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*Congenital Heart
Disease: Rehabilitation
Considerations for the Pediatric
Population*

Krista I. Gabello, DPT, PCS
PhysicalTherapy.com
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Objectives

- Identify at least three components of cardiopulmonary development in the newborn patient.
- List at least three common cardiac pathologies in the pediatric patient.
- Describe at least three components of medical management of the pediatric cardiopulmonary patient.
- List at least three appropriate therapeutic interventions for the pediatric cardiopulmonary patient.

CARDIOPULMONARY DEVELOPMENT

CARDIOPULMONARY DEVELOPMENT

- Pulmonary structures in **infants**
 - Narrow airway from nares to terminal bronchioles.
 - High position of larynx
 - Low compliance of newborn lungs
 - Immature alveolar structure and function
 - Less type I muscle fibers in the diaphragm

CARDIOPULMONARY DEVELOPMENT

- Biomechanical growth and development of the thoracic cavity.
 - Shape, the effects of gravity, function.
 - Triangular shape of newborn chest
 - Horizontal presentation ribs
 - Narrow intercostals spacing



CARDIOPULMONARY DEVELOPMENT

- Biomechanical growth and development of the thoracic cavity.
 - Anterior chest wall development
 - Increased extension
 - Upright against gravity:
 - Rectangular shape of chest
 - Angular presentation of ribs (>7years old)
 - Increased intercostals spacing

INFANT VS. ADULT THORAX



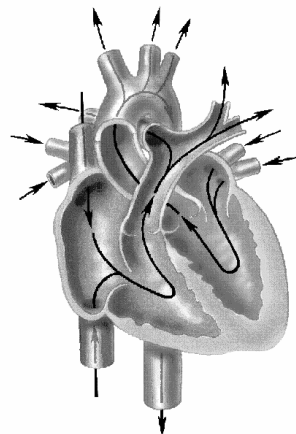
ONE VS. FOUR MONTHS





CARDIOPULMONARY DEVELOPMENT

CIRCULATION & PHYSIOLOGY



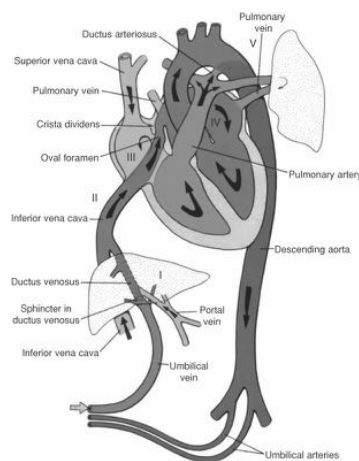
Quick Tip:

Tips to help understand heart *function* and *dysfunction*:

“Right Heart” = **low** saturations & pressure.

“Left Heart” = **high** saturations and pressure.

FETAL CIRCULATION



CONGENITAL HEART DEFECT

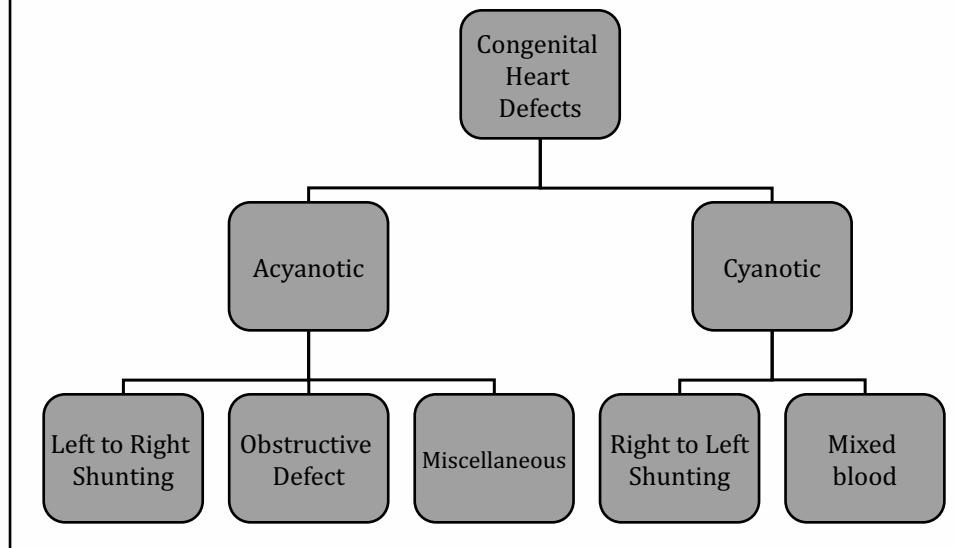
- Persistence of fetal cardiac structure
- Malformation of heart or great vessels
- Abnormal embryology
 - teratogens & genetics
- At least 15% of CHDs are associated with genetic conditions.
 - Some common genetic syndromes: Tiresome 21, DiGeorge, Marfans, Williams, Turners...

CONGENITAL HEART DEFECT

- Stats:
 - Most occur during the 4th-7th week of gestation.
 - 1% (~40,000) births/year in US
 - Increases to 2.5-4% if parent or sibling has CHD
 - ~20% to 30% of people with a CHD have other physical problems / developmental / cognitive disorders
 - Extra-cardiac anomalies occur in 20-45%
 - 16% have skeletal, muscle or skin involvement
 - 15% have GI anomalies
 - 15% have urogenital anomalies

• <http://www.cdc.gov/ncbddd/heartdefects/data.html>

CONGENITAL HEART DEFECT



ACYANOTIC DEFECTS

- Blood shunts from the LEFT → Right side of the heart
- Patients are PINK with normal O2 saturations.

CYANOTIC DEFECTS

- Patients are blue/dusky with O2 saturations 15-30% below normal
- Blood shunts from the RIGHT → Left side of the heart

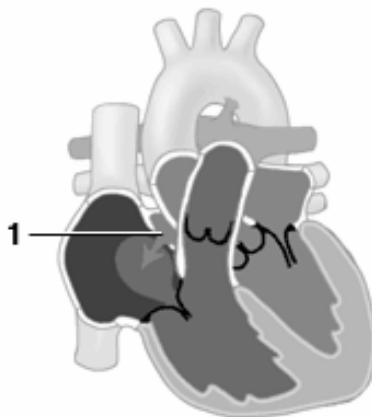
CONGENITAL HEART DEFECT

- | | |
|----------------------------|---------------------------------------|
| ▪ Acyanotic | • <u>Cyanotic</u> |
| ▪ ASD | • TOF |
| ▪ VSD | • HLHS |
| ▪ PDA | • Transposition of the Great Arteries |
| ▪ Coarctation of the aorta | • Tricuspid Atresia |
| ▪ Pulmonary stenosis | • Pulmonary Atresia |
| ▪ Aortic stenosis | |

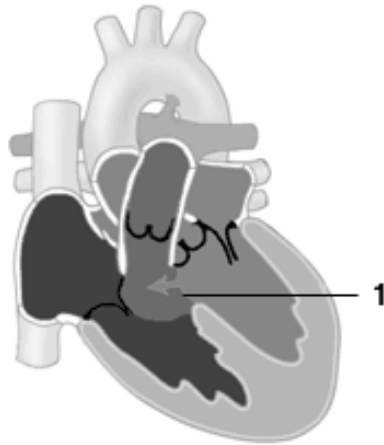
ACYANOTIC DEFECTS



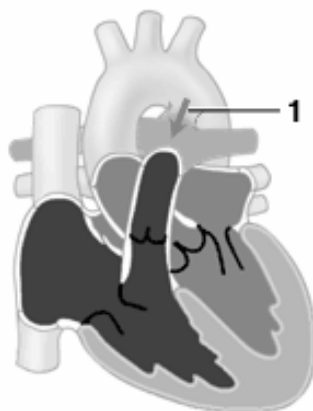
ATRIAL SEPTAL DEFECT (ASD)



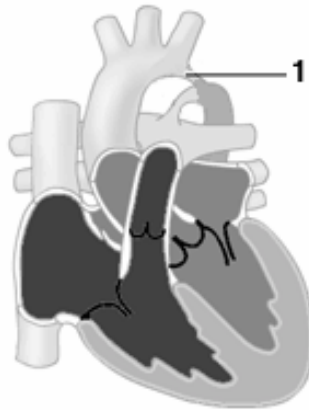
VENTRICULAR SEPTAL DEFECT (VSD)



PATENT DUCTUS ARTERIOSUS (PDA)

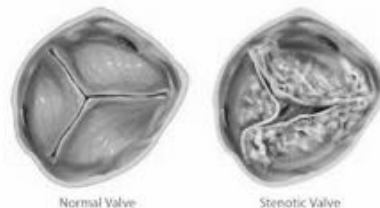


COARCTATION OF THE AORTA

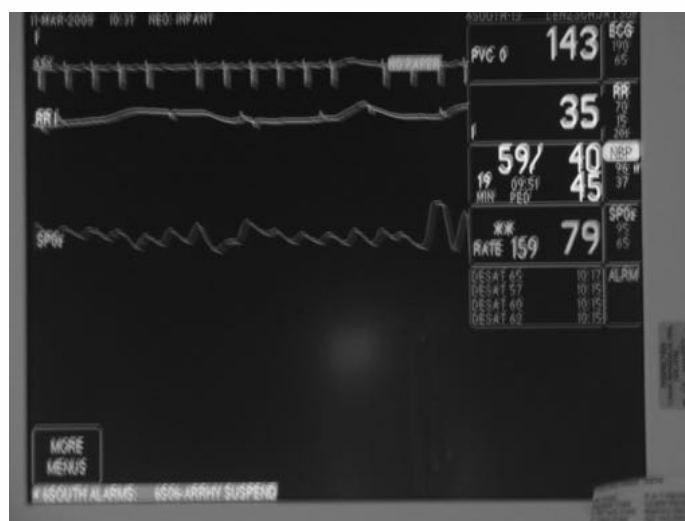


STENOSIS

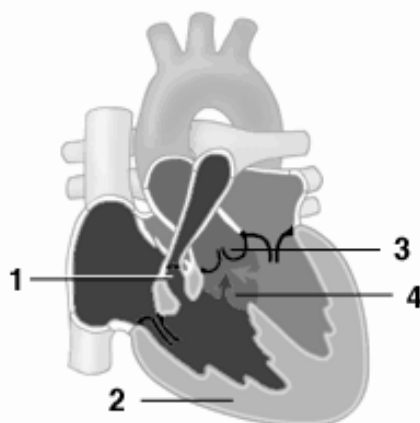
- Pulmonary Stenosis:
 - Narrowing of the RIGHT ventricular outflow tract.
- Aortic Stenosis:
 - Narrowing of the LEFT ventricular outflow tract



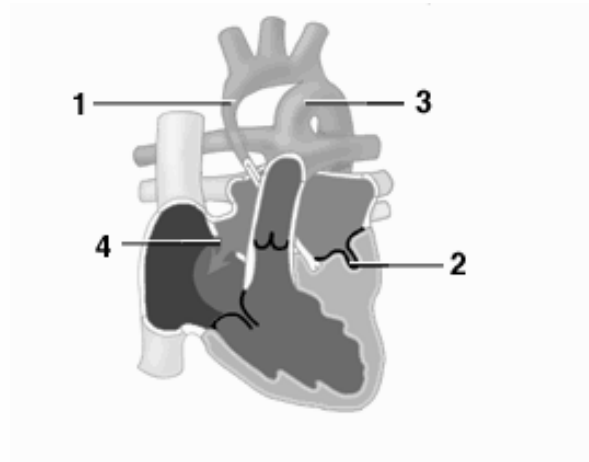
CYANOTIC DEFECT



TETROLOGY OF FALLOT (TOF)



HYPOPLASTIC LEFT HEART (HLHS)

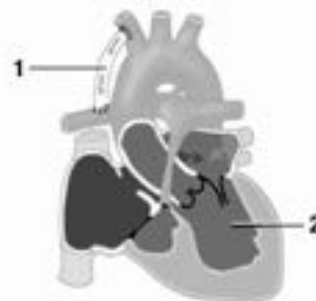


HLHS- FONTAN CIRCULATION

- 3 stage procedure used as intervention for most single ventricle defects
 - Stage I palliation: Sano or BT shunt
 - Stage II: Bi-directional Glenn or Hemi-Fontan
 - Stage III- Fontan completion

STAGE I: NORWOOD

- What do I need to know?
 - Goal: balanced blood flow between heart & lungs
 - O₂ saturations = variable but low
 - Numerous surgical options depending on the defect.

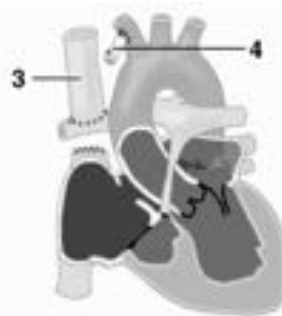


BT-shunt

- <https://www.youtube.com/watch?v=-87kq98I1kk>

STAGE II

- What do I need to know?
 - Bi-directional Glenn OR Hemi-Fontan Procedure
 - O₂ Saturations 75-85%
 - WHY? The blood from the inferior vena cava is bypassing the lungs

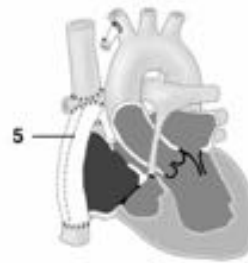


Bidirectional Glenn

- <https://www.youtube.com/watch?v=fUPVkkvPGe4>

STAGE III- FONTAN COMPLETION

- What do I need to know?
 - Extra cardiac or lateral tunnel
 - O2 Saturations = 90% and higher
 - Blood thinners are stopped.
 - Activity to tolerance- no restrictions



Extra -Cardiac

https://www.youtube.com/watch?v=mu_amCCD8gg

HLHS ANIMATIONS

- Stage I
 - <https://www.youtube.com/watch?v=-87kq98l1kk>
- Stage II
 - <https://www.youtube.com/watch?v=fUPVkkvPGe4>
- Stage III
 - https://www.youtube.com/watch?v=mu_amCCD8gg

FONTAN CIRCULATION

- Success rates are 90 % and higher
- Speculation that most single ventricle hearts will not function efficiently beyond 30 to 40 years
- Technique is just over 35 years old
- Known late complications: irregular rhythms and heart failure may occur

Surgical repair failure....

INDICATIONS

- Heart Failure:
 - congenital defect
 - cardiomyopathy
 - intractable arrhythmia
- Contraindications.....

CARDIOMYOPATHY

- Dilated
- Hypertrophic
- Restrictive

INTRACTABLE ARRHYTHMIA

- Atrial fibrillation
- Atrial flutter
- Wolff-Parkinson-White syndrome
- Ventricular tachycardia
- Ventricular fibrillation
- Prolonged QT interval

HEART FAILURE INTERVENTION :

- CHD, Cardiomyopathy & arrhythmia
 - Pharmaceutical Management
 - Pace makers
 - ICD

PHARMACEUTICAL MANAGEMENT

- Inotropic meds- affect myocardial contractility (+ or -)
 - Milrinone
 - Digoxin
 - Amrinone
 - Dopamine
 - Dobutamine

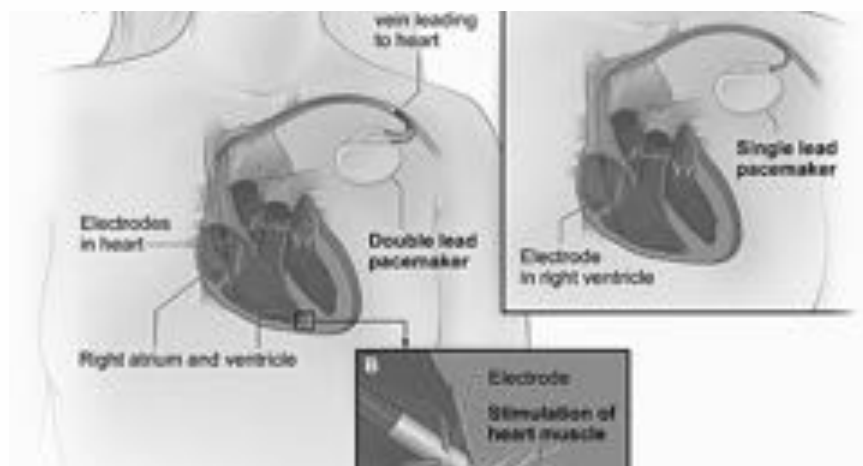
PHARMACEUTICAL MANAGEMENT

- calcium channel blockers
- anticoagulants
- diuretics
- digoxin
- vasodilators

PHARMACEUTICAL MANAGEMENT

- Anti-Arrhythmic
 - Lidocaine
 - Mexiletine
 - Amiodarone
- Pulmonary vasodilators
 - Prostaglandins
 - Oxygen
 - Sildenafil
 - Nitric oxide
- Take away information
 - www.nlm.nih.gov/medlineplus/druginfo

PACEMAKERS



PACEMAKERS

- use low-energy electrical pulses to overcome faulty electrical signaling
 - Speed up a slow heart rhythm.
 - Help control an abnormal or fast heart rhythm.
 - Make sure the ventricles contract normally if the atria are quivering instead of beating with a normal rhythm
 - Coordinate electrical signaling between the atria and ventricles

PACEMAKERS

- Pacemakers can monitor:
 - and record heart's electrical activity and heart rhythm.
 - blood temperature
 - breathing rate
- Pacemakers can adjust heart rate in response to activity

TYPES OF PACEMAKERS

- demand pacing
 - Monitors heart rhythm. It only sends electrical pulses if heart is beating too slow or if it misses a beat.
- rate-responsive pacing
 - speeds up or slows down heart rate depending on activity

PACEMAKERS AND ACTIVITY

- In most cases, a pacemaker will not limit sports and exercise, including strenuous activities.
- full-contact sports, such as football may be avoided.

IMPLANTABLE CARDIOVERTER DEFIBRILLATOR

- An ICD uses electrical pulses or shocks to help control life-threatening arrhythmias, especially those that can cause sudden cardiac arrest.

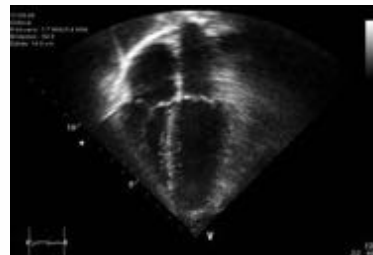
HOW DO WE CONFIRM FAILURE?

ASSESSMENT TOOLS

- “Primitive”
 - Visual observation
 - Palpation
 - Auscultation
 - Vital signs
 - Chest x-ray
 - ECG / EKG
- “Advanced”
 - Echo
 - Cardiac catheterization
 - Cardiac CT
 - Cardiac MRI
 - Radionuclide scans
 - Rhythm studies

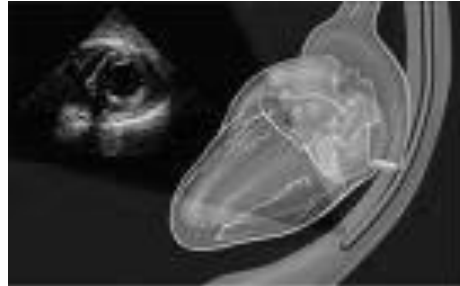
ECHO

- non-invasive imaging
- 3D ECHO
 - direct assessment of myocardial motion
- 2D doppler-
 - heart & great vessel anatomy
 - evaluates SF & EF
 - detects valve regurgitation, cardiac shunting, estimates pressure gradients.



TRANSESOPHAGEAL ECHO (TEE)

- Invasive
- Allows immediate revision when used intra-operatively
- Appropriate for patients with poor thoracic windows



CARDIAC CATHETERIZATION

- Invasive- usually the final definitive test
- Degree of shunting
- Severity of obstruction
- Coronary angiography
- Oxygen saturations at each site
 - Cardiac output ultimately calculated



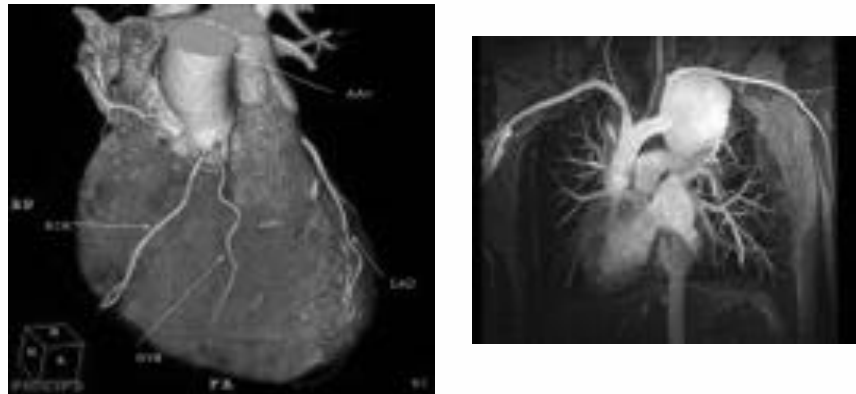
CARDIAC CT

- Imaging is comparable to catheterization
 - Advantages over cath: reduced sedation, short imaging time, can be used with pacemakers, stents & ICD
 - Disadvantages over cath: requires radiation, little or no info on cardiac function

CARDIAC MRI

- Non-invasive- cardiac structure, myocardial & valvular function, blood flow & myocardial perfusion in ONE test.
 - Dark blood imagining
 - Bright blood imagining
- Intubation in peds. Due to requirement of breath holding.

CARDIAC MRI



RADIONUCLEAR METHODS

- Non-invasive
- Determines percent of flow to right & left lungs
- Estimates degree of left to right shunting
- Evaluate ventricular function.
- Nuclear med. scans requiring injection of isotopes = small dose radiation

RHYTHM STUDIES

- Holter Monitor
- Electrophysiology

If conventional interventions are not successful.....

TRANSPLANT OVERVIEW

- Transplant waiting list
 - Heart Status:
 - 1A/urgent
 - 1B
 - 2
 - 7

• <https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3231539/>

TRANSPLANT OVERVIEW

- UNOS- United Network for Organ Sharing
 - Manages the *nationwide* organ donor & waiting list
 - Monitors every organ match
 - Develops policies & sets the standards

TRANSPLANT OVERVIEW

- Transplant waiting list
 - Allocation
 - Blood type
 - Degree of medical urgency
 - Geography
 - Size



26 year overview of Transplant by organ type....

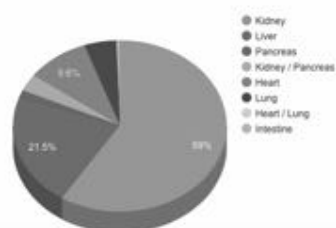


National data

Transplants By Organ Type January 1, 1988 - December 31, 2016

Based on OPTN data as of January 22, 2017

Organ	Transplants
Kidney	405,341
Liver	147,842
Pancreas	8,348
Kidney / Pancreas	22,122
Heart	65,700
Lung	33,363
Heart / Lung	1,199
Intestine	2,805
Total	686,798



BRIDGE TO TRANSPLANT

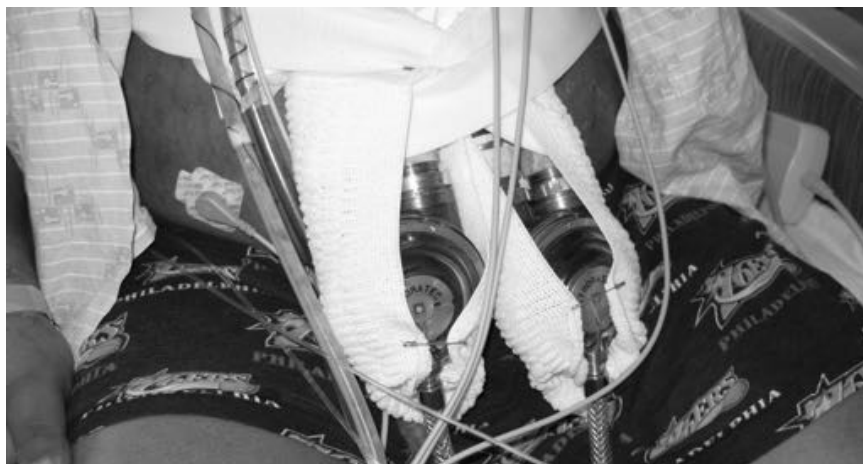
- VAD- ventricular assistive device.
 - Berlin Heart
 - SynCardia
 - HeartWear
- ECMO- Extra Corporeal Membrane Oxygenation.

VENTRICULAR ASSISTIVE DEVICE

- Mechanical pump that does the work for a failing heart
 - LVAD
 - RVAD
 - Bi-VAD
- High risk for bleed or clot
- What do I need to know?
 - The patient can & should be active
 - Do NOT kink the lines
 - Always support the lines



BI-VAD



Berlin Heart (EXCOR®)



HeartWear / HVAD® System



SYNCARDIA



VAD: What do we do for these kids?....



What do we do.....



What do we do.....



ECMO

- Used as a prolonged but still temporary support of the heart AND/OR lungs
 - Venoarterial
 - Venovenous
- High risk for bleed or clot
- What do I need to know?
 - No activity (usually!)
 - Help with position changes



ECMO: What do we do for these kids?....



HEART TRANSPLANT

- Median sternotomy
- Reanastomosis of atria to donor heart
- Great arteries reanastomosed
- Vagus nerve removed
- Cervical and thoracic sympathetic cardiac nerves are severed = denervated heart
 - Faster resting heart rate.

TRANSPLANT COMPLICATIONS

- Rejection
 - Immune mediated response (T cell driven) in which the body recognizes the transplanted organ as foreign.
 - Result- transplanted cell destruction & death
 - Graded depending on the severity
- Heart:
 - Cellular
 - Non-Cellular
 - Vascular

TRANSPLANT COMPLICATIONS

- “commandments” of rejection
 - 1) Everyone will reject
 - 2) Non-invasive way to diagnosis rejection are in R & D
 - 3) Rejection can be acute OR chronic
 - 4) You are never free of the possibility of rejection
 - 5) Most rejection can be treated completely
 - 6) Rejection can kill

TRANSPLANT COMPLICATIONS

- Immunosuppression
 - Agents provided to decrease the opportunity for rejection
- Increased risk of INFECTION
 - Wash your hands, toys, monitors!
 - Avoid or wear a mask in public place
 - No live virus vaccines

CONGENITAL HEART DISEASE: WHAT IS OUR ROLE AS THERAPISTS?

OUR ROLE AS THERAPISTS

1) Neurodevelopment issues may include problems affecting:

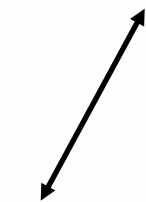
- Behavior
- Social interaction
- Gross and fine Motor skills
- Feeding
- Academic performance
- Speech and language development

2) Musculoskeletal

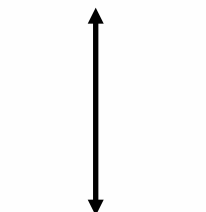
3) Integumentary

ROLE OF THERAPISTS

• Spans acute care  Life long health & wellness



Acute
Care /
rehab



Early
Intervention /
home care



Outpatient
/School
based

WHAT DO WE DO FOR THESE KIDS?

- **Pre-operative:**

- Positioning
- Exercise testing- ie: Six Minute Walk Test
- Functional mobility / developmental testing and facilitation
- Endurance training
- Pulmonary management- incentive spirometer
- Oropharyngeal Dysphasia evaluation- *videofluoroscopic swallowing study* (VFSS).
- Speech and language evaluation- receptive and expressive assessment

WHAT DO WE DO FOR THESE KIDS?

- **Pre-Operatively**

- Postural Education
- Strengthening & stretching
- Education- patient /family
- Referral- cardiac rehab., child life, play groups, sports, music & art therapies etc...

*GOAL = objective baseline assessment and optimal conditioning

WHAT DO WE DO FOR THESE KIDS?

- **Post-Operatively:**
 - Early mobilization
 - Sternal precaution education
 - Exercise testing- Six Minute Walk Test
 - Functional mobility / developmental testing & facilitation
 - Pulmonary management
 - Oropharyngeal Dysphasia evaluation
 - Speech and language evaluation
 - Postural Education

WHAT DO WE DO FOR THESE KIDS?

- **Post- Operatively**
 - Endurance training
 - Strengthen
 - Education- patient/family
 - **Scar management**
 - **Discharge planning**
 - Referral- cardiac rehab., child life, play groups, sports, music & art therapies etc...
 - * Community, school, vocational support service referrals

WHAT ARE STERNAL PRECAUTIONS?

- **Two weeks post-op:**
 - Patient may begin prone activities.
 - Patients may begin to weight bear through upper extremities into a walker.
- **For 1st 6 weeks post-op:**
 - Patients may assist with bed mobility as tolerated.
 - Patients may not be lifted under arms or pulled to sit by hands.
 - Patients may not push, pull, or lift greater than 10lbs.
 - Patients may not raise both arms simultaneously greater than 90 degrees shoulder flexion or abduction. Raising arm unilaterally greater than 90 degrees is allowed.
 - Patients must avoid distracting forces on sternum, such as reaching behind back with both arms simultaneously or twisting upper body.
- **continued...**
 - No driving
 - No bending down at waist (i.e. tying shoes)
 - Scar massage may begin after skin is completely healed (at least 6 weeks after closure with no complications).
- **OT application:**
 - Handling babies during OT sessions while observing precautions and parent ed
 - ADL re-training for the child/adult with sternal precautions with adaptive equipment, positioning, or body mechanics
- Of course, a thorough chart review will reveal any special precautions that need to be taken if the sternal closure was delayed or complicated by wound dehiscence or infection. As always, consult MD with any questions.

WHAT DO WE DO FOR THESE KIDS?

- Exercise implications:
 - 1) Warm up & cool down
 - 2) Higher resting heart- transplant
 - 3) Lower peak heart rate- transplant
 - 4) Sternal precautions with weight training, UE ROM, postural education
- BORG
- Monitor vitals throughout

WHAT DO WE DO FOR THESE KIDS?

- Physical / Speech & feeding implications
 - Hemi-diaphragm



What do we do for these kids?....

- Vocal cord Paralysis
 - A breathy quality to the voice
 - Noisy breathing
 - Loss of vocal pitch
 - Choking or coughing while swallowing food, drink or saliva
- The need to take frequent breaths while speaking
- Inability to speak loudly
- Loss of your gag reflex
- Ineffective coughing
- Frequent throat clearing
- Hoarseness

EXAMINATION

- History:
 - Birth: Apgars, pre-maturity
 - Prior surgical intervention(s)
- Systems Review & Referrals :
 - Recommend PM&R, Audiology, ophthalmology, orthopedics etc..
- Test & measures:
 - Developmental testing, Six Minute Walk, Incentive Spirometry, MMT, ROM, VFSS

STRIDOR



EVALUATION

- Observation is key!
- Environment
- Vital signs
- Skin integrity
- State control
- Respiratory status
- Posture/positioning
- Tone
- Strength
- Endurance
- Balance & coordination

DIAGNOSIS

- Musculoskeletal
- Neuromuscular
- Cardiopulmonary
- Integumentary
- Speech & language
- Feeding

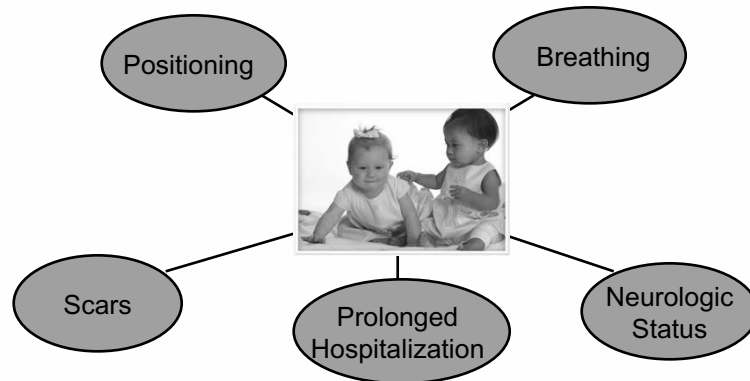
PROGNOSIS

- GOALS
- Frequency
- Discharge planning
 - Age
 - Medical status
 - Future surgeries
 - Medical complications
 - Compliance
 - Social-emotional status

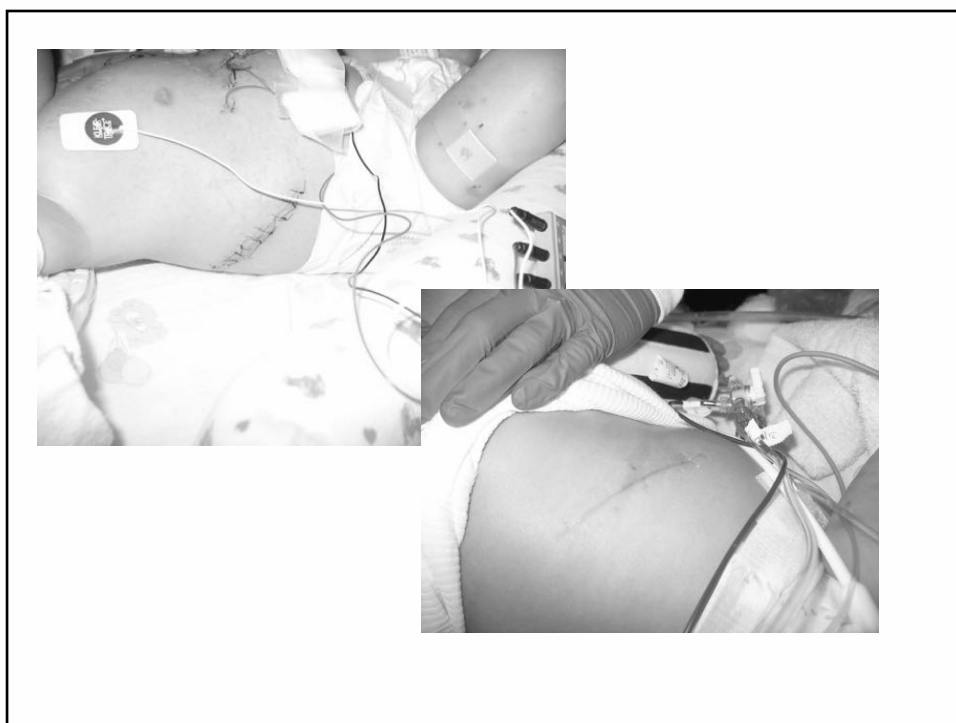
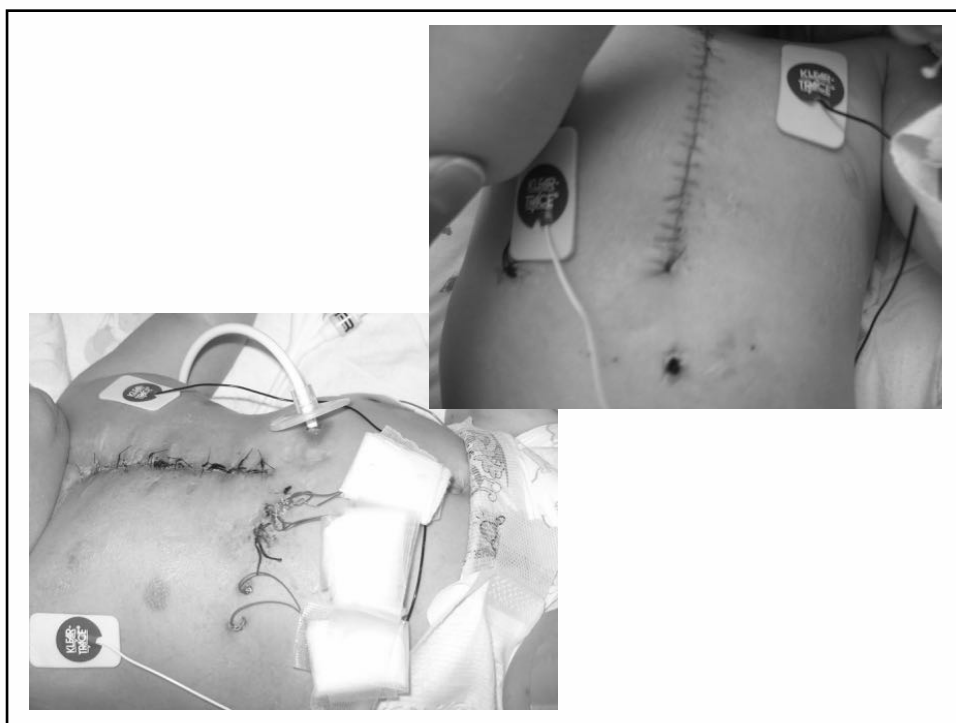
INTERVENTION

- Developmental through Geriatric
 - Positioning
 - Developmental facilitation / stimulation
 - Sensory integration
 - Aerobic conditioning
 - Strength training
 - Functional training
 - Respiratory training
 - Speech and language / feeding
 - Postural education
 - Scar management
 - Caregiver education
 - Discharge planning
 - **Life-long health and wellness**

Barriers To Development













PATIENT CASE

- AW is a 26 month old female with complex congenital heart defects.
 - * Heterotaxy syndrome, dextrocardia, pulmonary stenosis, congenital complete heart block, common atria, DORV, CHF
- Admitted to CHOP on 11/26/07 (14 mths) for transplant evaluation.
 - Boston, USC & PITT all turned her down for evals.
- Birth Hx: prenatal dx; full term, c-section

PATIENT CASE

- PMH (“in a nut shell”)
 - cardiac palliation (BT shunt)
 - PHTN
 - tracheobronchomalacia (vent & trach dependent)
 - Dual chamber pacemaker (multiple revisions)
 - FTT (g-tube)
 - three documented arrests
- AW waited from 11/26/07 until 9/5/08 for heart transplant
- AW discharged to UVA on 11/2/08:
 - **HME 6 hours/day**
 - **PO trials**



QUESTIONS?

CARDIORESPIRATORY PARAMETERS

	Heart Rate	Respiratory Rate	Blood Pressure	Temperature
Neonate	100-180	30-50	60/30-90/60	98.6 F
Infant	100-160	30-50	87/53-105/66	98.6 F
Toddler	80-110	24-40	95/53-110/69	98.6 F
Preschool	70-110	22-34	96/55-110/69	98.6 F
School age	65-110	16-24	97/57-112/71	98.6 F
Adolescent	60-90	12-20	112/66-128/80	98.6 F

Values courtesy of The Children's Hospital of Philadelphia

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