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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

- 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 intercostal 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 (>7 years old)
      - Increased intercostal 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

Congenital Heart Defects

- Acyanotic
  - Left to Right Shunting
  - Obstructive Defect
  - Miscellaneous
- Cyanotic
  - Right to Left Shunting
  - Mixed blood

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
  - ASD
  - VSD
  - PDA
  - Coarctation of the aorta
  - Pulmonary stenosis
  - Aortic stenosis

- Cyanotic
  - TOF
  - HLHS
  - Transposition of the Great Arteries
  - Tricuspid Atresia
  - Pulmonary Atresia
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.

  • [Link](https://www.youtube.com/watch?v=9Q5q9vrh1Ek)

  ![BT-shunt](image)

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

  • [Link](https://www.youtube.com/watch?v=fJpVk6vPGe4)

  ![Bidirectional Glenn](image)
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

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

Extra - Cardiac

HLHS ANIMATIONS

• Stage I
  • https://www.youtube.com/watch?v=-87kq98l1kk

• Stage II
  • https://www.youtube.com/watch?v=fUPVkJvPGe4

• 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
  - Prostoglandins
  - 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 imagining 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....
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

“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 decease 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.
  - 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

Positioning

Breathing

Scars

Prolonged Hospitalization

Neurologic Status
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

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Values courtesy of The Children’s Hospital of Philadelphia

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CITATIONS