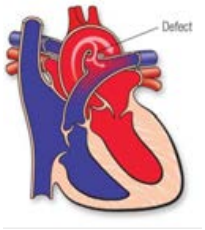
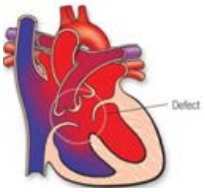
Arrhythmias and Pacemakers

Pacemakers

Positions	Describes how pacemaker functions and programmed • **Position 1**: The chamber being paced (A = atrium, V = ventricle, D = dual • **Position 2**: The chamber being sensed (A, V, D or O = no sensing). • **Position 3**: Response to a particular sensed event (I = a sensed event inhibits pacemaker output, T = a sensed event triggers pacemaker output, D = dual modes of response (i.e. a sensed event in the atrium inhibits pacemaker output in the atrium, but triggers ventricular pacemaker output w/ a programmed delay to mimic intrinsic AV delay), O = no response to sensed events).
Settings	Re-entry, enhanced automaticity, triggered activity • **AAI**: Atrial demand pacing and is an appropriate mode for patients w/ sinus node dysfunction, but should not be used for patients w/ AV node dysfunction • **VVI**: Ventricular demand pacing and is used quite commonly-- results in loss of AV synchrony and can result in a type of cardiomyopathy called pacemaker syndrome (signs and symptoms similar to heart failure) • **DDD**: Dual chamber pacing- provides more physiologic pacing w/ preserved AV synchrony and may be used in patients w/ both sinus node and AV node dysfunction. This mode of pacing can result in four different rhythms: ▪ Normal sinus rhythm (pacemaker does not fire) ▪ Atrial pacing w/ a native QRS (pacemaker provides atrial impulse only) ▪ AV sequential pacing (pacemaker provides atrial impulse w/ a programmed PR interval mimicking AV node function followed by ventricular impulse) ▪ Atrial sensing and ventricular pacing (pacemaker provides ventricular impulse only at intervals mimicking AV node function)

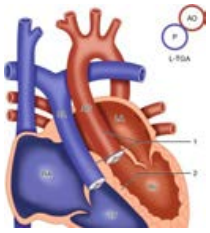
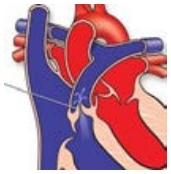
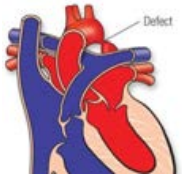
Acyanotic Heart Disease

Lesion	Basics	Hx/Exam	Studies	Treatment
Atrial Septal Defect	• Volume overload • 4 types based on location and embryologic origin. 1. Ostium primum: low in septum; can involve AV valve. 2. Ostium secundum: most common; near foramen ovale. 3. Sinus venosus: may involve connection w/ SVC, IVC, often associated PAPVC. 4. Coronary sinus (defect between CS and LA, not truly in atrial septum). • Amount of L→ R shunt depends on side of defect, SVR relative to PVR, relative LV and RV compliance • PAPVC has similar hemodynamic consequences as ASDs	**Hx**: often asymptomatic, may result in poor growth. When causing significant overcirculation, causes fatigue, dyspnea, CHF and can lead to pulmonary vascular disease (Eisenmenger syndrome). Paradoxical emboli **PE**: Fixed and widely split S2. SEM caused by increased flow across PV, not flow through septal defect. Diastolic rumble if significantly increased volume of flow across the tricuspid valve.	**EKG**: Enlargement of right-sided chambers, RBBB (complete or incomplete), RAD. Superior axis in primum ASD **CXR**: Overcirculation (increased pulmonary vascular markings). Cardiomegaly. 	• Secundum defects may close spontaneously • Surgery indicated if symptomatic or is Qp:Qs>2:1. Surgical or cath patch closure. • Surgical goal = close the defect and avoid development of irreversible pulmonary hypertension/ Eisenmenger's syndrome

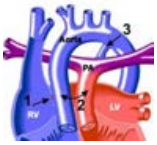
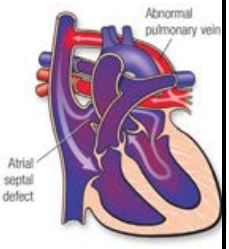
Acyanotic Heart Disease				
Lesion	**Basics**	**Hx/Exam**	**Studies**	**Treatment**
Ventricular Septal Defect	• Volume overload and possible pressure overload. • Opening in ventricular septum. - Occurs in one of four locations: inlet, outlet, membranous, muscular. • Degree of shunting determined by size of defect and relative SVR/PVR • If small in size (and restrictive) may not be hemodynamically significant. • If moderate in size, can cause pulmonary overcirculation and left-sided volume overload • If large can expose RV to systemic pressure in addition to volume overload	**Hx**: depends on size. Symptoms occur as PVR decreases during first weeks of life and flow across the defect increases. Sx of CHF include, tachypnea, poor growth, sweating, feed fatigue, dyspnea. **PE**: early or holosystolic regurgitant-type murmur. Smaller defects are louder because of higher pressure gradient across lesion. Large defects may cause very quiet murmurs. Volume overload can produce a left-sided heave.	**EKG**: normal or LAE, LVH, sometimes RVH if defect is large and RV is exposed to systemic pressure OR if pulmonary vascular disease has developed due to chronic overcirculation **CXR**: most often normal. +/- mild cardiomegaly or increased pulmonary blood flow.	• May spontaneously close on own, especially small muscular types. • Surgery if symptomatic or persistently elevated PVR. Otherwise, may observe. • Repair is surgical patch closure or cath device closure • Surgical/cath goal = close the defect. 
Patent Ductus Arteriosus	• Volume overload. • Common in premature newborns. • Can be asymptomatic. Can also cause pulmonary overcirculation, CHF and systemic hypoperfusion	**Hx**: Respiratory distress, feeding fatigue, poor growth, CHF. **PE**: continuous "machine-like" murmur at LUSB (though murmur can also be systolic only). Wide pulse pressure, bounding or palmar pulses.	**EKG** is often normal. Can have LVH or RVH. **CXR**: Nml +/- increased vascular markings. +/- cardiomegaly. 	• Indomethacin, ibuprofen or Tylenol in preemies. Less likely to be successful in non-preemies. • Surgical ligation or cath coiling in larger children • Surgical/cath goal = close the duct
AV canal Defects	Volume overload Components: 1. Primum ASD 2. Inlet VSD 3. AV valve defects Occurs on a spectrum: 1. Partial AV canal (ASD, single AV valve annulus w/ separate MV and TV orifices and cleft MV) 2. Transitional AV canal (Cleft MV, ASD and hemodynamically insignificant VSD) 3. Intermediate AV (Large ASD and VSD, single valve annulus, distinct TV and MV orifices) 4. Complete AV canal (ASD, VSD, common AV valve) Common in T21	**Hx**: presentation similar to that of VSD w/ CHF: poor growth, sweating, feed fatigue, dyspnea. Severity depends on type of defect. **PE**: Murmurs of ASD, VSD, MR +/- gallop.	**EKG**: Superior axis. +/- RVH, LVH. **CXR**: cardiomegaly +/- increased vasc markings. 	• Surgery often required before 1st birthday to prevent CHF. • Patch closure of septal defects, often involves valvuloplasty. • Surgical goal = closing defects and achieving AV valve competency • Complications: AV valve regurgitation and stenosis after repair

Acyanotic Heart Disease continued on next page →

Acyanotic Heart Disease

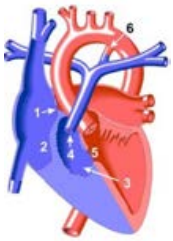
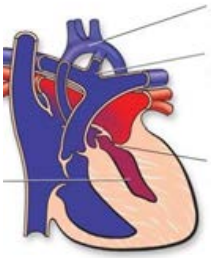
Lesion	Basics	Hx/Exam	Studies	Treatment
Congen. Corrected TGA	• Transposed great arteries (PA off LV, Ao off RV) • L-looped ventricles • Segmental anatomy is {S,L,L} or, less commonly, {I,D,D} • **Blood flow**: LA-->RV -->Aorta-->Body -->IVC/SVC -->RA-->LV-->PA-->Lungs-->Pulmonary veins-->LA • Often associated w/ other cardiac defects (often a VSD) • Often have coronary anomalies	**Hx**: No cyanosis unless other cyanotic defects present. Can present w/ right heart failure in early adulthood as RV cannot tolerate work load as systemic ventricle. **PE**: Dependent on associated defects. May have stigmata of right heart failure. May have loud S2 due to anterior position of AoV.	**EKG**: Q waves in right precordial leads, no Q waves in left-sided leads. Often have conduction system abnormalities including bradycardia and AV block. **CXR**: Dextrocardia or mesocardia are common. 	• Conventionally, only associated defects were repaired. • The newer anatomic approach involves the "double switch" operation, which involves an arterial and atrial level switch via baffling or a Senning-Rastelli procedure if significant PS is present. • Often "training" of the LV w/ PA banding before the LV is made the systemic ventricle is required, unless significant PS is present. • Timing of surgery is a major challenge
Pulmonary Valve Stenosis	• Pressure overload • Stenotic pulmonary valve, causing increased pressure on RV, TR, may be transmitted toRA • "Critical"if ductal patency required for pulmonary blood flow. These children require prostaglandins and early repair. ductus).	**Hx**: If mild/moderate, asymptomatic. If severe, w/ RV dysfunction and TR, hepatomegaly. If critical, can present w/ cyanosis. **PE**: SEM at LUSB, ejection click. +/-TR murmur. Often worsens in first few months of life, then stabilizes.	**EKG**: Normal to RAD, RVH. +/- RV strain pattern **CXR**: +/- ↓vasc markings 	• If critical start PGE • Repair is balloon valvuloplasty in cath lab. Surgical repair if severely thickened valve, or muscular subpulmonary stenosis. • Surgical/cath goal = relieve obstruction, will often have some degree of PR afterward
Aortic Stenosis	• Pressure overload. • Can be at level of valve, supravalvar or subvalvar. • LVOT obstructional LVH, systolic and diastolic dysfunction, CHF, MR. Severe LVOTO causes decreased CO • Critical if ductal patency required for systemic blood flow • Supravalvar stenosis common in William's Syndrome.	**Hx**: Infants often asymptomatic. Stenosis worsens w/ age, causing CHF or even cardiogenic shock. **PE**: Harsh SEM at base, radiating to neck. Ejection click w/ valvar stenosis. LV heave or tap.	**EKG**: LVH +/- strain pattern. **CXR**: normal to cardiomegaly. pulmonary edema possible 	• If critical PGE to maintain CO. • Repair is cath balloon valvuloplasty or surgical aortic valvuloplasty or valve replacement. • Surgical goal = relieve obstruction, avoid AR.
Coarctation of the Aorta	• Pressure overload. • Narrowing of the descending aorta in one of three locations: pre-ductal, juxtaductal (most common) or postductal (adult-type). Often worsens as PDA closes. • Common in Turner Syndrome.	**Hx**: In infants, often presents as PDA closes: poor growth, sweating, feed fatigue, dyspnea and can present as cardiogenic shock. Upper extremity hypertension, w/ drop in lower extremity BPs **PE**: SEM at LUSB radiating to back. BP gradient btwn right arm and legs. Brachiofemoral delay and/ or decreased/ absent femoral pulses.	**EKG**: RVH in infancy. LVH in children. **CXR**: Cardiomegaly. "3 sign", rib notching in older children (collateral vessels eroding bone). 	• **Infants**: PGE if signs of shock to maintain CO. • Repair is surgical coarct excision and anastomosis or cath balloon dilation and possibly stenting. • Surgical goal = relief of obstruction. • **Complication**: re-coarctation

Cyanotic Heart Disease

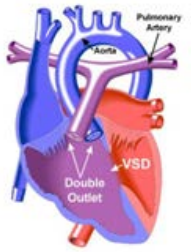
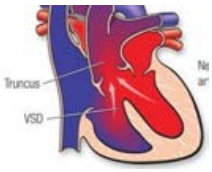
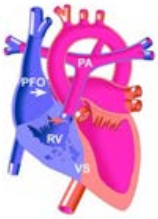
Lesion	Basics	Hx and Exam	Studies	Treatment
Tetralogy of Fallot	• Anterior malalignment of the conal septum, causing: 1.Large VSD. 2.RV outflow obstruction. 3.Overriding aorta. 4.RV hypertrophy. • Degree of cyanosis depends on amount of RVOT obstruction • "Pink Tets" have minimal RVOT obstruction (VSD-like physiology) and "Blue Tets" have significant RVOT obstruction. • Pulmonary Atresia and Major Aorto-Pulmonary Collateral Arteries (TOF/PA/MAPCAs) is the most severe variant • Hypercyanotic episode ("Tet Spell"): occurs 2/2 to • Dynamic worsening of RVOT obstruction • Increased PVR • Decreased SVR and results in cyanosis and, if persistent, acidosis 2/2 RàL shunting	**Hx**: May have "Tet Spells" Symptoms can range from severe cyanosis to predominantly pulmonary over circulation and volume overload resulting in heart failure depending on degree of RVOTO "Balanced" tets (moderate PS, Qp:Qs close to 1) may present only w/ a murmur **PE**: SEM at LUSB (2/2 RVOT obstruction, VSD does not cause murmur). Absent or soft P2.	**EKG**: RAD, RVH, RAE, RBBB **CXR**: "boot-shaped" heart. Decreased pulmonary markings. +/- right-sided aortic arch. Look for absent thymic shadow (seen in patients w/ 22q11 deletion). Coronary artery anomalies are common, may have absent ductus arteriosus 	• PGE if neonatal cyanosis to preserve ductal patency and pulmonary blood flow. • **Surgical repair**: patch closure of VSD and relieve RVOT obstruction (may require muscle bundle resection, patch augmentation of RVOT which may be valve-sparing or a transannular patch) • Unifocalization for TOF/PA/MAPCAs • Surgical goal = close VSD, relieve RVOT obstruction • Will often have PR after repair • Acute hypercyanotic episode: 1.Decrease PVR Supplemental O2 Morphine Bicarb 2.Increase SVR Knees to chest Alpha-1 agonists 3.Increase systemic venous return • Beta blockers may be used to prevent infundibular spasm
Transpos. of the Great Vessels	• Aorta arises from RV, pulmonary artery arises from LV w/ D-looped ventricles. • Results in two parallel circulations and severe cyanosis unless mixing occurs at the atrial or ventricular level (PDA alone is not sufficient)	**Hx**: Profound cyanosis and tachypnea at birth. If large VSD, can have comfortable dyspnea. **PE**: Often no murmur if no VSD. +/- single S2.	**EKG**: RAD, RVH **CXR**: "Egg on a string" heart. Increased pulmonary vascular markings. Right-sided aortic arch. 	• PGE in newborns. • Often emergent balloon atrial septostomy to ensure mixing of the two parallel circulations. • **Surgical repair**: arterial switch w/ transfer of the coronary buttons. Older surgeries involved atrial switch (i.e. Mustard, Senning) • Surgical goal = restore normal connections between ventricles and great vessels
Total Anomlous Pulmonary Venous Return	• Pulmonary veins do not return to LA • Four types: 1.Supracardiac 2.Intracardiac 3.Infracardiac 4.Mixed • Cyanosis due to mixing of oxygenated and deoxygenated blood or pulmonary edema is veins are obstructed (common in infracardiac type) • Must have mixing lesion to survive • Anomalous connection causes L--> R shunt and there is shunting of mixed blood R--> L at the atrial or ventricular level, causing cyanosis (net shunt is usually L--> R)	**Hx**: can mimic RDS if obstruction is present. Can present w/ signs of RV volume overload is obstruction is not significant (similar to other Là R shunt lesions). **PE**: If vein obstruction, single loud S2, If no obstruction, increased RV impulse, SEM at LUSB, diastolic TV rumble. +/- fixed split S2. No significant cyanosis if Qp:Qs is high and there is no obstruction	**EKG**: RAD, RVH, +/-RAE. **CXR**: if pulm vein obstruction, pulm edema (similar to RDS), "Snowman in a snowstorm" 	• Emergent surgery if severe vein obstruction: anastomose pulm venous confluence to LA and close ASD • Supportive care including O2, inotropes, mechanical ventilation, ECMO as needed • Consider PGE if cyanotic, though need to be judicious as this can increase pulmonary blood flow and worsen pulmonary edema if obstruction present • Surgical goal = connect pulm veins to LA and close mixing lesion.

Cyanotic Heart Disease continued on next page →

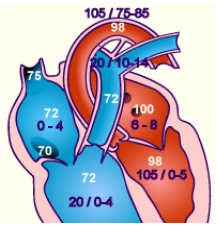
Cyanotic Heart Disease

Lesion	Basics	Hx and Exam	Studies	Treatment
Tricuspid atresia	• No outlet from RA-->RV. Supply to LA via PFO or ASD. • Classified based upon great arterial relationship (d-TGA in type II), presence of VSD and degree of PS • If no VSD, will have hypoplastic RV and pulmonary atresia • If + VSD, variable severity of RV and PA hypoplasia • Pulmonary blood flow may be PDA dependent	**Hx**: Variable timing (50% present on DOL 1), depending on size of VSD and degree of PS. Usually cyanotic by 2 months w/ cyanosis, tachypnea. **PE**: +/- VSD murmur. Single S2. 	**EKG**: RAE, LVH, LAD w/ superior axis (distinguishes TA from most other forms of cyanotic disease). **CXR**: Usually decreased pulmonary vascular markings. Can have increased if d-TGA	• PGE if cyanotic, to maintain pulm flow • Some neonates require atrial septostomy. • Manage CHF if present. • Surgical repair: staged palliation: BT shunt--> bidirectional Glenn-->Fontan. • Surgical goal = make two separate circulations w/ passive blood flow to the lungs and LV-driven systemic flow
Ebstein's Anomaly	• Tricuspid valve is inferiorly displaced into RV w/ leaflets adherent to RV wall, often associated w/ ASD/PFO and can have PS • Causes atrialization of the RV and RA enlargement • Impaired RV output 2/2 TR, RV dysfunction, possible RVOTO from redundant valve tissue. • Can cause a "circular shunt" in utero (Ao-->ductus-->retrograde PA -->RA--> PFO--> LA--> LV--> Ao) and hydrops • Frequently associated w/ WPW • Classically associated w/ maternal Li therapy	**Hx**: Variable presentation from cyanosis in delivery room and early right heart failure to adults w/ murmurs, arrhythmia or incidental EKG findings based upon degree of TV displacement **PE**: systolic murmur 2/2 TR. Often has gallop. 	**EKG**: RAE, RBBB. May have WPW and may present in AVRT. **CXR**: Cardiomegaly, which can be massive and box-like 2/2 RAE. Decreased pulmonary vascular markings can be normal.	• Consider PGE in neonates w/ severe cyanosis. • Improves as PVR falls • Surgical repair: Variable depending on severity, but may include TVplasty (Cone procedure) or replacement, reduction atrioplasty and ventricular plication. If severe, may require palliation down single ventricle pathway. • Surgical goal = improve RV function, reduce TR
Hypoplastic Left Heart Syndrome	• Group of left-sided obstructive anomalies characterized by underdevelopment of the left heart thought to be secondary to reduced in utero blood flow • Requires PDA and ASD for survival • Three types: • MS/AS • MS/AA • MA/AA • Further classified based upon presence or absence of unrestrictive atrial septal defect • If atrial septum is intact (IAS) or restrictive, outcome is poor	**Hx**: Presents w/ cyanosis secondary to left atrial hypertension and pulmonary edema if atrial septum intact or restrictive. Presents w/ cardiogenic shock and CHF if atrial septum unrestrictive as PDA closes. **PE**: Increased RV impulse, single S2, often no murmur, poor pulses, cool extremities	**EKG**: RVH, reduced left-sided forces. **CXR**: Cardiomegaly, ↑ pulm markings. 	• PGE to preserve ductal patency and systemic perfusion • Balloon atrial septostomy if IAS • Surgical repair: Three-stage univentricular palliation: • Atrial septectomy, creation of neoaorta, modified BT-shunt or Sano shunt v. Hybrid procedure • Bidirectional Glenn (superior cavopulmonary anastomosis) • Fontan (total cavopulmonary shunt) • May require heart transplant • Surgical goal = separation of pulmonary and systemic circulation w/ passive pulm return and RV-generated systemic flow

Cyanotic Heart Disease

Lesion	Basics	Hx and Exam	Studies	Treatment
Double Outlet Right Ventricle	• Family of lesions where both great vessels arise from RV • VSD always present • Three types : 1. TOF-type: oxygenated blood passing through VSD directed to aorta, PS present. 2. TGA-type: oxygenated blood directed through subpulmonic VSD to PA (Taussig-Bing heart). 3. VSD-type: normally-related vessels, no PS.	**Hx**: 1. TOF presents like TOF 2. TGA-type presents like TGA, but usually w/ better mixing 3. VSD type like VSD **PE**: variable, based on type of DORV	**EKG**: No hallmark EKG, because of variety of physiology types. **CXR**: Cardiomegaly and pulm flow depend on degree of PS present 	• Medical management determined by Qp:Qs. • Treat CHF if present • Surgical repair depends on physiology • Surgical goal = separation of pulmonary and systemic circulations versus single ventricle repair
Truncus arteriosus	• Failure of embryonic bulbar trunk to divide into PA and aorta. • Associated w/ a VSD, aortic arch and coronary anomalies • Several subtypes depending on how PAs come off the truncus. • Cyanosis is secondary to mixing • Both ventricles feed both arteries, pulmonary overcirculation worsens as PVR falls • Associated w/ 22q11 syndrome	**Hx**: CHF over first few weeks as PVR falls and dependent on degree of truncal valve regurgitation **PE**: loud single S2, ejection click. SEM at LUSB. Diastolic de-crescendo murmur from truncal regurgitation. Bounding pulses from diastolic runoff	**EKG**: LVH, RVH **CXR**: Cardiomegaly. Increased pulmonary vascular markings. +/- right-sided aortic arch. 	• Treat CHF if present • Surgical repair: Division of pulmonary arteries from truncus and placement of RV-PA conduit. Closure of VSD. • Surgical goal = establishing separated pulmonary and systemic circulations.
Pulmonary Atresia	• Fused pulm valve leaflets. Inability of flow from RVàPA. Malformed RV and TV w/ tricuspid regurg. • Pulm flow depends on PDA. • R-->L shunt via atrial or ventricular level. • PA w/ intact ventricular septum (PA-IVS) can result in a high pressure RV and RV-coronary fistulae à "RV-dependent coronary circulation"	**Hx**: Cyanosis at birth that worsens as PDA closes. **PE**: PDA murmur. 	**EKG**: Mild LAD from weak right side. RAE. **CXR**: ↓ pulm markings	• PGE in newborns • Surgical repair: surgical or cath valve repair. If RV cannot be grown by increase in flow. • Surgical goal = pulm valve integrity w/ normal circulation. If this not possible and RV remains non-functional, goal is Fontan physiology. • If coronary circulation is RV-dependent in PA-IVS, RV decompression may cause "steal" and massive ischemia

Catheterizations/Caring for the Post-Cath Child



Normal pressures/O2 sats

1. Inspect **access site (usually femoral) for bleeding** or hematoma formation.
2. Assess **distal pulses** and ensure they are intact and equal bilaterally
3. Compar**e lower extremity warmth, edema and skin color.** Signs of venous thrombus include edema, increased warmth and erythema. Signs of arterial thrombus include pain, pallor, paresthesia/numbness, poor pulses and cool extremities.
4. Listen t**o heart and lung sound**s and think about what you should be hearing given what procedures were performed
5. Most patients will require at least one **hemoglobin/hematocrit check to ensure they are not bleeding**
6. Some patients will require **a chest x-ray** to ensure they have not developed a pneumothorax and to ensure their device has not migrated