"We don't have to wait until we are very sick and know this might be 'it.' We can get support from the day we learn something could happen."

- Mattie Stepanek, February 2000
Session Objectives

• By the end of the session, participants should be able to:
  • Compare adult and pediatric palliative care
  • Identify common diagnoses found in pediatric palliative care
  • Explore ethical challenges unique to pediatrics
WHO Definition of Palliative Care for Children

• Palliative care for children is the active total care of the child's body, mind and spirit, and also involves giving support to the family.
• It begins when illness is diagnosed, and continues regardless of whether or not a child receives treatment directed at the disease.
• Health providers must evaluate and alleviate a child's physical, psychological, and social distress.
• Effective palliative care requires a broad multidisciplinary approach that includes the family and makes use of available community resources; it can be successfully implemented even if resources are limited.
• It can be provided in tertiary care facilities, in community health centres and even in children's homes.
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
- Children represent 25% US population

- United States 2.5 million deaths annually
- About 50,000 deaths are pediatric 0-19 (2.2%)

- 100% Adults die
- 0.06% Children die

- 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

Wolfe, NEJM, 342:5; 2000
Symptoms Children with Advanced Cancer

- Scores of high distress:
  - Pain (48%),
  - Fatigue (46%)
  - Drowsiness (39%)
  - Irritability (37%)

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

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

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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%)

www.nhpco.org, Facts & Figures on Pediatric Palliative Care and Hospice
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 epidermolysis bullosa
  • Chronic 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
  • 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
Survival function in the cohort of 515 patients who received pediatric palliative care consultation services and among patients with the 3 most prevalent conditions.

Role of Palliative Care

Primary Care  |  Palliative Care
---|---

Diagnosis  |  Death

Primary Care  |  Palliative Care

Comprehensive Care
San Antonio Outpatient Experience

- 359 enrolled children
- 4.3 visits per patient per year
- Death rate for clinic population is 4.3 deaths per 100 patients per year.
- 4.8% were enrolled in concurrent hospice at some point over the study period
Support

- Home Nursing: 59%
- VP Shunt: 12%
- Ventilation: 31%
- Tracheostomy: 25%
- Gastrostomy: 69%
Primary Diagnosis

- Congenital/Genetic: 38.2%
- Neuromuscular: 7.9%
- Cardiac: 36.8%
- Metabolic: 5.6%
- Gastrointestinal: 3.6%
- Malignant: 3.3%
- Heme/immuno: 2.3%
- Respiratory: 1.6%
- Renal: 0.3%
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.
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.
Indiana Baby Doe

• Born with Trisomy 21
• Tracheo-esophageal fistula
• Doctors gave conflicting medical opinions to parents

- 1982
Your Turn

• **Discuss:** What characteristics of the surgery or of baby’s situation lead you to believe that surgical intervention is or is not beneficial?

• How would you advise patient?
Real Life Decision

- Parents declined surgery
- Illinois courts upheld parent’s right to decide fate of their child
- Baby died on day 7 of life
Key Points

- While not risk-free, this surgery is a fairly standard procedure and likely to have a good outcome.

- Future quality of life of a child with Down syndrome, although uncertain, tends to be one that allows reasonable cognitive function, interaction with others, and contribution to society.
The Best Interest Standard

- Ultimately, caregivers must compare the burdens, consequences, and potential complications of the treatment itself against the burdens, consequences and potential complications of non-treatment; and they must compare the likely realities of life after treatment against the likely realities of life without treatment

President’s Council on Bioethics, 2005
Baby Jane Doe

• Born with spina bifida, microcephaly and hydrocephalus
• Doctors gave conflicting medical opinions to parents as to whether the myeloencephalocele should be repaired

Bowen vs. American Hospital Association, US Supreme Court, 1983
Your Turn

• **Discuss:** What characteristics of the surgery or of baby’s situation lead you to believe that surgical intervention is or is not beneficial?

• How would you advise patient?
Real Life Decision

- Parents declined surgery
- She survived, but neurologic status worsened by meningitis due, in part, to delayed treatment

Bowen vs. American Hospital Association, US Supreme Court, 1983
Lydia

- Holoprosencephaly
- Severe congenital brain malformation
- Cognitive impairment
- Quadriplegia
- Seizure disorder
- Pan-hypopituitarism
- Cleft lip and palate
What are our obligations?

- She is not nippling well.

- Should we place a gastrostomy?
Samuel

- 18 y/o male with osteosarcoma right distal femur.
- 12 y/o: limb salvage procedure of right knee with chemotherapy.
- 16 y/o: local recurrence, with amputation of right leg and another course of chemo.

- Parents have been very involved in decision-making and have sought care at nationally recognized cancer centers.

- Sammy is a quiet guy and rarely participates in discussions of his cancer.
• On a surveillance visit, studies show new metastatic lesions bilateral humerus and multiple pulmonary nodules.

• The parents discuss with the pediatric hematologist and request that Sammy not be told of metastases and poor prognosis.

• The nurses call and consult palliative care. Due to his age, it is directed to the adult team.

• What to do?
Julio

- Encephalocele
- Cleft Palate
- Rest of body perfect
- No surgical options for repair
What are our obligations?

- He has poor nippling.
- Should we place an NG tube?