Session Objectives

- Compare adult and pediatric palliative care
- Identify barriers to provide inter-professional comprehensive pediatric palliative care for pediatric patients with advanced illness
- Discuss assessment and management of pain and non-pain symptoms in pediatric patients
- Review processes to be followed when a child dies including bereavement

Definitions

- Neonate: Birth to 28 days old
- Infant: Birth to 1 year old
- Child: 1-18 years old

Statistics

- 81 million children in the United States
- United States 2.5 million deaths annually
- About 50,000 deaths are pediatric 0-19 (2.2%)
- Children represent 25% US population
- Half of childhood deaths are in first year of life
- Half of infant deaths are in the first month of life

Symptoms in Dying Children

- 89% suffered “a lot” or “a great deal” from at least one symptom in their last month of life
- Most common reported symptoms:
  - Pain
  - Fatigue
  - Dyspnea

Causes of Deaths

All Infants

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

Wolfe, NEJM, 342:5; 2000

www.nhpco.org, Facts & Figures on Pediatric Palliative Care and Hospice
Causes of Deaths
Infants with Complex Chronic Conditions

1. Cardiovascular (32%)
2. Congenital / genetic (26%)
3. Respiratory (17%)
4. Neuromuscular (14%)

www.nhpco.org, Facts & Figures on Pediatric Palliative Care and Hospice

Causes of Death Children 1-19

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

www.nhpco.org, Facts & Figures on Pediatric Palliative Care and Hospice

Causes of Death
Children 1-19 with Complex Chronic Condition

1. Malignancy (43%)
2. Neuromuscular (23%)
3. Cardiovascular (17%)

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Pediatric Palliative Care Diagnosis

- **Conditions for which curative treatment is possible but may fail**
  - Advanced or progressive cancer or cancer with a poor prognosis
  - Complex and severe congenital or acquired heart disease

- **Conditions requiring intensive long-term treatment aimed at maintaining the quality of life**
  - Human immunodeficiency virus infection
  - Cystic fibrosis
  - Severe gastrointestinal disorders or malformations such as gastroschisis
  - Severe epidermolysis bullosa
  - Severe immunodeficiencies
  - Renal failure in cases in which dialysis, transplantation, or both are not available or indicated
  - Chronic or severe respiratory failure
  - Muscular dystrophy


- **Progressive conditions in which treatment is exclusively palliative after diagnosis**
  - Progressive metabolic disorders
  - Certain chromosomal abnormalities such as trisomy 13 or trisomy 18
  - Severe forms of osteogenesis imperfecta

- Conditions involving severe, nonprogressive disability, causing extreme vulnerability to health complications
  - Severe cerebral palsy with recurrent infection or difficult-to-control symptoms
  - Extreme prematurity
  - Severe neurologic sequelae of infectious disease
  - Hypoxic or anoxic brain injury
  - Holoprosencephaly or other severe brain malformations

Diagnoses in Pediatric Palliative Care

- Genetic/Congenital (40%)
- Neuromuscular (40%)
- Oncologic (20%)
- Respiratory (12%)
- Gastrointestinal (10%)
- Cardiovascular (8%)

Some children with multiple diagnoses

[Graph depicting survival function in the cohort of 515 patients who received pediatric palliative care consultation services and among patients with the 3 most prevalent conditions.]

Dichotomous Care Process

- Mutually Exclusive
  - Health Care Providers
    - Prognostic Uncertainty
    - Family resistance
    - Challenge of changing course of care
    - Conflict around goals of care
    - Avoidance of emotionally charged situations
    - Relinquishing care to another team
    - Hope
  - Family
    - Forced Choices
    - No best option
    - Abandonment
    - Hope

Illness Trajectories

Role of Palliative Care
Models of care
- Inpatient consultation palliative care teams
- Inpatient palliative care
- Home hospice
- Perinatal and neonatal hospice
- Concurrent care
- Respite

Palliative Care Team
- Nurses
- Dietary
- Respiratory care
- Chaplains
- Bereavement counselors
- Social workers
- Child life therapists
- Primary care physicians
- Subspecialty physicians
- Consultants.

Concurrent Care for Children Requirement
- The CCCR provision (Section 2302 of the ACA) states that
  - Children under the age of 21
  - Diagnosed with a life-limiting illness
  - Eligible for Medicaid or the Children’s Health Insurance Program
- May receive all services that are related to the treatment of a child’s life-limiting illness.
- This allows these young people to have palliative and hospice care services while they are receiving other disease-modifying treatments.

Infants and toddlers (age 0-3)
- Interactions initially limited to sensory and motor actions. Needs a sense of trust and hope in others. Developing feelings of self-worth and love.
- No concept of death.
- Limited concept of reality and may simply sense something is wrong.

Developmental Stages
- Infants and toddlers (age 0-3)
Infants and toddlers (age 0-3)
- Care focused on providing maximal physical relief of suffering.
- Benefits from simple physical communication.
- Benefits from physical touch, maintaining routines, and safe environment.

Preschool age (age 3-6)
- Egocentric. Uses magical and animistic thinking. Understanding centers on interweaving fact and fantasy.
- Spirituality is magical and imaginative. Participation in rituals becomes important.
- Death is a temporary separation and reversible. Illness perceived as the result of a “contagion” or from contact with an object or person and benefit from reassurance.

Preschool age (age 3-6)
- May feel responsible for illness and misinterpret emotions (e.g., think sadness is disappointment). Benefits from clarification of misconceptions and reassurance.
- Use precise language (avoid euphemisms). Evaluate for feelings of guilt and anger at self or others.
- Benefits from minimizing separation from family and maintaining consistency in daily routine. Utilizes play, puppets, dolls, expressive therapies and story telling in coping and learning.

School-age (age 7-12)
- Begin to differentiate between self and others. Concrete thoughts with beginning transition to more logical thinking.
- Start connecting ritual with personal identity and accepting external interpretation as truth.
- Understand that death is irreversible and personal. Interested in physiology and details of death.
- May feel responsible and need reassurance. Begin to demonstrate mature understanding of illness.

School-age (age 7-12)
- Should be invited to share their emotions when they are ready. May exhibit stoic responses in an attempt to protect their parents and caregivers.
- Should have some participation in decision making. Benefits from open, honest communication and concrete details of treatments.
- Benefits from maintaining access to peers and fostering a sense of control/mastery over the illness. Benefits from maintaining usual activities as much as possible.

Adolescents (age 13-17)
- Searches for meaning, purpose, hope, and value of life. Evolution of relationship with higher power.
- Explores nonphysical explanations of death.
- Very self-conscious of physical change. Struggle with need for independence and physical dependence due to illness.
Adolescents (age 13-17)

- Should be helped to communicate feelings and invited to share their thoughts and sadness when they are ready. May turn to a non-parental adult to share sadness.
- Key participant in decision making. Benefits from clear, direct, honest communication.
- Benefits from reinforcement of body image and fostering self-esteem by providing privacy and promoting independence. Benefits from access to peers and support groups and utilizing creative outlets. Is at risk for developing depression and risk-taking behaviors.

Advanced Care Planning

- The child should participate to the fullest extent possible, given his or her illness experience, developmental capacities, and level of consciousness.
- Regardless of the prognosis, respect for the child requires that he or she be given a developmentally appropriate description of the condition along with the expected burdens and benefits of available management options, while soliciting and listening to the child's preferences.
Neurological Impairment

- Developmental
  - Cerebral palsy
- CNS insult
  - Hypoxic ischemic encephalopathy (HIE)
  - Anoxic encephalopathy
  - Traumatic brain injury
- Specific diagnosis
  - Genetic disorder (muscular dystrophy, spinal muscular atrophy)
  - Congenital anomaly (holoprosencephaly)
  - Structural brain malformation (lissencephaly)
  - Metabolic disorder (mitochondrial disease, Tay-Sachs)

Identifying presence of pain

- Behaviors seen in validated pain assessment tools for nonverbal children with neurologic impairment
  - Vocalizations (crying, moaning)
  - Facial expression (grimacing, fussy)
  - Consolability
  - Interactivity (withdrawn, less active)
  - Movement (pulls legs up)
  - Tone and posture (arching, stiffening)
  - Physiological responses (sweating)


Opioid dosing basics

- Dosage initially based on weight
- Same escalation principles as in adults
- No upper dose limits
- Taste can be a limiting factor

Morphine

- Gold standard for moderate or severe pain
- Increased half-life and diminished clearance in neonates.
- Starting doses for infants about ½ of older children.
- Infants more sensitive to respiratory depression.

Codeine

- Use in mild pain only, limited use in severe pain
- Maximum recommended dose (60mg) produces analgesia equal to 600mg aspirin
- Combination product with acetaminophen
- Highly variable metabolism makes it unreliable
- Not a preferred agent
Hydrocodone
- Only available in combination with acetaminophen, aspirin or ibuprofen
- Schedule 3 controlled substance makes it easier to prescribe
- Preferred over codeine

Fentanyl
- Used in anesthesia, procedural sedation
- Acute moderate to severe pain
- Patch has found use in some cancer and chronic non-malignant pain
- Patches are high enough dose that can’t be used in opioid non-habituated children based on weight

Methadone
- Used in chronic pain
- Long half-life therefore longer time to steady state
- Should not be used for breakthrough pain
- Only liquid long-acting formulation

Sustained-release Opioids
- Children often cannot swallow pills
- Even lowest dosages of sustained release products may be too high for children
- High rates of gastrostomy tubes in pediatric palliative care population necessitate liquid formulations

Co-Analgesics
- Antidepressants - amitriptyline, nortriptyline
- Anticonvulsants - valproic acid, phenytoin
- Anxiolytics - lorazepam, diazepam, midazolam
- Corticosteroids – dexamethasone
- Anesthetics - lidocaine, ketamine, propofol
- Barbiturates - phenobarbitol, pentobarbitol

SUMMING IT UP
Pediatric Palliative Care vs. Adult

- Smaller numbers of dying children than adults mean that there is less professional expertise and underrepresentation of children in palliative care protocols.
- The heterogeneity of illnesses, many rare, requires the involvement of many disciplines and specialists.
- Many children have genetic diseases so that there may be more than one affected child in a family.

- The time course of some illnesses is extremely variable; pediatric palliative care may extend over years, even decades. Prognosis is very difficult.
- A broad developmental spectrum is represented, including changes in the individual child through time.
- Pediatric hospice care tends to be more expensive, and palliative and/or curative oriented therapies may happen in concert with active end of life care.

- Kids have specific developmental needs which are dependent upon age, but also impact of disease, developmental capacity, etc.
- Family centered care: family as the unit of care
- Emotional intensity: tends to be a concern for adult specific providers, and is realistic.

- Legal and ethical issues of non-autonomous individuals
- Artificial nutrition and hydration
- Informed consent vs. assent
- Declaration of brain death difficult
- Baby Doe law

Not talked about, but can discuss

http://www.chionline.org/