Palliative Care in Pediatric Neurology

Canadian Neurological Sciences Federation Conference
Halifax NS
25 June 2018
Dr. C. Vadeboncoeur MD FRCPC
Founder, Palliative Medicine
Objectives

• What is pediatric palliative care
• What patients are common to PPC and neurology
• Therapeutic strategies
World Health Organization

• The goal of palliative care is the achievement of the best quality of life for patients and their families, consistent with their values, regardless of the location of the patient
Pediatric Palliative Care

Palliative care is an active and total approach to care, embracing physical, emotional, social and spiritual elements. It focuses on enhancement of quality of life for the child and support for the family and includes the management of distressing symptoms, provision of respite, and care following death and bereavement.

Joint Working Party of the Association for Children with Life Threatening or Terminal Conditions and their Families (ACT) and the Royal College of Paediatrics and Child Health (RCPCH)
Disease Trajectory

![Diagram of disease trajectory]

- **Child/Youth with illness** OR **Life**
- **Limiting conditions**
- **Family**
- **Caregivers**
- **ILLNESS TRAJECTORY** May extend over many years
- **Assessment Phase**
- **Palliation Phase**
- **Terminal Phase**
- **Bereavement Phase**

May be noted at birth or in antenatal period

To adult facility by age 18

Adapted from the Canadian Palliative Care Association.
Palliative Care in Children

• Support children from diagnosis (sometimes antenatally) until death and into the bereavement period.
Benefits of Palliative Care

• Communication
• Symptom management
• Increased length of survival with better quality of life
• Care coordination, continuity
• Health care resource utilization
Communication

• Adult and pediatric studies confirm that early involvement of PC leads to improved communication

• PPC approach is to live life for as long as possible for as well as possible (and provide support to family during the child’s life and following their death)
Setting Goals

• Acknowledgement of terminal illness is difficult
• Allows focus on tests and therapies directed at increasing comfort and quality of life, rather than maintaining life
• Some interventions indicated event though child is “palliative”
• Good medical care, doing as much as possible to meet agreed goals that serve to make the most of the life of the terminally ill child
Pain and Symptom Management

• Sometimes used as a way to introduce team to family
• Disparate diagnoses but similar issues (pain, secretions, sleep, constipation, feeding)
• Management throughout life, with specialized expertise at end of life
• Emphasis on support in addition to management of physical symptoms
Patients Common to PPC and Neurology

• Children with
  – Brain tumours
  – Neuromuscular diseases
    • SMA
    • Muscular dystrophy
  – Bad brains
    • Lissencephaly
    • Severe cerebral palsy
    • Intractible epilepsy
Problems to be Addressed

• Symptoms
  – (seizures)
  – Pain
  – Spasticity
  – Family support

• End of Life
Neuroirritability

Beyond normal infantile colic
Neuroirritability

• Hauer - children with severe neurological impairment (SNI) who have persistent or recurrent episodes with pain behaviors after assessment and management of potential nociceptive sources, suggesting the CNS as a source of persistent pain features
Pain Behaviours in Nonverbal Children with SNI

- Vocalizations - crying, whimpering
- Facial expression - grimace, frown
- Consolability – inability to be made comfortable
- Interaction – withdrawn, seeking comfort
- Sleep – disturbed, increased or decreased
Pain Behaviours in Nonverbal Children with SNI - continued

• Movement – increase in movement from baseline in arms and legs
• Tone – stiffening, resisting movement
• Physiologic – tachycardia, sweating, pallor
• Atypical features – laughter, self-injurious behaviours
Validated Pain Scales

• r-FLACC
  – 5 item pain assessment tool (score 0-10)
  – Includes pain behaviours specific to children with cognitive impairment
  – Parents can individualize with behaviours specific to their child

• Simplest to use, especially by people who are not familiar to the child
More Validated Pain Scales

• INRS – personalized for individual child
  – Individualized Numeric Rating Scale – numbers assigned by parents to correspond with judged level of pain

• NCCPV-PC – 27 item tool with validated cut off for moderate to severe pain (post operative version)
  – Non-communicating Children’s Pain Checklist

• NCCPV-R – above tool revised, 30 item tool, validated cut off for moderate to severe pain

• PPP – 20 item tool – ratings for “on a good day”, “most troublesome pain” etc.
  – Pediatric Pain Profile
Approach to Management

• History
• Physical exam including eyes, ears, shunt catheter site, gastrostomy tube, skin, extremities, range of motion of joints
• Judicious use of investigations according to findings on history and/or physical exam and known sources of pain in SNI
Empiric Management (off label use)

• Trial of gabapentinoids ie gabapentin or pregabalin

• Second line ie low dose tricyclic antidepressants (Amitriptyline, Nortriptyline), opioids, clonidine

• Third line ie methadone, canabanoids
End of Life Considerations

- Unpredictable prognosis
- Physical changes seen as end of life approaches
Physical Changes

- In the days/weeks prior to death:
  - Weight loss/gain
  - Respiratory changes
  - Increasing somnolence
  - Less interactivity (walking/speaking)
Physical Changes

• In the days/weeks prior to death:
  – Labored breathing (may be more disturbing to parents/visitors than to the child)
  – Secretions that cause a gurgling sound
  – The last breath
Questions/Scenarios to be prepared for . . .

• If my child is given opioids, can’t this hasten his/her death?
• Isn’t it cruel to stop providing at least a little nutrition or fluid?
• How will I know if he/she is suffering?
• Can he/she hear me?
• Can’t you just give her something to stop her suffering (hasten his/her death)?
Questions/Scenarios to be prepared for

- If only tissue is taken from my child after his/her death, is that considered an autopsy?
- My husband and I (or family members) can’t agree about the care directive. What should I do?
- If my child starts to get better (miracles happen) can we change the care directive?
How parents can provide comfort to a child who is dying

- Continue to bathe, change the position of his/her body, and watch for signs of discomfort
- Familiar voices, sounds, and touch that will comfort (read favorite books, psalms, listen to music, soothing smells)
- Complementary therapies such as massage and Reiki are often welcomed by families as hands on healing of the child.
Interventions to consider discontinuing...

- Monitoring including vital sign assessment, non-essential medications and blood work
- Comfort for the child actively dying  
  Examples include:
  - Constipation
  - Bedsores
  - Anemia
Communication and actions at child’s end of life

• Staying present, but be allowed to take care of oneself
• Giving permission to let go
• Reassuring the child that he/she is not alone
Questions/Comments
1. Parents' Experiences and Wishes at End of Life in Children with Spinal Muscular Atrophy Types I and II.  

2. Palliative care in neonatal neurology: robust support for infants, families and clinicians.  

3. Challenges and opportunities to advance pediatric neuro-oncology care in the developing world.  

4. Observational study of spinal muscular atrophy type I and implications for clinical trials.  

