Pediatric Palliative Care: For Practitioners Who Take Care of Adults - Update

Dr. Kimberly Case
2/25/2011
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%)
<table>
<thead>
<tr>
<th>Causes of Death for Children (1-19 years)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Accidents</td>
</tr>
<tr>
<td>2. Assault</td>
</tr>
<tr>
<td>3. Malignancy</td>
</tr>
<tr>
<td>4. Suicide</td>
</tr>
<tr>
<td>5. Congenital malformations, deformations</td>
</tr>
<tr>
<td>6. Chromosomal anomalies</td>
</tr>
<tr>
<td>7. Heart disease</td>
</tr>
<tr>
<td>8. Cerebrovascular</td>
</tr>
</tbody>
</table>
# Cancer Death Rates* in Children 0-14 Years by Sex, US, 2001-2005

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

*Per 100,000, age-adjusted to the 2000 US standard population.

ONS = Other nervous system

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
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 in children with Down’s syndrome or DiGeorge Syndrome
Normal Heart

- Atrial septum
- Right atrium
- Left atrium
- Left ventricle
- Right ventricle
- Ventricular septum

Tetralogy of Fallot

- Stenotic pulmonary valve
- Ventricular septal defect
- Thickened muscle
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
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

- Small mouth, small jaw, short neck
- Shield chest, or short and prominent sternum; and wide-set nipples
- Clenched hands with overlapping fingers
- Flexed big toe; prominent heels
- Occiput, or back part of the skull, is prominent
- Dysplastic, or malformed ears

[Image: Illustration of abnormalities associated with Trisomy 18]
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 6 months-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

**Visual Analog Scale**

- **No Pain at All**
- **Worst Pain Imaginable**

**Happy Face - Sad Face Scale**

- 0: No Hurt
- 2: Hurts Little Bit
- 4: Hurts Little More
- 6: Hurts Even More
- 8: Hurts Whole Lot
- 10: Hurts Worst

- No pain
  - Mild
  - Moderate
  - Severe

- No pain
  - Pain as bad as it could be
<table>
<thead>
<tr>
<th></th>
<th>DATE/TIME</th>
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<tbody>
<tr>
<td><strong>Crying</strong> - Characteristic cry of pain is high pitched.</td>
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<tr>
<td>0 - No cry or cry that is not high-pitched</td>
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<tr>
<td>1 - Cry high pitched but baby is easily consolable</td>
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<tr>
<td>2 - Cry high pitched but baby is inconsolable</td>
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<tr>
<td><strong>Requires O₂ for SaO₂ &lt; 95%</strong> - Babies experiencing pain manifest decreased oxygenation. Consider other causes of hypoxemia, e.g., oversedation, atelectasis, pneumothorax)</td>
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<tr>
<td>0 - No oxygen required</td>
<td></td>
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<tr>
<td>1 - &lt; 30% oxygen required</td>
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<tr>
<td>2 - &gt; 30% oxygen required</td>
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<tr>
<td><em><em>Increased vital signs (BP</em> and HR</em>)** - Take BP last as this may awaken child making other assessments difficult</td>
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<tr>
<td>0 - Both HR and BP unchanged or less than baseline</td>
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<tr>
<td>1 - HR or BP increased but increase in &lt; 20% of baseline</td>
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<tr>
<td>2 - HR or BP is increased &gt; 20% over baseline.</td>
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<tr>
<td><strong>Expression</strong> - 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.</td>
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<tr>
<td>0 - No grimace present</td>
<td></td>
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<tr>
<td>1 - Grimace alone is present</td>
<td></td>
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<tr>
<td>2 - Grimace and non-cry vocalization grunt is present</td>
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<tr>
<td><strong>Sleepless</strong> - Scored based upon the infant’s state during the hour preceding this recorded score.</td>
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<tr>
<td>0 – Child has been continuously asleep</td>
<td></td>
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<tr>
<td>1 – Child has awakened at frequent intervals</td>
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</tr>
<tr>
<td>2 – Child has been awake constantly</td>
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<tr>
<td><strong>TOTAL SCORE</strong></td>
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</table>
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 $\frac{1}{4}$ - $\frac{1}{2}$ 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?

Your nose isn't wet, you sick?

...there, I fix that!