

## What Every Anesthesiologist Must Know About Congenital Heart Disease

Review Article

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**Received:** August 01, 2013**Accepted:** August 24, 2013**Published:** August 28, 2013**Citation:** Skidmore K, Phebe Ko (2013) What Every Anesthesiologist Must Know About Congenital Heart Disease. Int J Anesth Res. 1(2), 8-11. doi: <http://dx.doi.org/10.19070/2332-2780-130003>**Copyright:** Skidmore K<sup>©</sup> 2013. This is an open-access article distributed under the terms of the Creative Commons Attribution License, which permits unrestricted use, distribution and reproduction in any medium, provided the original author and source are credited.

In Canada, Europe and the United States, more than five million adults suffer complications of congenital heart disease (CHD), which recently surpassed the number of children with CHD. I provided anesthesia during open-heart surgery for children undergoing repair of CHD for six years at University of California, San Francisco and for three years at Children's Hospital of Orange County. The usual anesthesia residency experience only includes a few adults with CHD undergoing general surgery. Fellowships in cardiac anesthesia were not formalized according to the Accreditation Council for Graduate Medical Education (ACGME) until the last several years. Although "Adult Cardiothoracic Anesthesiology" is now an ACGME fellowship, it only allows two months of electives overall, and merely strongly encourages two weeks of CHD. An ACGME pediatric anesthesia fellowship year requires two months of CHD surgery. With this limited experience dealing with CHD, there is discussion in some hospitals of requiring a second year of fellowship, totaling six years of training after medical school, to conduct anesthesia for CHD.

The orphans are the adults with CHD undergoing general surgery who are lost to follow-up because they do not have access to a pediatric hospital. In the United States, approximately half a million adults have CHD but only 30,000 receive care in tertiary centers with an adult CHD program. Case reports exist of fatal Cesarean sections that resulted from pulmonary hypertension when a ventricular septal defect (VSD) went undiagnosed until after intubation, cardiopulmonary resuscitation, and transesophageal echocardiography (TEE). A thorough preoperative evaluation, revealing erythrocytosis, right ventricular hypertrophy (RVH) on EKG, cyanosis, and tricuspid regurgitation (TR) gradients on echocardiography (allowing an estimation of pulmonary artery systolic pressure) might have prevented these deaths. Anesthesiologists will undoubtedly encounter patients with CHD in their practice.

Thus they must know the hemodynamic goals during general surgery. The purpose of this review is to simplify the hemodynamic goals according to three categories of CHD, describe the flow and pressures in the anatomic variants, and suggest resources for training.

**Preoperative History**

Ideally, the patient should be evaluated on a preoperative day. The anesthesiologist should ask whether strenuous exercise has been, in recent months, more limited by symptoms of dyspnea, wheezing, palpitations, nausea, diarrhea, abdominal pain, peripheral swelling, sweating, angina, seizures, or recent upper respiratory infections. Ask which medications and herbal supplements the patient actually takes and when they last took them. Ask what surgeries occurred for the heart, and when and where the last catheterization or echocardiography was performed. Obtain all relevant records from hospitals where the patient had care.

Fasting guidelines should include allowing medications with sips of water and unlimited water, clear apple juice, or pedialyte until 2 hours before surgery to avoid dehydration. Stop other food or drinks for 8 hours preoperatively.

In addition to a physical exam (blood pressure on all extremities, pulse oximetry on both hands after walking with the patient, edema, murmurs, crackles, etc), the doctor should consider obtaining a chest x-ray, 12 lead EKG, chem-7, Magnesium, Calcium, Phosphorous, AST, ALT, albumin, CBC, PT, PTT, urine pregnancy for women of childbearing age, and a transthoracic echocardiogram within the last year, especially if the surgery involves major fluid shifts. The echo or cath report may indicate pulmonary hypertension, a risk factor for postoperative morbidity. Erythrocytosis may indicate chronic hypoxemia and the patient may benefit from intravenous hydration or preoperative iron as well as invasive perioperative monitoring of fluid status and blood gases.

A list of the types of heart surgeries and catheterization procedures, with corresponding diagnostic diagrams, should be in the chart preoperatively. The anesthesiologist should draw the catheterization diagram. The vena cava, right atrium, right ventricle, pulmonary artery, pulmonary vein, left atrium, left ventricle, and aorta, are each drawn as blocks that contain a PRESSURE and a SATURATION. CHD lesions are drawn in as shunts or narrowings, with obvious in and out arrows indicating direction of blood flow. MRI may be useful in trending the size of the right ventricle and aortic aneurysms. If the patient has poor exercise tolerance, despite an unchanged echocardiogram, then an exercise test, Holter electrophysiology study, or therapeutic catheterization can allow optimization prior to noncardiac surgery.

Even in minor surgery, anesthetics often reduce cardiac output by 20%, a reduction magnified in CHD, such that cardiovascular collapse during induction is one hundred fold higher in CHD, especially with outflow tract obstruction, pulmonary hypertension, arrhythmias, myocardial failure, or severe cyanosis. The anesthesiologist must prepare the hemodynamic milieu via the optimization of intravascular volume, inotropes, and systemic and pulmonary vascular resistance (SVR, PVR). A pre-induction arterial line, central venous line, and fluid or inotrope loading may be required during a very gradual induction. Planning for overnight observation postoperatively may include the intensive care unit, even when the surgery is minor, because hypercapnea, hypovolemia, and arrhythmias unnoticed can cause sudden death.

## Volume Overload Work

Systolic dysfunction and low ejection fraction are understood by every anesthesiologist. The left ventricle eccentrically dilates when pumping extra volume backwards, such as in mitral or aortic regurgitation (MR, AR). Although the ejection fraction is deceptively maintained "normal" in MR and AR, the contractility is depressed. Similarly, ventricular septal defects (VSD) create left ventricular dilatation with systolic congestive heart failure (CHF). However, VSD create a high Qp:Qs, i.e. pulmonary to systemic blood flow ratio. Qp is pulmonary blood flow while Qs is cardiac output (into the aorta and body), which are data available on echo or cath. In MR and AR the Qp:Qs is 1:1.

Therefore a large VSD, but not MR or AR, causes over-circulation to the lungs, leading rapidly to pulmonary vascular occlusive disease (PVOD) and irreversible pulmonary hypertension (Eisenmenger's Syndrome), sometimes in infancy. MR and AR can cause pulmonary edema and secondary pulmonary hypertension, but Qp:Qs is 1:1. Thus, pulmonary hypertension occurs more slowly than with a VSD. If PVR approaches 8 Woods Units despite challenge in the catheterization laboratory with inspired 100% oxygen and nitric oxide 20 ppm, then postoperative outcomes are dismal.

The right ventricle eccentrically dilates when pumping extra volume, such as in tricuspid or pulmonic valve regurgitation (TR, PR), but also with atrial septal defects (ASD). PVOD takes longer to develop in ASD than with VSD, because the amount of volume overload is lower.

Anatomically, there are four types of VSD and four types of ASD. Physiologically, there are two types of each: large and small. When Qp:Qs is more than 1.5:1, a VSD is large and should be closed promptly. The most common type of VSD is perimembranous, often associated with a tricuspid valve (TV) "aneurysm" of tissue covering part of the VSD. Doubly committed---otherwise known as supracrystal---VSD are near the pulmonic and aortic valves. Muscular VSD are often multiple and may close on their own. Inlet VSD are perimembranous and near the TV.

The most common type of ASD is secundum, which is at the inferior portion of the membranous septum. Primum ASDs are alternatively called "partial Atrioventricular Canal" because they always have an associated adjacent cleft in the regurgitant mitral valve, but no VSD. Complete Atrioventricular Canal (when "balanced" i.e. without a diminutive left or right ventricle) is a primum ASD combined with a perimembranous VSD and a combined atrioventricular valve often with 5 leaflets that include an anterior bridging leaflet. The normal TV sits about one cm lower than the MV. The hallmark of complete AV Canal is that the TV and MV

are on the same level. Unroofed coronary sinus is a rare form of ASD. Sinus venosus ASD are usually near the superior vena cava but can be near the inferior vena cava in Scimitar Syndrome. Sinus venosus ASD almost always involve partial anomalous pulmonary venous return because pulmonary veins are emptying into both atria near the ASD.

Patent ductus arteriosus or other left to right shunts (surgically created, or major aortopulmonary collateral arteries beginning in utero in cyanotic states) cause LV volume overload work and pulmonary HTN.

The hemodynamic goals for volume overload hearts are "fast, full, and forward." Sinus rhythm is not as essential as for the pressure overload lesions. Too full a preload should be avoided, so as not to reduce cardiac output as on the descending limb of Frank Starling curves. Forward includes two components: low systemic vascular resistance (SVR) and high contractility.

## Pressure Overload Work

Diastolic dysfunction raises serum brain natriuretic peptide and causes pulmonary edema and arrhythmias, just as systolic CHF, but maintains a normal ejection fraction and a lower end diastolic volume (no dilatation). Pressure overload work is less well tolerated than volume overload work, because of the concentric hypertrophy causing arrhythmias and ischemia. The left ventricle concentrically dilates when pumping extra pressure forward, such as in severe aortic stenosis (AS) with a peak gradient over 70mmHg, mean gradient over 40mmHg, or valve area under 1cm<sup>2</sup>. CHD includes a spectrum of AS such as a subaortic membrane obstruction, excessive muscular bundles like idiopathic hypertrophic (hence, dynamic) subaortic stenosis, bicuspid valves (present in 3% of the population), supra-aortic narrowing at various places along the aorta, interrupted aortic arch or a discrete coarctation (near the PDA). Associated syndromes include Williams, Shones, and DiGeorge (the last of which is associated with immune deficient problems and hypocalcemia).

The right ventricle concentrically dilates when pumping extra pressure forward, such as in severe pulmonic valve stenosis (PS) with a peak gradient over 64mmHg (or over 4m/s as seen with continuous wave Doppler in echocardiography). CHD includes a spectrum of PS including tetralogy of Fallot (TOF). In utero, TOF patients may develop major aorto pulmonary collateral arteries (MAPCAS) that can be occluded in the cath lab or surgically "unifocalized" into a PA conduit. Approximately every ten years, the former TOF patient requires another "conduit change" i.e. a pulmonic valve replacement usually due to PR more so than PS. The valve can be replaced percutaneously in the catheterization lab by a "Melody Valve" or via redo sternotomy, usually with a bovine jugular (Contegra) or in large adults, a bioprosthetic valve such as normally placed in the aortic position. Double chamber RV has an hourglass appearance because muscle bundles block the RV outflow tract and can usually be resected and corrected.

Hemodynamic goals for RV or LV pressure overload include slow sinus rhythm, full preload, normal contractility, and high SVR (to perfuse the thick subendocardium). If pulmonary HTN is the primary problem, it should be reduced, or else the RV fails.

## Cyanotic CHD

Prototypical single ventricle physiology is hypoplastic left heart

syndrome (HLHS), requiring Norwood, Glenn, and Fontan procedures, at approximately ages 1 week, 6 months, and 5 years, respectively. A Rashkind in the cath lab is creation of an ASD to increase pulmonary blood flow; often a PDA stent is inserted as well.

A Sano is a RV to aorta conduit (instead of a Blalock Taussig subclavian to pulmonary artery conduit). A Norwood is aortic arch augmentation with circulatory arrest and using pulmonary artery tissue. The Damus Kaye Stansel is similar to Norwood operation. A Glenn is connection of the superior vena cava (SVC) to the pulmonary artery, and is a prophylactic measure to prevent ventricular systolic dysfunction from volume overload work.

A Fontan is performed when the saturations are falling, often near age four years, to increase pulmonary blood flow by connecting the inferior vena cava (IVC) to the pulmonary artery. Ideally, transpulmonary gradient of 7mmHg results, meaning for example that a vena cava pressure is 13mmHg while the common atrial pressure is 6mmHg. The goals pre-Fontan are to maintain the hematocrit above 40%, the arterial saturation no greater than mid 80s, sinus rhythm, and the Qp equal to Qs. During anesthesia, this is achieved by using 21% inspired oxygen and normal ventilation. Gradually check while spontaneous ventilation is taken over with positive pressure ventilation whether the blood pressure or saturation drastically changes. If saturation is too high, blood pressure is too low, the anesthesiologist must ventilate less and with lower oxygen, and vice versa.

After a Fontan, the hematocrit can be allowed to drift down to near 30-32%, the saturation should climb to low 90's, and the PVR and SVR must be maintained low, using drugs such as milrinone (PDE 3 inhibitor), to avoid SVC syndrome, ascites, peripheral edema, protein losing enteropathy, or pulmonary edema. Fenestrated Fontans have a small connection created between the SVC/IVC-to-PA conduit and the common atrium, to allow a pop off of high SVC/IVC pressures, costing some points in the pulse oximetry, but gaining cardiac output. Many patients are maintained on anticoagulants to avoid stagnant flow and clotting of the pulmonary arterial tree. Nitric oxide 20ppm usually reduces PA pressures by 20% and can be delivered by nasal prongs, face mask, or endotracheal tube. Many patients require pacemakers since sinus rhythm is important to maintaining cardiac output.

Other single ventricle patients who may require a palliative Glenn and Fontan include: Ebsteins anomaly (atrialization of the right ventricle), double inlet left ventricle (diminutive right ventricle), double outlet right ventricle (diminutive left ventricle), unbalanced atrioventricular canal (diminutive right or left ventricle), and tricuspid atresia (diminutive right ventricle). If there is a VSD, and the diminutive ventricle is fairly large and functional, then a 1 and 1/2 ventricle repair means that there is no need for Fontan after Glenn.

Prior to complete surgical correction, truncus arteriosus (where a new pulmonary valve is inserted and the truncal valve becomes the neo-aortic valve, sometimes containing one to 5 leaflets), total anomalous pulmonary venous return, dextro-transposition of the great vessels (corrected via Arterial Switch Operation since 1980, but Mustard and Senning intracardiac atrioventricular baffles prior to 1980; or Rastelli repair if creating a conduit from each ventricle to artery), pulmonary atresia (corrected if two ventricle repair is possible), and aortic coarctation behave as a single ventricle and are dependent on patent ductus arteriosus and/or ASD/

VSD shunting. Residual defects in fully corrected lesions tend to mimic general adult heart disease, such as valve regurgitation or stenosis, CHF and pulmonary HTN. Additional collaterals and obstructions are sometimes noted on the cath report.

## Pulmonary Hypertension (PHTN)

Pulmonary HTN is the most deadly comorbidity during induction and emergence. PHTN is defined as a PVR greater than 3 Woods Units, or a mean PA greater than 25mmHg. Ohms Law,  $V=IR$ , voltage difference = current times resistance, translates to Mean Arterial Pressure minus central venous pressure = cardiac output times SVR. PVR is mean pulmonary artery pressure minus wedge pressure, divided by cardiac output. Division by 80 yields Woods Units, rather than dynes second per cm<sup>5</sup>.

If 20 ppm nitric oxide and 100% inspired oxygen reduce mean PA to under 50mmHg, pulmonary hypertension is considered "reactive" and more likely to respond to medications and surgical corrections. PHTN can be inherited, due to CREST syndrome, CHD, HIV, liver failure, methamphetamine, sickle cell disease, obstructive sleep apnea, thromboembolic disease, left heart failure or other etiologies.

The same anesthetic goals apply whether the pulmonary HTN is from CHD or non-CHD etiologies. Administer bicarbonate or THAM intravenously until the pH is 7.45 to 7.55. Keep the patient warm and pain-free, hyper-oxygenated, and hyperventilated. "Flolan" (epoprostenol) intravenously or a similar prostanoid "remodulin" (treprostinil) subcutaneously are approved by FDA for home use, along with "Viagra" (sildenafil, a PDE5 inhibitor) tablets. The best pulmonary vasodilator is oxygen. Inhaled nitric oxide, or nebulized PGI2 (prostacyclin, a less expensive alternative), might be required. Maintain a normal viscosity of blood by hemodiluting to a hematocrit near 30% and do not ventilate with high airway pressures. Central venous pressure monitoring is useful when volume shifts occur during invasive surgeries, because diuretics (with calcium, magnesium and potassium replenishment) are important but avoidance of dehydration is equally important.

Arterial, central venous pressure, pulmonary artery pressure, and TEE monitors can be helpful, but in some cases are fraught with more risk than benefits, especially if the surgery creates few volume shifts. A stat-mode on the noninvasive cuff, a large peripheral intravenous catheter, gradual volume loading and inotropes titrated during the taking over of positive pressure ventilation, while incrementally inducing unconsciousness is more wise than a rapid intravenous induction followed by paralysis. An intravenous line must be present in pulmonary HTN, like for AS, HLHS or TOF inductions, even if placement requires intramuscular ketamine sedation. Dobutamine, milrinone, norepinephrine, and epinephrine are often needed. PHTN patients are often maintained on Coumadin to an INR of 1.5 to 2, or if positive for anticardiolipin or lupus anticoagulant, an INR of 2.3 to 3, in order to prevent pulmonary clotting in areas of sluggish flow. Atrial septal defects are planned and created in the cath lab in patients with high right pressures, to allow a "pop-off" and reduce symptoms of refractory PHTN.

## LVAD

Left ventricular assist devices such as HeartMate or HeartWare are placed in adults as destination therapy or bridge to transplant. Left ventricular blood is drained, pumped and returned to the

aorta. Noninvasive blood pressure cuffs may not always yield information, and ultrasound may be required to place an arterial catheter, or to see what systolic pressures are (as color reappears) as the cuff deflates. Since cardiac output is fixed, often hypotension is the result of hypovolemia or low SVR, easily treated with phenylephrine and fluids.

## Transplants And Pacemakers

Some patients have defibrillators, pacemakers, or heart or lung transplants. Noncardiac surgery may require placement of defibrillator pacer pads before prepping the skin, reprogramming the device, placing the return pad of the Bovie far from the device, or a magnet on the device.

## Training

The National Board of Echocardiography offers a basic or an advanced Perioperative TEE Testamur, or Diplomate certification. The tests are multiple choice, taken at a computer center, and presented to credentialing groups at hospitals. The American Society of Anesthesiologists (ASA) publishes indications for insertion of a TEE. Real-time ultrasound for central line placement is rewarded by an additional billing unit, listed in the ASA Relative Value Guide, when an image is placed in the permanent medical record. Transthoracic ultrasound at the bedside can be useful to show tamponade, poor LV or RV function or filling or major valve obstruction.

Multidisciplinary quality improvement meetings should occur regularly. Case logs should be maintained for anesthetics provided to various ages and concurrent CHD, not only stating the general surgery performed. Case logs and Continuing Medical Education are required for Diplomate Perioperative TEE certification, and at many hospital committees.

Thanks to publications such as this one in Open Access, more peer-reviewed articles, reviews, and expert opinions are freely available. The American Heart Association in 2007 published new guidelines, listing which patients and which antibiotics prevent subacute bacterial endocarditis. Dental care and X rays must be routine. The Canadian website reference for adult CHD is very useful and lists other resources.

Every anesthesiologist must take it upon him/herself to learn

the didactic cardiovascular physiology, read review chapters and articles, and then seek out these cases while being mentored by experienced anesthesiologists. Without adequate knowledge and experience of anesthesiologists, morbidity and mortality will continue to rise in noncardiac surgery for those patients with CHD. Many children have had CHD surgery but later succumb to irreversible CHF or pulmonary HTN because they did not have access to pediatric cardiologists. For example, pulmonary artery stents to enlarge a narrowing (or surgically placed bands, to limit flow) can not grow and patients may not realize they need serial clinic visits with specialists. The patient may compensate by leading a sedentary life, and by the time symptoms develop, irreversible CHF has developed.

Anesthesiologists must be both preoperative evaluation clinic physicians and postoperative intensive care unit physicians, adept at directing optimal intraoperative and postoperative monitors and therapies. In addition, they must encourage patients to have follow-up care (such as serial echocardiograms) with a pediatric cardiologist, or a cardiologist who has experience with adult CHD. Presentation at noncardiac surgery is an opportunity to catch those patients lost to follow-up and delay their elective surgery until the proper workup is completed,

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