Pediatric Palliative Care:

For Practitioners Who Take Care of Adults-Update

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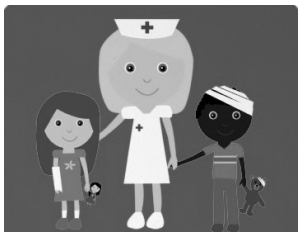
Objectives

- Learn the basics of the common conditions that effect children in hospice and palliative care, focusing on the diagnoses of our current/recent patients
- Review medications and doses for children
- Learn how to communicate with children of different ages



Causes of Death for Infants (Birth-1 year)

1. Congenital malformations (19.5%)
2. Short gestation /LBW (16.5%)
3. Sudden Infant Death Syndrome (7.4%)
4. Maternal complications (6.3%)
5. Complications of placenta, cord, or membranes (4%)
6. Accidents/unintentional injury (4%)

Causes of Death for Children (1-19 years)

1. Accidents
2. Assault
3. Malignancy
4. Suicide
5. Congenital malformations, deformations
6. Chromosomal anomalies
7. Heart disease
8. Cerebrovascular

Cancer Death Rates* in Children 0-14 Years by Sex, US, 2001-2005

Site	Male	Female	Total
All sites	2.7	2.3	2.5
Leukemia	0.8	0.7	0.8
Acute Lymphocytic	0.4	0.3	0.4
Brain/ONS	0.8	0.7	0.7
Non-Hodgkin lymphoma	0.1	0.1	0.1
Soft tissue	0.1	0.1	0.1
Bone and Joint	0.1	0.1	0.1
Kidney and Renal pelvis	0.1	0.1	0.1

*Per 100,000, age-adjusted to the 2000 US standard population.
ONS = Other nervous system
Source: Surveillance, Epidemiology, and End Results Program, 1975-2005, Division of Cancer Control and Population Sciences, National Cancer Institute, 2008.

Current Diagnosis at HPCC

- Cerebral Palsy
- Congenital Heart Disease
  - Tetralogy of Fallot
  - Hypoplastic Heart Syndrome
- Cerebral Hemorrhage at Birth
- Trisomy 13

Cerebral Palsy

- Name for a number of neurological disorders that permanently affect body movement and muscle coordination caused by injury or abnormal development in the immature brain, most often before birth
- Not a progressive disease
- Incidence is significantly higher in pre-term infants
- Problem with the area of the brain that affects muscle coordination
- Wide array of symptoms and disability
- Now 90% of patients survive to adulthood

Cerebral Palsy - Symptoms

- Very variable!
- Often they have other conditions related to developmental brain abnormalities, such as intellectual disabilities, vision and hearing problems, or seizures
- It is often these other conditions that cause a lot of the morbidity

Cerebral Palsy - Symptoms

- Variations in muscle tone - too stiff or too floppy
- Stiff muscles and exaggerated reflexes (spasticity)
- Stiff muscles with normal reflexes (rigidity)
- Lack of muscle coordination (ataxia)
- Tremors or involuntary movements
- Slow, writhing movements (athetosis)
- Delays in reaching motor skills milestones
- Difficulty walking, such as walking on toes, a crouched gait, a scissors-like gait with knees crossing or a wide gait
- Excessive drooling or difficulty with swallowing
- Difficulty with sucking or eating
- Delays in speech development or difficulty speaking
- Difficulty with precise motions, such as picking up a crayon or spoon

Cerebral Palsy - Treatment

- Physical, Occupational and Speech Therapy
- For isolated spasticity- Botox injections
- For more generalized spasticity- muscle relaxers such as Valium and Baclofen
- Surgical interventions for patients with severe contractures

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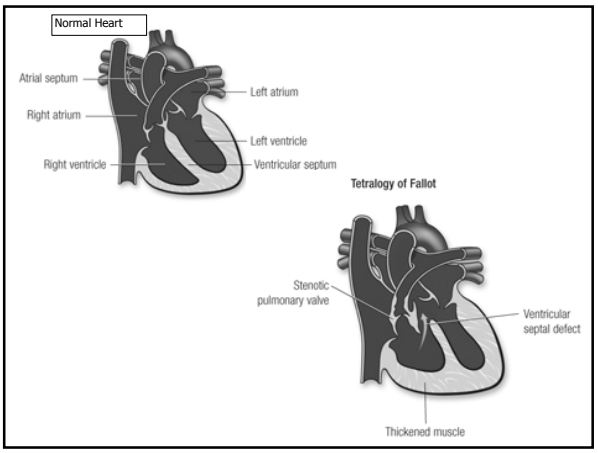
Tetralogy of Fallot

- Tetralogy of Fallot refers to a combination of cardiac abnormalities
  - A ventricular septal defect (a hole between the ventricles)
  - Obstruction of blood flow from the right ventricle to the lungs (either pulmonary stenosis or atresia)
  - The aorta lies directly over the ventricular septal defect
  - The right ventricle develops hypertrophy (thickened muscle)
- The cause isn't known but it is more common is children with Down's syndrome or DiGeorge Syndrome



Tetralogy of Fallot - Symptoms

- Most babies at birth are cyanotic and remain cyanotic unless the defect is repaired
- Very limited endurance because of the lack of oxygen perfusion
  - Low oxygen saturations are normal
- Can be treated with surgical repair, either a temporary shunt or complete repair
- Normal life expectancy if repaired
  - Surgery has risks and mortality itself
  - Still a higher risk of arrhythmias once repaired

Hypoplastic Heart Syndrome

- The heart's left side is underdeveloped
  - The aorta, aortic valve, left ventricle and mitral valve
  - Ductus arteriosus remaining patent is the only thing keeping oxygenated blood pumping to the body
- Cause isn't known
- Can be present with other abnormalities or isolated

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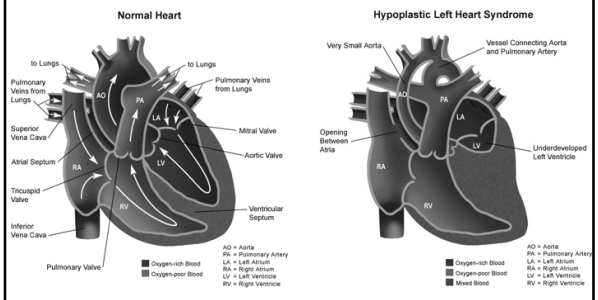

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Hypoplastic Heart Syndrome-Symptoms

- Baby appears normal at birth but after a couple days once the ductus closes and baby quickly decompensates
  - Become ashen
  - Have rapid and difficult breathing
  - Have difficulty feeding
- Ductus Arteriosus can be kept patent with medications
- This defect isn't able to be 'fixed' but surgeries can help, most of the time multiple surgeries in stages
  - Heart transplant is an option, but has its own risks
- Children are advised to limit activity as cardiac activity will never be normal

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Children with Tetralogy of Fallot exhibit bluish skin during episodes of crying or feeding.

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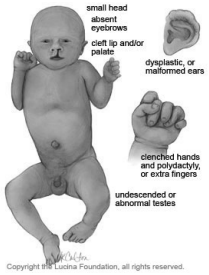
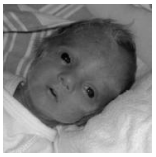
Trisomy 13 - Patau syndrome

- Trisomy means a baby is born with 3 copies instead of the normal 2 copies of a chromosome
- Down Syndrome is caused by Trisomy 21 and is the most common Trisomy
- Most trisomies result in an early miscarriage
- Trisomy 13 and 18, while they can result in a live birth, are 80-90% fatal within the first couple months of life
  - They are fatal because of the multiple congenital defects, including cardiac defects
- Can be recognized early by amniocentesis and at birth by the obvious abnormalities
- No treatment because of so many systems are involved

Trisomy 13



Trisomy 18



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Medications
Tylenol

- Uses
  - Pain
  - Fever
- Dosing for children <12
  - 10-15mg/kg q 4-6 hours
  - Not to exceed 5 doses in 24 hours
- Children >12
  - 325-650mg q 4-6 hours
  - 4000mg was/is maximum recommended dose in 24 hours, however FDA has recently (6/2009) advised decreasing this maximum 24hr dose but they have not given a new number
- Formulations available
  - Liquid 80mg/2.5mL or 160mg/1.6mL
  - Chewable tabs 80mg
  - Adult tabs

Medications Ibuprofen

- Uses
  - Pain
  - Fever
- Doses for children <12
  - 5-10mg/kg q 6-8 hrs
  - Max 40mg/kg/day
- Doses for children >12
  - Adult dosage
- Formulations available
  - Liquid 50mg/1.25ml or 100mg/5mL
  - Chewable tabs
  - Adult tabs

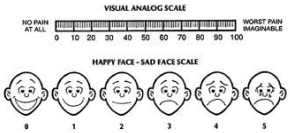
Medications Morphine

- Uses
  - Pain
  - Shortness of Breath
- Dosage for infants <6 months
  - 0.1mg/kg po q 3-4 hours
  - 0.05-0.2mg/kg IV/SQ/IM q 4 hours
- Dosage for children 6months-12 yrs
  - 0.2-0.5mg/kg po q 4-6 hours
  - 0.1-0.2mg/kg IV/SQ/IM q 2-4 hours
- Dosage for children >12 yrs
  - Adult dosage
- Formulations
  - Liquid
  - Tabs
  - Parenteral

Pain Scales

- Need to be geared towards the child's understanding
- Children past infancy will be able to point to where it hurts
- Children as young as 3 years old can use pain scales
- Observation scales have been developed for non verbal children or infants

Visual Scales



CRIES

DATE/TIME						
Crying - Characteristic cry of pain is high pitched. 0 – No cry or cry that is not high-pitched 1 - Cry high pitched but baby is easily consolable 2 - Cry high pitched but baby is inconsolable						
Requires O_2 for SaO_2 < 95% - Babies experiencing pain manifest decreased oxygenation. Consider other causes of hypoxemia, e.g., oversedation, atelectasis, pneumothorax) 0 – No oxygen required 1 – < 30% oxygen required 2 – > 30% oxygen required						
Increased vital signs (BP* and HR*) - Take BP last as this may awaken child making other assessments difficult **0** – Both HR and BP unchanged or less than baseline **1** – HR or BP increased but increase in < 20% of baseline **2** – HR or BP is increased > 20% over baseline.						
Expression - The facial expression most often associated with pain is a grimace. A grimace may be characterized by brow lowering, eyes squeezed shut, deepening naso-labial furrow, or open lips and mouth. **0** – No grimace present **1** – Grimace alone is present **2** – Grimace and non-cry vocalization grunt is present						
Sleepless - Scored based upon the infant's state during the hour preceding this recorded score. 0 – Child has been continuously asleep 1 – Child has awakened at frequent intervals **2** – Child has been awake constantly						
TOTAL SCORE						

Medications Valium

- Uses
  - Seizures
- Dosages
  - Weight and age based dosage
  - Can give times 1 PR and recommendation is not more than q 5days
- Formulations
  - Diastat-AcuDial system 10mg/20mg, delivers at 2.5mg increments
  - Custom suppositories (less expensive)
- Ativan can be used for seizures as well

Medications Miscellaneous

- Ativan
  - 0.05mg/kg q 4 hours, max 2mg/dose
- Scopolamine Patch
  - Frequently used for neurologically impaired children
- Oxygen
  - Start lower ¼- ½ liter and can deliver by "blow by"
- EMLA cream
  - Can apply to skin to numb prior to intervention (IV or SQ site)

A person's a person, no matter how small.

Dr. Seuss

Phases of Children's Comprehension of Death

- The Separation Phase
  - 0-3 years old
  - May not understand death as any different from temporary separation
  - Crying, separation anxiety and attachment to PCG
- The Structural Phase
  - 3-6 years old
  - Death is reversible and not permanent
  - Closely associated death with sleeping or going on a trip
  - Fear of sleeping and separation
  - Magical thinking, no thoughts that they could die

Phases is Children's Comprehension of Death

- The Functional Phase
  - 6-12 years old
  - Starting to realize finality of death
  - Later some realization that they can die but unlikely as it is old people that die
  - Recognizing external, but not internal causes of death
  - Fascination with specific details
  - Need to have control and as much information as possible
- The Abstract Phase
  - 12 years old and older
  - Adult understanding that death is final, universal and permanent
  - Realize that they can die as well and how this will affect the world around them
  - Anger about loss of a future and acting out

Need for Communication

- Children can feel isolated from the medical staff and caregivers
- When given the choice most children want to be a part of decision-making process
- Can use many ways of communication: verbal, art, or music
- Find out what they know and understand, realize they don't always need an adult understanding of death
- Avoid euphemisms because they can be confusing for children

Ethical Issues

- Potential conflicts in decision making exist
  - Parents and the child
  - Parents and the medical team
  - Mother and Father
- Goal is shared decision making
- Treatment should be in the best interest of the child
- The "reasonable parent standard" is similar to determining if an adult is able to make decisions
- While parental permission/consent is required, the child's assent should be obtained
- Problems arise when the child dissents

Legal Issues

- Emancipated minors
  - Pregnant or a parent
  - Married
  - In the military
  - Declared so by the court system
- Law enforcement gets involved when parents are clearly not acting in the best interest of the child
  - Cases of child abuse or neglect
  - Medical neglect (example is religious groups not providing basic treatment to children)
- Most organizations have Ethics Committees that deal with cases of conflict
  - Not actually a legally binding decision, just a suggestion

What is Available in our Community

- HPCC
  - Home Health and Pediatric Hospice
  - Carousel Center
  - Med Staff (available for Palliative Care Consults)
  - KBR
  - Grief Counseling Center/Camp Carousel
- Pediatric Community Alliance
- WFUBMC
  - Complex Case Management Team
  - Beds available on the Palliative Care Unit
- Heartstrings Infant Loss Support Group
- Maternal Fetal Medicine/Perinatal Group at Forsyth/Baptist
- 2 schools for Disabled Children
- Now I Lay Me Down to Sleep-pictures
- Victory Junction Gang Camp

Any Questions?