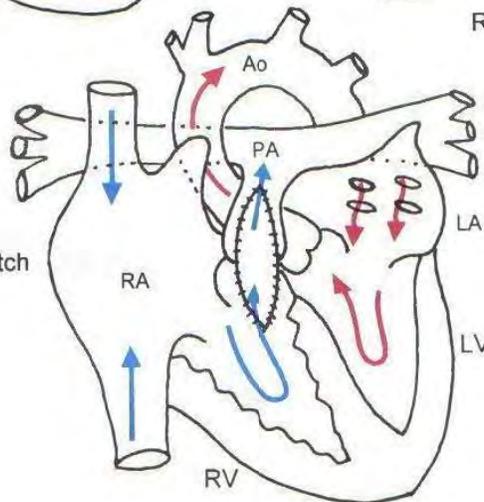


Figure 9: Surgical repair of pulmonary stenosis (*Pediatric Heart Surgery, A Ready Reference for Professionals, 3<sup>rd</sup> Ed. May, LE. Reprinted with permission*)

## Aortic Stenosis

The pathophysiology of aortic stenosis (AS) is the same in children as adults. The usual principles of hemodynamic management apply (maintenance of preload, age appropriate heart rate, maintenance of afterload to preserve coronary perfusion pressure, preservation of contractility). Inhalation induction without intravenous access creates the situation of deep anesthesia while lacking the ability to administer medications or a volume bolus and is generally not recommended. Intravenous access, facilitated by local anesthetic cream if necessary, should be established prior to induction which can then be either inhalational or intravenous. Any combination of intravenous drugs or a slow inhalational induction can be successfully used keeping in mind the principles of aortic stenosis. What is vastly more important is how the induction is achieved rather than what drugs are used. In the child with very difficult intravenous access, another option is intramuscular ketamine to

establish a hemodynamically stable plane of anesthesia, followed by placement of an intravenous catheter. In the neonatal period the echocardiogram should be carefully reviewed because the qualitative description of the echocardiographer is the best assessment of left ventricular function. Extrapolating from valve areas and gradients used in adult patients will not work. The gradient of severe AS in a newborn has to be appreciated in the context that a normal blood pressure would be 70/40. Furthermore, in utero severe AS may have impaired left ventricular development, causing varying degrees of hypoplasia. In this scenario, severe AS may occur with gradients that appear quite benign.

If the left ventricle is severely hypoplastic it cannot function as the systemic ventricle and single ventricle surgical palliation is required. When left ventricular function is adequate, initial treatment with balloon dilation can be performed in the cardiac catheterization laboratory. This is not a simple and benign procedure, especially in the child with severe disease. The stressed left ventricle may be quite susceptible to arrhythmia from catheter manipulation. If the arrhythmia does not promptly resolve by withdrawing the catheter, immediate cardioversion or defibrillation is required. Resuscitation in the setting of severe aortic stenosis is extremely high risk. The most worrisome complication of balloon dilation is sudden, severe aortic insufficiency (AI). If severe AI results, the patient will almost certainly go into pulmonary edema because of the sudden volume load on a previously pressure loaded ventricle. Leaving the child intubated would be prudent for the management of pulmonary edema while various treatment options are considered. Because the consequences of severe AI are so damaging, pediatric interventional cardiologists will err on the side of caution and opt to leave the patient with some residual AS rather than repeatedly attempting balloon valvuloplasty at the risk of causing severe AI. There are three surgical options for AS, either as the initial treatment or after balloon valvuloplasty failure. Depending on valvular anatomy and surgical experience, valve repair may be considered. As in adults, valve replacement is an option but has significant problems in childhood. Bioprosthetic valves will need multiple revisions over the life of the patient and are thus rarely used. Mechanical valves will last longer but will still require re-operation. Mechanical valves require anticoagulation with warfarin which is difficult for children because of the need for monitoring with INR. Despite anticoagulation there is still a risk of thrombosis and anticoagulation itself carries an inherent risk of bleeding.

A unique procedure designed to get around these problems is the Ross operation. The stenotic aortic valve is resected en bloc with the aortic root and the patient's own pulmonary valve root is excised and implanted in the aortic position. The coronary arteries are then re-implanted into the "neo-aortic" root. A right ventricle to pulmonary artery (RV-PA) connection provides pulmonary blood flow. The RV-PA connection is made with homograft (cadaveric tissue) or a conduit (synthetic material). A preoperative echocardiographic assessment of the pulmonary valve is vital to ensure that it is anatomically normal and appropriately sized to become the new aortic valve. The RV-PA connection may require revision (surgically or percutaneously) in the future but the Ross operation may provide a better long term solution to aortic valve disease than valve replacement. The use of the Ross operation is quite specific to the institution with some centers being "believers" and others less convinced.

The Ross-Konno procedure is done when there is significant obstruction in the LVOT, in addition to valvular aortic stenosis. The Konno modification involves extending an incision through the ventricular septum. The LVOT is then enlarged with a patch and the patient's own pulmonary valve becomes the neo-aortic valve as in the standard Ross operation.

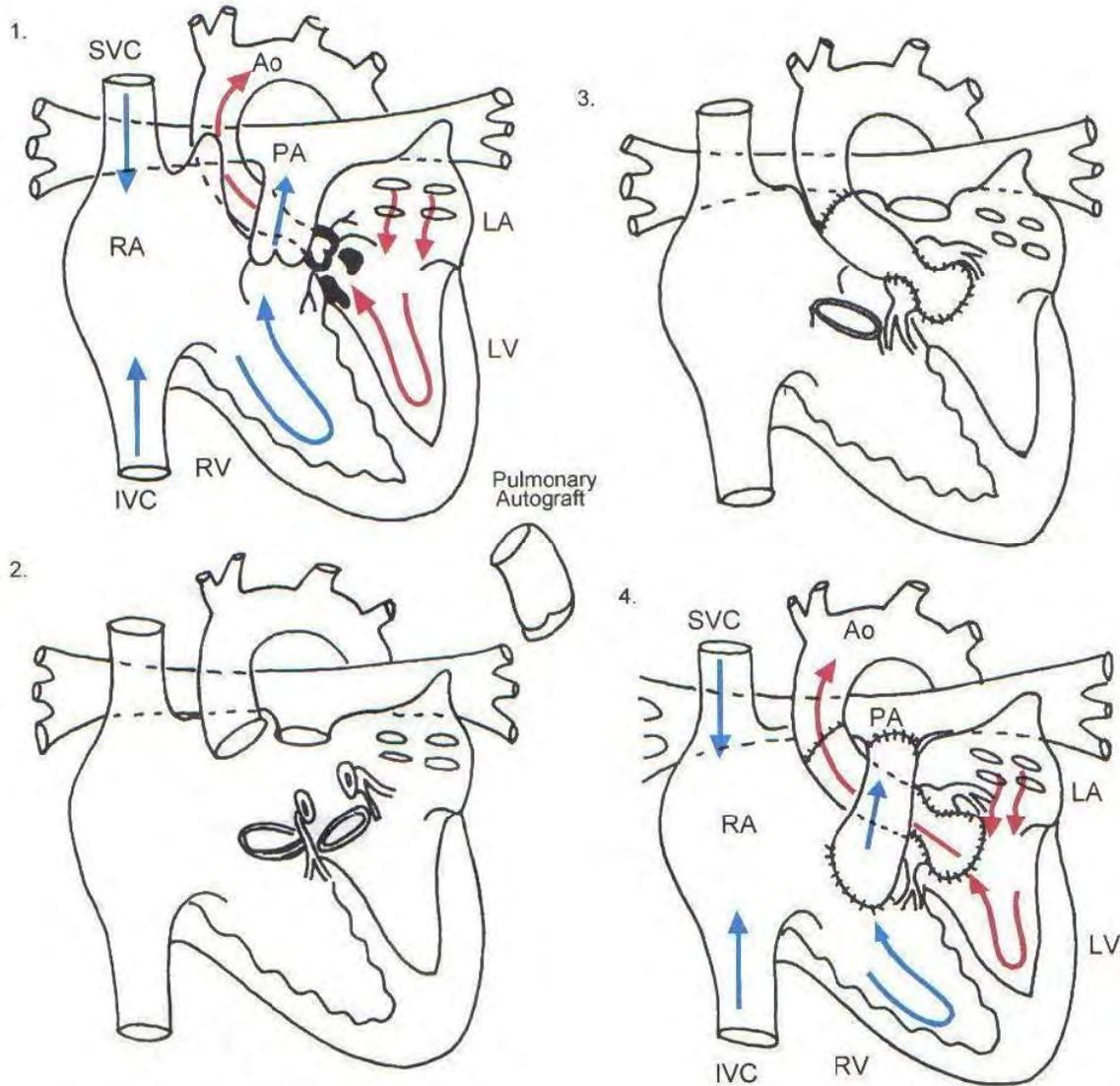


Figure 10: Ross operation for aortic stenosis (*Pediatric Heart Surgery, A Ready Reference for Professionals, 3<sup>rd</sup> Ed. May, LE. Reprinted with permission*)

## Interrupted Aortic Arch

Interrupted aortic arch (IAA) exists when there is a complete defect or interruption in the aortic arch. Classification of IAA depends on the site of the interruption.

IAA-A: distal to the left subclavian artery (25%)

IAA-B: between the left subclavian artery and the left common carotid artery (70%)

IAA-C: between the left common carotid artery and the innominate artery (5%)

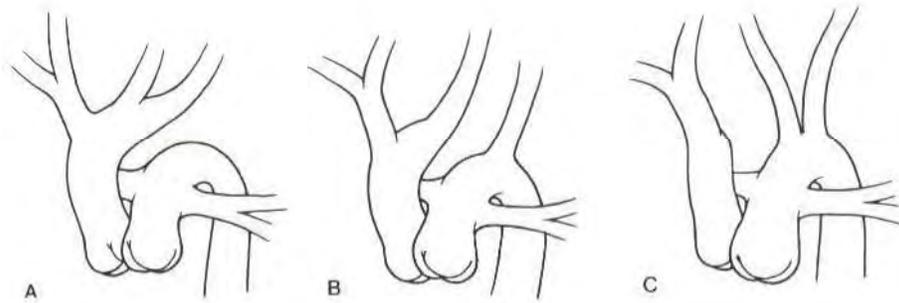


Figure 11: Classification of interrupted aortic arch (*Pediatric Cardiac Intensive Care* Chang AC, Hanley FL, Wernovsky G, Wessel DL. Reprinted with permission)

Perfusion beyond the interruption is through the PDA which merges into the descending thoracic aorta. There is a high incidence of other cardiac lesions with IAA. The most common type of IAA is Type B which is almost always associated with a VSD. Type A-IAA is clinically equivalent to a critical coarctation presenting in the newborn period.

Stabilizing the patient requires PGE1 to maintain ductal patency. Closure of the ductus arteriosus causes severe distal hypoperfusion with lactic acidosis and shock. The diagnosis is suspected when the SpO<sub>2</sub> is greater in the right hand compared with other extremities. Proximal to the interruption, the SpO<sub>2</sub> will be 100% reflecting the fully oxygenated blood ejected from the left ventricle. Distal to the interruption, the SpO<sub>2</sub> will be reduced. Perfusion distal to the interruption is from the right ventricle via the PDA. Right ventricular output is mainly deoxygenated blood mixed with some fully oxygenated blood from the left ventricle (left-to-right shunt across the VSD).

IAA-A: upper extremity SpO<sub>2</sub> > lower extremity SpO<sub>2</sub>

IAA-B: right hand SpO<sub>2</sub> > left hand & lower extremity SpO<sub>2</sub>

IAA-C: right hand SpO<sub>2</sub> > left hand & lower extremity SpO<sub>2</sub>

Surgery for IAA is done in the newborn period with deep hypothermia and circulatory arrest. This results in a bloodless field to repair the aortic arch. Some surgeons prefer to try and perfuse the brain selectively during circulatory arrest to the body while others use total circulatory arrest. The aortic arch defect is repaired using a patch of homograft. The PDA is ligated and the VSD is closed. After the repair, the differential diagnosis for oliguria must include significant stenosis at the site of the arch repair along with the other usual causes. Blood pressure and oxygen saturation should be measured in the right upper extremity and either of the lower extremities. Long term complications of surgery are

iatrogenic coarctation of the aortic arch which may be amenable to percutaneous balloon dilation without or without stenting.

## **Blalock-Taussig Shunt**

The Blalock-Taussig (BT) shunt is an operation and not a congenital heart defect but it is included here because it is the initial surgical palliation for certain types of cyanotic lesions. It also occupies a pioneering place in the history of surgery for congenital heart disease.

Dr. Helen Taussig, a pediatric cardiologist at Johns Hopkins, treated many patients with cyanotic CHD. The most common diagnosis was Tetralogy of Fallot (TOF). She observed the clinical deterioration of patients when the ductus arteriosus would close which confirmed the pathophysiology of insufficient pulmonary blood flow. This observation led to the idea of creating an “artificial ductus”. She presented the idea to Dr. Robert Gross at Children’s Hospital Boston because he had done the first surgical ligation of a PDA in 1938. When he proved reluctant, she discussed her idea with the chief of surgery at Johns Hopkins, Dr. Alfred Blalock. By fortunate coincidence, Dr. Blalock’s earlier research involved creating an animal model of pulmonary hypertension. The method he used to increase pulmonary blood flow to achieve pulmonary hypertension was the creation of a subclavian artery to pulmonary artery anastomosis. Dr. Blalock’s surgical assistant, Vivien Thomas, devised the instruments needed to perform the operation and perfected it on animals before teaching it to Dr. Blalock. Vivien Thomas, an African-American without a college education was Dr. Blalock’s indispensable assistant for 34 years. His contribution to the BT shunt was vital and retrospectively he has been acknowledged as a surgical pioneer in his own right.

On November 29, 1944 the first BT shunt was done on a 15 month old patient with TOF who weighed only 9 pounds and was deteriorating from progressive cyanosis. The success of the operation was immediately apparent, ushering in a new era in CHD. A year later the patient’s shunt stopped working and Dr. Blalock attempted another BT shunt on the contralateral side. The patient briefly improved but died soon after. Word of the successful BT shunt quickly spread and the operation became standard treatment for TOF and other cyanotic CHD lesions. There was excellent longitudinal follow up of the early BT shunt patients showing that many survived and went on to have families of their own. In others, the BT shunt provided crucial palliation prior to complete repair later in life when they were better able to withstand the stress of surgery.

### **Classic BT shunt**

The original operation by Dr. Blalock is now called the classic BT shunt. The subclavian artery is divided and directly anastomosed (end to side) to the ipsilateral pulmonary artery. The arm is then perfused by collateral circulation. Constructing the shunt from the patient’s own subclavian artery allows it to grow so there is no need for revision. It is important for the anesthesiologist to suspect a classic BT shunt in older adult survivors of congenital heart disease or in patients who had their operation in the developing world. After classic BT shunt, the pulses in the ipsilateral arm will be decreased or non palpable.

An arterial line may be difficult to place and even if successful, the blood pressure may not reflect true central pressure and must be measured at another site.

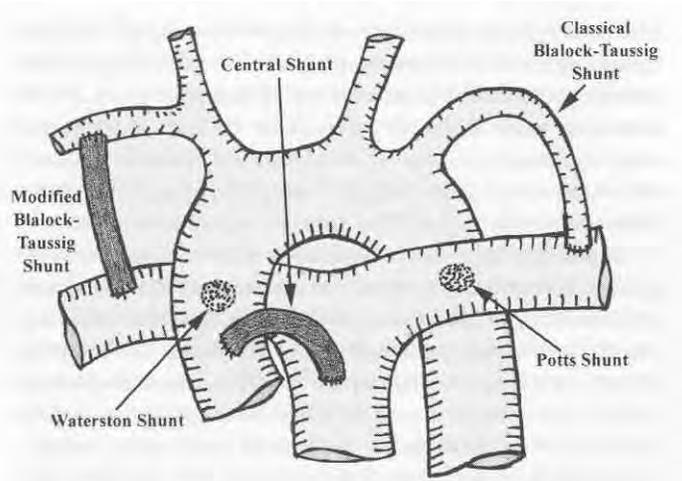


Figure 12: Types of systemic to pulmonary shunts (*Anesthesia for Congenital Heart Disease. Andropoulos DA, Stayer SA, Russell IA. Reprinted with permission*)

### **Potts and Waterston shunts**

To avoid dividing the subclavian artery and to further increase pulmonary blood flow, direct aortic to PA anastomoses (side to side) were tried. The Potts shunt connected the left PA to the descending thoracic aorta and the Waterston shunt connected the right PA to the ascending aorta. These shunts fell into disfavor because the very high volume of pulmonary blood flow led to pulmonary hypertension and CHF. Also, when the shunt was taken down at the time of definitive repair, there was often residual stenosis at the site of the pulmonary anastomosis.

### **Modified BT shunt**

In the 1970s, high quality synthetic material (PTFE, Goretex) allowed the development of the modified BT shunt. An end-to-side anastomosis is completed between the shunt and the subclavian and pulmonary arteries. Blood pressure in the ipsilateral arm still reflects true central pressure. Artificial material used for the shunt does not grow with the patient. In the modern era of congenital heart surgery, the BT shunt is a newborn operation followed by removal of the shunt and definitive repair in the first year of life.

Dr. Blalock's first BT shunt was done via left thoracotomy. Today the modified BT shunt is usually done in the newborn period via sternotomy. Many of these patients have such tenuous pulmonary blood flow that they will not tolerate the temporary clamping of the left or right PA. A sternotomy approach provides the option of briefly going on cardiopulmonary bypass to safely complete the operation. After modified BT shunt, it is vitally important to maintain perfusion pressure through the shunt. Hypotension leads to sluggish shunt flow and possibly thrombosis which will further decrease pulmonary blood flow, often to critical levels. An emergency trip to the cardiac catheterization laboratory or operating room for shunt revision is the next step. Surgeons will usually only partially reverse heparin with protamine in an effort to avoid shunt thrombosis.

## Central shunt

When there are technical reasons that preclude a modified BT shunt, a central shunt can be placed from the ascending aorta to the main PA. Synthetic material is used as in a modified BT shunt. Surgical approach is via sternotomy to allow the possibility of cardiopulmonary bypass if necessary. The post operative considerations are the same.

## Tetralogy of Fallot

The classic findings described by Etienne Fallot in 1888 are:

1. VSD (anterior malalignment)
2. Right ventricular outflow tract obstruction
3. Right ventricular hypertrophy
4. Over riding aorta

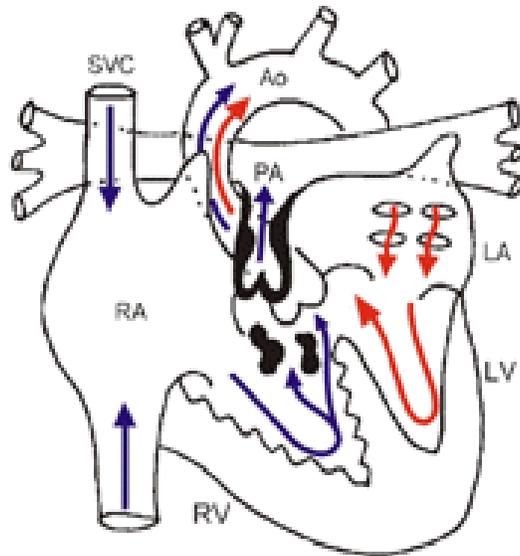


Figure 13: Tetralogy of Fallot (*Pediatric Heart Surgery, A Ready Reference for Professionals, 3<sup>rd</sup> Ed. May, LE. Reprinted with permission*)

The most significant defect is the VSD because of its anterior malalignment which narrows the RVOT and leads to the three other classic findings. The VSD and resulting RVOT obstruction causes a right-to-left shunt and cyanosis. Repair is usually in the first six months of life because relieving the RVOT obstruction and allowing the right ventricle to remodel reduces the incidence of long term right ventricular dysfunction and arrhythmia. Neonates with hypercyanotic spells who are too small for definitive repair are palliated with a Blalock-Taussig shunt and have a formal repair when they are older.

## **Tetralogy of Fallot Spells**

Hypercyanotic spells or “Tet spells” occur when there is a sudden increase in right-to-left shunting and the patient becomes intensely cyanotic. The cause is spasm of the infundibular muscle in the RVOT, usually provoked by pain or anxiety. Treatment involves maneuvers that either increase SVR or relax the infundibular muscle. Initial treatment is 100% O<sub>2</sub> and morphine, which provides sedation and usually is sufficient to relax the infundibular spasm and break the spell. For more severe spells, SVR can be augmented with phenylephrine and the infundibular muscle can be relaxed with beta-blocker. Raising SVR reduces the large right-to-left shunt and increases pulmonary blood flow. Other important aspects of treatment are bicarbonate to correct a rapidly developing acidosis and supporting the circulating volume with a fluid bolus of crystalloid.

Prophylaxis for Tet spells is with beta-blocker, usually propranolol. Of historical interest is that before repair was done in infancy, children with TOF would instinctively learn ways to raise SVR and treat Tet spells. This was done by squatting which became a characteristic finding of children with unrepaired TOF.

Repair of TOF is one of the most common CHD operations. Traditionally, it was performed at 4-5 years of age when the child was believed to be strong enough to survive the procedure. Over the past 20 years there has been recognition that early relief of cyanosis is beneficial for systemic and neurologic development. Early repair (3-4 months old) also improves the ability of the hypertrophied right ventricle re-model, decreasing the long term incidence of arrhythmia and right ventricular dysfunction.

### **Induction**

Children older than six months or younger infants with a history of Tet spells are generously premedicated to ease separation anxiety and lessen the chance of a Tet spell on induction. Historically, mask induction with halothane was recommended because its negative inotropic properties were believed to reduce the risk of infundibular muscle spasm and decrease the incidence of Tet spells. In actual practice, mask induction with sevoflurane does not appear to have any increased risk of Tet spells. After induction and intubation, a Tet spell is rare if an adequate level of anesthesia is maintained. Should a Tet spell occur, it can be rapidly treated by raising SVR with a bolus of phenylephrine.

### **Surgical repair**

The VSD is closed with a patch. It is usually approached from the right atrium but may be approached from the right ventricle. The surgeon then addresses the degree of RVOT obstruction post-VSD patch. If the RVOT is sufficiently patent nothing further is required but frequently the degree of obstruction is unacceptable. Obstructing right ventricular muscle bundles if present, are resected. Should the RVOT remain significantly obstructed, an incision is made above and below the pulmonary valve annulus and a patch is sutured in. This “trans-annular patch” relieves the RVOT obstruction at the cost of creating free pulmonary insufficiency (PI). About 10% of patients will have the left coronary artery originating anomalously from the right coronary artery. This might be seen on preoperative echocardiography. The left coronary artery courses across the

RVOT in the location where a trans-annular patch incision would be made. Cardiac catheterization is not necessary because the surgeon can make diagnosis by direct inspection intraoperatively. Patients with an anomalous origin of the left coronary artery require a RV-PA connection to relieve RVOT obstruction.

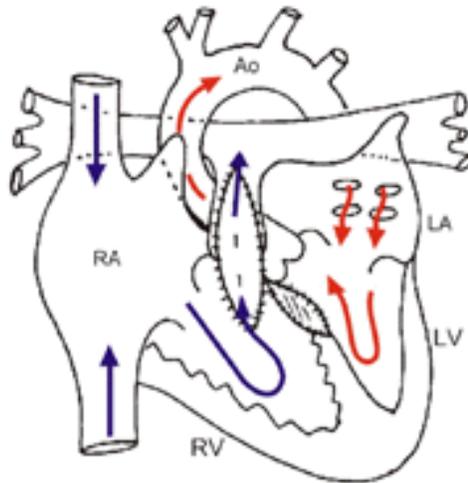


Figure 14: Surgical repair of Tetralogy of Fallot with trans-annular patch (*Pediatric Heart Surgery, A Ready Reference for Professionals, 3<sup>rd</sup> Ed. May, LE. Reprinted with permission*)

### Postoperative

The hypertrophied right ventricle has poor compliance which worsens in the immediate post op period due to the right ventriculotomy. Right ventricular filling pressures must be maintained with adequate volume. The PFO is left open or a small ASD is created that becomes a “pop-off valve” if right sided pressures increase due to transient postoperative right ventricular dysfunction. Cardiac output is maintained at the expense of modest systemic desaturation from the right-to-left shunt at the atrial level. The conduction system can be damaged in the repair, leading to heart block.

### Long term follow-up

The free PI that results from a trans-annular patch repair for TOF was believed to be a benign condition. Unlike aortic insufficiency, the “back pressure” causing regurgitation into the right ventricle is pulmonary diastolic pressure which is much lower than systemic diastolic pressure. This is why the right ventricle can cope with free pulmonary insufficiency for a long period of time. It is true that free PI is usually well tolerated but over time the volume load on the right ventricle eventually leads to hypertrophy, dilation and failure. The time frame may be as much as 20-30 years. If there is significant coexisting tricuspid regurgitation, the volume load on the right ventricle becomes severe and the progression to right sided heart failure is accelerated. Long term follow up in TOF adults reveals universal right ventricular dysfunction. There may be a ventricular focus for arrhythmia. This occurs even when TOF repair is done in infancy. Right ventricular dysfunction is usually due to dilation from free pulmonary insufficiency. In children with a RV-PA connection there may be right ventricular dilation from free PI as

the valve within the homograft fails or right ventricular hypertrophy if the homograft develops calcific stenosis.

### **Percutaneous pulmonary valve replacement**

A truly exciting development in the management of TOF patients is percutaneous pulmonary valve replacement. The consequences of free PI in the long term are now well appreciated but surgical replacement of the pulmonary valve involved the risk of cardiac re-operation. Consequently the surgery was delayed as long as was safely possible. Now the pulmonary valve can be replaced with a percutaneous technique and the right ventricle spared the effects of free PI. The caveat is that only patients with a RV-PA homograft connection are candidates for percutaneous valve replacement. As mentioned earlier when there is an anomalous left coronary artery traversing the RVOT, a trans-annular patch is contraindicated and a RV-PA homograft (cadaveric tissue) is required. The native valve within the homograft usually fails within a few years with resultant free PI. The fact that the valve fails within the tubular confines of the homograft means that an appropriate cylindrical, annular “landing zone” exists for deployment of the percutaneous valve. When a trans-annular patch repair has been done, the “patched” area of the RVOT becomes aneurysmal over time, precluding the safe deployment of a valve in this area. Hopefully the technical restrictions of an aneurysmal RVOT can be overcome for percutaneous valve replacement because the vast majority of patients with TOF have a trans-annular repair. Additionally, any other patients with a RV-PA homograft connection (post-Ross surgery, post-Truncus Arteriosus repair) are candidates for percutaneous pulmonary valve replacement.

### **Tetralogy of Fallot variants**

Classic TOF covers the spectrum from the “pink Tet” with minimal right-to-left shunting to the severely cyanotic child with critical right ventricular obstruction and the need for a newborn BT shunt. Two other TOF variants have unique characteristics that deserve mention.

### **Tetralogy of Fallot with Absent Pulmonary Valve**

The lack of a pulmonary valve leads to free PI in utero with a very large volume of pulsatile blood flowing back and forth through the developing branch pulmonary arteries. The increased volume can lead to enormous dilation of the left and right pulmonary arteries such that there is marked compression of surrounding structures, particularly the main stem bronchi. The compression of the main stem bronchi by the enlarged branch PAs leads to severe bronchomalacia. The airways beyond the mainstem bronchi are often abnormal as well. These patients usually require intubation and positive pressure to stent open their bronchi. Surgery consists of repair in the standard fashion with volume reduction (plication) of the left and right pulmonary arteries. Morbidity is usually from respiratory causes. These patients often require tracheostomy and long term ventilation.

## **Tetralogy of Fallot with Pulmonary Atresia**

There is a plate like pulmonary valve which is completely atretic. Pulmonary blood flow is supplied by major aorto-pulmonary collateral arteries (termed MAPCAs). Pulmonary blood flow through the MAPCAs may be abundant, especially as the infant grows older and PVR falls. Alternatively, the child may become very cyanotic because MAPCA flow is inadequate due to tortuosity and the small size of the collateral vessels. These patients cannot have Tet spells since they have no antegrade flow from the right ventricle to the pulmonary circulation. The major problem is that the MAPCAs usually do not flow into a normal pulmonary artery configuration. Surgical repair involves trying to establish normal antegrade pulmonary flow by constructing an adequate main pulmonary artery, right and left PA and then dividing the MAPCAs off the aorta and anastomosing them to the reconstructed pulmonary artery system. A RV-PA connection establishes flow from the right ventricle to the pulmonary artery. This operation is called unifocalization and is one of the most technically challenging in CHD.

## **Truncus Arteriosus**

This lesion results from failure of the truncus arteriosus to divide into a discrete aorta and pulmonary artery. Embryologically, this lesion is a classic cono-truncal abnormality and is associated with certain genetic syndromes. A VSD is present, with right and left ventricular output into the truncus. Pulmonary blood flow originates from the truncus. There is a truncal valve with between three and six leaflets, because it is the amalgamation of the aortic valve and the pulmonary valve. Leaflet tissue is often abnormal leading to truncal valve insufficiency. The anatomic relationship of how the main pulmonary artery or branch pulmonary arteries connect to the truncus defines the classification of subtypes. Truncus arteriosus can lead to a very high Qp:Qs. Some centers will consider a neonatal repair for this reason, while others are content to wait and treat CHF medically. The latter option is justified only as long as the infant can continue to grow and develop adequately. Significant failure to thrive is an indication for surgery.

Complete mixing at the ventricular level leads to SpO<sub>2</sub> of 75-80% with a balanced Qp:Qs in the newborn period. It would be expected that the SpO<sub>2</sub> would rise in the first few months of life as the PVR falls.

Surgery consists of:

- closure of the VSD
- division of the pulmonary arteries from the truncus
- placement of an RV-PA connection
- attempted repair of truncal valve insufficiency if possible

### Truncus Arteriosis Repair

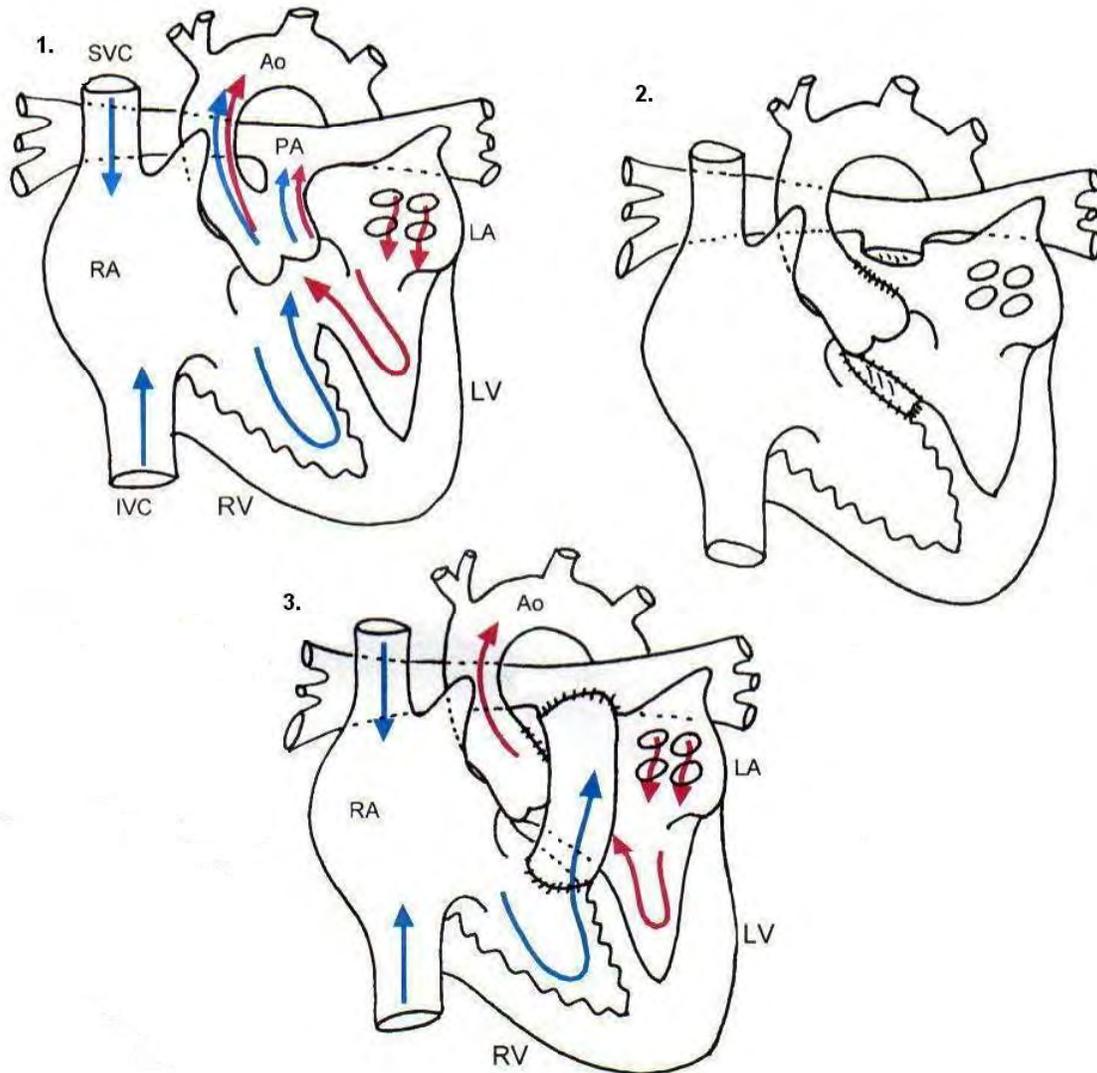


Figure 15: Surgical repair of truncus arteriosus (*Pediatric Heart Surgery, A Ready Reference for Professionals, 3<sup>rd</sup> Ed. May, LE. Reprinted with permission*)

### Transposition of the Great Arteries (D-TGA)

Transposition of the Great Arteries (TGA) is defined strictly as a right ventricle to aorta connection and a left ventricle to PA connection. The common term D-TGA refers to the Van Praagh description. The segmental Van Praagh notation for D-TGA would be S,D,D. This refers to normal abdominal situs, D-looped ventricles (the normal position) and an aorta that is in the “D” position relative to the PA. In the “D” position the aorta is to the right and anterior of the PA. The “S” or normal aortic position is to the right and posterior to the PA. The patient with D-TGA has two circulations in parallel rather than in series with deoxygenated venous blood delivered systemically from the right ventricle

to the aorta and oxygenated pulmonary venous blood is returned directly to the pulmonary artery by the left ventricle. For this arrangement to be compatible with life there must be a site or sites where blood from the two parallel circulations can mix. Sites of mixing are a PDA, ASD or VSD. A sufficient ASD or VSD are the most stable locations for mixing. A PDA is not always a reliable site for mixing. Flow through the PDA must be bidirectional to achieve adequate mixing.

In the setting of D-TGA with inadequate mixing, the newborn is critically ill with deeply compromised oxygen delivery to the heart and brain. Even though the PDA is not providing a site of sufficient mixing, the small amount of bidirectional shunting may be the only thing keeping the newborn alive. Therefore PGE1 to maintain some ductal patency is a vital temporizing measure. An emergency balloon atrial septostomy (BAS) may be done at the bedside under echocardiographic guidance or in the catheterization laboratory. Institutional practice varies. When the newborn is brought to catheterization laboratory they may be accompanied by the intensive care staff or an anesthesiologist. Once again institutional practice varies. One might ask what further can be done when the newborn is already intubated? While the situation is critical it is not futile. Optimization of oxygen delivery is the goal which involves increasing supply and decreasing demand. The newborn is sedated with full neuromuscular blockade to decrease oxygen consumption. Ventilation with  $FiO_2$  allows for the highest dissolved fraction of oxygen. The severe hypoxia induces a progressive cardiovascular deterioration which is the scenario one must avoid. Hypoperfusion combined with severe hypoxia is lethal. Therefore support of the circulation with cautious fluid boluses (usually albumin) and vasopressor (usually dopamine) are required. In a severely acidotic environment, cardiovascular function will deteriorate. Sodium bicarbonate should be given. The newborn normally has a hematocrit well over 40%, but if for any reason the hematocrit is in the 30% range transfusion is justified to increase oxygen carrying capacity. Once the patient is stabilized with the creation of an adequate ASD, the  $SpO_2$  on room air should be between 75-80% reflecting a balanced Qp:Qs and free mixing at the atrial level.

The first operation for D-TGA was the Senning procedure. Baffles were created in the atria to route systemic venous blood in the right atrium across the mitral valve and into the left ventricle where it would be pumped to lungs. Pulmonary venous blood in the left atrium was baffled across the tricuspid valve into the right ventricle where it would be pumped systemically. The Senning procedure used the patient's own atrial tissue. The Mustard procedure was similar but used pericardium to construct the baffles. Problems with the Mustard/Senning operation were the following:

- baffle stenosis led to obstruction of venous return either systemic or pulmonary
- the extensive atrial suture lines led to long term problems with atrial dysrhythmias and sinus node dysfunction
- the systemic right ventricle was at risk of failing over the long term

Nevertheless, the operation proved life saving for patients who had no other option and there are a number of long term survivors. The Mustard/Senning operation is only done today in circumstances where the coronary anatomy does not permit an arterial switch.

## **Arterial switch (Jatene)**

This operation was first done by the Brazilian surgeon Dr. Jatene in 1975. It overcame the problems with the Mustard/Senning by avoiding atrial level baffles altogether and switching the great vessels into their correct location and re-implanting the coronary arteries. The operation is performed in the first few days of life or as soon as possible after the patient has been stabilized with a balloon atrial septostomy. The steps of the Jatene arterial switch are:

- division of the aorta and PA at the supravalvular level
- excision of the left & right coronary arteries with a surrounding “button” of aorta
- the Lecompte maneuver which moves the aorta posterior and leftward of the PA from its initial position of being anterior and rightward
- anastomosis of the aorta to the proximal stump of the PA
- re-implantation of the coronary arteries
- anastomosis of the PA to the proximal stump of the aorta
- closure of the ASD and/or VSD and ligation of the PDA if present

Blood now flows from the right ventricle across the morphologic aortic valve into the PA and from the left ventricle across the morphologic pulmonary valve into the ascending aorta. Critical aspects of the operation are the extensive arterial suture lines which make bleeding a problem and the quality of the coronary re-implantation because any kinking or obstruction leads rapidly to myocardial ischemia. In the early days of the arterial switch the operation carried a very high mortality. The primary reason was technical problems with coronary artery re-implantation. Only by the mid to late 1980s did surgeons begin to understand and master this technically demanding aspect of the procedure. Presently, the operation carries a very low mortality if coronary artery anatomy is normal. Unfortunately, there continue to be coronary artery anatomic variants that are unfavorable for successful re-implantation. Mortality continues to be significant in this sub-group.

One other scenario that deserves mention is the patient who presents for repair late in infancy. The SpO<sub>2</sub>, if mixing continues to be adequate, will be between 75-80% but of more importance is that the left ventricle will have become “de-conditioned”. As PVR falls in the first few months of life the left ventricle adjusts to pumping against the lowered pulmonary resistance. An arterial switch done at this point suddenly subjects the left ventricle to systemic afterload, leading to acute left ventricular failure. The left ventricle is “prepared” by placing a band around the pulmonary artery (PA band). This artificially elevates the afterload of the left ventricle and prepares it to be the systemic ventricle. The arterial switch operation is performed at a later date. The PA band is a constricting tie placed around the PA via median sternotomy.

### Arterial Switch Operation

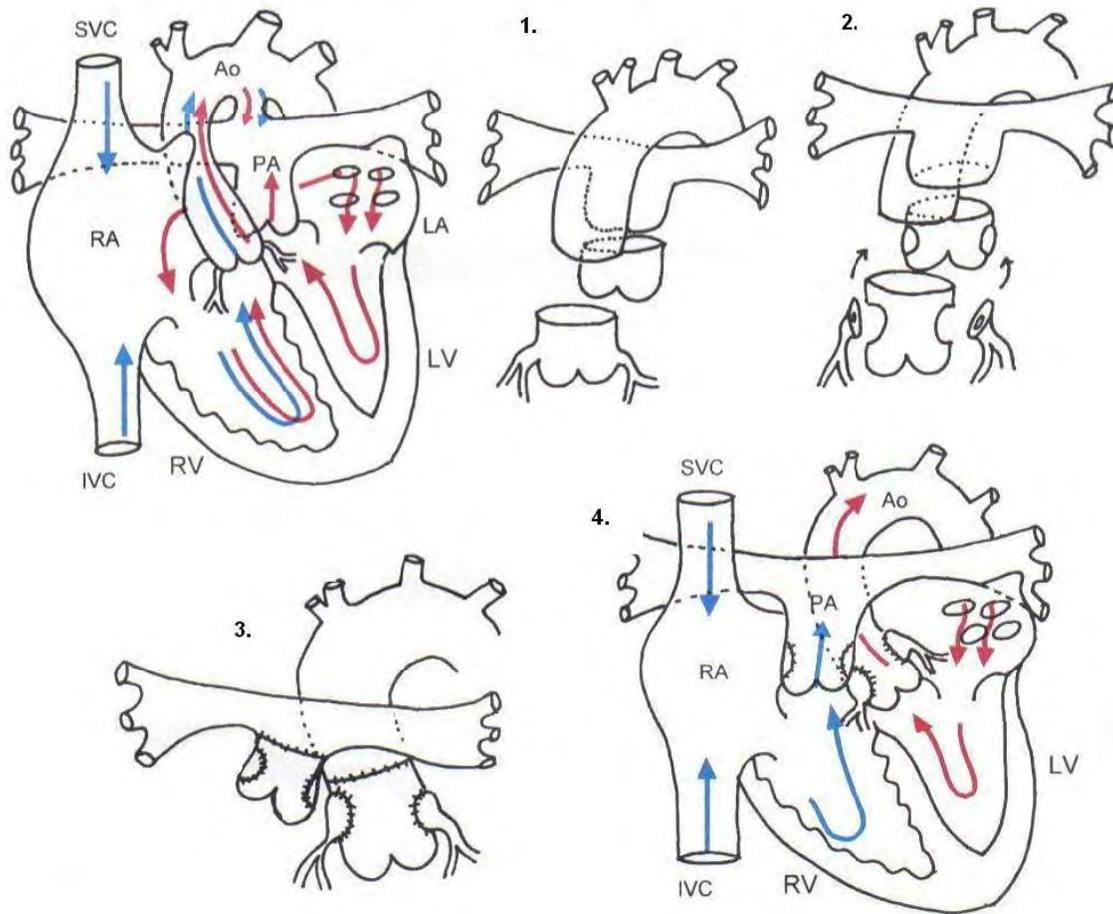


Figure 16: Arterial switch operation for D-Transposition (*Pediatric Heart Surgery, A Ready Reference for Professionals, 3<sup>rd</sup> Ed. May, LE. Reprinted with permission*)

### Rastelli Operation

The Rastelli operation was first performed to treat D-TGA with VSD and pulmonary stenosis. In D-TGA with a VSD, pulmonary stenosis protects the neonate from excessive pulmonary blood flow. The left ventricle which connects to the PA may be obstructed proximal and distal to the pulmonary valve. An arterial switch with closure of the VSD would not be suitable because of the resulting LVOT obstruction. The Rastelli operation closes the VSD in such a way that left ventricle blood is directed out both the aorta and the PA. The PA is then ligated just distal to the pulmonary valve and an RV-PA connection is created.

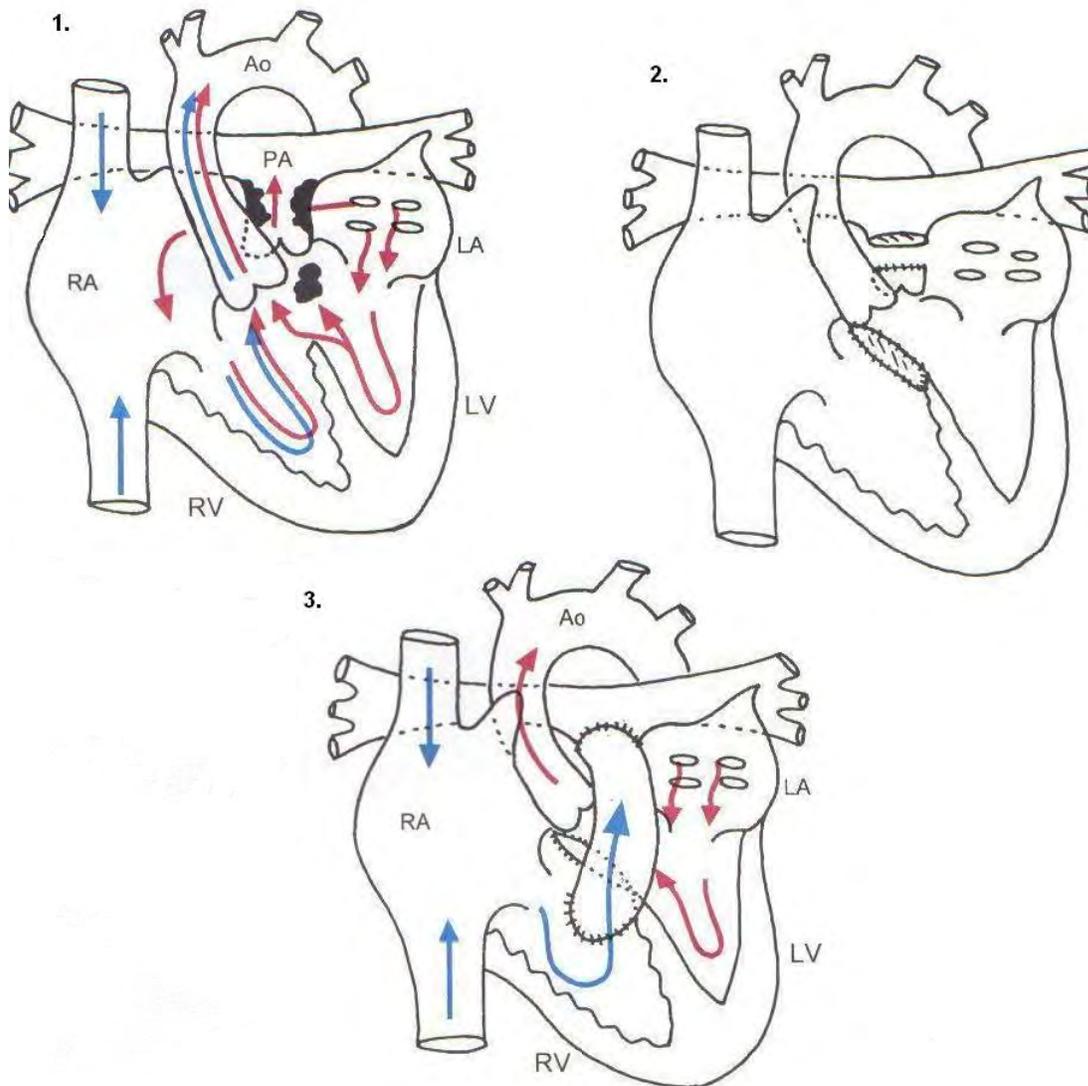


Figure 17: Rastelli operation for D-transposition with VSD and pulmonary stenosis  
*(Pediatric Heart Surgery, A Ready Reference for Professionals, 3<sup>rd</sup> Ed. May, LE. Reprinted with permission)*

## Hypoplastic Left Heart Syndrome

Hypoplastic left heart syndrome (HLHS) is the prototype of a single ventricle lesion requiring three staged palliative surgeries:

- Stage I or Norwood
- Stage II or Bidirectional Glenn
- Stage III or Fontan completion

Norwood first performed the operation that now bears his name in 1979 at Children's Hospital Boston. While enormously high risk, their landmark paper showed that a surgically viable option existed for this condition that otherwise would be fatal in the first

few weeks of life.<sup>12</sup> Glenn and Fontan operations were devised as palliative surgeries for other lesions but became part of the staged palliation for HLHS once the Norwood operation was created. Other than compassionate care, the only other option for HLHS is neonatal transplantation. Pioneered by Bailey and colleagues at Loma Linda University<sup>13</sup> in the early to mid 1980s using primate hearts and later orthotopic grafts, the procedure has the obvious advantage of rendering the infant cardiovascularly “normal” with a two ventricle heart. Technical obstacles to transplantation in a neonate with a severely hypoplastic ascending aorta have been overcome. The limitation to neonatal transplantation is the short supply of organs with many potential recipients dying before a suitable donor is found. In present day practice, the Norwood operation remains by far the most common surgical procedure for HLHS.

In HLHS, the entire left side from the mitral valve to the aortic arch is hypoplastic. An ASD is present and systemic and pulmonary venous return mixes in the right atrium. The right ventricle provides pulmonary blood flow. Systemic blood flow is from the pulmonary artery via the PDA. Therefore, systemic blood flow is said to be ductal dependent. With a high Qp:Qs the newborn may look surprisingly well oxygenated. It is only when the PDA closes in a few weeks that the neonate presents in shock because systemic perfusion is severely reduced. Resuscitation consists of volume, bicarbonate to correct the severe metabolic acidosis, PGE1 to re-open the ductus and inotropic support if necessary. This scenario is much less common today because many cases are diagnosed in utero and immediately after delivery umbilical lines are placed and PGE1 started to maintain ductal patency. PGE1 is adjusted to the lowest possible dose that provides good systemic perfusion as evidenced by warm, well perfused extremities and the absence of metabolic acidosis. If the Qp:Qs is balanced with good mixing at the atrial level, the SpO<sub>2</sub> should be 75-80% on room air. Usually there is excess pulmonary blood flow and the infant will have a SpO<sub>2</sub> greater than 85%.

The Norwood operation is completed under deep hypothermic circulatory arrest (DHCA) or deep hypothermia with selective cerebral perfusion. The operation proceeds in five steps:

1. Ligation of the PDA
2. Atrial septectomy to create a common atrium
3. Division of main PA at its point of bifurcation and amalgamation of the main PA to the hypoplastic ascending aorta in a side to side fashion
4. Augmentation of the hypoplastic ascending aorta and aortic arch with homograft
5. Establishment of pulmonary blood flow with either a BT shunt or RV-PA connection

Steps 3 & 4 comprise the bulk of the operation

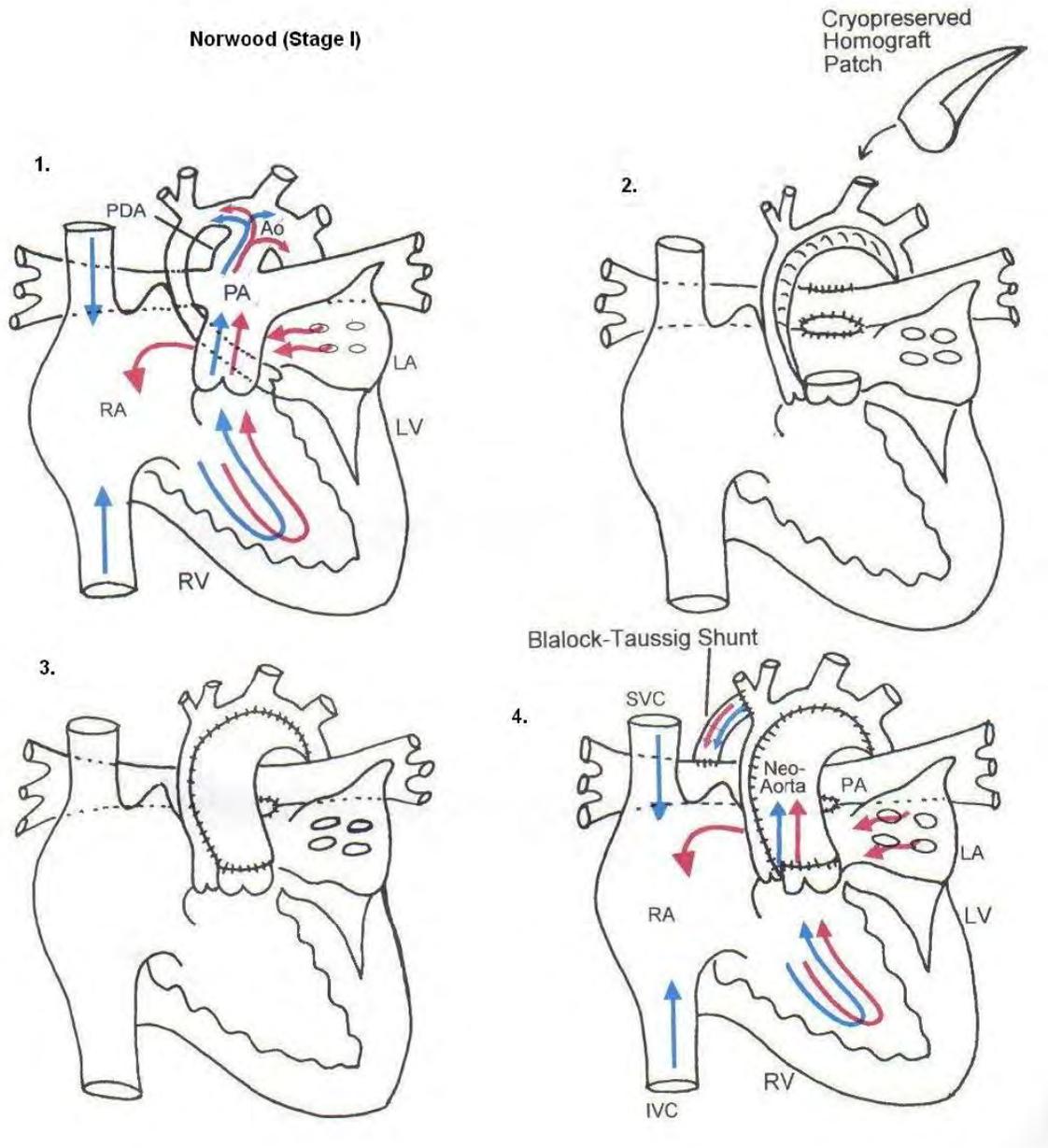


Figure 18: Norwood operation for hypoplastic left heart syndrome (*Pediatric Heart Surgery, A Ready Reference for Professionals, 3<sup>rd</sup> Ed. May, LE. Reprinted with permission*)

After the Norwood operation, the patient's anatomy consists of

1. Common atrium where systemic and pulmonary venous return mix freely
2. Systemic right ventricle
3. Native pulmonary valve has become the "neo-aortic valve"
4. An ascending aorta and aortic arch, called the "neo-aorta", that has been constructed from main PA and homograft
5. Pulmonary blood flow via a BT shunt or RV-PA connection

## **Post-operative Norwood operation (Stage I)**

The above description belies that fact that the Norwood operation is a massive surgical insult inflicted on a 3 kg neonate who is less than one week old. Despite the complexity of the operation, none of the physiological demands of single ventricle physiology have been substantially lessened from the preoperative state. Simply put, the operation consists of a technically exacting rearrangement of “plumbing”.

Key concerns postoperatively are balancing Qp:Qs and ventricular function. A balanced Qp:Qs results in a SpO<sub>2</sub> of 75-80%. If the SpO<sub>2</sub> is higher than 85%, the Qp:Qs is greater than 2:1. The pulmonary over circulation will result in a greatly increased myocardial work. Milrinone provides both pulmonary and systemic vasodilation in addition to augmenting inotropy through a  $\beta$ -receptor independent mechanism. Milrinone is now used almost routinely and is usually combined with some vasopressor pharmacologic support (low dose epinephrine or dopamine). The use of intense  $\alpha$ -1 blockade with phenoxybenzamine (non-competitive inhibition) or phentolamine (competitive inhibition) has been used in the past but has fallen out of favor today. A SpO<sub>2</sub> lower than 70% reflects a Qp:Qs less than 1 with inadequate pulmonary blood flow due to problems with the BT shunt (clot, kinking) or inadequate oxygenation due to lung disease (atelectasis, pneumonia, effusion). It is imperative to distinguish between the two causes because technical problems with the BT shunt or RV-PA connection require correction in the cardiac catheterization laboratory or operating room.

In addition to the above physiologic challenge, the inotropic state of the heart may be reduced due to the prolonged period of ischemia during of aortic cross clamp. Profound vasodilation can occur as a result of huge inflammatory response induced by such a complex operation with a long duration of cardiopulmonary bypass and deep hypothermia with circulatory arrest. With HLHS anatomy, the pulmonary vasculature has been “over circulated” both in utero and postnatally prior to surgery, putting the neonate at risk for increased pulmonary vasoreactivity and even a pulmonary hypertensive crisis. Milrinone is widely used to prevent such an occurrence. Persistent low oxygen saturations without a respiratory (pneumonia, pleural effusion) or technical reason (thrombosed or kinked BT shunt, obstructed RV-PA connection) are presumed to due to pulmonary hypertension. Inhaled nitric oxide is usually started in this scenario. Lastly, the neonate is at increased risk for all the usual types of postoperative complications such as sepsis, pneumonia, etc. It is not surprising that 30 day mortality after the Norwood operation varies between 7-19%. Within the entire HLHS cohort, higher risk neonates are those with other congenital anomalies, genetic syndromes, other cardiac lesions especially anomalous pulmonary venous return, low birth weight or prematurity.

## **BT shunt v. Sano modification**

In the early days of the Norwood operation, the pioneering surgeon and colleagues experimented with different anatomic arrangements for pulmonary blood flow. They abandoned the RV-PA connection because of pulmonary over circulation and favored the BT shunt. Other surgeons followed suit but the problems with pulmonary blood flow via a BT shunt became evident. The BT shunt provides continuous pulmonary blood flow throughout systole and diastole. The continuous flow into the lower resistance pulmonary

circulation reduces systemic diastolic pressure and can compromise coronary perfusion. Recall that the right ventricle is the systemic ventricle and is only perfused during diastole according to the relationship.

$$\text{Coronary perfusion pressure} = \text{Diastolic BP} - \text{RV end diastolic pressure}$$

Combined with pulmonary blood flow that often exceeds systemic, ( $Q_p:Q_s > 1$ ) an environment is created where myocardial work is increased in the setting of potential coronary ischemia. Systemic perfusion particularly mesenteric and renal is also reduced with a BT shunt in the same manner as a PDA. This imbalance between pulmonary and systemic blood flow with accompanying coronary ischemia is cited as a major cause of perioperative mortality. Additionally, among those infants who survive to hospital discharge, further 5-15% mortality occurs prior to Stage II (Bidirectional Glenn). This phenomenon, known as “interstage death” is particularly devastating to parents who believe their child has survived the most dangerous part of the three stage surgical palliation process.

Searching for a method to overcome these problems, Sano and colleagues from Japan returned to the concept of the RV-PA connection. Synthetic conduits better sized for neonates provided a more controlled and physiologic volume of pulmonary blood flow. Their encouraging results<sup>14</sup> published in 2003 garnered widespread attention and were quickly adopted in many centers. In the setting of the Norwood operation, the RV-PA connection is now known as the “Sano modification”. However, a clear outcome benefit and longer term follow-up for the Sano modification did not exist and very serious theoretical concerns existed for this approach. The more optimal balancing of  $Q_p:Q_s$  occurred at the price of a right ventricular incision. We know from the early days of TOF repair that a ventriculotomy can lead to long term deterioration in right ventricular function and is also substrate for arrhythmia. These are serious consequences for a child with TOF but potentially fatal in HLHS with a single, systemic right ventricle. The physiologic advantages and disadvantages of the BT shunt and RV-PA are summarized as follows:

### **BT shunt**

#### Advantages

1. Higher  $Q_p:Q_s$  results in larger branch pulmonary arteries (beneficial for Stage II/III)
2. Increased  $SpO_2$  makes it less likely that Stage II will have to occur early
3. No right ventriculotomy

#### Disadvantages

1. Lower diastolic BP creates “coronary steal”, possible myocardial ischemia
2. Higher  $Q_p:Q_s$  increases myocardial work
3. Lower diastolic BP may reduce mesenteric/renal perfusion
4. Risk of thrombosis of shunt (usual size 3.5 mm) with dehydration or other illness

## RV-PA connection (Sano modification)

### Advantages

1. Higher diastolic BP preserves coronary perfusion
2. Balanced Qp:Qs reduces myocardial work
3. Higher diastolic BP preserves mesenteric/renal perfusion
4. Less likely to have critical thrombosis of RV-PA connection (usual size 5-6 mm) with dehydration or low cardiac output state

### Disadvantages

1. Right ventriculotomy
  - negative effect on right ventricular function (immediate and long term)
  - substrate for arrhythmia (medium and long term)
2. Non-valved conduit results in free PI and volume load on right ventricle
3. Lower Qp:Qs results in smaller branch pulmonary arteries (limitation for Stage II/III)
4. Lower SpO<sub>2</sub> may require Stage II to be done earlier than usual

As one can see the advantages of the BT shunt are the disadvantages of the RV-PA connection and vice-versa. Valid reasons exist to pursue either option. To answer the question of which is better the Pediatric Heart Network Investigators conducted one of the most ambitious studies ever undertaken in CHD.<sup>15</sup> A remarkable 549 neonates were randomized to BT shunt or RV-PA at 15 North America congenital heart centers. The results are summarized below:

Outcome: death or transplant	BT shunt	RV-PA connection
1 year	36%	26%
30 days	14%	10%
30 days-Stage II (interstage)	22%	12%
Stage II-1 year	5%	7%
After 1 year	4%	8%

The improvement in survival during the first year was statistically and clinically significant for the RV-PA group. Although the primary outcome of the trial was death or transplant-free survival at 1 year, follow-up continued for a mean of 32 months (range 12-52 months). The survival advantage of the RV-PA connection disappeared after 1 year from the original Norwood operation. As noted, the trial was a hugely ambitious effort which included both angiographic and echocardiographic measures of right ventricular function and branch pulmonary artery size at different time points.

## **Summary points of the trial**

1. The hypothesis of improved survival through preserved coronary perfusion and a more optimal balancing of Qp:Qs was borne out with increased survival in the first year after the Norwood operation.
2. The benefit of the RV-PA connection was seen during perioperative period after the Norwood operation and more impressively during the interstage period before Stage II (Glenn)
3. After Stage II (Glenn) the survival benefit of the RV-PA connection disappeared with a non-statistically significant trend to improved outcomes in the BT shunt group.
4. Right ventricular ejection fraction was statistically better in the RV-PA group after the Norwood operation and before Stage I (Glenn). The difference disappeared when echocardiograms were repeated at 14 months of age.
5. When measured prior to Stage II (Glenn), the right PA diameter was larger in the BT shunt group. The left PA dimensions were equal in the two groups. The clinical significance of this finding is uncertain.
6. Improved outcomes were seen in the RV-PA group but only if only if the surgeon did less than 15 Norwood operations/year and the institution did less than 25 Norwood operations/year. More experienced surgeons and higher volumes centers appear to have better results with the BT shunt.
7. A higher number of interventions were required in the RV-PA group and these consisted of balloon dilation and stenting of the conduit.

The trial did not definitively answer the question it posed. More experienced surgeons and higher volume centers report seem more adept at managing BT shunt patients. Clear evidence of deterioration in right ventricular function in the RV-PA group was not seen but follow-up extended only to an average of 32 months. Perhaps the most striking finding is that 30 years after the first Norwood operation, with an entire cohort of physicians (surgeons, anesthesiologists, intensivists, cardiologists) who have been specifically trained to manage these high risk infants, the 1 year mortality or need for transplant is still approximately 30%.

## **Hybrid procedure in HLHS**

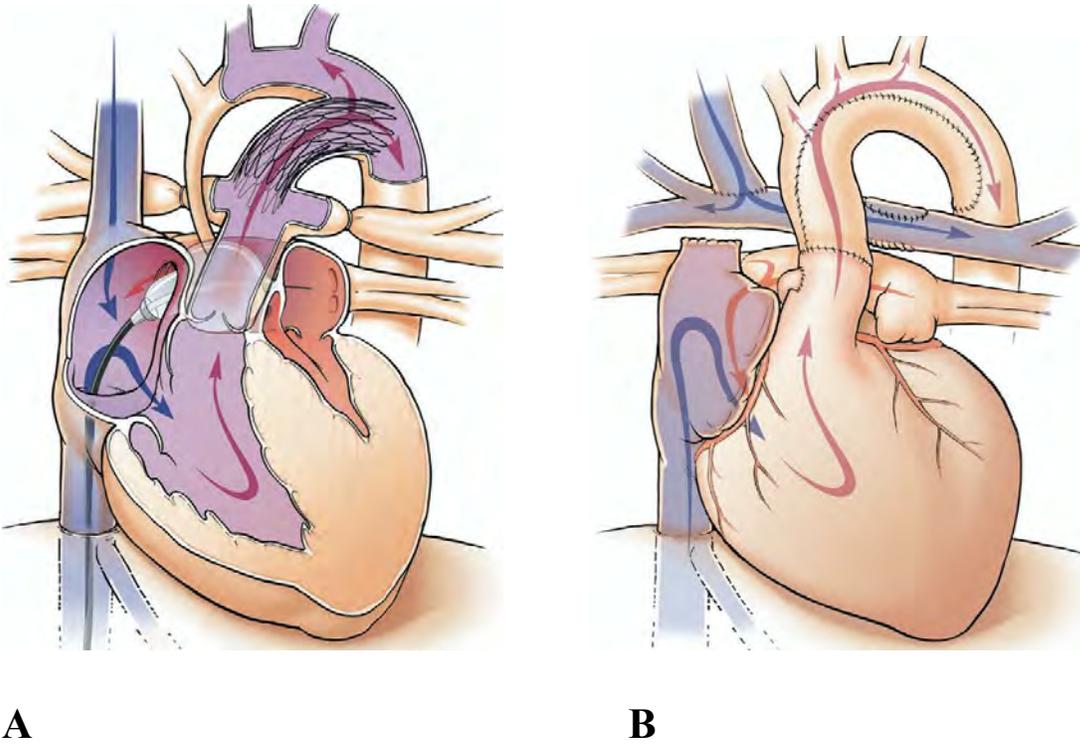
“Hybrid procedures” is a term for a combined approach utilizing both surgery and interventional cardiology. The procedure is performed in a specially designed “hybrid operating room” which has full angiography and surgical capability. Surgical exposure could be full sternotomy, hemi-sternotomy or some form of thoracotomy allowing the interventional cardiologist to gain vascular access directly into a major vessel or the heart itself. The usual benefit is that cardiopulmonary bypass is avoided or the surgical incision is greatly reduced or both. This working definition of “hybrid procedures” may change in the future as the scope and complexity of these interventions increases.

The hybrid procedure that has generated the most interest occurs in HLHS. While almost all HLHS infants are candidates for the procedure, it was initially used for neonates thought too high risk (low birth weight, significant prematurity) to withstand the stress of a full surgical repair with a prolonged duration of CPB and deep hypothermic circulatory arrest. Some centers are now extending the procedure to healthier patients with standard HLHS anatomy. At the forefront of this technique is Nationwide Children's Hospital (Columbus, OH). Their experience shows promise but one cautions that centers adopting this technique may not be able to initially duplicate their good results.<sup>16</sup>

The premise of the hybrid procedure in HLHS is that the condition is compatible with life for a short period of time as long as the ductus remains open. In the interval between birth and the Norwood operation PGE1 is the life saving medication to keep the ductus open and allows ductal dependent systemic circulation. There is antegrade flow into the descending aorta and retrograde flow into the aortic arch and ascending aorta. Retrograde flow through the hypoplastic arch and ascending aorta must be sufficient for cerebral and coronary perfusion. If one could mechanically "open" the ductus with a device such as a stent, PGE1 could be stopped and neonate would be able to survive with the same ductal dependent systemic circulation. Technically speaking, as long as the PDA is not extremely tortuous, it can be reliably stented using a percutaneous technique. However, ductal stenting only solves the most critical problem of HLHS. Recall that the right ventricle is in continuity with the pulmonary artery and that even in the newborn period PVR is lower than SVR. Consequently, the pulmonary vasculature is "over circulated". Clinically we observe this when the SpO<sub>2</sub> of a neonate with HLHS exceeds 80%. In fact, most neonates with HLHS have a SpO<sub>2</sub> well above 80% reflecting a Qp:Qs of at least 3:1. Thus the second part of the hybrid procedure is a technique to limit pulmonary blood flow. Lastly, to ensure a widely patent atrial communication, the final part of procedure is an atrial septostomy.

The hybrid procedure for HLHS occurs in specialized hybrid suite that is a combination operating and interventional cardiology suite. The goals of the hybrid stage 1 palliation are the following:

1. Unobstructed systemic output through the patent ductus arteriosus (PDA)
2. Improved balance of the pulmonary and systemic circulations
3. An unobstructed atrial septal defect



**A** **B**

Figure 19: Hybrid procedure for HLHS. A: Hybrid stage I with branch pulmonary artery banding, PDA stent and balloon atrial septostomy. B: Comprehensive stage II with Norwood type reconstruction of the ascending aorta and aortic arch, removal of branch pulmonary artery bands, excision of PDA and stent and completion of SVC-right PA anastomosis (Glenn). (*Galantowicz et al. Hybrid approach for hypoplastic left heart: intermediate results after the learning curve. Ann Thorac Surg 2003;85(6):2063-70*)

The procedure is done in the neonatal period at the same time as a conventional Norwood would be scheduled. A median sternotomy is performed, followed by branch pulmonary artery banding. Pulmonary artery banding is somewhat subjective and the most physiologically crucial part of the procedure because the other two components are purely anatomic. The goal is to achieve a balanced circulation ( $Q_p:Q_s$  of 1) reflected in a  $SpO_2$  of 75-80% on room air. Prior to banding the neonate should be hemodynamically stable and ventilated with a  $FiO_2$  as close to room air as possible. In the experience of Galantowicz,<sup>16</sup> appropriate banding usually results in a rise of 10 mmHg in systolic blood pressure and a drop of 10% in oxygen saturation. Following banding, the PDA is stented and a balloon atrial septostomy is completed. The hybrid procedure does not require CPB. If the atrial communication is adequate, some centers defer the balloon atrial septostomy until prior to discharge. Using this approach, centers experienced in the hybrid procedure are able to extubate the majority of neonates immediately after the procedure or within 24 hours of ICU admission. Length of stay in ICU and hospital is shorter and overall perioperative mortality is also improved.

Stage II which would normally be a Glenn procedure now becomes a “comprehensive stage II” consisting of a combination of the Norwood and Glenn operations. The comprehensive stage II is done between 4-6 months of age. The pulmonary artery bands are removed, the PDA excised with stent in situ, a Norwood reconstruction of the

ascending aorta and arch is done and a SVC-right PA superior cavo-pulmonary anastomosis is completed. This is obviously a major surgery requiring CPB but circulatory arrest is not necessary.

The purported benefits of the hybrid stage I-comprehensive stage II approach are:

1. Hybrid stage I has better perioperative outcomes than Norwood because it is physiologically so much less demanding.
2. Qp:Qs is more balanced than with Norwood-BT shunt because branch pulmonary artery banding reduces pulmonary blood flow.
3. Ventricular function is better preserved than in Norwood with Sano modification because a right ventriculotomy is avoided.
4. Cardiopulmonary bypass and deep hypothermic circulatory arrest are not inflicted on a fragile neonate. The avoidance of CPB during the neonatal period might result in better long term neuro-developmental outcomes.
5. The most physiologically stressful comprehensive stage II procedure is done at 4-6 months of age when the child is much more physically robust.
6. Circulatory arrest is not usually required for comprehensive stage II.
7. The Norwood aortic reconstruction might be technically easier when done at 4-6 months of age rather than during the neonatal period.
8. The most physiologically stressful operations are Norwood or comprehensive stage II. After comprehensive stage II, the child has more stable "Glenn physiology" with a series circulation rather than the more vulnerable parallel circulation that results after the Norwood procedure.

The hybrid-HLHS procedure has many attractive features but its role remains undefined. Despite better initial perioperative outcomes, the interstage mortality rate is still significant. In small series the interstage mortality is approximately 10% which is roughly the same rate quoted for Norwood with Sano modification but slightly better than that of Norwood-BT shunt. There are other theoretical concerns. Even though comprehensive stage II is done between 4-6 months when PVR is low, such a complex operation with a long duration of CPB might result in pulmonary hypertension that is problematic for the passive pulmonary blood flow of Glenn circulation. Also, the end goal for these patients is Fontan circulation, a basic requirement of which is normal caliber branch pulmonary arteries. Pulmonary artery banding in the neonatal period during the hybrid procedure could lead to stenosis in the long term.

## **Bidirectional Glenn Operation**

The Bidirectional Glenn (BDG) is anatomically termed a superior cavo-pulmonary anastomosis and is the second stage of palliation for single ventricle lesions. The first procedure was done in 1957 by a Russian surgeon Dr. Galankin in a patient with Tetralogy of Fallot. Dr. Glenn of Yale did the first American superior cavo-pulmonary anastomosis in 1958. This first operation involved suturing the SVC to the right PA after the right PA was divided from the main PA. The main PA was left in continuity with the right ventricle. This created “unidirectional” flow from the SVC to only the right PA and antegrade pulmonary blood flow from the right ventricle to left PA. For its time the operation was revolutionary, not in its technical challenge but in conceiving of the idea that systemic venous return could flow passively into the pulmonary circulation. The operation was not part of any staged palliation as it is today. The operation was later modified to anastomose the SVC to the right PA without dividing the right PA from the main PA thus creating SVC flow “bidirectionally” into both PAs.

Today the BDG is done as the second stage of palliation for single ventricle lesions. The timing of the BDG depends on PVR, which must fall from neonatal levels to allow central venous pressure to drive pulmonary blood flow. This usually occurs between 4-6 months of age.

### **Pre-Glenn Hemodynamic Cardiac Catheterization**

Hemodynamic catheterization prior to BDG is required to obtain the critical information about Qp:Qs and PVR which cannot be gained non-invasively using echocardiography or MRI. After Norwood the following data is needed to assess suitability for BDG:

1. Qp:Qs
2. Trans-pulmonary gradient
3. Pulmonary vascular resistance
4. Ventricular function
5. Atrio-ventricular valve regurgitation

The information needed for the calculations is:

1. Hgb
2. O<sub>2</sub> saturation measured in the SVC, pulmonary vein, and aorta
3. Pressures measured in the atrium, pulmonary artery, pulmonary vein
4. Cardiac output

All measurements are ideally done on room air, adjusting the anesthetic or sedation to achieve a normal blood pressure and normocarbica with or without controlled ventilation. Access to the PA in a patient post Norwood with a BT shunt is problematic. Crossing the shunt with even a small catheter may significantly decrease pulmonary blood flow. Also, placing a catheter through the shunt alters flow and can result in PA pressures that do not reflect the patient’s true physiology. To get around the problem, the PA pressure can be measured using a retrograde technique. A catheter is advanced into a pulmonary vein from the common atrium and the balloon is inflated to determine pulmonary vein wedge

pressure. This is exactly the same principle as floating a Swan-Ganz catheter into the PA. Just as the PA wedge pressure is assumed to be equal to the pulmonary vein pressure, the pulmonary vein wedge pressure is assumed to be equal to the mean PA pressure. Using tables to obtain an age adjusted estimate of  $VO_2$ , the  $Q_p$  and  $Q_s$  are determined by the Fick method and then the ratio of  $Q_p:Q_s$  is determined.

$$Q_p = \frac{VO_2}{Hgb \times 10 \times 1.34 (SpvO_2 - SpaO_2)} \quad Q_s = \frac{VO_2}{Hgb \times 10 \times 1.34 (SaO_2 - SvO_2)}$$

- $Q_p:Q_s$  can also be determined by the quicker method using only  $O_2$  saturation

$$Q_p:Q_s = \frac{SaO_2 - SvO_2}{SpvO_2 - SpaO_2} \quad \text{where } SaO_2: \text{Aorta, } SpaO_2: \text{PA} \\ SvO_2: \text{SVC, } SpvO_2: \text{Pulm vein}$$

The next calculation is the trans-pulmonary gradient which is defined as the pressure drop across the pulmonary circulation: PA pressure – pulmonary vein pressure. Pulmonary vein wedge pressure is used for PA pressure. After BDG, the  $Q_p$  will be upper body venous return which is about 60% of total venous return in a six month old due to the high cerebral blood flow. The  $Q_p:Q_s$  therefore will be 0.6. As pulmonary blood flow falls from its level post Norwood, the trans-pulmonary gradient will fall correspondingly.

Sample calculation:

Atrial pressure: 7                       $SaO_2$ : 80,  $SvO_2$ : 55  
 PV wedge pressure: 11               $SpvO_2$ : 100  
 PV pressure: 7                          $SpaO_2$ : 80 (not measured but assumed to be equal to  $SaO_2$ )

$$Q_p:Q_s = \frac{SaO_2 - SvO_2}{SpvO_2 - SpaO_2} = \frac{80 - 55}{100 - 80} = \frac{25}{20} = 1.25$$

Trans-pulmonary gradient = PV wedge – PV pressure = 11-7 = 4

$$\text{Glenn pressure (predicted)} = \text{Trans-pulmonary gradient} \times \frac{\text{Future } Q_p:Q_s}{\text{Present } Q_p:Q_s} + \text{Atrial pressure} \\ = 4 \times \frac{0.6}{1.25} + 7 = 9$$

$$\text{Pulmonary Vascular Resistance (PVR)} = \frac{\text{Trans-pulmonary gradient}}{\text{Pulmonary cardiac index}}$$

Pulmonary cardiac index is the  $Q_p$  from the Fick equation, divided by the body surface area (BSA). The result is quoted in Wood units and should be less than 2-3. From the

calculations, high predicted Glenn pressure can result from a high trans-pulmonary gradient or high atrial pressures. High trans-pulmonary pressures suggest pulmonary hypertension, either primary or secondary to lung disease. High atrial pressure suggests significant ventricular dysfunction or AV valve regurgitation. Ventricular function and AV valve regurgitation are assessed subjectively. When the predicted Glenn pressures are high (>15 mmHg), the patient is a poor candidate for BDG because there will be venous engorgement of the upper body and insufficient pulmonary blood flow. It is important to realize that the calculation of a predicted Glenn pressure is a rough approximation only.

Two other unique circumstances deserve mention. A left SVC (LSVC) which drains directly to the coronary sinus is a common abnormality. If it has not already been confirmed by echocardiography, it can be visualized in the cath lab. The surgeon will now have to do a bilateral BDG suturing the LSVC to the left PA in addition to the usual anastomosis of SVC to right PA. Failure to do so would mean only half the upper body venous return would circulate to the lungs ( $Q_p:Q_s$  would be approximately 0.3) and the patient would be very cyanotic. Decompressing veins is a term describing veins that flow between the SVC and IVC. After BDG, the SVC pressure will exceed IVC pressure. The decompressing vein provides a low resistance pathway for blood to flow from the SVC system to the IVC system. The result is that some SVC blood bypasses the lungs lowering the  $Q_p$ . Large decompressing veins, which are usually part of the azygous system should be coil occluded in the cardiac catheterization lab prior to surgery.

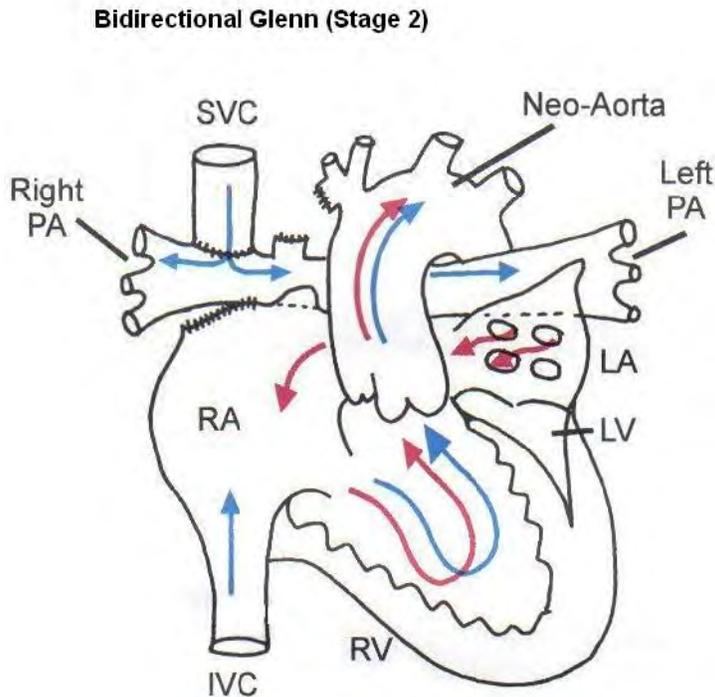


Figure 20: Bidirectional Glenn operation (*Pediatric Heart Surgery, A Ready Reference for Professionals, 3<sup>rd</sup> Ed. May, LE. Reprinted with permission*)

The BDG operation is usually well tolerated as the volume load on the ventricle is reduced. Surgery may be done without CPB but if CPB support is required cardioplegia and aortic cross clamping are rarely necessary. Vasoactive support of the heart post

operatively is usually minimal. A central line in the SVC now becomes a direct measure of pressure in the Glenn circuit. If there is no central line the surgeon can temporarily put a needle in the SVC and connect it to a transducer to measure Glenn pressure. The pulmonary vasculature is extremely sensitive to oxygen, carbon dioxide, temperature and pH. Abnormalities in the above variables can elevate pulmonary artery pressures which has the effect of raising Glenn pressure, causing venous engorgement of the upper body and lowering pulmonary blood flow. Moderate hyperventilation with 100% oxygen and correction of any acid-base problems is required. Narcotic dose is reduced to facilitate early extubation as pulmonary blood flow through the Glenn circuit is aided by the negative pressure generated with spontaneous breathing.

### **Hemi Fontan**

The BDG is an intermediate step between Norwood and Fontan operations. In Fontan physiology, all venous return bypasses the heart and returns directly to the lungs by passive flow. Hemi-Fontan achieves the same physiologic result as the BDG but configures the anatomy in a slightly different way. The BDG is an end to side anastomosis between the SVC and right PA. Following the BDG, the next surgery is an extra cardiac Fontan in which a conduit is sutured from the IVC to the inferior surface of the right PA. The Hemi-Fontan is technically more complex involving an atrial baffle in addition to the SVC-PA anastomosis. After the Hemi-Fontan the next surgery is a lateral tunnel Fontan in which a tunnel is created within the right atrium so that venous return from the IVC flows through the tunnel and into the pulmonary artery via the previously made connection between the SVC and right PA.

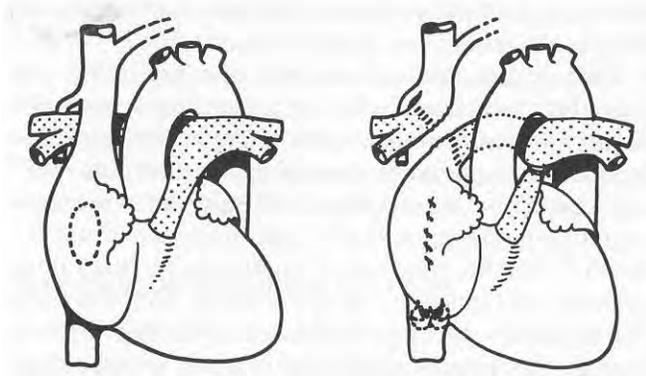
## **Fontan Operation**

The Fontan operation is anatomically a total cavo-pulmonary anastomosis and has become the third and final stage of palliation for single ventricle disease (HLHS type lesions). The first operation was done in 1968 by Dr. Fontan in France for a patient with tricuspid atresia. The initial criteria for those patients suitable for Fontan surgery was much more stringent than today. This reflected concern about the viability of totally passive pulmonary blood flow. Over time, various technical modifications have been developed to avoid or minimize some of the long term complications of Fontan circulation.

### **Classic Fontan and Atrio-pulmonary Fontan**

The first Fontan operation consisted of a Glenn type connection between the SVC and right PA combined with a right atrial to PA anastomosis. The right and left PAs were divided such that SVC flow from the upper body was solely directed to the right lung and lower body blood flow from the IVC exclusively perfused the left lung. Flow to the right lung would be purely passive while the left lung would have the benefit of atrial contraction to augment pulmonary blood flow. This anatomic arrangement was the original Classic Fontan which was very soon modified to a single right atrial-PA anastomosis with the belief that atrial contraction would help to assist pulmonary blood flow to both lungs. This variation of the original operation is known as an atrio-

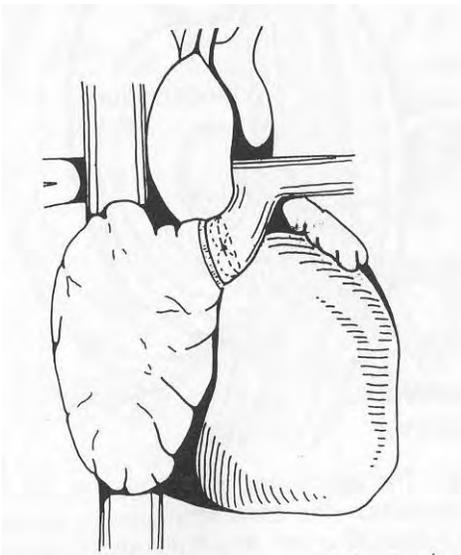
pulmonary Fontan. The terms Classic Fontan and atrio-pulmonary Fontan are sometimes used interchangeably but the anatomic arrangement is almost always a single right atrial-PA anastomosis.



Tricuspid atresia

Original Fontan operation

Figure 21: Original or Classic Fontan for tricuspid atresia. Unidirectional Glenn shunt from SVC to right PA and right atrial-PA anastomosis. (Fontan et al. *Surgical repair of tricuspid atresia. Thorax 1971;26:240-48.*)



Atrio-pulmonary Fontan

Figure 22: Single right atrial-pulmonary artery anastomosis (Kreutzer et al. *An operation for the correction of tricuspid atresia. J Thorac Cardiovasc Surg 1973;66:613-21.*)

The Glenn operation had set the stage for the Fontan procedure but in the early Glenn operation there was still antegrade pulmonary blood flow from the right ventricle into the

pulmonary circulation in addition to the SVC-pulmonary artery connection. In Fontan physiology, the total venous return would have to flow passively into the pulmonary circulation. Thus, the eligibility criteria were very strict when compared to today and it was felt that anything that could be done to assist venous return into the lungs would be beneficial.

Original criteria for Fontan operation:

Over 4 years of age  
Normal sinus rhythm  
Normal systemic venous return  
Right atrium of normal volume  
Mean PA pressure < 15 mmHg  
PVR < 4 Woods  
PA diameter at least 75% of aortic diameter  
Normal ventricular function  
No AV valve regurgitation

Many of these criteria have been liberalized and others obviously abandoned as the Fontan operation has evolved into the final stage of palliation for various single ventricle lesions.

### **Long term complications of Fontan circulation**

The theoretical advantage of an atrio-pulmonary Fontan did not last as the right atrium soon lost its contractile function and ended up as a greatly dilated chamber, providing a source for both thrombosis and dysrhythmias. Patients with Fontan operations can develop three major complications:

1. Thrombosis
2. Atrial tachyarrhythmia and sinus node dysfunction
3. Protein losing enteropathy

The dilated, non-contractile atrium served as reservoir for blood to pool and a ready source of thrombus. Atrial thrombus can embolize to the pulmonary vasculature, raising PA pressures and seriously impairing the passive blood flow of Fontan circulation. Atrial thrombus may also embolize paradoxically through residual right to left shunts. Stasis provides the conditions for venous thrombo-embolism but Fontan patients also satisfy another of Virchow's triad and are at risk for arterial thrombosis, secondary to a mild hypercoagulable state. There is no consensus on the appropriate form of thromboembolic prophylaxis.

Fontan patients show a steady increase in atrial tachyarrhythmias with an incidence of over 50% at 20 years. Fontan patients tolerate tachycardia very poorly and acute episodes usually require urgent treatment using medical therapy to control ventricular rate or cardioversion. The most common tachyarrhythmia is right intra-atrial reentrant tachycardia. Over time, episodic attacks of tachycardia become more frequent. Atrial fibrillation may occur with the loss of atrio-ventricular synchrony resulting in decreased

effort tolerance. Therapy for chronic atrial dysrhythmias consists of medication, catheter ablation or surgery. Given the complex anatomy, dilated atrium and atrial scar with suture lines from prior surgeries, it is not surprising that atrial arrhythmias may become refractory to standard treatment in many patients. Catheter ablation typically has high initial success rates that are not maintained.

Bradyarrhythmias, caused by sinus node ischemia, are common. Progressive fibrosis and scar around the sinus node, caused by prior surgical dissection, eventually leads to ischemia and clinical sinus node dysfunction. If accompanied by premature atrial contractions, sinus or junctional bradycardia may precipitate an intra-atrial reentry tachycardia. Thus, sinus node dysfunction also serves as a risk factor for the development of atrial tachyarrhythmias. Clinically significant bradyarrhythmias require pacing. Pacemakers pose special problems in the Fontan patient because the altered anatomy may preclude transvenous placement. Thus, Fontan patients who require pacing end up with epicardial leads placed via repeat sternotomy with all the risks that entails. Even though atrio-ventricular synchrony can be achieved with pacing, it is still not as good as intrinsic sinus rhythm.

Protein losing enteropathy (PLE) is a poorly understood complication in which patients develop hypoalbuminemia despite normal renal and hepatic function. Protein loss from the gut was initially thought to be due to high pressure in the Fontan pathway leading to hepatic congestion. The cause however, is more complex and not fully understood as some patients with low mesenteric venous pressure develop PLE and others with higher pressures do not. The significance of PLE is that it portends a poor prognosis even if the patient receives a heart transplant.

### **Modern Fontan Operation**

The theoretical benefits of an atrio-pulmonary Fontan did not materialize prompting the search for a better surgical option. Elegant studies of fluid dynamics revealed that the streams of SVC and IVC blood collided in the atrium, losing vital energy and forward propulsive force. Pulmonary blood flow was produced by the low pressure driving force of blood swirling in the dilated atrium. This pressure is even lower than a normal central venous pressure. The atrial dilation combined with scar from prior suture lines provided the ready substrate for thrombosis and arrhythmia. Lastly, atrio-pulmonary Fontan physiology led to sub-optimal pulmonary blood flow which resulted in problems for the systemic ventricle (either right or left morphology). Pulmonary blood flow becomes the preload for the systemic ventricle. Chronic “under loading” of the systemic led to ventricular dysfunction. The summary of all these complications is after 20 years of experience with the atrio-pulmonary Fontan, there was an urgent need to technically refine the operation and create a more efficient method of pulmonary blood flow, albeit within the confines of central venous pressure being the driving force. The solution was to directly connect the respective cava to the pulmonary circulation. (see Figure 23) The modern Fontan operation has reduced but not eliminated the major complications of Fontan physiology described earlier.

## Extra-cardiac Fontan

This modification consists of a conduit sutured from the IVC to the pulmonary circulation. The conduit is separate from the heart and is therefore “extra-cardiac”.

## Lateral tunnel Fontan

A baffle is constructed within the right atrium to direct IVC blood into the pulmonary circulation. The baffle forms a tunnel in the lateral part of the right atrium leading the operation to be called a “lateral tunnel Fontan”.

Both operations achieve the same physiologic result but the extra-cardiac Fontan results in less atrial suture lines. It is hoped that this will lead to fewer atrial dysrhythmias in the long term but this has not yet been clearly demonstrated.

## Fenestrated Fontan

The final modification of the Fontan was the creation of a fenestration. The fenestration is a hole (usually 4 mm) between either the extra-cardiac conduit or the lateral tunnel and the right atrium. The fenestration creates a right-to-left shunt which serves the purpose of being a “pop-off valve” for the Fontan circuit. Elevated venous pressures in the Fontan result in decreased pulmonary blood flow and a fall in cardiac output due to a reduction ventricular preload. At the expense of a small right-to-left shunt the fenestration preserves cardiac output in the setting of high venous pressures in the Fontan pathway. This fulfills the pediatric cardiologists’ motto, “It is better to be blue (decreased SpO<sub>2</sub>, good cardiac output) rather than gray (normal SpO<sub>2</sub>, decreased systemic perfusion).”

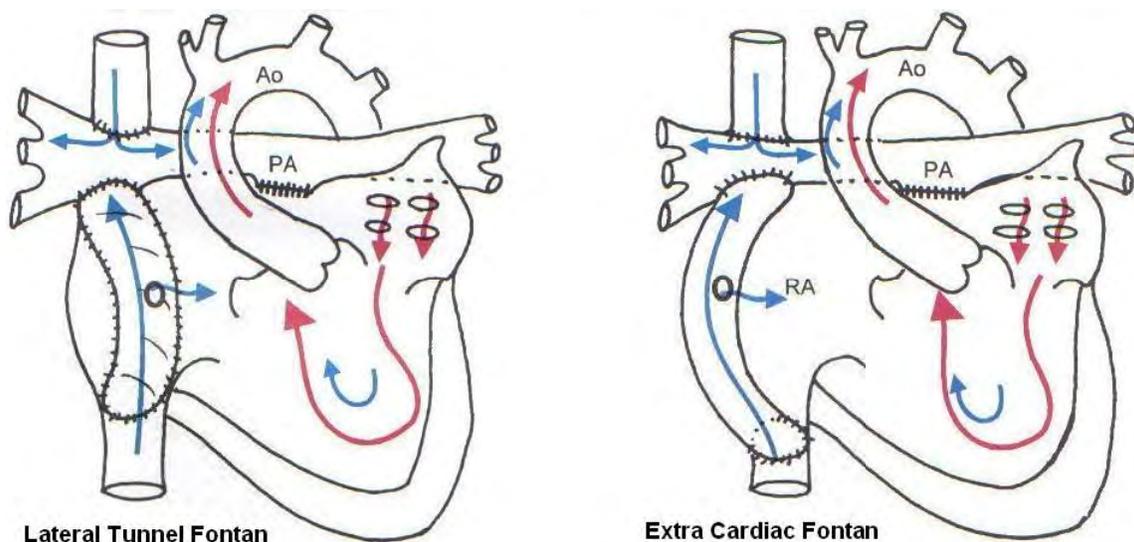


Figure 23: Modern Fontan operation in its two forms (with fenestration) (*Pediatric Heart Surgery, A Ready Reference for Professionals, 3<sup>rd</sup> Ed. May, LE. Reprinted with permission*)

## Pre-Fontan Hemodynamic Cardiac Catheterization

The principles involved and information required prior to the Fontan operation are the same as those needed before the Glenn operation. The major difference is that the pulmonary artery pressures and oxygen saturations can be measured directly because there is no modified BT shunt. A catheter in the SVC can be advanced into the pulmonary artery for direct measurement. The following data is needed:

1. Qp:Qs
2. Trans-pulmonary gradient
3. Pulmonary vascular resistance
4. Ventricular function
5. Atrio-ventricular valve regurgitation

The information needed for the calculations:

1. Hgb
2. O<sub>2</sub> saturation measured in the SVC, PA, pulmonary vein and aorta
3. Pressures measured in the atrium, pulmonary artery, pulmonary vein
4. Cardiac output

In Glenn physiology the Qp:Qs is approximately 0.6 but will rise to unity or 1 after the Fontan operation. It is critical to obtain an estimate of the pressures in the Fontan circuit after surgery. This information can only be determined by hemodynamic catheterization. Qp:Qs was reduced in the conversion from Norwood to Glenn but will now rise after completion of the Fontan.

Sample calculation:

$$Qp:Qs = \frac{SaO_2 - SvO_2}{SpvO_2 - SpaO_2}$$

where SaO<sub>2</sub>: Aorta, SvO<sub>2</sub>: SVC  
SpvO<sub>2</sub>: Pulm vein, SpaO<sub>2</sub>: Pulm artery

SaO<sub>2</sub>: 78, SvO<sub>2</sub>: 55, SpvO<sub>2</sub>: 100, SpaO<sub>2</sub>: 55 (O<sub>2</sub> saturation in the SVC is the same as PA)

$$Qp:Qs = \frac{78 - 55}{100 - 55} = \frac{23}{45} = 0.5$$

Trans-pulmonary gradient is the pressure drop across the pulmonary circulation which is:

Mean PA pressure – Pulm vein pressure

Since pulmonary blood flow will increase after the Fontan operation the trans-pulmonary gradient will similarly increase. The equation to estimate the Fontan pressure after surgery is:

$$\text{Fontan pressure (predicted)} = \text{Trans-pulmonary gradient} \times \frac{\text{Future Qp:Qs}}{\text{Present Qp:Qs}} + \text{Atrial pressure}$$

Sample calculation:

Mean PA pressure 10, Pulmonary vein pressure 6, Atrial pressure 6

$$\text{Fontan pressure (predicted)} = (10-6) \times \frac{1.0}{0.5} + 6 = 14$$

Similar to the pre-Glenn cath if the predicted pressures are high it can be due to either an elevated trans-pulmonary gradient or high atrial pressures. If the trans-pulmonary gradient is high the PVR can be determined simply by the following equation:

$$\text{PVR} = \frac{\text{Trans-pulmonary gradient}}{\text{Pulmonary cardiac index}}$$

If the predicted Fontan pressure is high but the trans-pulmonary gradient is normal then the atrial pressures must be high. This can be due to either significant atrio-ventricular valve regurgitation or poor ventricular function. When predicted Fontan pressures are high (greater than 20 mmHg), it signifies a poor prognosis and a search for any reversible causes must be undertaken. Similar to the pre-Glenn catheterization, the calculation of a predicted Fontan pressure is only a rough estimate. The true hemodynamics of this entirely passive pulmonary blood flow configuration can only be determined after surgery in a spontaneously breathing patient.

### Operative management

The Fontan procedure is completed on CPB but usually does not require aortic cross clamping. With the heart continuously beating there should not be any deterioration in ventricular function after separation from CPB. Optimizing Fontan physiology requires an understanding of the trans-pulmonary gradient. Conceptually, the trans-pulmonary gradient describes, in the form of an equation, the forces that determine pulmonary blood flow. In the cardiovascular system blood can only flow from point A to point B if a pressure gradient exists between those two points with point A having a higher pressure than point B. In a Fontan circulation, both vena cava are in continuity with the pulmonary vasculature. Therefore a central line in the internal jugular vein or femoral vein measures both central venous pressure (CVP) and PA pressure which is actually the same thing. The driving force for pulmonary blood flow or “point A” is CVP/PA pressure. The resisting pressure or “point B” is atrial pressure. Recall, that there is a common atrium in single ventricle anatomy that is disconnected from the vena cava. This requires the surgeon placing a trans-thoracic atrial line to measure atrial pressure.

Trans-pulmonary gradient = CVP/PA pressure --- Atrial pressure

The usual CVP/PA pressure to achieve successful separation from CPB with Fontan circulation is between 15-20 mmHg. This should result in a trans-pulmonary gradient of approximately 10 mmHg. When a CVP/PA pressure of over 20 mmHg is required to achieve a SpO<sub>2</sub> of at least 85-90% with an acceptable blood pressure two very different scenarios may exist. Consider these two examples

CVP/PA pressure 23 -- atrial pressure 7 = trans-pulmonary gradient 15 mmHg

CVP/PA pressure 23 -- atrial pressure 14 = trans-pulmonary gradient 9 mmHg

In the first scenario the high CVP/PA pressure is a result of elevated PVR. The second scenario of elevated CVP/PA pressure occurs with a normal trans-pulmonary gradient and is due to raised atrial pressure. It should be noted the trans-pulmonary gradient is a mechanistic oversimplification of the forces governing pulmonary blood flow in Fontan circulation. It does not take into account the intrinsic resistance of the pulmonary capillary network and the complexity of venous physiology but it does serve to illustrate the relevant hemodynamic and clinical principles for successful management.

Therefore the keys to Fontan circulation both in the immediate post-CPB period and long term are low PVR and good ventricular function. Optimizing PVR begins with getting the physiologic variables right: oxygen, carbon dioxide, acid-base status, temperature and lung recruitment. In anticipation of separation from CPB the anesthesiologist must ensure there is modest hyperventilation with FiO<sub>2</sub> 1.0, normal acid-base status, normothermia and full recruitment of the lungs. Recruitment of the lungs after ventilation was stopped during CPB is crucial and often under appreciated. The PVR is lowest when functional residual capacity (FRC) is restored to normal. Therefore, applying a small amount of positive end-expiratory pressure (PEEP) of 5 cmH<sub>2</sub>O, after full recruitment of the lungs is beneficial in maintaining FRC. Contrary to what has been preached in the past, low dose PEEP does not impede passive pulmonary blood flow. Rather it enhances pulmonary blood flow by maintaining FRC and lowering PVR. Any respiratory factors that impair ventilation (secretions, atelectasis, pleural effusion, bronchospasm) must be identified and corrected.

When the CVP/PA pressure is high with a normal trans-pulmonary gradient, the explanation is ventricular dysfunction. The cause is either reduced contractility or valve disease. Since there was no intra-cardiac surgery, valve function should be no different than preoperatively. Similarly, the lack of aortic cross clamping should preserve contractility and any deterioration post-CPB would be unusual. Ventricular function is easily assessed with TEE during separation from CPB. Frequently, there are small elevations of PVR which are presumed to occur as a result of inflammatory mediators induced by CPB. Milrinone is usually started during CPB and has the twin benefits of pulmonary vasodilation and enhanced inotropy. It is often combined with low dose adrenergic support (dopamine, epinephrine, norepinephrine).

The coronary sinus is not included in the Fontan pathway leading to a small obligatory right-to-left shunt. Further, the creation of a fenestration increases the right-to-left shunt

while protecting cardiac output. Therefore a systemic O<sub>2</sub> saturation in the high 85%-90% range is to be expected in the postoperative period. Narcotic doses for surgery are reduced to aim for early extubation. Some centers use neuraxial opioids administered after induction to facilitate early extubation. Postoperative pain is significant because the Fontan operation is usually the third sternotomy. The negative pressure generated by spontaneous breathing augments flow through the Fontan circuit. Obviously, Fontan physiology is compatible with positive pressure ventilation because this is the method of ventilation during separation from CPB. However, the general trend is to aim for extubation in the operating room or within the first few hours of admission to the ICU. Even with ideal Fontan hemodynamics, the obligatory central venous hypertension (approximately 15-20 mmHg) results in significant postoperative effusions for many children. Lymphatic drainage must eventually return to the venous circulation. Raised venous pressure impedes the normal lymphatic flow and results in ascites and pleural and pericardial effusions. This incidence has been reduced but not eliminated by routine fenestration of the Fontan pathway.

### **The "Failing Fontan" and Fontan Conversion**

The "failing Fontan" is a descriptive term for an extremely sick group of patients. Their Fontan physiology is "failing" because of some combination of cyanosis, effort intolerance, arrhythmia, ventricular dysfunction, thrombosis or the poorly understood phenomenon of protein losing enteropathy. Not all of these problems are amenable to intervention. A cardiac catheterization is indicated to assess the hemodynamic state and determine whether any therapeutic options exist. Cyanosis may be due to intra-cardiac shunting that may be amenable to a catheter based closure device. Elevated PVR may require creation of a fenestration that functions as a small right-to-left shunt, preserving ventricular preload at the expense of systemic desaturation. Relief of mechanical obstructions in the Fontan pathway with dilation and stenting may be another therapeutic option. Gradients of only a few millimeters of mercury can be very significant and debilitating in Fontan physiology.

Physiologically these patients are fragile. Failing Fontan physiology is more likely but not confined to patients with an atrio-pulmonary connection. As explained in an earlier section the dilated atrium of an atrio-pulmonary Fontan is ready substrate for arrhythmia and thrombosis. The loss of forward energy as blood swirls in the dilated atrium leads to a chronically "underloaded" ventricle and low cardiac output state. In this setting atrial tachyarrhythmias cause rapid decompensation whereas atrial fibrillation with ventricular rate control results in a more chronic deterioration in cardiovascular performance. Pacing is problematic in Fontan patients. A pacing wire in a dilated atrium has a considerable risk of thrombosis. Fontan anatomy does not allow trans-venous pacing access to the ventricle.

If failing Fontan physiology occurs in either of the modern Fontan iterations (lateral tunnel or extra-cardiac) few therapeutic options exist aside from percutaneous interventions to relieve mechanical obstruction or pacing. Atrio-ventricular pacing is the most efficient configuration but requires epicardial leads which are placed surgically. A full sternotomy (usually the patient's fourth) is required although a more limited sub-xiphoid approach may be successful. The final option, should none of the previous

therapies prove successful, is of course transplantation. In the present era, with the dearth of organs, patients may be “bridged to transplant” with the Berlin Heart ventricular assist device (VAD). This is a VAD in multiple sizes designed for pediatric patients from infant to teenagers.

A surgical option for the failing atrio-pulmonary Fontan is Fontan conversion. Mavroudis and colleagues have been leaders in the surgical conversion of the atrio-pulmonary Fontan to an extra cardiac connection.<sup>17</sup> The procedure involves multiple phases. There is revision of the atrio-pulmonary connection to the more energy efficient extra-cardiac Fontan. As the procedure has been refined by Mavroudis’ group, sophisticated intra-operative electrophysiological mapping is done to precisely guide ablation of the atrial tachyarrhythmia pathways. Lastly, epicardial atrio-ventricular pacing leads are placed. This is likely the highest risk surgery in the adult CHD population involving a cohort that is uniquely compromised by failing Fontan physiology. In the hands of an experienced perioperative team it can be done with a low mortality and morbidity. Experience has shown that results are best when surgery is done earlier in the disease process rather than repeatedly trying medical therapy or percutaneous interventions in the hope of avoiding a high risk surgery.

## **Tricuspid Atresia**

Tricuspid atresia (TA) occurs when there is complete absence of the tricuspid valve and no connection between the right atrium and right ventricle. All systemic venous blood in the right atrium must pass across either a PFO or ASD and enter the left atrium. This creates a complete right-to-left shunt at the atrial level. With no flow into the right ventricle, there is severe right ventricle hypoplasia. There is a VSD in most cases (90%). Tricuspid atresia exists in two major forms. There are normally related great arteries (70% incidence) and transposition in the remainder of the cases (30%). Despite the VSD, the underdeveloped right ventricle is usually inadequate to provide pulmonary blood flow. Therefore, PGE1 is required to maintain pulmonary blood flow via the PDA. In TA with transposition, the situation is reversed. The small right ventricle leads to the aorta and the left ventricle leads to the pulmonary artery. There is some antegrade flow from the left ventricle across the VSD and into the aorta but this is generally not enough to meet systemic demands. Thus, PGE1 is required to maintain systemic blood flow across the ductus from the pulmonary circulation.

Presentation is in the neonatal period with cyanosis. Once the diagnosis is made, PGE1 must be started to stabilize the patient. The SpO<sub>2</sub> reflects the degree of pulmonary blood flow. A Qp:Qs of unity results in a SpO<sub>2</sub> of 75-80%. Increases in SpO<sub>2</sub> above 80% result from a rising Qp:Qs and there is progressive left ventricular volume overload. Regardless of whether there is transposition, all patients with tricuspid atresia will have single ventricle physiology. In the newborn period the operation performed to establish pulmonary blood flow is a modified BT shunt. This allows PGE1 to be discontinued. When the PVR begins to fall later in infancy, the patient will proceed to Bidirectional Glenn and Fontan operations on the same timetable as patients with HLHS.

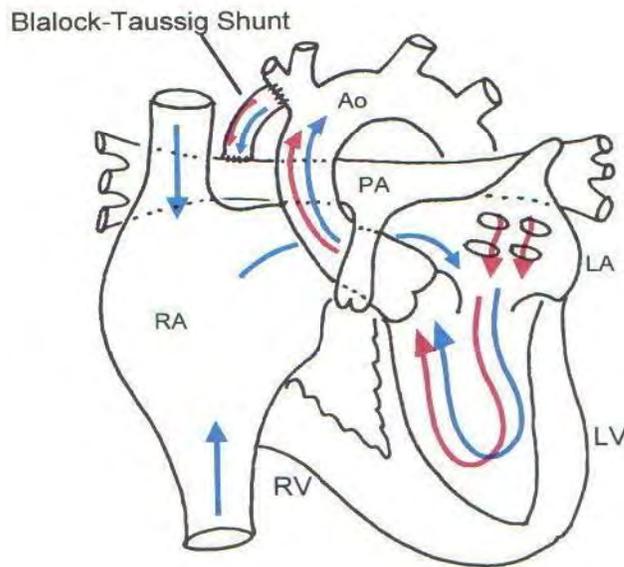


Figure 24: BT shunt for tricuspid atresia in newborn period (*Pediatric Heart Surgery, A Ready Reference for Professionals, 3<sup>rd</sup> Ed. May, LE. Reprinted with permission*)

## Pulmonary Atresia with Intact Ventricular Septum

Pulmonary atresia with intact ventricular septum (PA/IVS) is a rare lesion that has unique features affecting surgical management. Atresia of the pulmonary valve results in a complete right-to-left shunt at the atrial level. The right ventricle is small and variably hypoplastic. Pulmonary blood flow is ductal dependent. The infant is cyanotic within the first few hours of life. The diagnosis is made by echocardiography and PGE1 is started to maintain ductal patency and provide pulmonary blood flow. Multiple surgical options now exist depending of the severity of right ventricle hypoplasia and a unique feature of PA/IVS know as right ventricular dependent coronary circulation (RVDCC).

The echocardiogram report will comment on the tricuspid valve. A competent tricuspid valve raises suspicion for RVDCC and a cardiac catheterization is required. When the tricuspid valve is competent, the small right ventricle generates very high pressures during ventricular systole. This pressure leads to the formation of sinusoidal channels between the right ventricle cavity and the coronary arteries. These are called coronary-cameral fistulae. The presence of these fistulae is confirmed angiographically with contrast injections into the right ventricle and the coronary arteries. The next step is to look for stenoses in proximal segments of the major coronary arteries. When these stenoses are severe, all myocardium distal to the stenoses is supplied retrograde from the right ventricle. The myocardium has some degree of chronic ischemic because it receives deoxygenated blood at a high perfusion pressure from the right ventricle. The coronary circulation is now referred to as right ventricle dependent (RVDCC).

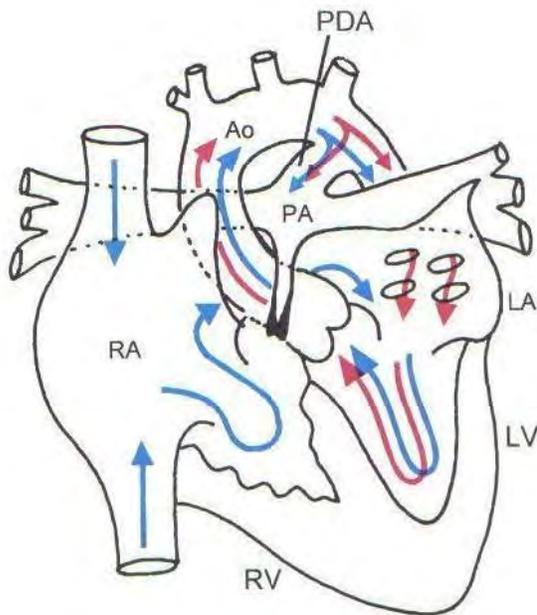


Figure 25: Pulmonary atresia with intact ventricular septum (*Pediatric Heart Surgery, A Ready Reference for Professionals, 3<sup>rd</sup> Ed. May, LE. Reprinted with permission*)

A procedure that decompresses the right ventricle by relieving the pulmonary atresia (pulmonary valvotomy, RV-PA connection) will decrease right ventricular systolic pressure dramatically. The right ventricle will not be able to generate enough pressure to perfuse the coronary-cameral fistulae. Myocardium dependent on retrograde flow via the coronary-cameral fistulae will develop severe ischemia or frank infarction. The possibility of RVDCC must be ruled out as the first step in the surgical decision making.

The next step is to assess the right ventricle for possible two ventricle repair which depends on the degree of hypoplasia. The echocardiogram and the cardiac catheterization will provide the required information. Lastly, the tricuspid valve is assessed for the degree of regurgitation. In some the cases the tricuspid valve will have an Ebstein like deformity with severe regurgitation. The right ventricle function, degree of tricuspid regurgitation and presence or absence of RVDCC results in the following surgical options:

RVDCC: These patients obviously cannot have the right ventricle decompressed and will need single ventricle surgical palliation. The newborn operation is a modified BT shunt to provide pulmonary blood flow. Subsequently, Glenn and Fontan operations will be needed.

No RVDCC, good right ventricle function: These fortunate patients are candidates for a two ventricle repair. They will have either a pulmonary valvotomy or a trans-annular patch. The caveat here is that the tricuspid valve cannot have significant regurgitation.

No RVDCC, poor right ventricle function: Despite the absence of RVDCC, these patients do not have a right ventricle that is adequate for two ventricle repair. They will be treated as if they have RVDCC and require single ventricle surgical palliation.

No RVDCC, moderate right ventricle function: These patients are candidates for the so called “one and a half ventricle repair”. The right ventricle will not be able to handle the entire systemic venous return. A Bidirectional Glenn operation off loads the right ventricle and provides a secure source for pulmonary blood flow. The right ventricle is decompressed with a pulmonary valvotomy or trans-annular patch allowing it to pump venous return from the lower body to the lungs.

In many cases, it is not clear cut which option is best. The presence or absence of RVDCC can be easily defined, but the assessment of right ventricle function and tricuspid valve integrity is much harder and it is these factors which have a critical bearing on the eventual outcome.

## **Total Anomalous Pulmonary Venous Return**

Total anomalous pulmonary venous return (TAPVR) occurs when the entire pulmonary venous return of oxygenated blood enters the right atrium. The pulmonary veins draining each lung flow together, forming a confluence posterior to the left atrium. Blood from the confluence can take four routes to eventually reach the right atrium, giving rise to the various anatomic subtypes.

Supracardiac TAPVR (45%): A vertical vein from the confluence drains superiorly to one of the major veins of the upper body (innominate, SVC, azygous). The most common arrangement is for a left vertical vein to drain to the innominate vein.

Cardiac TAPVR (25%): The confluence drains via a connecting vein to the coronary sinus.

Infracardiac TAPVR (25%): A vertical vein from the confluence drains inferiorly, passing through the esophageal hiatus. Usually the vertical vein empties into the portal vein. Other possibilities include draining into the IVC, ductus venosus or hepatic veins.

Mixed TAPVR (5%): In the mixed type of TAPVR, the left sided and right sided veins drain separately without coming together to form a confluence. The most frequently occurring arrangement is for the left sided veins to drain superiorly to the innominate vein and the right sided veins to drain directly to the right atrium or coronary sinus.

The pathophysiology of TAPVR is complete mixing of the systemic and pulmonary venous return causing cyanosis. A communication at the atrial level (ASD/PFO) must be present to permit a right-to-left shunt and ensure left ventricular output and systemic perfusion. In addition to cyanosis, there is significant right sided volume overload leading to right heart failure. When the atrial level communication is restrictive, the right sided volume overload is exacerbated since less blood can pass into the left atrium. The right heart is maximally stressed and the left heart does not have sufficient preload to meet systemic demands. As a temporizing measure these patients may require a balloon atrial

septostomy to enlarge a restrictive PFO and relieve some of the right side volume overload prior to surgery. Alternatively, they may undergo definitive repair as an urgent procedure. A mildly restrictive PFO is well tolerated but a severe restriction demands urgent intervention.

The term “obstructed total veins” refers to patients in whom the pulmonary venous return is obstructed somewhere along its pathway to the right atrium. Infracardiac TAPVR is the most common subtype to present with obstruction because of two reasons. The long course of the vertical vein from the confluence to the abdomen puts it at risk to become obstructed or kinked. This is a mechanical obstruction to flow. Also, when an infracardiac lesion drains to the portal vein, pulmonary venous blood must pass through the liver before returning to the right atrium. The liver has an intrinsic resistance which can impair pulmonary venous drainage. The result is that pulmonary venous pressure rises leading to pulmonary edema and pulmonary arterial hypertension. These patients are cyanotic by virtue of mixing in the right atrium but they decompensate rapidly because the obstructed pulmonary veins lead to pulmonary edema. Pulmonary venous blood is now no longer fully oxygenated which creates increased cyanosis. Pulmonary arterial hypertension increases right ventricular and right atrial pressure which worsens the pulmonary venous congestion. A critical cycle ensues of pulmonary venous obstruction leading to pulmonary edema, worsening hypoxemia, right heart failure and further pulmonary venous obstruction. This is one of the rare situations in modern congenital heart surgery where definitive surgery with CPB is performed as an emergency.

Surgery for TAPVR consists of dividing the vertical vein and anastomosing the confluence to the left atrium. In the case of cardiac TAPVR, the coronary sinus is baffled to the left atrium since there is no vertical vein to ligate. Patients with obstructed TAPVR are extremely sick. Every effort must be made to correct acidosis and support the circulation with inotropes if necessary. After the repair, pulmonary circulation remains very labile because of the prior obstruction. The in utero obstruction leads to a pulmonary vasculature which is small, underdeveloped and reactive. This, combined with the inflammatory insult of CPB, can lead to significant problems with pulmonary hypertension. Pulmonary hypertensive crises are the most feared development after surgical repair of obstructed TAPVR. Initial management consists of the usual steps to lower pulmonary artery pressures. Hyperventilating with 100% oxygen, avoiding hypercarbia, correcting acidosis, maintaining temperature and ensuring an adequately deep level of anesthesia are all essential. Milrinone's effects on the pulmonary vasculature are beneficial as long as the systemic vasodilation is well tolerated. Inhaled nitric oxide is the last line of defense against a pulmonary hypertensive crisis. It is often started empirically along with all the other measures described, in an attempt to do everything possible to ensure a smooth separation from CPB.

# Cardiac Catheterization Laboratory

Anesthesia for pediatric cardiac catheterization provides multiple challenges:

1. The location is remote from the operating room.
2. The spectrum of disease varies from healthy to critically ill.
3. Children with single ventricle physiology or PHTN are high risk patients with unique cardiovascular limitations.
4. Cardiovascular effects of anesthetic drugs, the mode of ventilation,  $FiO_2$ ,  $PaCO_2$  and acid-base status all must be integrated in such a way that diagnostic studies truly reflect the child's cardiac status.
5. Higher risk interventional procedures now outnumber diagnostic studies by 2:1

## Diagnostic studies

Hemodynamic catheterization quantifies the gradients, shunts, pulmonary blood flow, PVR, systemic blood flow and SVR upon which clinical decision making is based. The following indications exist for diagnostic studies.

### A. Hemodynamic assessment

The clinician, especially a trainee, should pay close attention to the calculations performed because it enhances the pathophysiologic understanding of CHD. Understanding the hemodynamic data makes one a physician who comprehends the pathophysiology of CHD rather than merely a technician. Refer to the section on Hemodynamic Calculations for a more detailed explanation. The most common scenario for diagnostic catheterization is prior to the Glenn and Fontan procedures.

### B. Examination of complex anatomy

The complexity of certain CHD conditions and/or poor echocardiographic windows may necessitate a diagnostic catheterization.

### C. Evaluation of myocardial function and pulmonary hypertension

Hemodynamic indices are often measured in children with heart failure or with pulmonary hypertension. Refer to later sections on Anomalous Origin of the Left Coronary Artery from the Pulmonary Artery and Pulmonary Hypertension for a more detailed description of these conditions.

### D. Evaluation after heart transplant

Standard follow up requires endomyocardial biopsies at predetermined intervals after the transplant. Children range from those with excellent heart function having an elective procedure to those who present in severe heart failure from rejection.

### E. Electrophysiological studies

These are usually initially diagnostic studies to induce and map the arrhythmia followed by therapeutic intervention if suitable.

## **Catheterization laboratory environment**

The catheterization laboratory is an unfamiliar location remote from the operating room. The environment is cramped and access to the anesthesia machine, drug carts, intravenous poles, monitors and most critically, the patient, is restricted. Even in the new so called "hybrid labs" designed for combined percutaneous and surgical procedures, the extra space is entirely devoted to the surgical scrub table along with room for a CPB pump or extracorporeal membrane oxygenator (ECMO) circuit. The lesson here is that it is crucial to spend a few extra moments organizing the "workspace". Monitoring and intravenous lines, along with the anesthesia machine and breathing circuit need to be organized, labeled and easily accessible. Other equipment should not restrict access to the child unless absolutely necessary. All standard emergency cardiac medications should be available in doses appropriate for the child and placed in an easily accessible location. Placing medications on the anesthesia cart as is our usual habit is not sensible if the anesthesia cart is trapped behind other equipment. Trying to resuscitate a patient cannot be done competently when one cannot even locate the injection port of the intravenous tubing. Lastly, in an anticipated difficult airway, consideration should be given to inducing anesthesia and managing the airway in the operating room where equipment and other anesthesia personnel are readily available. After the airway is secured, the child can be transferred to the catheterization laboratory.

## **Anesthetic goals & techniques**

The presence of an anesthesiologist ranges from near universal to rare, depending on institutional practice and the comfort of the cardiologist in managing sedation. The issue of who manages sedation and anesthesia for cardiac catheterization is relevant in light of the recent findings of the Pediatric Perioperative Cardiac Arrest Registry.<sup>18</sup> Fully one-third of cardiac arrests happened in children with CHD and most of these occurred during non-cardiac surgery (50%) or cardiac catheterization (20%).

The fundamental art and challenge of anesthesia for pediatric cardiac catheterization is to provide a safe anesthetic that accurately reflects the child's true physiology. During the early days of pediatric cardiac catheterization the "lytic cocktail" or "CM3" (chlorpromazine, meperidine and promethazine) was given by intramuscular injection. This produced a child with stable cardiorespiratory parameters at the expense of prolonged deep sedation and lack of titrability. Today the wide array of sedatives and anesthetics come with far greater risk cardiorespiratory depression, but can be titrated to effect and are relatively short acting. Light or "conscious sedation" as used for most adults has minimal cardiorespiratory effects when compared to general anesthesia but is not feasible for most children. Almost all children receive general anesthesia or deep sedation with the difference between the two defined by whether the airway has been instrumented. This is a semantic point because one can have a fully anesthetized child breathing spontaneously with a natural airway. Anesthetic techniques range from spontaneous ventilation with a natural airway and intravenous sedation with intermittent ketamine/midazolam/fentanyl, or continuous infusions of propofol or dexmedetomidine to intubation, positive pressure ventilation, use of muscle relaxants and maintenance with volatile anesthesia. The technique must be suited to the child, their CHD, their cardiovascular reserve and the planned procedure.

A hemodynamic study must accurately reflect the true physiologic state of the child. Given that there are no hemodynamically neutral anesthetic drugs, this requirement is challenging. Furthermore, there is no surgical stimulation to offset the effect of anesthetic drugs, making the maintenance of baseline hemodynamics even more difficult. The cardiovascular effects of anesthetic drugs in children with CHD have been reviewed in an earlier section. Different ventilatory modes affect venous return and the loading conditions for the heart. The ventilatory mode,  $\text{FiO}_2$ ,  $\text{PaCO}_2$ , temperature and acid-base status all critically affect hemodynamic measurements.

A brief mention here is necessary on the subject of emergence. Large sheaths have been placed in the femoral artery/vein or the internal jugular vein. Pressure is held at the site at the conclusion of the procedure and a compressive dressing is applied. Hemostasis at the site is tenuous and can easily be disrupted by rises in arterial pressure from agitation or increases central venous pressure from coughing and straining during emergence. If bleeding resumes, then pressure must be re-applied at the site. This is poorly tolerated in an awake child. Deep extubation is unfortunately not a solution to the problem of emergence agitation or delirium because it does not prevent the problem. It simply transfers it to an event that will occur in PACU instead of the catheterization laboratory. Therefore, supplementation of deep extubation with further intravenous agents is required to reduce the incidence of emergence agitation and prolong the time to full awakening. Multiple drugs have been successfully used to prevent emergence agitation with no one being clearly better than another. However, given the lack of post-procedure pain, opioids are not the best choice. Small doses of propofol (0.5-1 mg/kg) will ensure a quiet child for approximately the first 15-30 minutes in PACU. Dexmedetomidine (0.5mcg/kg) is another attractive choice in this setting.

### **Interventional Procedures**

While not universal, pediatric cardiologists are more likely to request anesthesia for interventional procedures because they have more potential for hemodynamic disturbance and complications. The children cover the spectrum from those with well compensated conditions to those who are critically ill. Given the possible hemodynamic disturbance, many anesthesiologists opt to secure the airway although for most procedures this is not strictly necessary. If one thinks along the lines of the "ABCs" of trauma resuscitation, when the airway is secured with an endotracheal tube, "A" and "B" are taken care of, in case the circulation or "C" deteriorates. The most likely complication is vascular injury which can occur at the access site allowing direct compression to stop bleeding. Bleeding may also involve the great vessels or the heart resulting in life threatening hemorrhage or cardiac tamponade. Blood must be available and interventional procedures which use bigger vascular access sheaths and have a higher rate of complications should only be performed in centers with cardiac surgery and ECMO capabilities. Great vessel bleeding during balloon angioplasty can be temporized by inflation of the balloon. Resuscitation, rapid transfusion and arterial monitoring are best done through the vascular access sheaths. Cardiac tamponade can be treated by needle pericardiocentesis, guided fluoroscopically or echocardiographically. Major vascular injuries most often will require surgical repair or ECMO. Arrhythmias are usually iatrogenic and respond to catheter withdrawal. Rarely heart block ensues and temporary

pacing is necessary. Other rare events include thrombo-embolism, air embolus and embolism of device parts such as coiling wires or stent material. Hemodynamic compromise may result from the cardio-respiratory effects of anesthesia. The highest risk groups are single ventricle infants and children with PHTN.

### **Electrophysiological (EP) studies**

It is difficult to make generalizations about anesthetic management for children having EP studies. By far and away the most common diagnoses are teenagers with pre-excitation (Wolff-Parkinson White) or atrio-ventricular nodal re-entry tachycardia (AVNRT). Both of these occur in the setting of a structurally normal heart. Ideally, EP mapping and potential ablation would require light or conscious sedation. This would prevent suppression of the arrhythmia by anesthetic agents. While most anesthetics do not have a direct anti-arrhythmic effect, arrhythmias may be more difficult to induce under general anesthesia. Even in teenagers, the duration of EP studies usually requires general anesthesia. A standard induction is well tolerated because most children have a structurally normal heart however there are a few key points to remember. Intubation, mechanical ventilation, neuromuscular blockade and a reduced depth of anesthesia will cause the least suppression of the arrhythmia and allow the most successful mapping of the lesion. Because the procedure is not painful, narcotics are rarely indicated as they most often lead to hypotension. Blood pressure should be kept in the high normal range by appropriately titrating the depth of anesthesia. This will allow the child to better tolerate the hemodynamic consequences of inducing the tachyarrhythmia. Volume should be restricted especially during long procedures as the continual flushing of the EP catheters can deliver significant fluid. The total volume is kept track of by the EP technician. Transcutaneous patches are routinely placed on all patients for cardioversion or defibrillation. Immediate complications with ablation are rare but may lead to complete heart block especially in AVNRT because the ablation pathway is so close to the AV node. Pacing can be achieved using the existing catheters. Lastly, perforation resulting in tamponade can occur but is extremely rare.

### **Miscellaneous lesions**

#### **Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA)**

Anomalous origin of the left coronary artery from the pulmonary artery is known by its acronym ALCAPA. The left coronary artery originates from the pulmonary artery, supplying deoxygenated blood to the majority of the left ventricle. Collateral flow from the right coronary is usually insufficient to alleviate coronary ischemia. At birth, the infants are usually asymptomatic. Indeed, it is not the deoxygenated blood supply to the left ventricle that causes problems because there are many cyanotic congenital heart lesions in which the left ventricle is supplied with desaturated blood. Symptoms develop later in infancy. Coronary perfusion to the left ventricle is: diastolic BP-LVEDP

As PVR drops in infancy, the pulmonary diastolic BP falls resulting in a critical decrease in coronary perfusion pressure. The presence of large collaterals may be harmful because

they allow blood to flow from the high pressure in the right coronary artery into the low pressure left coronary artery and then into the pulmonary artery. In effect the left coronary acts as a fistula between the right coronary and the pulmonary artery. This creates a steal phenomenon. The ischemic heart fails and the infant presents in CHF. The initial echocardiogram reveals a structurally normal heart with severe hypokinesis. The differential diagnosis of an infant with a structurally normal heart presenting in CHF is:

#### ALCAPA

Dilated (idiopathic) cardiomyopathy

Post-viral cardiomyopathy

Cardiac catheterization may be required to rule in or out ALCAPA. The distinction is critical as the treatment is surgical. Post viral cardiomyopathy has a better prognosis for recovery than dilated cardiomyopathy but both conditions are managed medically. The surgical management of ALCAPA is to reimplant the anomalous left coronary artery from the PA to the aorta. If this is not technically possible, the left coronary artery is ligated to prevent it from being a fistulous connection between the right coronary artery and the PA.

These infants have profound decreases in left ventricular function. The echocardiographic appearance is usually more severe than the clinical presentation suggests. This is because infants are by definition sedentary. When they left have left ventricular dysfunction severe enough to be tachypneic at rest and diaphoretic with feeds, the ejection fraction is 20% at best. Their degree of cardiovascular reserve is minimal. Usually these infants have already been medically stabilized with milrinone +/- inotropes and have sufficient intravenous access and usually, invasive monitoring. If already intubated, the anesthetic is "plug and play" consisting of vigilance, +/- neuromuscular blockade and continuation of sedative infusions (usually fentanyl/midazolam). Those infants not already intubated present a major challenge. Spontaneous ventilation with a natural airway is an attractive option. Ketamine and midazolam given as cautious intermittent boluses or infusion is a common method employed and is very cardiovascularly stable. Propofol's effect on SVR and dexmedetomidine's vagotonic effect on heart rate are major limitations of these drugs. Generally these patients will not hemodynamically tolerate the depth of volatile anesthesia required to breathe spontaneously through an LMA or ETT. Therefore if the airway must be secured, controlled ventilation with an ETT is usually chosen and combined with neuromuscular blockade and low dose volatile anesthesia or some combination of ketamine/midazolam/fentanyl. Options for induction are ketamine, etomidate or fentanyl/midazolam. Critical points to remember are that the sicker the heart, the less it can tolerate hypoxia. Therefore at all cost, one must not lose the airway. The consequences of laryngospasm or airway obstruction, regardless of the type of anesthesia chosen, can be severe. Aggressive positive pressure ventilation will decrease preload and is poorly tolerated. Lastly, ventricular dilation makes the heart prone to arrhythmia which may occur spontaneously or in response to catheter manipulation. Aggressive diuresis may result in abnormal electrolytes (potassium, magnesium) and increase the propensity for arrhythmia. Defibrillation equipment must be immediately available along with cardiac resuscitation medications, inotropic infusions and antiarrhythmic drugs.

## Vascular ring

Vascular ring is a rare lesion of the aortic arch causing circumferential entrapment of the trachea and esophagus. The most common abnormality is a double left sided aortic arch. The arch splits, encircling and compressing the trachea and esophagus. The next most common lesion is a right aortic arch with an aberrant retro-esophageal left subclavian artery. With a right arch the aberrant left subclavian artery is the last rather than the first major branch of the aortic arch. The aberrant left subclavian artery traces a path from right to left and posterior to the trachea and esophagus. The vascular ring is created by the ligamentum arteriosum (ductal remnant) which connects the aberrant left subclavian artery and the left PA. Numerous other anatomic variants of vascular ring exist.

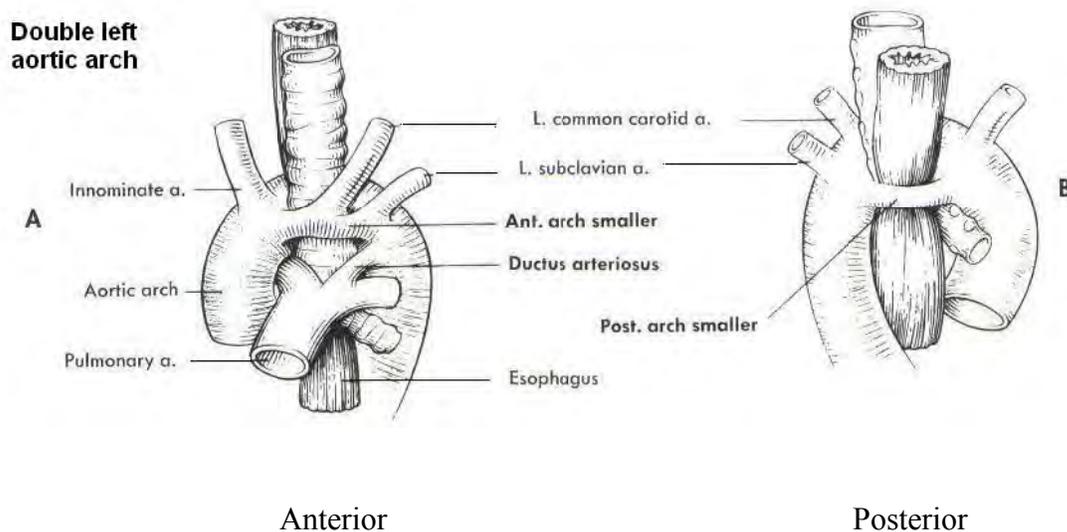


Figure 26: Double left aortic arch (*Smith's Anesthesia for Infants and Children, 6<sup>th</sup> Ed* Motoyama EK, Davis PJ. Reprinted with permission)

Patients present with either respiratory or gastrointestinal symptoms. Feeding difficulties with frequent spitting up or vomiting may initially be interpreted as reflux. Wheezing, stridor or recurrent respiratory infections may be part of the initial presentation. Barium swallow may show esophageal compression. The diagnosis is confirmed with MRI or contrast CT scan of the chest. Surgical approach is via thoracotomy with the exact procedure dependent on the type of vascular ring.

## Ebstein's anomaly

This congenital malformation of the tricuspid valve was first described by Ebstein in 1866. It consists of the inferior displacement of the septal and posterior leaflets of the tricuspid valve. The anterior leaflet is abnormally large and redundant. The features of the lesion are:

1. Tricuspid regurgitation
2. An enlarged right atrium
3. Hypoplastic right ventricle

The inferior displacement of two of the valve leaflets results in the atrium being enlarged with tissue that is morphologically right ventricular in origin. This is known as an “atrialized right ventricle”.

Presentation is variable in severity, ranging from neonates with critical right ventricle failure to adults who are only mildly symptomatic. When the right ventricle is severely hypoplastic, the patient will need single ventricle surgical palliation. Those patients with a moderately hypoplastic right ventricle may be candidates for the “one and a half ventricle” repair which involves a superior cavo-pulmonary anastomosis (Bidirectional Glenn) to relieve some of the right ventricle volume load. Those patients deemed to have good right ventricle function will have definitive repair consisting of tricuspid valvuloplasty and reduction of the atrialized right ventricle segment.

### **Aorto-Pulmonary Window**

Aorto-pulmonary window consists of a direct communication between the ascending aorta and the main pulmonary artery. The pathophysiology is a large left-to-right shunt. The aorto-pulmonary window acts as a huge PDA. Similar to other left-to-right shunt lesions the newborn period is one of relative stability because PVR is high. As PVR falls later in infancy, the volume of blood shunting left-to-right increases. Presentation is usually an infant in heart failure due to pulmonary over circulation. Auscultation may be normal because the unrestrictive aorto-pulmonary window may not have sufficiently turbulent flow to generate a murmur.

After medical stabilization with diuretics, the patient requires surgery. An aorto-pulmonary window is not like a PDA that can simply be ligated and divided. There is actually a fusion of the great vessels over a small discrete area. Usually the main PA is incised to expose the window and the defect patched from the PA side, obviating the need for an aortic suture line.

### **Cor triatriatum**

Cor triatriatum is Latin for “three atria”. The lesion consists of a left atrium which is divided into two compartments which communicate via a narrow orifice. Pulmonary venous return flows into one chamber, across the narrow orifice, into the second chamber and then across the mitral valve. The narrow orifice between the two compartments of the left atrium provides an obstruction to pulmonary venous inflow. The pathophysiology is the same as that of valvular mitral stenosis. Over time the pulmonary venous hypertension leads to pulmonary edema and pulmonary arterial hypertension. Surgical management consists of resecting the membrane which divides the left atrium. If the lesion occurs in the right atrium the correct terminology is cor triatriatum dexter.

### **Shone’s Complex**

This rare congenital lesion involves left sided obstruction at multiple levels. The four classic sites are:

1. Supravalvar mitral ring
2. Parachute mitral valve
3. Subaortic stenosis
4. Coarctation

The supravalvar mitral ring divides the left atrium in a different anatomic configuration than a Cor triatriatum but has the same clinical effect of mitral stenosis. In parachute mitral valve, the appearance of the mitral valve is similar to that of a parachute with chordae from both leaflets attaching to one central papillary muscle instead of two. The clinical effect is also that of mitral stenosis. Subaortic stenosis and coarctation is self explanatory. There is a high incidence of bicuspid aortic valve and VSD associated with Shone's Complex. The presentation and surgical management depend on where the most critical lesion is located.

### **Congenitally Corrected Transposition (L-TGA)**

Congenitally corrected transposition is the condition in which there is discordance at the atrio-ventricular connection and the ventriculo-arterial connection. Using the Van Praagh system the atrial situs is solitus (S), there is L-looping of the ventricles (L) and the aorta is to the left of the PA. Therefore the nomenclature is S,L,L transposition which is often known as L-TGA. Other names are corrected TGA, ventricular inversion, double TGA, discordant TGA and inverted TGA.

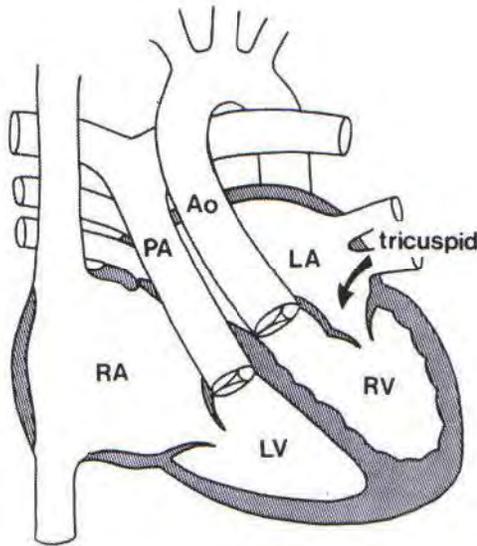


Figure 27: Congenitally corrected transposition (*The natural and "unnatural" history of congenitally corrected transposition. Lundstrom U, Bull C, Wyse RKH, Somerville J. American Journal of Cardiology. Reprinted with permission*)

The L-looping of the ventricles is associated with several other abnormalities:

1. Ventricular septal defect
2. LVOT obstruction
3. Congenital heart block
4. Ebstein's deformity of the tricuspid valve with regurgitation

In the absence of any of the above conditions, the patient will be asymptomatic at birth. The natural history is progressive right ventricle dysfunction due to the burden of being the systemic ventricle. There is also an increasing incidence of heart block over time.

Those patients who present in infancy or as newborns do so secondary to a severe form of one of the associated abnormalities listed above. Surgical management is directed to fix the specific problem. For example, closure of the VSD, relief of the LVOT obstruction or tricuspid valve repair. The patient would still be left with a systemic right ventricle and at risk for long term deterioration of ventricular function.

A controversial option is the “double switch” procedure. The aim is to create a series circulation with the left ventricle as the systemic ventricle. This is achieved by a Senning or Mustard procedure combined with the Arterial Switch.

## **Pulmonary Hypertension**

Pulmonary hypertension (PHTN) is defined by a mean pulmonary artery pressure of 25 mmHg in the presence of a normal pulmonary wedge pressure. PHTN is not synonymous with increased PVR. The equation for PVR is:

$$\frac{\text{Mean PAP—PA Wedge pressure}}{\text{Pulmonary Cardiac index}}$$

The result is expressed in Wood units (normal < 3). In the case of a large left-to-right shunt, the pulmonary artery pressures may be high but the right sided cardiac output may also be increased by a commensurate amount. Therefore the PVR is not elevated, signifying that the pulmonary vasculature is able to accept the higher flow and the resistance is not yet increased. If untreated, eventually the increased flow will lead to pulmonary vascular changes. Pulmonary pressures remain elevated but the PVR will rise and the pulmonary blood flow will decrease.

Pulmonary hypertension may be idiopathic, known as primary pulmonary hypertension (PPH) or secondary to a wide array of other conditions. If PHTN is secondary to CHD, the usual cause is an unrepaired large left-to-right shunt. The PVR increases as changes in the pulmonary vasculature develop, the shunt becomes bidirectional and then eventually right-to-left, causing systemic desaturation. Fixed or very high PVR with right-to-left shunting is classically described as Eisenmenger’s Syndrome but is now very rare in developed nations. Despite cyanosis, Eisenmenger’s syndrome has a better prognosis than PPH because cardiac output is maintained at the expense of systemic hypoxemia. In PPH, increasing PVR causes right heart strain and then frank failure. Systemic cardiac output falls which is more poorly tolerated than well perfused cyanosis.

This is the highest risk subset of children with CHD and has been recently well reviewed.<sup>19</sup> An estimate of the degree of pulmonary pressures can be gained echocardiographically and should be classed as sub-systemic, systemic or

suprasystemic. Children with PHTN require a hemodynamic catheterization to calculate PVR and determine the response to pulmonary vasodilators to guide future therapy. Hemodynamic catheterization requires the measurement of pulmonary artery pressures, cardiac output and PVR. The measurements are first done with a  $\text{FiO}_2$  0.21. The measurements are then repeated on  $\text{FiO}_2$  1.0 and then  $\text{FiO}_2$  1.0 with nitric oxide 40 parts per million. Prognosis and treatment is dependent on the degree of PVR responsiveness to a high  $\text{FiO}_2$  and nitric oxide. Patients with significant improvement are usually placed on the phosphodiesterase V inhibitor, sildenafil (Viagra™) which has some selectivity for the pulmonary vasculature. Epoprostenol is used for those with more severe disease thought to be unresponsive to sildenafil. Epoprostenol is a pulmonary vasodilating prostaglandin. It is also known by its other names prostacyclin or PGI<sub>2</sub> and marketed as Flolan™. Epoprostenol must be given by a continuous infusion through a secure long term IV catheter. Another option is bosentan, an endothelin receptor antagonist with the benefit of oral administration. Its role in pediatric practice is evolving. In children with PHTN secondary to left-to-right shunts, the findings of the diagnostic catheterization are needed to determine the appropriateness and timing of future surgery as well as the need for specific pulmonary vasodilators perioperatively.

The anesthetic principles for PHTN are to limit the physiologic derangements that elevate PVR and to prevent rises in PVR from potent stimuli. The physiologic abnormalities that elevate PVR are acidosis, hypercarbia, hypoxia, hypothermia and significant atelectasis. Hypercarbia, if accompanied by an adequate compensatory metabolic response to prevent acidosis, does not significantly elevate PVR. However, most children do not retain  $\text{CO}_2$  at baseline and thus hypercarbia causes acidosis. A very potent stimulus is laryngoscopy and endotracheal intubation. This raises the question of why endotracheal intubation is the most common form of airway management. There does not appear to be a good reason other than that most anesthesiologists seem nervous with anything less than a fully secured airway in the child with PHTN. While it may be difficult to administer iNO and different levels of  $\text{FiO}_2$  through a tight fitting facemask, a LMA provides an excellent airway with the ability to vary  $\text{FiO}_2$  and deliver iNO. PVR is lowest when patients ventilate to a normal FRC. However the preservation of FRC does not mandate intubation and positive pressure ventilation. A physiologic amount of PEEP can be added to spontaneous ventilation with an LMA with excellent preservation of lung volumes. Positive pressure ventilation can also be delivered through an LMA with effective tidal volume and low peak pressures. Spontaneous ventilation augments venous return when compared to positive pressure ventilation. This is important for the hypertrophic right ventricle which is preload dependent. Lastly, at the end of the case an LMA can be smoothly removed at a light plane of anesthesia, avoiding the elevations in PVR secondary to coughing and straining on the endotracheal tube. The LMA offers the following benefits: avoidance of stimulating laryngoscopy, a smooth emergence, the ability to alter  $\text{FiO}_2$  and administer iNO, spontaneous ventilation at a depth of anesthesia that usually prevents hypercarbia, preservation of right ventricular preload and the ability to deliver positive pressure ventilation if necessary. An LMA obviously may not be suitable for all children or in all situations but it clearly has a role and is an underused method of airway management in PHTN. What is most critical for children with PHTN is meticulous airway management regardless of the device chosen.

Based on preoperative echocardiography the severity of PHTN should have been

estimated as subsystemic, systemic or suprasystemic. General experience has found that when pulmonary artery (PA) pressures are less than 50% systemic, children are low risk and tolerate anesthesia well. As PA pressures approach systemic, risk increases considerably. No ideal drug for PHTN exists. Most commonly used are sub-anesthetic doses of multiple drugs in the attempt to minimize the hemodynamic and respiratory effects of one drug given in full anesthetic dose. The crucial cardiovascular determinant is the status of the right ventricle. Therefore a recent echocardiogram must have been done and reviewed. In the setting of PHTN the right ventricle develops compensatory hypertrophy with preserved function. Only at the end stage of PHTN does the right ventricle dilate and fail. Thus most children with PHTN will have a preload dependent, stiff and non-compliant right ventricle with relatively preserved function. As long as venous return can be supported with intravenous fluids, children will tolerate a cautious titrated induction of anesthesia with any combination of the available agents. The preceding statement only precludes a mask induction with deep levels of volatile anesthesia, followed by placement of an intravenous catheter. Intravenous access prior to induction also allows the prompt treatment of laryngospasm rather than waiting for the onset of intramuscular succinylcholine. In young children in whom intravenous access is presumed to be difficult, intramuscular ketamine is an option followed by an intravenous catheter. A frequent question regarding ketamine is whether its sympathomimetic effect will exacerbate pulmonary hypertension. It appears that as long as hypercarbia and hypoxia are avoided ketamine does not have deleterious effects on PVR.

A management plan for a pulmonary hypertensive crisis must be formed prior to the induction of anesthesia. The best prevention for trouble is skilled airway management. Laryngospasm, breath holding, airway obstruction and bronchospasm are all potentially lethal events in children with PHTN. As mentioned earlier, skilled airway management is not synonymous with endotracheal intubation. One must remember that children with PHTN can become hypoxic for all the same reasons that healthy children become hypoxic. Therefore before frantically calling for iNO, ensure there is an appropriate  $\text{FiO}_2$ , confirm the circuit or endotracheal tube is not kinked or blocked with secretions, make sure the LMA has not become dislodged and check the carbon dioxide absorbent is not depleted. If a pulmonary hypertensive crisis develops the management principles are to reverse the inciting event should one be identified, administer pulmonary vasodilators and support right heart function. In the catheterization laboratory the inciting event may be something as simple as allowing the child to breathe room air while hemodynamic parameters are assessed. If there is an intra-cardiac shunt, at least initially, systemic blood pressure will be maintained but profound cyanosis may occur. If the heart is structurally normal, there will be both hypotension and cyanosis. The child should immediately be switched to enriched oxygen ( $\text{FiO}_2$  1.0) and ventilation controlled. Depending on the rapidity at which the pulmonary hypertensive crisis develops, options are intravenous milrinone or iNO. Both should be combined with epinephrine to support right heart function. Rapid echocardiographic assessment will reveal the intracardiac effects (septal bowing to the left, pulmonary insufficiency, tricuspid regurgitation) of the sudden rise in PA pressure. Epinephrine will usually offset systemic vasodilation induced by milrinone. Initially, epinephrine can be given through a peripheral intravenous in small bolus doses (1 mcg/kg). If necessary, an infusion can be started through a central catheter already in place for the procedure. Unless the pulmonary hypertensive crisis is due to a reversible respiratory event (laryngospasm, airway obstruction) it usually does

not quickly resolve. Therefore, children remain intubated with ventilation controlled, FiO<sub>2</sub> 1.0 and iNO. Continued vasopressor support is usually required. If all standard resuscitative measures have failed, ECMO is an option of last resort.

Surgery for patients with significant pulmonary hypertension carries a high perioperative risk. The anesthetic plan is directed at meticulous attention to airway management, ventilation, preservation of lung volumes and provision of deep anesthesia prior to airway manipulation. Acute rises in PVR secondary to hypoxia, hypercarbia, acidosis, hypothermia and light anesthesia must be prevented at all cost. Nitric oxide should be available. Right heart decompensation secondary to a pulmonary hypertensive crisis is treated with cautious volume expansion and inotropic support (epinephrine and milrinone). Isoproterenol is used by some centers as a single agent because its beta-1 effect supports the myocardium and the beta-2 stimulation provides pulmonary vasodilation. In the event of hypotension or desaturation not promptly responding to standard therapy, a TEE is recommended. Right heart volume status, contractility and a measurement of the degree of pulmonary hypertension can all be rapidly accomplished using TEE.

## **Cardiopulmonary Bypass**

Significant differences exist in the management of cardiopulmonary bypass (CPB) between adult and pediatric patients.

### **Temperature**

Pediatric patients, especially neonates undergoing complex repair are subjected to great extremes of temperature. The majority of adult CPB is usually warm or mildly hypothermic (34 C). Older children and infants with simple lesions are managed with mild to moderate hypothermia (34 C to 28 C) but complex neonatal repairs are usually done under deep hypothermia (18C) with circulatory arrest (DHCA). The DHCA induces profound changes in blood viscosity, microcirculatory flow and oxygen consumption.

### **Hemodilution and CPB Prime**

In relation to total blood volume, infants suffer marked hemodilution on CPB. The prime volume of the CPB pump for an adult is roughly 33% of blood volume. Even with the smallest CPB circuits, the prime volume far exceeds the blood volume of an infant. To avoid severe anemia, the CPB circuit must be primed with packed RBC. Depending on the institution various other combinations of crystalloid, colloid and blood products (FFP) are added. Some centers use fresh whole blood for CPB prime. There is no consensus on what type of CPB prime is best.

### **Blood gas management**

With the extremes of hypothermia used in pediatric CPB, role of blood gas management takes on greater importance. Alpha-stat management does not correct blood gases to account for the patient's temperature. It is preferred for adult patients because it preserves

enzyme function and maintains cerebral autoregulation. Cerebral autoregulation is important in limiting cerebral blood flow for adult patients because neurologic injury from CPB is primarily embolic. Increased cerebral blood flow places the patient at higher risk of an embolic injury (air, plaque).

Pediatric patients are at risk for a different type of neurologic injury based on their very high cerebral oxygen consumption ( $CMRO_2$ ). Correcting blood gases to the patient's temperature is pH-stat management. It provides cerebral blood flow that is far in excess of the brain's metabolic needs during severe hypothermia. The abundant cerebral blood flow helps to ensure even and complete cooling of the brain which is critically important prior to circulatory arrest.

### **Ultrafiltration**

Despite priming with blood components, the pediatric patient undergoes significant hemodilution on CPB. In addition, the large volume of non-endothelialized circuit tubing induces a profound inflammatory response with multiple end organ effects. In an effort to minimize this response, two ultrafiltration techniques were devised. Conventional ultrafiltration occurs while the patient is being rewarmed on CPB. Modified ultrafiltration filters the patient's blood volume once CPB has been terminated. Both techniques serve to reduce positive fluid balance, decrease total body water, remove inflammatory mediators, improve hemodynamics and raise hematocrit.

### **Hematocrit**

Adults and older children tolerate a hematocrit in the low 20s without difficulty on CPB. In adults, a low hematocrit (approximately 20%) is often accepted as the price for avoiding RBC transfusion which is associated with worse outcomes perioperatively and in the short to medium term. Due to their higher metabolic rate infants and in particular newborns, require a higher hematocrit. Oxygen delivery, even in a hypothermic environment with markedly reduced  $VO_2$ , may be insufficient with a "normal" hematocrit. The safest lower limit of hematocrit for infants undergoing complex repair has not been determined. The most comprehensive work in this area from Children's Hospital Boston initially suggested the hematocrit should be 30%. Levels substantially below 30% were associated with worse neurodevelopmental outcomes. A revisiting of this work now suggests the lower limit of hematocrit may be closer to 25%.<sup>20</sup> These results are restricted to neurodevelopmental outcomes only in the absence of major stroke. The incidence of thromboembolic or hemorrhagic stroke is not related to the level of hematocrit on CPB.

### **Circulatory arrest**

For complex repair in neonates involving repair of the ascending aorta and arch, the surgeon must have a bloodless field. The traditional approach has been deep hypothermia (18C) with total circulatory arrest. With DHCA, surgical exposure is ideal because the field is bloodless and free of cannulae. Once the period of DHCA is begun the surgeon is "on the clock". Controversy exists as to the safe length of cerebral ischemic time. Less

than 30 minutes appears safe. Between 45 and 60 minutes is a gray area and neurologic injury goes up significantly when DHCA time exceeds 60 minutes.

Another option is deep hypothermia with selective cerebral perfusion. Selective cerebral perfusion involves perfusing the brain only via a graft sutured into the innominate artery. This approach maintains cerebral perfusion. Only low flows are required because the brain's oxygen consumption at 18C is very low. Another advantage is that the surgeon has more time to complete the repair. Outcome studies suggest that avoiding DHCA leads to improved neurodevelopmental results but this remains an area of controversy. In the hands of an experienced surgeon with DHCA time of less than 30 minutes, it would be difficult to show a benefit of one technique over the other.

### **Reoperation and antifibrinolytics**

Compared to conventional adult heart surgery, a much larger percentage of congenital heart surgery involves reoperation. It is well described that reoperation leads to increased blood loss. The surgical trauma caused by dissecting through dense adhesions and scar tissue activates the coagulation system and subsequently the fibrinolytic cascade with conversion of plasminogen to plasmin. The large amount of circulating plasmin overwhelms the body's ability to neutralize it with  $\alpha$ -2 anti-plasmin. Circulating plasmin dissolves stable clots and impairs platelet adhesion through inhibition of von Willebrand Factor. Fibrinogen degradation products (FDP) are released by the dissolution of stable clots. These FDP are natural anticoagulants, inhibiting both platelets and clotting factors.

Two classes of drugs with antifibrinolytic action are used to decrease blood loss. Epsilon amino caproic acid (EACA) and tranexamic acid (TA) are lysine analogues. They bind to the lysine site of plasminogen, preventing its conversion to plasmin. In the US, EACA is widely used with TA being preferred in Europe. Both drugs have proven benefit, with modest reductions in blood loss, low cost and a very favorable side effect profile. Aprotinin is a serine protease inhibitor extracted from bovine lung. Numerous enzymes have a serine site that is inhibited by aprotinin. This explains in part, aprotinin's diverse action on the coagulation, complement and inflammatory systems. Aprotinin's antifibrinolytic action is mediated via direct inhibition of plasmin. Despite aprotinin's inhibition of plasmin, its exact mechanism of decreasing blood loss is not fully understood. Aprotinin is effective in reducing blood loss, especially in the patient with multiple prior sternotomies but it is very costly and can lead to severe anaphylactic reactions upon repeat exposure.

Aprotinin has generated a large amount of controversy. Uncontrolled studies in adults undergoing conventional heart surgery note increases in renal failure, stroke and myocardial infarction. Aprotinin use fell dramatically and the manufacturer has now withdrawn the drug from the market. However, its use in pediatrics remains uncertain. Pediatric patients are very different from the usual adult cardiac surgery patient who has hypertension, hyperlipidemia, diabetes and widespread vascular disease. It would be very inappropriate to extrapolate this adult data to the pediatric population. Furthermore, many centers advocate its use in complex newborn procedures (Norwood, Arterial Switch) because of its anti-inflammatory properties. Lastly, in comparison to the dosing of the lysine analogues which is well standardized, aprotinin dosing is hugely variable. There

are high and low dose regimens and some centers dose based on body weight while others use body surface area (BSA). Doses based on BSA tend to be much larger. More studies are clearly needed in this area if aprotinin returns to the market.

## **Syndromes with Congenital Heart Defects**

For additional information on these syndromes or information on additional syndromes, see Baum VC and O'Flaherty JE: Anesthesia for Genetic, Metabolic and Dysmorphic Syndromes of Childhood. Lippincott, Williams and Wilkins 2007

### **Down syndrome**

Down syndrome or trisomy 21 has a high incidence of congenital heart disease. The most common defect is a VSD. Down syndrome patients comprise the greatest percentage of those with CAVC, giving the erroneous impression that CAVC is the most common congenital cardiac defect in trisomy 21. Another feature of congenital heart disease in Down syndrome is the propensity to develop pulmonary hypertension. With classic left-to-right shunt lesions such as VSD and CAVC, Down syndrome children display an accelerated progression to pulmonary hypertension.

### **DiGeorge syndrome**

Dr. DiGeorge, in 1965, described this constellation of findings in infancy:

- Congenital heart disease with cono-truncal abnormalities
- Velo-pharyngeal incompetence
- Immune dysfunction with absent thymus or thymic hypoplasia
- Parathyroid dysfunction with hypocalcemia
- Renal abnormalities
- Distinctive facies

The story of DiGeorge syndrome is an interesting one. Approximately 25% of patients with a clinical suspicion of the syndrome were found to have a deletion of the long arm of Chromosome 22. The deletion is called 22q11 signifying its location on the q arm of the Chromosome 22. The deletion was seen by standard microscopy. The remaining 75% of patients had no visible chromosomal lesion. More precise testing known as Fluorescence In Situ Hybridization (FISH) with DNA probes for specific genetic foci revealed that all DiGeorge syndrome patients have a 22q.11 micro-deletion.

Meanwhile another group of patients were described as having Velo-Cardio-Facial syndrome. These patients were generally older children and came to medical attention primarily for problems with velo-pharyngeal incompetence resulting in speech difficulties. They had distinctive facies and congenital heart disease. Sophisticated chromosomal testing with FISH showed that they had exactly the same 22q11 micro deletion. Thus, 22q11 is a genetic abnormality with different phenotypic manifestations.

The cono-truncal abnormalities suspicious for DiGeorge syndrome are Tetralogy of Fallot (especially with right aortic arch), Truncus Arteriosus, Interrupted Aortic Arch and VSD in the suprasternal region. Two features of DiGeorge syndrome are of special relevance to the anesthesiologist. All blood products must be irradiated to kill donor leukocytes which can incite a serious graft versus host response in the DiGeorge patient with depressed immune function. Secondly, parathyroid dysfunction causes hypocalcemia. This is very significant in the newborn period. The neonatal myocardium requires an adequate serum level of ionized calcium because intracellular calcium sequestration and release is immature. When weaning from CPB it is crucial to ensure a normal serum ionized calcium. Requirements of exogenous calcium may be very large and an infusion is often required.

## **Heterotaxy**

Heterotaxy is synonymous with situs ambiguous. Situs solitus is the normal arrangement and situs inversus is the mirror image. Heterotaxy exists when the normal “sidedness” of thoraco-abdominal viscera is lost. There is a very high association of congenital heart disease. CAVC is very common followed by other complex lesions particularly of the single ventricle variety that are often associated with abnormalities of systemic and pulmonary venous return. Heterotaxy exists in two main subtypes although there is considerable overlap. Surgery is directed to the specific CHD lesion but abnormalities of the IVC significantly affect single ventricle children who will eventually require Fontan completion. The worst prognostic indicator in Heterotaxy is congenital complete heart block. Immune function may be compromised even in the polysplenia variant. Skeletal, neurologic and urologic abnormalities may also occur.

Asplenia or Right Isomerism:

1. Two morphologically right lungs (each has three lobes)
2. Two morphologically right atria
3. Midline liver
4. Absent spleen
5. Variable stomach location
6. Abdominal malrotation
7. Normal IVC

Polysplenia or Left Isomerism:

1. Two morphologically left lungs (each has two lobes)
2. Two morphologically left atria
3. Midline liver
4. Multiple small spleens
5. Variable stomach location
6. Abdominal malrotation
7. IVC interruption with azygous continuation to the atrium

## Williams syndrome

First described in 1961, Dr. Williams noticed the following association:

Developmental delay  
Characteristic facial appearance (previously known as “elfin” facies)  
Growth retardation  
Friendly nature (so called “cocktail party personality”)  
Hypercalcemia  
Mandibular and dental abnormalities  
Congenital heart disease

We now know the syndrome occurs because of a deletion on Chromosome 7. The cardiac lesions of Williams syndrome are characterized by stenosis at multiple levels due to defects in elastin:

1. Supravalvar aortic stenosis
2. Branch pulmonary stenosis
3. Valvar pulmonary stenosis
4. Coronary stenoses (ostial and in the major epicardial vessels)
5. Stenoses of the aorta and major branches

Supravalvar AS is progressive but branch PA stenosis tends to improve over time.

The most feared problem in Williams syndrome is sudden death. The etiology of sudden death is believed to be severe myocardial ischemia leading to left ventricular dysfunction +/- arrhythmia. The supravalvar AS is clinically equivalent to valvar AS in adults with one key difference. The coronary ostia are located between the native aortic valve and supravalvar stenosis. With significant supravalvar AS the coronary arteries are chronically exposed to high systolic pressures which induce tortuosity and a premature atherosclerotic type lesion. The correlation between the degree of supravalvar AS and the severity of coronary artery disease is poor. The supravalvar AS leads to left ventricular hypertrophy with increased myocardial oxygen consumption. The hypertrophied myocardium is vulnerable to ischemia in the presence of coronary stenoses. Children with an associated ventricular outflow tract obstruction are at highest risk.

A recent review describes the challenge of these children.<sup>21</sup> There is a clear risk of perioperative death due to coronary ischemia. While hypotension from any cause (anesthesia, sedation, hemorrhage, sepsis) is obviously dangerous in Williams syndrome with supravalvar AS, what is confusing is that hypotension is not always the precipitant of coronary ischemia. Coronary ischemia in the setting of supravalvar AS will lead to hypotension which worsens the ischemia and sets in motion the “death spiral” well described in aortic stenosis. The fact that coronary ischemia may result from causes other than systemic hypotension makes prevention difficult. There is much we do not understand about sudden death in Williams syndrome. Predictably if cardiac arrest occurs, the response to resuscitation is poor as would be expected in the setting of severe AS. The adult corollary of these children would be a patient with good ventricular function, AS and significant coronary artery disease (CAD). The physiologic challenge is

delivering an anesthetic that combines the management principles of AS while optimizing the balance of myocardial oxygen supply and demand.

Aortic stenosis: maintain preload, preserve afterload to maintain coronary perfusion pressure, avoid negative inotropes

Coronary artery disease:

Increase myocardial oxygen supply: relative bradycardia, adequate oxygen content of the blood, maintain coronary perfusion pressure

Decrease myocardial oxygen demand: relative bradycardia, decrease wall stress, decrease contractility

These children do not routinely get coronary angiograms so the nature of their CAD is unknown but it should be presumed to be significant. The severity of the supraaortic AS and branch PA stenosis should be known from the most recent echocardiogram. The key physiologic goals are maintenance of blood pressure and heart rate control. An intravenous induction is preferred because it offers the ability to titrate anesthetic drugs, control heart rate with beta blocker and support intravascular volume with fluid. Multiple attempts at securing intravenous access in an infant with poor peripheral veins may do more harm than good. The child may become very irritable and tachycardic and while future intravenous sites are being lost due to failed attempts. In this scenario there are two unattractive possibilities: intramuscular ketamine or mask induction with volatile anesthesia and placement of an intravenous catheter as soon as possible. Ketamine will maintain blood pressure but may induce tachycardia. The period of time under deep volatile anesthesia without intravenous access may result in considerable hypotension. Clearly neither option is ideal. The risk of sudden death mandates observation in monitored setting. If anesthesia was only required for procedural sedation, overnight observation is sufficient. After surgery the child must be monitored in an appropriate setting for the duration of the hospital admission.

## **Noonan Syndrome**

In 1963, Dr. Noonan published her observations of the syndrome that now bears her name:

Hypertelorism

Micrognathia

Webbed neck

Short stature

Pectus excavatum/carinatum

Bleeding diathesis

Congenital heart disease

The common congenital heart defects with Noonan's Syndrome are valvar pulmonary stenosis, ASD, VSD and hypertrophic obstructive cardiomyopathy. Prior to surgery, patients require a hematologic work up. Difficult airway considerations apply.

## Turner Syndrome

Dr. Turner, in 1938, described the syndrome we now know carries the karyotype XO:

Short stature  
Webbed neck  
Shield chest (widely spaced nipples)  
Congenital heart disease

The common congenital heart defects with Turner Syndrome are coarctation, bicuspid aortic valve and aortic dissection.

## Long QT syndrome

Long QT syndrome is a disorder of cardiac ion channels ( $K^+$ ,  $Na^+$ ) leading to a delayed repolarization which manifests on electrocardiogram (ECG) as a prolonged QT segment. The significance of delayed repolarization is a propensity to develop the ventricular arrhythmia “Torsades de Pointes”, which can deteriorate into ventricular fibrillation. Patients who are otherwise healthy present with syncope and tragically sometimes sudden death.

An ECG is required for any patient who has suffered a syncopal episode. Approximately 10% of patients will have a normal ECG despite having Long QT syndrome. If there is a high degree of suspicion, these patients may be diagnosed by Holter testing or an EP study. A careful history will often reveal a family member who died suddenly or “passed out” without explanation.

Long QT exists in acquired and congenital forms. Acquired forms are due to a very long list of medications that interfere with cardiac ion channel function. The FDA “black box” warning about droperidol was issued because high doses have been shown to prolong the QT interval and possibly lead to ventricular arrhythmia. These doses are far in excess of standard anti-emetic dosing. Chronically malnourished patients such as alcoholics with hypomagnesemia are the other group of patients with acquired Long QT syndrome. Congenital Long QT syndrome has two forms:

Romano Ward syndrome: autosomal dominant

Jervell Lange Nielson syndrome: autosomal recessive associated with deafness

Usually a premature beat during the prolonged repolarization period catches the myocardium at a vulnerable time and precipitates Torsades de Pointes. Conditions causing tachycardia (stress, anxiety, exercise) are the biggest risk factor. Tachycardia results in a shorter R-R interval but the QT interval does not shorten correspondingly. Thus the chance of a beat occurring during the vulnerable period of ventricular repolarization is increased. The mainstay of treatment is tachycardia prevention with beta blocker. A small number of patients fail beta blocker therapy and require an implantable defibrillator.

## **VACTERL**

The VACTERL syndrome consists of Vertebral, Anus (imperforate), Cardiac, TracheoEsophageal fistula, Renal anomalies and Limb abnormalities (more commonly absent radius). Common cardiac lesions are VSD and Tetralogy of Fallot. The term VACTERL has replaced the previous VATER because it more completely describes the syndrome.

## **CHARGE**

The CHARGE syndrome consists of Coloboma, Heart defects, Atresia (choanal), Retardation, Genital hypoplasia and Ear abnormalities. The most common cardiac defects are also VSD and Tetralogy of Fallot.

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