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

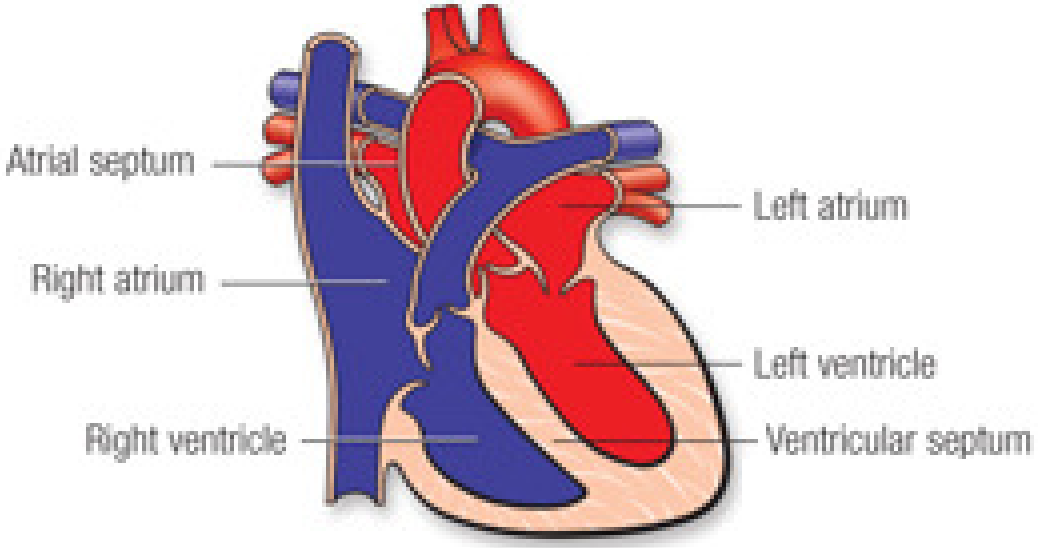
- Physical, Occupational and Speech Therapy
- For isolated spasticity- Botox injections
- For more generalized spasticity- muscle relaxers such as Valium and Baclofen
- Surgical interventions for patients with severe contractures


SPEAK

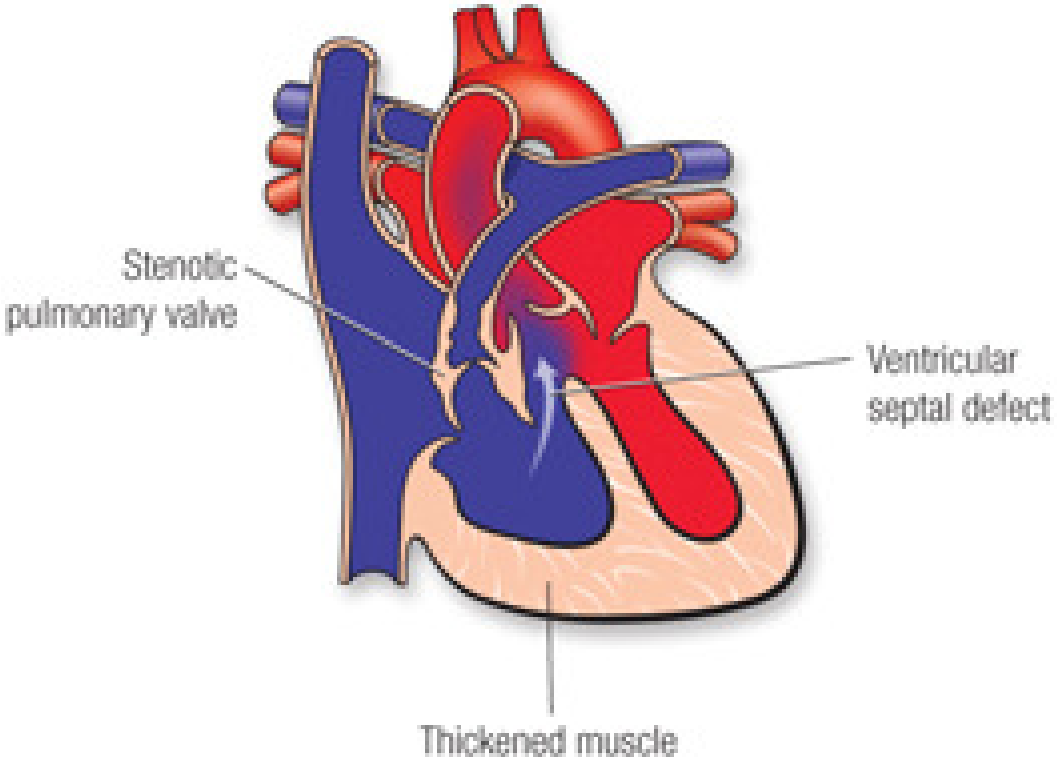
Tetralogy of Fallot

- 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

Normal Heart



Tetralogy of Fallot

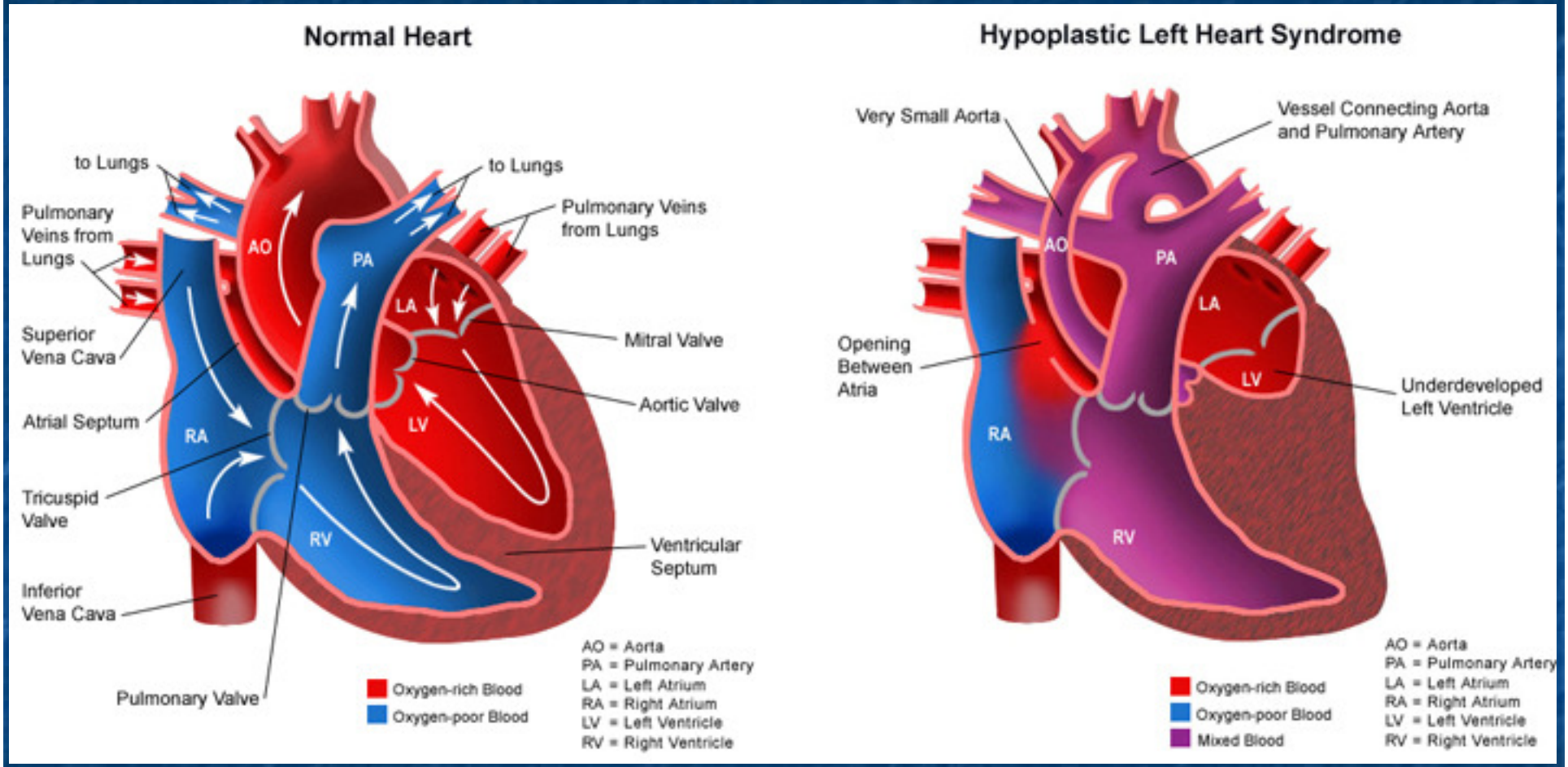


Tetralogy of Fallot - Symptoms

- 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


Normal Heart
to Lungs
to Lungs
Pulmonary Veins from Lungs
Pulmonary Veins from Lungs
AO
PA
LA
Superior Vena Cava
Mitral Valve
Atrial Septum
Aortic Valve
RA
LV
Tricuspid Valve
RV
Ventricular Septum
Inferior Vena Cava
Pulmonary Valve
Oxygen-rich Blood
Oxygen-poor Blood
AO = Aorta
PA = Pulmonary Artery
LA = Left Atrium
RA = Right Atrium
LV = Left Ventricle
RV = Right Ventricle
Hypoplastic Left Heart Syndrome
Very Small Aorta
Vessel Connecting Aorta and Pulmonary Artery
AO
PA
LA
Opening Between Atria
LV
Underdeveloped Left Ventricle
RA
RV
Oxygen-rich Blood
Oxygen-poor Blood
Mixed Blood
AO = Aorta
PA = Pulmonary Artery
LA = Left Atrium
RA = Right Atrium
LV = Left Ventricle
RV = Right Ventricle

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

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

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


small head
absent eyebrows
cleft lip and/or palate
dysplastic, or malformed ears
clenched hands and polydactyly, or extra fingers
undescended or abnormal testes
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Trisomy 18


occiput, or back part of the skull, is prominent
small mouth, small jaw, short neck
dysplastic, or malformed ears
shield chest, or short and prominent sternum; and wide-set nipples
clenched hands with overlapping fingers
flexed big toe; prominent heels


COMIX

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

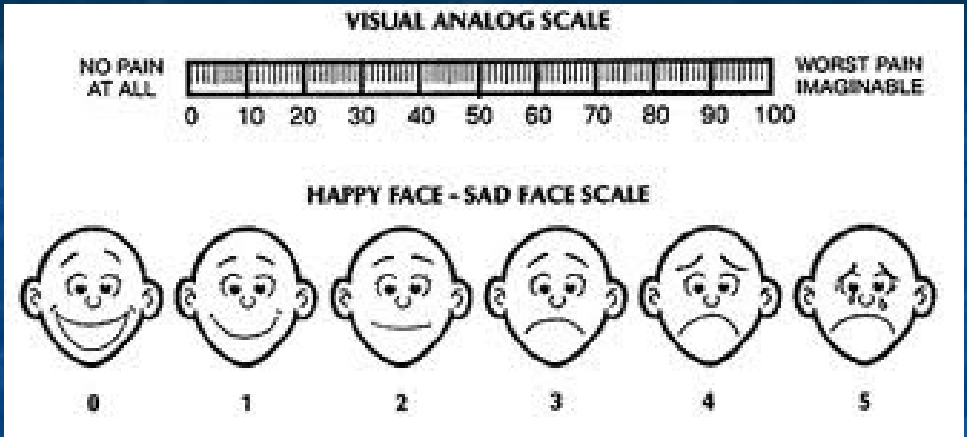
Medications
Morphine

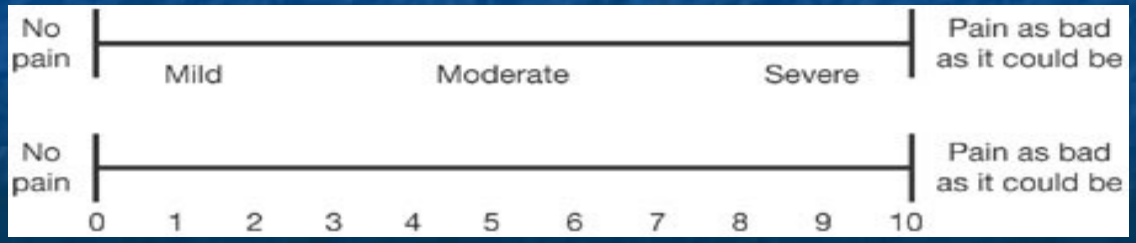
- Uses
  - Pain
  - Shortness of Breath
- Dosage for infants <6 months
  - 0.1mg/kg po q 3-4 hours
  - 0.05-0.2mg/kg IV/SQ/IM q 4 hours
- Dosage for children 6months-12 yrs
  - 0.2-0.5mg/kg po q 4-6 hours
  - 0.1-0.2mg/kg IV/SQ/IM q 2-4 hours
- Dosage for children >12 yrs
  - Adult dosage
- Formulations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





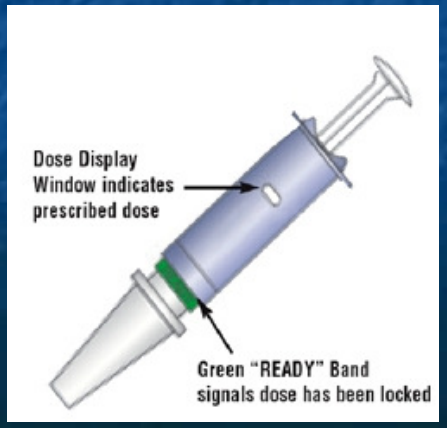
CRIES

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)

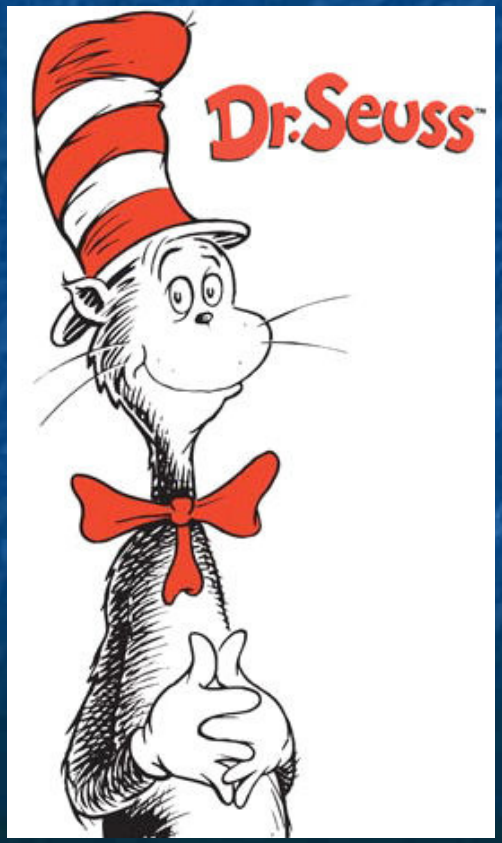
- Ativan can be used for seizures as well



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?