MODULE: END-OF-LIFE DECISION-MAKING IN PEDIATRIC PALLIATIVE CARE

Detailed Case Scenario

Daniel is a $3^1/_2$-year-old who presents with a 5-week history of weight loss, anorexia, limping, and intermittent abdominal pain. He has always been a “picky” eater and had a preceding history of “failure to thrive”, having fallen off his growth curves.

Daniel lives about 1 hour’s drive away from the tertiary care center where he receives his care. He lives with his 6-month-old sister, Maria, and his parents, Keith and Teresa. Daniel loves books. He is able to describe his pain as an “owie” and will tell his parents when he is hurting. He had been sleeping poorly and was irritable. These symptoms resolved with regular doses of acetaminophen and codeine.

Investigation of an abdominal mass confirms neuroblastoma (a cancer originating in the neuro-endocrine cells above the adrenal gland, typically presenting in early childhood at an advanced stage). A metastatic work-up shows stage IV disease with involvement of parietal bones, pelvis, right hip, eighth rib, multiple vertebral bodies, and bone marrow. In children older than 1 year with advanced stage neuroblastoma, the average long-term survival rate is 15-25%. For those who survive to undergo bone marrow transplant, there is a 40-50% survival rate. Treatment is associated with significant short-term morbidity and can be associated with long-term morbidity.

Daniel receives treatment with chemotherapy, radiation, and surgery. This is followed by a bone marrow transplant (BMT) when he is 4 years old. He requires hospitalization for neutropenia-related infections, nausea and vomiting, and mucositis. He requires artificial nutritional support with naso-gastric feeds and hearing aids.

In the year following his BMT, Daniel enjoys going to the petting zoo, picking strawberries, bird-watching, doing crafts, and working on his computer.

At 5 years of age, has returns to the hospital on Christmas Eve with increasing hip pain. Work-up shows a recurrence of his cancer. At this point, Daniel’s parents decide to shift the goals of his care, knowing that cure is no longer attainable. They wish to pursue measures that are intended to help Daniel live as long as possible with a good quality of life.

Escalating pain is managed with opioids, focal radiotherapy, and multiple co-analgesics. With progression of his disease, Daniel has nausea controlled with a variety of medications. He continues to enjoy eating small amounts of his favourite foods.

Daniel’s mom, Teresa, modifies his stroller, enabling him to be quite mobile. It has a steering wheel and horn attached to it within Daniel’s reach, so he seems very much in control. He dislikes being in the hospital for any extended period of time although he visits happily enough.

With quality of life as their main goal, Daniel’s oncologist and parents discuss treatments that may be life prolonging but do not have significant toxicity. They also consider his previous treatment, his response to it, and his current tumor burden. One option is oral daily chemotherapy that does not involve swallowing large numbers of pills and does not generally cause severe neutropenia, so that hospitalization for febrile neutropenia would not be expected. Another option is to travel to another province for systemic radiotherapy. The radiopharmaceutical MIBG is used for scans but with higher doses can kill neuroblastoma cells. This option is better established outside of Daniel’s home province and would require a 9-day outpatient stay in the treatment region. The possibility of not pursuing either of these treatments is also included as an equally reasonable option.

Daniel’s parents opt for palliative focal radiotherapy and therapeutic MIBG. This treatment is generally well tolerated. However, in children like Daniel, who have already received extensive myelosuppressive treatments, protracted myelosuppression, particularly of platelets, may be an inadvertent consequence.

Daniel’s family embraces the trip for treatment as an opportunity to spend some time together. They incorporate fun, family time into the travel, visiting special places, swimming in the wave pool, and buying some special treats, like a cowboy hat for Daniel and a special gift for his sister, Maria.

Over the 6 months following MIBG treatment, Daniel enjoys doing crafts and watching the family cat have her kittens. He then develops shortness of breath due to a large pleural effusion, compounded by anemia. His parents and health care providers must decide whether to use thoracentesis to drain the effusion while implementing other measures to ensure Daniel’s comfort. This option is not pursued.

Daniel is transfused because his anemia could be contributing to the dyspnea. His sensation of breathlessness is relieved with systemic opioids and anxiolytics, and he is positioned to ensure maximal comfort. Daniel dies at home 1 week later.

Teaching Instructions for Opinion Leaders

1. Distribute the case scenario. Allow participants a couple of minutes to read the information or have one of the participants read the scenario.
2. Ask participants to identify the issues and questions about decision-making in pediatric palliative care that are raised by this case scenario. Write these up on a flip chart or overhead. These issues and questions may include (but are not limited to):
   - ☐ decision-making on behalf of young children when the options are limited
   - ☐ involving young children in decision making
   - ☐ the effect of remitting-relapsing illnesses on family and child functioning and decision-making
   - ☐ discussing new therapeutic options with limited supporting data

  - changing discussions and decisions when the goals of care change
  - involvement of sibling children

3. Participants may have personal or professional experiences of dealing with decision-making for and with children. Ask participants to reflect on these experiences and discuss what skills and methods are useful to health care professionals when faced with such situations.
4. Ask participants to discuss how they would have approached this case differently if Daniel had been 14 years old at the time of presentation.
5. Ask participants to write down, and then share with the group, the wording they would use to tell Daniel’s parents that there are no further treatment options that could be hoped to extend his life.
6. Ask participants to write down, and then share with the group, the wording or methods they would use to tell Daniel that there are no further treatment options that could be hoped to extend his life.
7. Daniel’s parents may have preferred to him not to undergo BMT during the early course of his illness. Given the survival statistics with and without BMT, this preference could well create disagreement between the medical staff and Daniel’s parents. Ask participants to discuss what their approach to this situation would be.
8. Review the issues and questions generated by the group and address any that have not yet been discussed.