Lucile Packard Children's Hospital

Stanford University Medical Center

Department of Anesthesia & Pain Management

Pediatric Anesthesia & Pain Management

(650) 723-5728

Anesthesia in Children with Congenital Heart Disease

Elliot Krane, M.D.

Introduction

The evolution of medicine to managed care and contracted medical services has often had the effect of shifting a sicker and more complicated population of patients from specialized urban medical centers and children's hospitals to regional and community medical centers. The general practice anesthesiologist, therefore, may find him/herself in a position to care for children who would primarily have been operated upon in tertiary medical centers in years gone past. More commonly, you may be examining a previously well child for surgery, and be the first to hear a heart murmur. The rapid definition of a murmur as innocent or functional, or indicative of congenital heart disease, will reduce the need to postpone surgery and order expensive diagnostic evaluations.

In this lecture, one of the more common complex pediatric disease states will be reviewed: congenital heart diseases. We will discuss the evaluation of the heart, how to listen to and classify heart murmurs, and the anesthetic implications of congenital heart disease.

The Newborn Heart: A special case

While the normal newborn's heart cannot be said to be an example of congenital heart disease, its physiology is quite distinct from that of the older child or adult, therefore it is worthwhile reviewing some of the salient differences that have importance to an anesthesiologist.

The newborn's heart has ventricular tissue that contains fewer myocytes and a greater proportion of connective tissue, and the right ventricle is relatively hypertrophied. The heart's compliance is therefore diminished, and the heart is more sensitive to preload. Furthermore the newborn functions very much at the peak of its Starling curve, meaning that the stroke volume is relatively fixed, and making cardiac output very heart rate dependant.

Another salient difference is that the newborn myocardial cell derives a very high fraction of activator calcium (the ionized calcium that initiates muscle contraction by interacting with the actinomyosin unit) from the extracellular calcium pool, rather from the intracellular calcium pool (e.g. sarcoplasmic reticulum) as is the case in the mature heart. The consequence of this physiology can be disastrous if a newborn is administered a calcium channel blocking agent, that interferes with the ingress of ionized calcium into the myocardial cell.

The cardiovascular system of the preterm infant also demonstrates a heightened sensitivity to the depressant effects of inhaled halogenated anesthetic agents, and the response of the heart and vasculature to endogenous and exogenous catecholamines is attenuated.

These features mean that blood pressure varies with and is an excellent indicator of volume status. Another unique feature of the premature infant is the relatively high pulmonary pressure present at birth and persisting for some time after birth. Furthermore, the pulmonary vasculature is far more sensitive to those stimuli that increase pulmonary vascular tone, such as hypoxia, acidosis, and hypercarbia. A marked increase in pulmonary vascular resistance will lead to right-to-left shunting of blood at the atrial septal level and through a patent ductus arteriosus and hypoxemia.

The History & Physical Examination of the Pediatric Heart

History taking will tell you a lot about the child's level of function and reserve, and will indicate the presence or absence of CHF. Questions to ask are:

Is the child blue at rest? Does he turn blue when he cries? (Not all children with cyanotic heart disease are obviously cyanotic. Crying increases PVR and will shunt blood away from the lungs, increasing cyanosis).

Does the child loose consciousness? (This is not a good sign.)

Does the infant sweat when he nurses? (a sure fire sign of CHF).

Are the eyes puffy in the morning? (Children don't get ankle swelling.;. they get eye swelling. They spend much of their day recumbently, and the eyes have loose tissue that swells rapidly when edema is present. Ankle edema is practically unheard of).

Does the child stop playing and squat? (A good sign of diminished pulmonary blood flow; the child learns that squatting increases SVR and shunts more blood to the lungs).

Examination of the child's heart is a simple task if done in an organized manner and with certain rules in mind.

Ten Rules about the Physical Examination:

Rule #1: **You can't auscultate a crying child**. The heart is too fast and the sounds obscured by wailing.

Rule #2 (Corollary of Rule #1): **Examine the heart first**. After you're done with the stethoscope you can palpate, tongue-blade, or do whatever else that may make the child cry. Always get to the child before the phlebotomist.

Rule #3: As distracting as it might be, **start by ignoring the murmur**. First, define the nature of the first and second heart sounds. Are they louder than normal? Are they split? Is the splitting of the second sound normal, unusually wide, or paradoxical?

Rule #4: **Most systolic murmurs will be benign**. Start by describing systolic murmurs as contiguous with the first heart sound (pansystolic) or arising after the first heart sound (ejection). The combination of this information with the nature of the second heart sound will define whether a systolic murmur is functional and benign, or pathological. If you hear a systolic murmur in an asymptomatic child, the odds are on your side. Even if a lesion is present, it is probably not of physiologic significance for most surgery (exception of note: ASD + planned sitting craniotomy).

Rule #5: **All pansystolic murmurs are pathological.** They almost always are VSDs.

Rule #6: In a child with a VSD, **the louder the murmur, the smaller the shunt**. The softer the murmur, the larger the shunt, and the sicker the child.

Rule #7: **All diastolic murmurs are pathological**.

Rule #8: **Children develop bi-ventricular failure.** Palpate the liver. If a child has CHF, the liver will be enlarged and often tender. Obviously eliciting a history of LV failure by asking about orthopnea, PND, and exertional dyspnea is difficult and unnecessary in a two-year-old. All you need to do is palpate the liver.

Rule #9: **All murmurs that radiate are pathological.** Common examples: PDA's radiate to the back, aortic lesions radiate to the neck and/or back.

Rule #10: **Take upper and lower extremity BP** in a child with a murmur. Coarctation of the aorta is often occult, and accompanies many common cardiac lesions.

Congenital Heart Diseases

The anesthesiologist must understand the anatomy and physiology of congenital lesions of the heart in order to develop an effective plan for a general anesthetic, and to plan for intraoperative contingencies. (7)

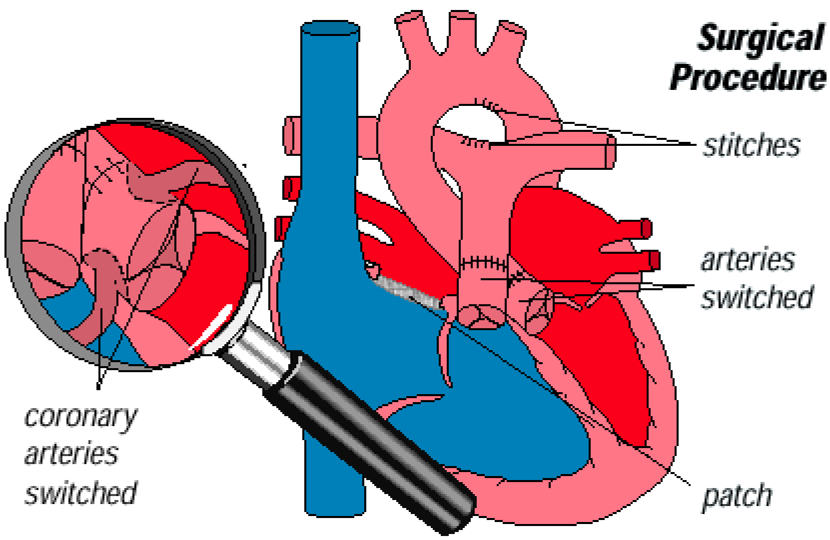
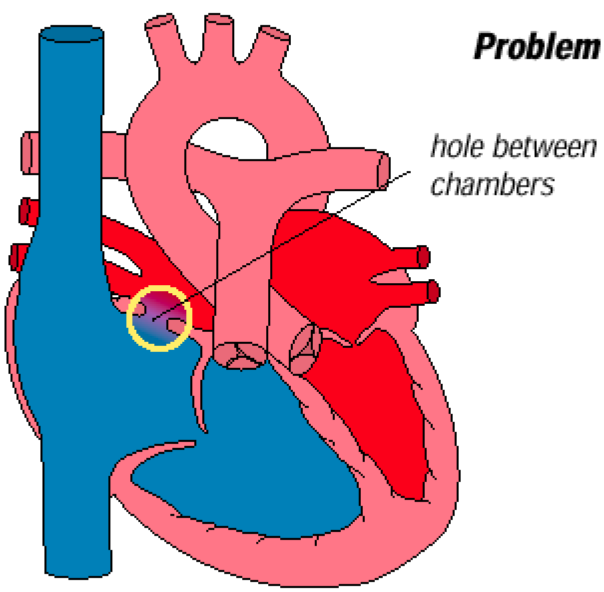
Congenital heart disease may be conveniently divided into two broad classes, cyanotic and acyanotic lesions. The latter are divided into obstructive and non-obstructive lesions.

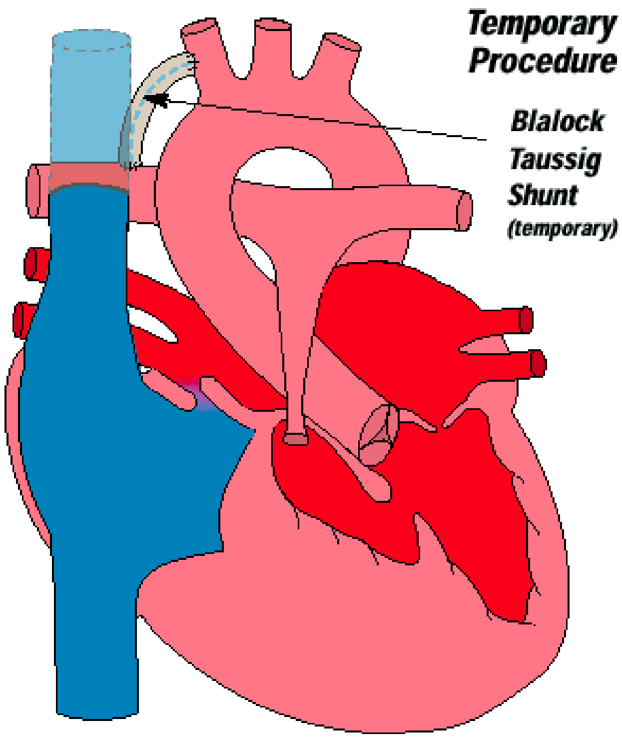
Table 1. Classification of congenital cardiac lesions.

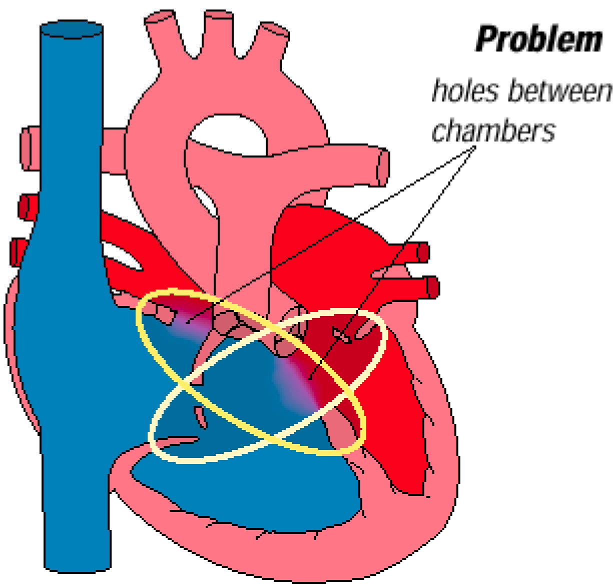
Classification	*Physiology*	*Effect*	*Examples*
Cyanotic: Normal pulmonary blood flow	Mixing of arterial and venous blood in common cardiac chambers	Cyanosis Polycythemia in older children	Single Ventricles Hypoplastic RV Double Outlet RV Transposition of the Great Arteries with an ASD or VSD
Cyanotic: Decreased pulmonary blood flow	Obstruction of pulmonary blood flow leads to shunting at the atrial and/or ventricular level	Cyanosis + CHF Polycythemia in older children	Tricuspid atresia Tetralogy of Fallot Pulmonary Atresia
Acyanotic: Increased pulmonary blood flow	Left to right shunt at the atrial, ventricular, or great vessel level leads to preferential flow to the low resistance pulmonary bed	PDA & ASD: Clinically normal VSD: CHF if severe	ASD VSD PDA Aortopulmonary window
Acyanotic: Obstructed blood flow	No shunt exists, but blood flow is obstructed	CHF	Pulmonary Stenosis Aortic Stenosis Coarctation of the Aorta

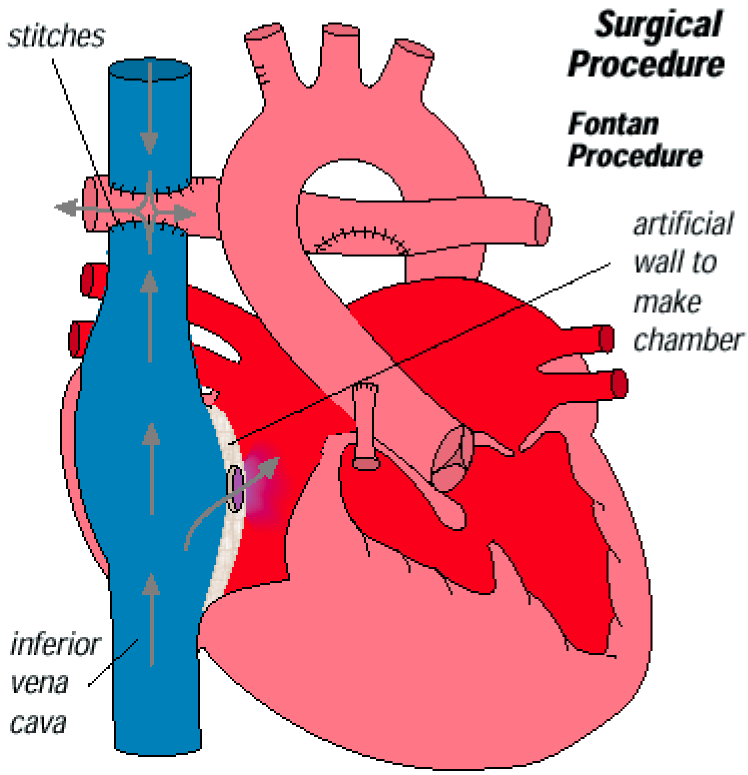
The anesthesiologist also must be familiar with the surgical management of the cardiac lesion up to that point in time, that is, whether the lesion has been corrected or palliated, and understand the present anatomic flow of blood through the heart and the potential effect of anesthetic management on that flow. Table 2 lists a glossary of common acronyms and eponyms, and describes the surgical repairs commonly performed on cardiac lesions.

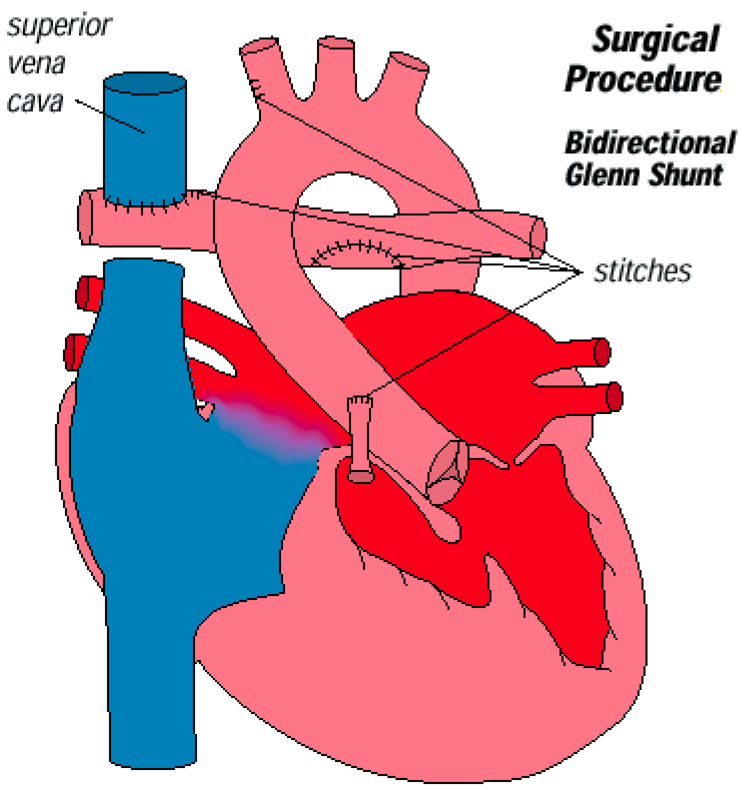
Table 2. A glossary of common acronyms and eponyms.

Term	Definition	Example
Arterial Switch	Surgical procedure to correct TGA; the aorta and PA are removed from the heart and reattached to the appropriate ventricular outflows. The coronary ostea are re-transplanted as well. 	
ASD	Atrial Septal Defect 	

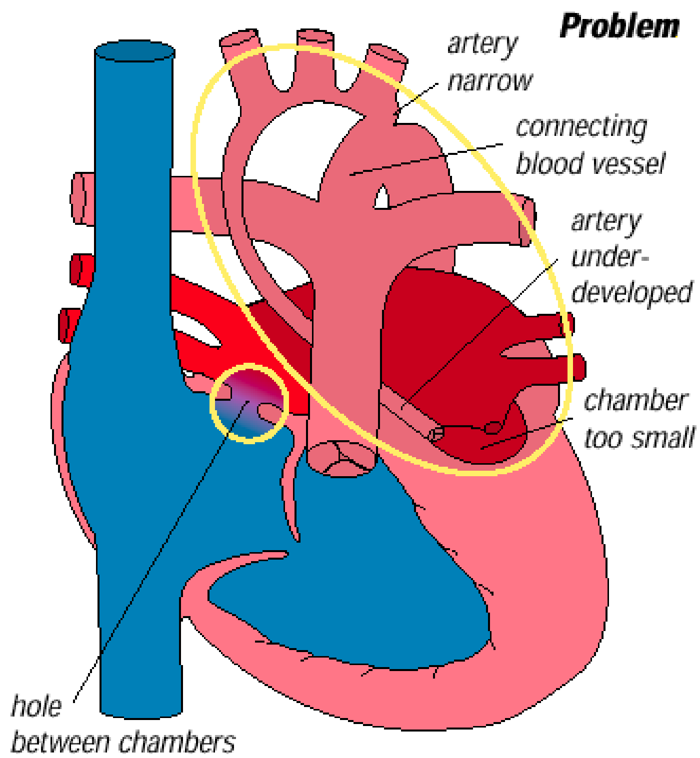
Term	Definition	Example
BTS	Blalock Taussig Shunt: classically a right subclavian to pulmonary artery anastamosis via a thoracotomy incision to augment PBF. Currently usually performed as a Gortex shunt from aorta or subclavian to PA via sternotomy. 	
Damus-Stansel-Kaye	An alternative to the switch for TGA + VSD. The MPA is divided and the proximal stump is sewn to the ascending aorta, and a valved conduit carries blood from the RV to the distal MPA.	
DORV	Double outlet right ventricle. May be similar to TOF, in that an aortic outflow tract sits atop a VSD, or similar to a TGA if the pulmonary outflow sits atop the VSD.	

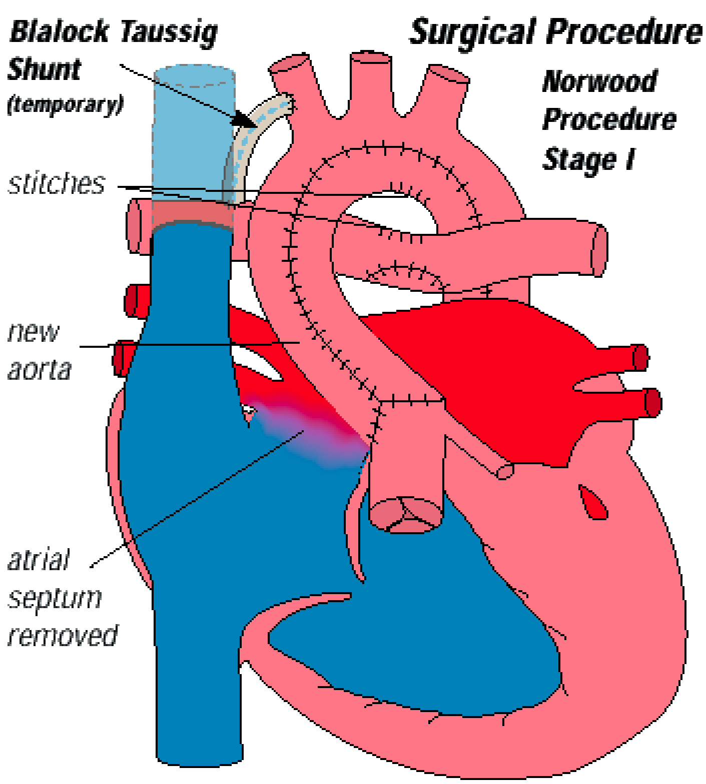
Term	Definition	Example
ECD	Endocardial cushion defect. Associated with Trisomy 21, a primum ASD with deformation of the mitral valve leading to severe MR and CHF. Predominant blood shunt is from LV to RA.  	

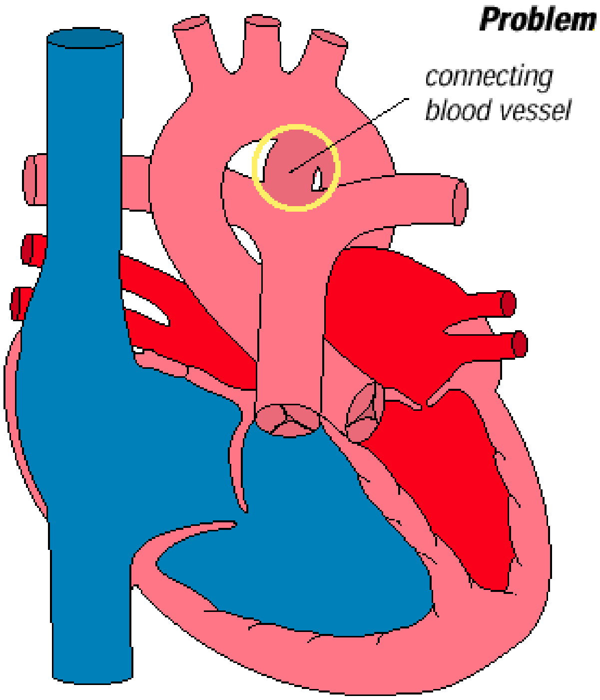
Term	Definition	Example
Fontan	Anastamosis of the right atrium to the PA to provide PBF past an obstructed tricuspid or pulmonic valve. A "fenestrated" Fontan is a Fontan with a "popoff" into the LA through an ASD (illustrated at right) 	

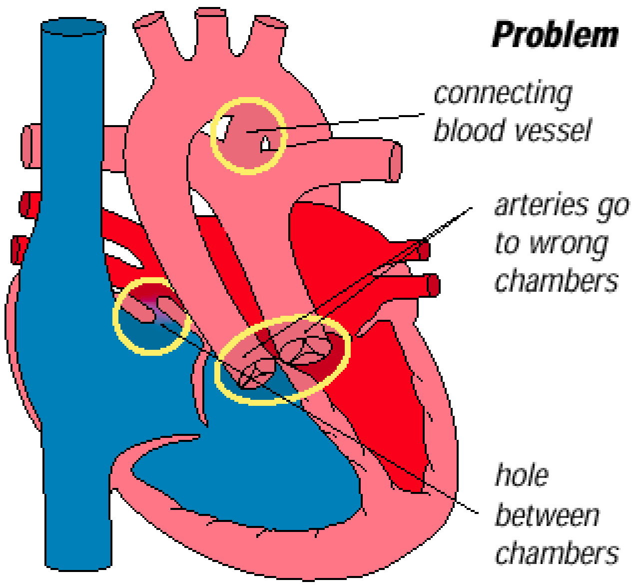
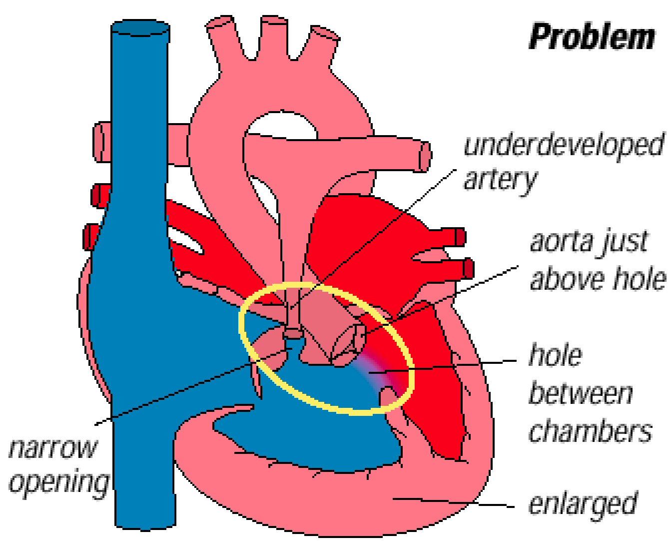
Term	Definition	Example
Glenn	Division of the SVC from the heart and anastamosis of the SVC to the PA, to augment pulmonary blood flow  	

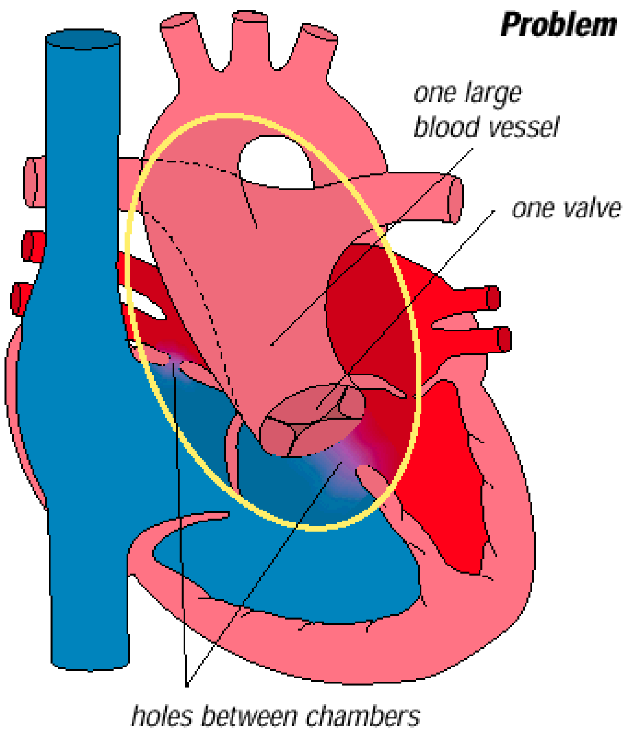
Term	Definition	Example
	Hypoplastic left heart syndrome. Severely hypoplastic LV, aorta, VSD.	





Term	Definition	Example
Mustard	Performed for infants with TGA; creation of an intra-atrial baffle using pericardium, to baffle caval blood to the LA and pulmonary venous blood to the RA.	
PDA	Patent Ductus Arteriosus 	
Rastelli	Synthetic conduit from the RV to the PA, typically performed in older children with TOF to bypass the pulmonic valve	
Senning	Same idea as a Mustard procedure, but associated with a lower incidence of heart block.	
TAPVR	Total Anomolous Pulmonary Venous Return: pulmonary venous drainage into a number of locations instead of or in addition to (PartialAPVR) the LA, including the hepatic vein, the SVC, the azygous vein, the coronary sinus, etc. May be present without or with obstruction, producing pulmonary congestion and CHF.	

Term	Definition	Example
TGV or TGA	Transposition of the great vessels, or, transposition of the great arteries. The aorta arises from the RV, and the PA from the LV. 	
TOF	Tetralogy of Fallot: (1) Pulmonic stenosis; (2) VSD; (3) An aortic outflow tract that overides, or sits atop of the VSD allowing both RV and LV output to enter; (4) RVH 	

<table>
<tr><th>Term</th><th>Definition</th><th>Example</th></tr>
<tr><td>TA</td><td colspan="2">Truncus Arteriosus. One large blood vessel leaves the heart. This vessel has branches that go to the lungs, the body, and the coronary arteries. There is almost always an associated large VSD. The drawing shows Type 1 TA, the most common type (60%).

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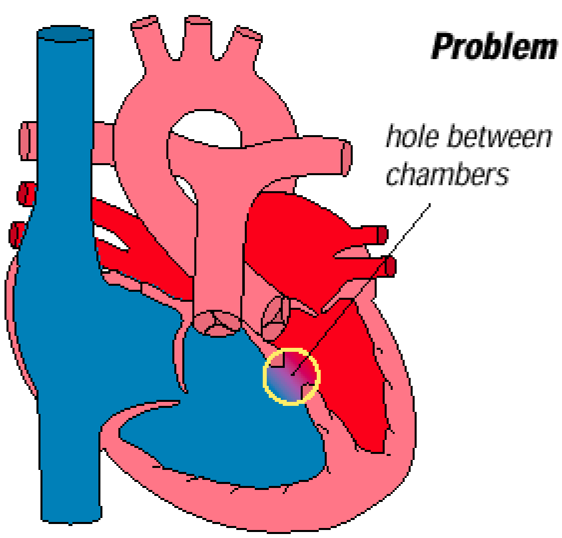
<tr><td>VSD</td><td colspan="2">Ventricular Septal Defect

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Anesthetic management should be guided by the prediction of effects of anesthetic agents and techniques on PVR, SVR, and ventricular function, the goal generally being to preserve ventricular function and SVR while maintaining oxygenation by reducing PVR. Table 3 lists salient anesthetic implications of lesions and their repairs.

Table 3. Some anesthetic implications of CHD.

Classification	*Lesion*	*Implications*
Cyanotic: Normal pulmonary blood flow	Single ventricles lesions and DORV s/p Fontan procedure TGA s/p atrial septostomy	PBF is preload dependant Sinus rhythm promotes PBF Increased PVR will dramatically decrease CO Venous air bubbles will embolize to the systemic circuit Faster effect of intravenous agents
Cyanotic: Decreased pulmonary blood flow	Tetralogy of Fallot s/p BTS TGA s/p BTS	PBF depends upon systemic BP; increased SVR usually improves oxygenation Systemic and pulmonary blood flow depend upon relative resistance of PVR and SVR Increased PVR increases R □♦ L shunting and cyanosis Reduced uptake and excretion of halogenated anesthetics Faster onset of effect of intravenous agents
Acyanotic: Increased pulmonary blood flow	ASD VSD PDA APW	RV overload, poor tolerance of negative inotropes Increase in PVR can reverse shunt from L □♦ R to R □♦ L Slower onset of effect of intravenous agents
Acyanotic: Obstructed blood flow	PS AS HLHS Coarctation	Ventricular failure, poor tolerance of negative inotropes Tachycardia poorly tolerated Rapid uptake of halogenated anesthetics

References

1. Baum, V.C., The adult patient with congenital heart disease, Journal of Cardiothoracic and Vascular Anesthesia, 10 (1996) 261-82.
2. Burrows, F.A., Anesthesia for cardiac surgery in neonates and infants, Advances in Cardiac Surgery, 6 (1995) 103-29.
3. Findlow, D. and Doyle, E., Congenital heart disease in adults, British Journal of Anaesthesia, 78 (1997) 416-30.
4. Greeley, W.J., Pediatric cardiovascular anesthesia, Anesthesia and Analgesia, Suppl (1998) 45-56.
5. Javorski, J.J., Hansen, D.D., Laussen, P.C., Fox, M.L., Lavoie, J. and Burrows, F.A., Paediatric cardiac catheterization: innovations, Canadian Journal of Anaesthesia, 42 (1995) 310-29.
6. Mendelson, M.A., Pregnancy in the woman with congenital heart disease, American Journal of Cardiac Imaging, 9 (1995) 44-52.