Congenital Heart Disease

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http://rc-edconsultant.com/

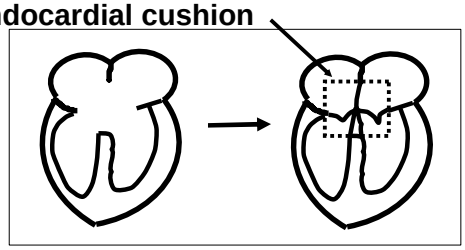
Learning Objectives:

- Identify common etiologies and risk factors for congenital heart defects.
- Describe clinical manifestations and diagnostic methods for congenital heart defects.
- Explain the pathophysiology, manifestations, diagnosis and management of acyanotic congenital cardiac anomalies.
- Explain the pathophysiology, manifestations, diagnosis and management of obstructive congenital anomalies.
- Explain the pathophysiology, manifestations, diagnosis and management of cyanotic congenital anomalies.
- Explain the implications of cardiac anomalies for respiratory care.

Development of the Cardiovascular System

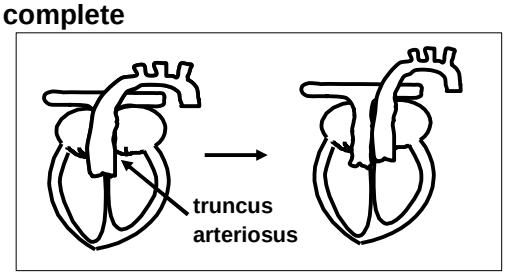
Development of the heart

- Parallel tubes convolute to form chambers
- Septa and valves form from endocardial cushion



Development of the heart

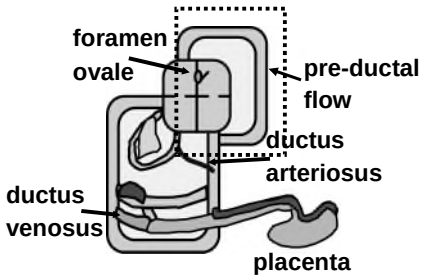
- Single artery (truncus arteriosus) divided by aorticopulmonary septum
- At eight weeks, fetal circulation is complete



Fetal circulation- anatomy

- Includes placental circulation- low resistance circuit
- Foramen ovale-- window between atria
- Ductus arteriosus-- vessel connecting aorta to pulmonary artery
- Ductus venosus- bypasses liver

Fetal circulation- anatomy



Click to see a diagram of fetal circulation
http://clem.mscd.edu/~haysc/public_html/bio232/fetalcirc.jpg

Fetal circulation- physiology

- High pulmonary vascular resistance
- Left side includes low resistance placental circuit
- Venous admixture at all shunts
- Pre-ductal blood with highest PaO2 to upper body

Changes at Birth

- Removal of placental circuit increases left-sided resistance
- Increased PaO2 lowers pulmonary vascular resistance
- Foramen ovale functionally closed- resistance on left > right
- Ductus closes due to increased PaO2, etc., about 15 hours postpartum

Congenital Heart Disease

Etiologic Factors

- maternal infections- rubella, syphilis
- maternal metabolic dx- diabetes
- maternal drug ingestion
  - phenytoin (Dilantin)
  - thalidomide
  - sex hormones

Medical history

- failure to thrive
- retarded growth, development
- decreased exercise tolerance
- squatting
- fainting

Medical history

- chronic pulmonary infections
- chronic cough
- feeding difficulties
- headaches
- epistaxis (nosebleeds)
- 'noisy breathing'

Physical examination

- small stature, underdeveloped
- color- may be cyanotic
- clubbing

Physical examination

- color- may be cyanotic
- clubbing
- heart murmurs- abnormal
  - blood flow
  - valve activity

Click for information on the physiology of heart murmurs
http://www.wilkes.med.ucla.edu/Physiology.htm

Physical examination

- cyanosis
- clubbing
- heart murmurs
- displaced point of maximal impulse (PMI)
- precordial bulge

Physical examination

- wheezing- CHD often mistaken for asthma
- tachypnea
- tachycardia

Physical examination

- wheezing- CHD often mistaken for asthma
- tachypnea
- tachycardia
- blood pressure greater in arms
- weak femoral pulses
- epistaxis

Diagnosis

- Radiography
  - chest radiograph
  - angiography
- Echocardiography- replaced catheterization for many defects

Click for information on echocardiography and CHD
http://www.echoincontext.com/advanced/chd_01.asp

Diagnosis

- Electrocardiography
- Blood gases and/or oximetry
  - pre, post-ductal SO2
  - SO2 in various compartments
- Cardiac catheterization
  - diagnostic
  - therapeutic

FYI - Click for article on diagnostic cardiac catheterization and CHD
http://www.ncbi.nlm.nih.gov/pmc/articles/PMC479386/

Categories

- Acyanotic CHD
- Obstructive defects
- Conduction defects
- Cyanotic CHD
- Miscellaneous
  - Dextrocardia
  - Vascular rings

Acyanotic Cardiac Anomalies

Acyanotic Anomaly Types

- Persistent fetal structures
  - patent ductus arteriosus
  - patent foramen ovale
- Septal defects
  - ventricular septal defects
  - atrial septal defects
  - endocardial cushion defects

Acyanotic Anomaly Types

- Obstructive defects
  - coarctation of aorta
  - aortic stenosis
- Conduction defects

Persistent Fetal Structures

- Types
  - patent ductus arteriosus
  - patent foramen ovale
- May persist asymptomatically, through adulthood.
  - exacerbated by pulmonary hypertension (hypoxemia)
  - shunt may change to right-to-left with PEEP, worsening hypoxemia

Persistent Fetal Structures

- Normal pulmonary vascular resistance
  - left-to-right shunt
    - no effect on arterial blood gases
    - elevated mixed venous PO2
  - increased LV work
    - LV failure
    - CHF

Persistent Fetal Structures

- Increased pulmonary vascular resistance
  - right-to-left shunt
  - hypoxemia, refractory to supplemental O2

Click to see persistent fetal circulation
http://www.kumc.edu/instruction/medicine/pedcard/cardiology/pedcardio/pfcdiagram.gif

Patent Ductus Arteriosus

- Second most common anomaly in term infants
- Etiologic factors
  - neonatal asphyxia, hypoxemia
  - maternal viral infections, e.g., rubella
  - low socioeconomic status- nutrition
- Note- patent ductus is necessary for survival in patients with ductal-dependent anomalies

Patent Ductus Arteriosus

- Complications
  - excessive workload on left ventricle
  - pulmonary artery disease (Eisenmenger's)
  - chronic pulmonary infections

Patent Ductus Arteriosus

- Manifestations
  - persistent murmur
  - decreased lung compliance ==> increased work of breathing
  - cardiomegaly
  - diagnosed by echocardiogram

Click for more information and pictures of PDA
http://www.ptcd.org/?id=patentductus1

Patent Ductus Arteriosus

- Management
  - Medical
    - ibuprofen (Advil) to close ductus
    - indomethacin to close ductus
    - intubate and ventilate with PEEP to improve oxygenation
  - Surgical
    - ligation (sometimes done in NICU)
    - division- requires thoracotomy

FYI - Click for article on PDA closure
http://pediatricct.surgery.ucsf.edu/conditions--procedures/patent-ductus-arteriosus.aspx

Septal Defects

- Normal pulmonary vascular resistance (PVR)
  - left-to-right shunt
    - no effect on arterial blood gases
    - elevated mixed venous PO2
  - increased LV work
    - LV failure
    - CHF

Septal Defects

- Normal PVR
  - left-to-right shunt
  - increased LV work
  - excessive pulmonary blood flow
    - causes chronic pulmonary infections
    - causes remodeling of pulmonary vasculature (Eisenmenger's complex)

Click to see evolution of Eisenmenger's complex
http://img.medscape.com/slide/migrated/editorial/cmecircle/2006/6423/images/landzberg/24.jpg

Septal Defects

- Increased pulmonary vascular resistance
  - right-to-left shunt
  - hypoxemia, refractory to supplemental O2

Septal Defects

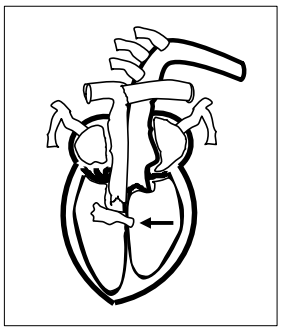
- Small VSD (less than diameter of aortic valve)
  - left-to-right shunt if VSD < 50% aortic diameter
  - RV & LV pressures normal
  - May close spontaneously

Small VSD

- Manifestations
  - may be asymptomatic
  - only clinical sign may be murmur
  - other data normal

Click to hear VSD murmur
http://www.wilkes.med.ucla.edu/Systolic.htm

Small VSD- Left-to-Right Shunt

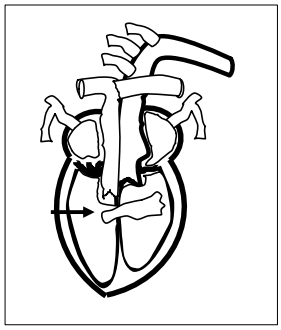


Large VSD

- VSD diameter > aortic valve
- Hemodynamics
  - excessive PA flow ==> vascular remodeling ==> increased PVR ==> right-to-left shunt (hypoxemia)
- Prolonged left-to-right shunt that causes remodeling of pulmonary vessels necessitates a heart & lung transplant

VSD- Right-to-Left Shunt

- venous admixture to left ventricle



Large VSD

- Manifestations
  - Murmur
  - CHF
  - Cyanosis with pulmonary hypertension
- LV hypertrophy

Click for more information and pictures of VSD
http://www.pted.org/?id=ventricularseptal1

Large VSD

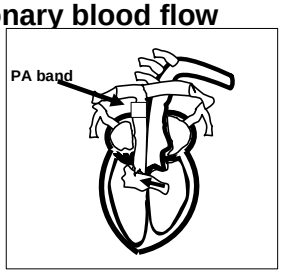
- Diagnosis
  - Echocardiography
  - Heart catheterization
  - Angiography

Large VSD

- Management
  - Palliation, to reduce pulmonary blood flow
    - PA banding
    - subambient FIO2- causes pulmonary vasoconstriction
  - Correction- Gortex patch closure

Pulmonary Artery Banding

- Palliative procedure to reduce pulmonary blood flow



Click for article about Flowatch PA band with illustrations
http://circ.ahajournals.org/content/110/11_suppl_1/II-158.full

Atrial Septal Defect

- Categories- based on position of the defect on atrial wall
  - ostium primum
  - ostium secundum
  - sinus venosus

Atrial Septal Defect

- Manifestations
  - murmur
  - may be otherwise asymptomatic for 20-30 years
  - normal PVR ==> left-to-right shunt ==> elevated RA and RV PO2

Click to hear murmur with ASD
http://www.wilkes.med.ucla.edu/Systolic.htm

Atrial Septal Defect

- Manifestations
  - murmur
  - may be otherwise asymptomatic for 20-30 years
  - normal PVR ==> left-to-right shunt ==> elevated RA and RV PO2
  - first sign may be right ventricular failure
  - may follow pathophysiology of VSD

Atrial Septal Defect

- Diagnosis
  - ECG - Right axis deviation
  - Echocardiography- detected with bubble test
  - Heart catheter- elevated RA, RV SO2

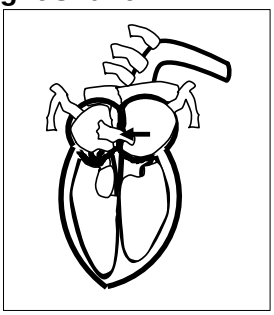
Atrial Septal Defect

- Diagnosis
  - ECG - Right axis deviation
  - Echocardiography- detected with bubble test
  - Heart catheter- high RA, RV SO2
- Treatment- closure in catheterization lab.

Click to see video on ASD repair (4 min)
http://www.youtube.com/watch?v=PzKJ_chafEU

Atrial Septal Defect

- left-to-right shunt



Click for more information and pictures of ASD
http://www.pted.org/?id=atrialseptal1

Endocardial Cushion Defect

- Pathogenesis- incomplete development of ECD
- Associated with trisomy 21 (Down's syndrome)
- Defects- permutations of:
  - ASD
  - VSD
  - Cleft mitral, tricuspid valve leaflets

Endocardial Cushion Defect

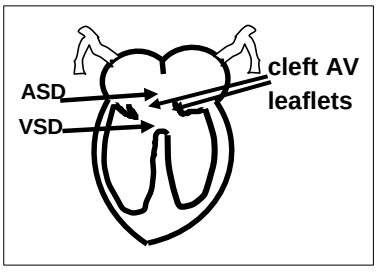
- Hemodynamics-- depend on specific defects
  - ASD- L to R shunt
  - VSD- L to R shunt ==> left ventricular hypertrophy
  - Mitral regurgitation ==> left atrial hypertrophy
  - Increased PA flow ==> vascular remodeling & increased PVR

Endocardial Cushion Defect

- Manifestations
  - May be asymptomatic
  - May develop severe CHF & pulmonary edema
- Diagnosis
  - ECG- left axis deviation
  - Heart catheter- increased SaO2 in RA & RV
  - Echocardiography

Endocardial Cushion Defect

- Complete AV canal



Click for more information and pictures of AV canal
http://www.pted.org/?id=atrioventricularcomplete1

Endocardial Cushion Defect

- Management
  - palliative PA banding
  - heart failure managment
    - diuretics
    - digitalis
  - surgical correction
    - septal defect closure- Dacron patch
    - valvuloplasty- technically difficult

Obstructive Anomalies

Aortic stenosis

- Narrowed aortic outflow tract
- Hemodynamics- increased resistance to LV outflow ==> increased LV work ==> hypertrophy ==> LV failure

Aortic stenosis

- Manifestations
  - Ejection systolic murmur
  - Left ventricular hypertrophy
  - CHF, sudden death (severe)
- Management
  - Valvotomy, balloon valvuloplasty
  - Valve replacement

Click for more information and pictures of aortic stenosis
http://www.pted.org/?id=aorticstenosis1

Coarctation of the Aorta

- Narrowing of portion of aorta
- Hemodynamics
  - aortic obstruction
  - severity dependent on degree of narrowing
- Associated with chromosomal abnormality- Turner's syndrome

Coarctation of the Aorta

- Manifestations
  - reduced pulses, blood pressure in lower extremities
  - headaches
  - epistaxis
  - leg cramps

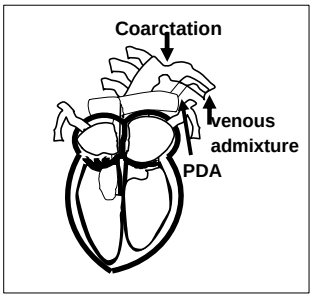
Coarctation of the Aorta

- Manifestations
  - left ventricular hypertrophy
  - CHF, pulmonary edema
  - neonates- lower body cyanosis
    - pre-ductal coarctation
    - in presence of PDA

Click for more information and pictures of coarctation
http://www.pted.org/?id=coarctation1

Coarctation of the Aorta

- Pre-ductal- coarctation proximal to ductus arteriosus



Coarctation of the Aorta

- Management
  - avoid heavy exercise
  - balloon dilatation with stent
  - resection- may require graft

Click to see surgical repair of coarctation (1.5 min)
http://www.youtube.com/watch?v=-9qguK2kZZw

Conduction defect

- Wolff-Parkinson-White syndrome
- Impulse aberrantly conducted through bundle of Kent
- Manifestations
  - PR interval < 0.12s
  - paroxysmal atrial tachycardia (PAT)
- Treatment
  - Medical- antidysrhythmics
  - Electrophysiology- ablation

Cyanotic Anomalies

Cyanotic Anomalies

- Categories:
  - increased pulmonary blood flow
  - decreased pulmonary flow

Cyanotic Anomalies

- Requirements for arterial desaturation
  - Communication between systemic & pulmonary circulation
    - abnormal vessels
    - septal defects
  - PVR > SVR
- Desaturation due to intracardiac shunt is unresponsive to increased FiO2

Cyanotic Anomalies

- Conditions with low pulmonary flow
  - Tetralogy of Fallot
  - Pulmonary atresia
  - Tricuspid atresia
- Bicuspid atresia, AKA hypoplastic left ventricle

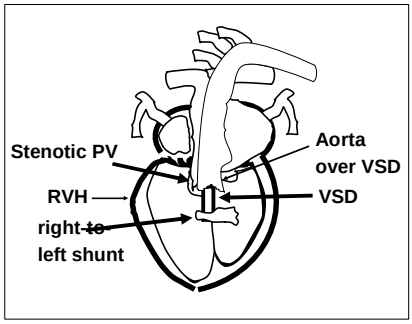
Cyanotic Anomalies

- Conditions with high pulmonary flow
  - Transposition of great vessels
  - Persistent truncus arteriosus
  - Total anomalous pulmonary venous return

Tetralogy of Fallot

- Defects (tetra = four)
  - Pulmonary stenosis
  - Ventricular septal defect (VSD)
  - Overriding aorta-- aorta straddles both ventricles
  - Right ventricular hypertrophy

Tetralogy of Fallot



Tetralogy of Fallot

- Hemodynamics
  - Pulmonary stenosis
    - Determines PA resistance to flow
    - Regulates resistance to right ventricular flow
    - Determines right to left shunt
    - Leads to RV hypertrophy
    - Degree of stenosis determines urgency of surgical intervention

Tetralogy of Fallot

- Hemodynamics
  - VSD- channel for shunt
    - Will be left-to-right with low pulmonary resistance
    - Usually large

Tetralogy of Fallot

- Hemodynamics
  - Overriding aorta
    - Carries outflow from both ventricles
    - Contributes to severity of shunt
  - RV hypertrophy
    - Chronic elevated flow resistance
    - Very large VSD- equalizes pressures in LV and RV

Tetralogy of Fallot

- Spectrum from "pink tets" to emergent cases in neonatal stage
- May not appear until closure of PDA, then pulmonary blood flow declines

Click for more information and pictures of TOF
http://www.pted.org/?id=tetralogyfallot1

Tetralogy of Fallot

- Manifestations
  - cyanosis- "tet spells" with exertion
  - squatting to relieve exertional spells
  - clubbing
  - growth retardation
  - systolic ejection murmur

Click to hear pulmonary stenosis murmur
http://www.wilkes.med.ucla.edu/Systolic.htm

Tetralogy of Fallot

- Chest xray- 'boot-shaped' heart
- ECG-- right axis deviation
- Echocardiography- usually definitive
- Catheterization

Click to see 'boot-shaped' heart on xray
http://www.bcm.edu/radiology/cases/pediatric/text/3a-desc.htm

Tetralogy of Fallot

- Management of tet spells
  - fetal positioning
  - morphine
  - oxygen- an exception for supplemental O2
  - bicarbonate
  - propanolol
  - vasoconstrictors

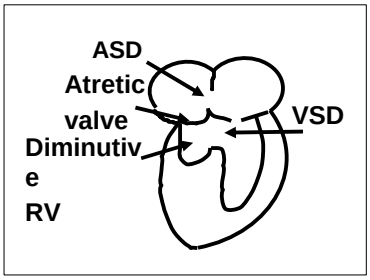
Tetralogy of Fallot

- Palliation- arterial to pulmonary artery shunts
  - bypass stenotic pulmonary valve
  - increase pulmonary blood flow
- Total correction
  - Excision of PV obstruction
  - Patch closure of VSD

Tricuspid atresia

- Defects
  - Atretic tricuspid valve- does not open, so blocks blood flow from atrium to ventricle
  - Diminutive (small) RV
  - VSD & ASD

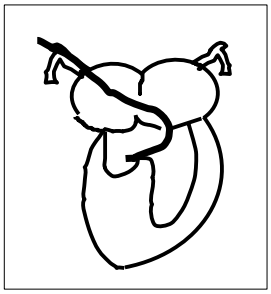
Tricuspid atresia



Tricuspid atresia

- Blood flow

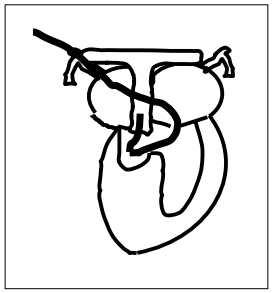
  Vena cava to RA to ASD to LA to LV to RV (via VSD)



Tricuspid atresia

- Blood flow

  Vena cava to RA to ASD to LA to LV to RV (via VSD) to PA



Click for more information and pictures of tricuspid atresia
http://www.pted.org/?id=tricuspidatresia1

Tricuspid atresia

- Signs
  - early cyanosis (from birth)
  - worsening, death on closure of ductus arteriosus
  - growth retardation
  - squatting
  - clubbing

Tricuspid atresia

- Diagnosis
  - ECG- left axis deviation
  - Echocardiography
    - diminutive right ventricle
    - absent tricuspid echoes
  - Catheterization-- catheter will not enter RV

Tricuspid atresia

- Palliative procedures- to increase pulmonary blood flow
  - Maintain PDA
    - subambient FIO2
    - alprostadil
    - stent placement
  - Waterston shunt-- aorta to RPA
  - Blalock-Taussig (BT) shunt- from subclavian artery to PA

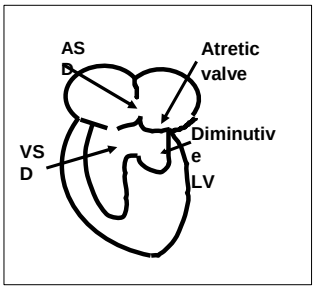
Tricuspid atresia

- Management
  - Corrective- Fontan
    - high risk, high failure rate
    - bypass RV by directing blood from RA to PA
    - pulmonary blood flow becomes dependent on passive venous return.

Click to see pictures of the Fontan procedure
http://www.pted.org/?id=fontan1

Bicuspid atresia- hypoplastic LV

- Defects
  - Atretic bicuspid valve
  - Diminutive LV
  - VSD & ASD



Hypoplastic LV

- Signs
  - early cyanosis
  - shock
  - worsening, death with DA closure

Hypoplastic LV

- Echocardiogram
  - diminutive left ventricle
  - absent bicuspid echo

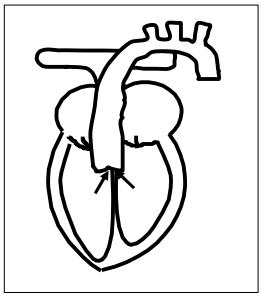
Click to see hypoplastic LV
http://www.pted.org/?id=hypoplasticleft1

Hypoplastic LV

- Maintain PDA
- Surgical management
  - Norwood- multiple stage procedure
  - Fontan
  - Blalock-Taussig (BT) shunt

Persistent truncus arteriosus

- Defects
  - Single artery for LV & RV
  - VSD



Persistent truncus arteriosus

- Hemodynamics
  - truncus carries blood to PA & aorta
  - flow is dependent upon resistance to flow at each side
    - increased SVR ==> increased pulmonary flow
    - increased PVR ==> increased systemic flow

Persistent truncus arteriosus

- Decreased PVR ==> excessive pulmonary blood flow ==>
  - high output LV failure (CHF)
  - pulmonary vascular dx
- Increased PVR ==> reduced in pulmonary blood flow ==> hypoxemia

Persistent truncus arteriosus

- Manifestations
  - Cyanosis
  - CHF

Click for more information and pictures of truncus arteriosus
http://www.pted.org/?id=truncusarteriosus1

Persistent truncus arteriosus

- Diagnosis
  - CXR- cardiomegaly
  - ECG- combined hypertrophy
  - Echocardiogram
    - visualize vessel origins
    - one semilunar valve
  - Catheterization- equal LV & RV pressures

Click to see chest xray of patient with truncus arteriosus
http://cardiopedia.wdfiles.com/local--files/truncus-arteriosus/truncus%20arteriosus%20type%201.jpg

Persistent truncus arteriosus

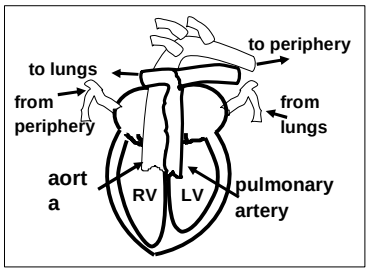
- Management
  - Heart failure
    - digoxin
    - diuretics
  - Palliative- reduce PA flow
    - PA banding
    - subambient FIO2

Persistent truncus arteriosus

- Corrective surgry
  - main trunk moved to left
  - creation of outflow tube from RV to PAs
  - closure of VSD

Transposition of great arteries (TGA)

- Defects
  - Aorta arises from RV
  - Pulmonary artery arises from LV
  - ASD and/or VSD, PDA (increase chance for survival)



TGA

- Hemodynamics
  - Separate circulations
  - Pulmonary venous blood to LA to LV through PA to lung
  - Systemic venous return to RA to RV to aorta to system
  - Without septal defect, life impossible
  - With VSD, there is mixing

Click for more information and pictures of TGA
http://www.pted.org/?id=transpositiond1

TGA

- Signs
  - diabetic mother- high risk
  - early cyanosis
  - CHF

TGA

- Diagnosis
  - CXR-- cardiomegaly
  - Echocardiogram- visualize vessels
  - Catheterization- catheter enters aorta from RV

Click for more information and pictures of TGA
http://www.pted.org/?id=transpositiond1

TGA

- Management
  - Palliative
    - maintain PDA
    - balloon septostomy

TGA

- Management
  - Corrective
    - Mustard-- baffle in atria
    - Jatene (switch)- vessels switched to correct ventricles

Therapeutics

Maintaining a PDA

- Indication- ductal dependent cardiac anomaly; e.g.:
  - transposition of great arteries
  - tricuspid atresia
  - mitral atresia
- Methods
  - stent
  - alprostadil (Prostin)
  - subambient O2

Subambient O2 Therapy

- goals
  - increase pulmonary vascular resistance to reduce pulmonary blood flow
    - large VSD
    - endocardial cushion defect
    - persistent truncus arteriosus
  - prevent closure of ductus arteriosus
    - transposition of great arteries
    - tricuspid atresia

Subambient O2 Therapy

- methods
  - bleed-in nitrogen to ventilator circuit
  - obtain premixed subambient mixture in cylinder
  - titrate FIO2 to SaO2 80-85%
- problem- some O2 analyzers may not measure subambient FO2

Post-surgical Considerations

Procedure	Response	Action
DA closure PIP PA band	Increase CL	Decrease
A-PA shunts PIP DA patency	Decrease CL	Increase

Special issues

- Transplant organs difficult to obtain.
- Patients' hearts can outgrow. synthetic structures, like valves.
- Oxygen therapy can kill patients with ductal dependent anomalies.

Summary & Review

- Development of the cardiovascular system
  - endocardial cushion
  - truncus arteriosus
  - fetal circulation with shunts
  - changes at birth

Summary & Review

- Congenital heart disease
  - etiologic factors
  - historical manifestations
  - physical manifestations
  - diagnostic procedures
  - categories
    - acyanotic
    - cyanotic
    - obstructive
    - conduction defects

Summary & Review

- Acyanotic defects
  - types
    - persistent fetal structures
    - ventricular septal defects
    - atrial septal defects
    - endocardial cushion defects
  - complications
    - remodeling of pulmonary vessels
    - left ventricular failure

Summary & Review

- Acyanotic defects
  - management
    - palliation with PA bands
    - total correction

Summary & Review

- Obstructive defects
  - types
    - aortic stenosis
    - coarctation of the aorta
  - manifestations
  - management
    - limit exercise
    - surgical repair

Summary & Review

- Conduction defect- WPW syndrome
  - abnormal conduction pathway
  - ECG- decreased P-R interval
  - management
    - medications for PAT
    - ablation of bundle of Kent

Summary & Review

- Cyanotic defects
  - types- high, vs. low pulmonary blood flow
  - ductal dependence
  - manifestations
    - cyanosis
    - tetralogy spells
    - ventricular failure

Summary & Review

- Cyanotic defects
  - tetralogy of Fallot
  - tricuspid atresia
  - hypoplastic left ventricle (mitral atresia)
  - Persistent truncus arteriosus
  - Transposition of great arteries

Summary & Review

- Subambient oxygen therapy
- Postoperative expectations
- Issues in congenital heart disease

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