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INDUSTRY DISCLOSURE

I have no relationships to disclosue

Outline

- 1. discuss how ~~when is it appropriate~~ to take a palliative approach in severe neo-natal brain injury

- 2. describe how to keep a palliative focus for adolescents with chronic and degenerative neurologic diseases, whilst still transitioning to an adult service model

- 3. describe how patients with delirium and their families can benefit from a palliative approach to care

- 4. identify how ~~when it is appropriate~~ to take a palliative approach in dementia

PALLIATIVE CARE

- Minimize discomfort
- Maximize comfort
- Provide support to family

Case 1: Jenna, with severe neonatal encephalopathy

- Jenna was born at 38 weeks by urgent c-section for fetal distress, after an unremarkable pregnancy
- APGAR scores: 1, 0, 0
- Required resuscitation for 25 minutes, including intubation and compressions
- Transferred to NICU, passive cooling as per protocol, antibiotics, anticonvulsants, full investigative work up
- Palliative care consulted on Day 4 of life after MRI confirmed extensive hemorrhage in cortical and subcortical structures, consistent with severe hypoxic ischemic injury

How can you provide care to Jenna and her family in your community?

Neonatal Encephalopathy

- Neonatal encephalopathy is a heterogeneous, clinically defined syndrome characterized by disturbed neurologic function in the earliest days of life in an infant born at or beyond 35 weeks of gestation, manifested by a subnormal level of consciousness or seizures, and often accompanied by difficulty with initiating and maintaining respiration and depression of tone and reflexes[1]
- Infants with severe encephalopathy have a 75 percent risk of dying in the neonatal period, and among survivors, an almost universal risk of sequelae exists
- Incidence 2 – 9 per 1000 term births
- Approximately 6 cases per year referred to Canuck Place Children's Hospice

Palliative Approach: Miminize discomfort

- Avoid unnecessary procedures, tests, monitoring
- This does not mean you should not provide routine, regular baby care
  - Family wants to see that you are still interested and concerned about the baby
- Discuss a plan for feeding
  - This is a complicated, emotional issue that warrants discussion early on
- Avoid taking baby away from family unnecessarily
  - A newborn is most comfortable with its parents

Palliative Approach: Maximize comfort

- If baby is uncomfortable, do something about it
- Model comforting behaviours for parents
- If babe is not settled with regular non-drug measures, then
- There is no need to withhold medications for pain or discomfort in an infant who is suffering

Morphine 0.3 mg/kg po/NG q 30 mins prn
Morphine 0.1 mg/kg sc / IV q 5 mins prn
Midazolam 0.1 mg/kg sc/IV/IN q 5 mins prn

- Your active management of distressing symptoms will make a significant difference in parent's experience

Palliative Approach: Provide support to family

- Use you team members
  - Social work, pastoral care, nurses
- Provide ample opportunity for parents and extended family to ask you questions
- Validate the family's choices
- Tell family what to expect
  - Breathing changes
  - Colour changes
  - Expected time line
- If able to discharge home, be prepared
  - DNAR, Emergency contact numbers
  - Discuss case with public health nurse

Canuck Place Children’s Hospice is a provincial resource

- Toll free number: 1.877.882.2288
- Other examples of neurologic diagnoses on our program:
  - Metabolic and Biochemical disorders manifested by progressive neurologic deterioration
  - Severe cerebral palsy with declining functional status
  - Severe epileptic encephalopathy, refractory to treatment
  - Malignant brain tumours
  - Neuromuscular diseases

Case 2: Peter, with Duchene Muscular Dystrophy

- You have rarely seen Peter, as he has been seeing his pediatric specialists

- He comes to see you because he was told he "should start seeing his family doctor now that he is 18."

- He says everything is fine and he just needs his referrals to his specialists and his medications refilled.

What kind of care can you offer Peter in your office?

Duchene Muscular Dystrophy

- X-linked mutation in the gene that produces “dystrophin,” a protein on cytoplasmic side of muscle cell membranes
- Found in skeletal muscle and cardiac muscle
- 1 / 3500 – 1/ 6000
- Progressive weakness of skeletal muscle, development of cardiomyopathy, orthopedic complications
- Life expectancy has changed over the past 40 years with the regular use of corticosteroids and non-invasive ventilation
- Life expectancy used to be into the mid teen years, and can now be in the the 30’s with case reports of survivors into their 40’s

Palliative Approach: Minimize discomfort

- Peter’s medications:
  - Deflazacort 36mg
  - Ramipril 5mg
  - Calcium + Vitamin D
  - Pamidronate q 12 weeks
- He is on bipap at night
- Peter has appointments every couple of weeks
  - Cardiology
    - ECG, echo
  - Neurology
  - Endocrinology
    - Lab work, bone density
  - Respirology
    - PFTs

Palliative Approach: Minimize discomfort

- Is there anything you can do to minimize his discomfort – does he need all these medications, appointments, test?
- The better question is does he *want* all of this?
- At this point in his disease trajectory, Peter probably does want all of these interventions
- It is important to discuss what Peter wants for himself as he starts to take more control of his health

Palliative Approach: Maximize comfort

- Ask about comfort, is Peter having pain or other symptoms on a day to day basis?
- People with neuromuscular disorders often have pain and discomfort that isn't treated
- Encourage Peter to be comfortable making decisions
  - promote his independence, encourage him to take on the responsibility of managing his health
  - form a collaborative relationship
- Discuss advance care planning early and often
  - Peter needs to be prepared for disease progression, and needs to be aware of choices he may need to make in the future
  - He needs to identify who can speak for him

Palliative Approach: Provide support to family

- Peter's family may need reassurance that Peter is able to make some of his own decisions now
- As a young adult with medical challenges, he still needs his family's assistance, but also needs to experience some independence
- Discuss advance care planning early and often
  - Peter's family needs to be prepared for disease progression
  - They need to identify who will act at substitute decision maker, and ideally, document this
  - Some advance care planning resources:
    - My Voice - http://www.health.gov.bc.ca/library/publications/year/2013/MyVoice-AdvanceCarePlanningGuide.pdf
    - Nidus - http://www.nidus.ca/
    - Tyze Network - http://tyze.com/

Case 3: Janice with delirium

- Janice is a 68 year old woman with advanced breast cancer brought to the hospital by her family for weakness and “strange behaviour”
- Her family reports that for the past 3 nights, Janice has been restless and seems to be talking to people that aren’t there
- She has been confined to bed for the past 24 hours, but previously was only getting to the bedside commode
- There are no other new symptoms, there have been no recent medication additions or deletions
- Lab results are all within normal limits and head CT is unremarkable

You determine that Janice is experiencing delirium and suspect she is approaching end of life. How do you manage this?

Delirium

- Delirium has been defined as a transient organic brain syndrome characterized by the acute onset of disordered attention and cognition, accompanied by disturbances of cognition, psychomotor behaviour and perception[2]
- Prevalence estimates very widely 25-85% for patients with advanced cancer
- 50% of delirium cases may be reversible[3]

- Suggested reference

http://www.fraserhealth.ca/media/07FHSymptomGuidelinesDelirium.pdf

Palliative Approach: Minimize discomfort

- Consider whether you need to do frequent blood work, vital signs, and more investigations
- Maintain a quiet calm environment, as much as is possible in a hospital setting
- Create a plan with family how to manage agitated behaviour
  - Can family stay with Janice overnight?
  - Are family willing to be called back to hospital?
  - Can family provide context to what Janice is talking about when she is agitated?
  - What can staff say that is reassuring to Janice?
- Discuss whether "re-orienting" statements are helpful or harmful to Janice
  - "Janice, you are in the hospital, not on your farm right now."
  - Or
  - "Janice, you are in a safe place, your husband is looking after the farm for you."

Palliative Approach: Maximize comfort

- If Janice is able to be redirected and reassured, then we don't need to medicate her
- If Janice is experiencing some pleasant memories, then we don't need to medicate her
- If Janice's experiences are frightening and upsetting for her, then medication to help calm and sedate her is necessary
- If Janice is at risk of hurting herself or staff, then medication is necessary
- Starting doses

Haloperidol 1-2 mg po/sc q 8 h regularly
Haloperidol 0.5 – 1mg po/sc q 30 mins prn

Methotrimeprazine 6.25 – 12.5 mg sc q 6h regularly
Methotrimeprazine 3.125 – 6.25 mg sc q 30 mins prn

Palliative Approach: Provide support to family

- Delirium is an extremely distressing symptom for family
- It is hard to see a loved one confused and disoriented
- It is hard to be with a loved one and not be able to interact with them properly, especially when time is limited
- Family may be able to cope better if they have an understanding of what is happening with their loved one

Explaining Delirium

Medical

- We have tests to determine kidney, liver and heart failure
- The main way we can can tell someone’s brain is failing is by their behaviour
- If all of the other organs are shutting down, then sometimes the brain starts to shut down as well
- Sometimes delirium can be a person’s defense mechanism to prevent them from experiencing discomfort, but it doesn’t always work

Holistic

- Dying is a process, a journey that a person must make
- Part of the process means detaching from loved ones, and sometimes they are trying to prepare us
- Many people review past experiences, sometimes in a dream like state
- Some of what a person says is metaphorical or symbolic
- Don’t take everything literally, but listen for recurrent themes

Case 4: Richard with advanced dementia

- Richard is 86 years old and has just been admitted to a residential care facility after a prolonged hospitalization for a hip fracture
- He was diagnosed with dementia 6 years ago
- Prior to be admitted to hospital, he was living with his wife, but they were having difficulty coping
- He has rheumatoid arthritis and his wife asks if there is anything you can give him for the pain other tylenol or ibuprofen, but she is scared to give him morphine, because after his hip surgery he "went crazy" with morphine.
- Richard can mobilize to the bathroom independently, but he is very unsteady
- He rarely eats regular meals, but will eat ice cream or cookies if his wife brings them

Advanced Dementia

Recommended references:

The Clinical Course of Advanced Dementia, Mitchell SL, Teno JM, Kiely DK, Shaffer ML, Jones RN, Prigerson HG, Volicer L, Givens JL, Hamel M, N Engl J Med 361:1529, October 15, 2009

This Changed My Practice UBC CPD: By Dr. Amanda Hill on August 23, 2010

http://thischangedmypractice.com/the-natural-history-of-severe-dementia/

Palliative Approach: Minimize discomfort

- Review Richard’s ability to swallow and take pills, and review all the medications he is on right now.
  - Are there any medications that can be discontinued?
  - Are his rheumatoid arthritis medications optimized right now?
- Is it necessary to send him for the x-rays and blood work that his rheumatologist recommended a year ago?
- Review transferring procedures and mobility aids with staff at the care home
  - Do they take Richard’s joint pain and stiffness into account?
- Are there any other factors contributing to Richard’s pain
  - Anxiety
  - Difficulty sleeping
  - Mood

Palliative Approach: Maximize comfort

- Find out what has helped with RA flare ups in the past
- Analgesics have side effects, but this doesn't mean Richard shouldn't have access to pain control
- Consider a short course of steroids
- Consider low dose opioids after discussion with family about circumstances of the use of morphine after surgery
- Suggestions

  Hydromorphone 0.5 – po q 1 h prn
  Oxycodone 2.5 po q 1 h prn

- In this situation, a low dose, long acting opioid may also be appropriate and cause less side effects. Try this in consultation with your local palliative team

Palliative Approach: Provide support to family

- Take time to listen to the caregiver’s concerns, they know the patient best
- If a caregiver doesn’t feel heard, their distress can escalate, as they feel the burden of being the advocate for their loved one
- Always inquire about what the patient would tell us if they could, and what the patient would want for themselves
- Family will often accept some level of side effect if the end result is seeing their loved one more comfortable

Palliative Care Everywhere

Minimize your patient's discomfort by avoiding unnecessary medications and investigations

Maximize your patient's comfort by actively treating their symptoms, including pain

Support your patient's family by helping them understand what is happening with their loved one

After today, hopefully you don’t think this is how we talk in palliative care

QUESTIONS AND
DISCUSSION

References

1. Neonatal encephalopathy and neurologic outcome, second edition. Report of the American College of Obstetricians and Gynecologists' Task Force on Neonatal Encephalopathy. Obstet Gynecol. 2014 Apr;123(4):896-901.
2. Oxford Textbook of Palliative Medicine. 3rd ed. Oxford, England: Oxford University Press; 2004, paperback 2005. p. 703-26
3. Lawlor PG, et al. 2000 Arch Intern Med 160: 786