

Pediatric Advanced End-of-Life Support

A series of workshops for professional development

Pediatric Palliative Care Team
CS Mott Children's Hospital
University of Michigan
Ann Arbor, MI

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The resources and videos used in this workshop can be found on line at:

open.michigan

<http://open.umich.edu/education/med/resources/palliative-care/2010/>

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Print this Document double-sided and then trim into individual case cards to distribute to Workshop participants.

PICU

Jason is a previously healthy 16 year old who was transferred to our ICU in septic shock 5 days ago. He has had two prolonged cardiac arrests with a severe ischemic injury to the brain and an encephalopathic EEG with seizures. He has concurrent acute lung injury and acute renal failure. Parents express the understanding that he will likely not survive this injury and if he did would certainly have a profoundly altered quality of life. Because of their strong religious views, they cannot bring themselves to agree with stopping any of the care he is getting, including the ventilator support, antibiotics or degree of blood pressure support he is on.

Questions:

1. What strategies for limiting/withdrawing care are available?
2. How do we help these parents through the decision-making?
3. In what ways may Jason or his family be suffering?

1.

PICU

Angela is a 2 year old girl with a metastatic neuroblastoma whose course was complicated by oliguric renal failure. She was started on CRRT while the oncologists re-evaluated. Her tumor is no longer responsive to any anti-neoplastic therapy, and the family understands that she has a terminal cancer. She has been requiring daily infusions of PRBC's and platelets. She is on methadone for pain, but has intact neurologic and pulmonary function. Grandma (primary caretaker) has offered to take Angela home to "be as normal as possible for as long as she has left." Grandma and the care team agree that goals of care will be re-directed to comfort.

Questions:

1. How does one approach comfort care planning with the family?
 2. What will happen if CRRT is stopped?
 3. What fluid/nutrition orders should be discussed?
 4. What is the role of blood products?
 5. Where is the best location for a child who is removed from support?
- 2.

PICU Cardiac ICU

Josh is a 5 year old boy with severe congestive heart failure. He is on the transplant list, but has developed fungal sepsis and multi-organ failure resulting in unlisting. He is only intermittently awake/responsive on fentanyl and midazolam infusions, but does not follow commands. He is additionally on broad spectrum antibiotics, ventilator support, and high dose inotropic drips. Five days ago he suffered an episode of prolonged hypotension, followed by a seizure. His EEG and MRI show evidence of anoxic brain injury and he is less responsive to stimuli, although he intermittently receives bolus sedation and neuromuscular blockade during unstable periods. The parents ask, "Is there any realistic chance now that he will be able to get his transplant and be at all normal? If not, we are ready to stop putting him through more of this."

Questions:

1. How can we best address the parents' question?
2. If the parents are ready to change the goals of care to comfort, what are our options for minimizing further suffering and their pros/cons?
3. What symptoms should be monitored as support is withdrawn?
4. Is "de-medicalizing" care important? Removal of tubes, holding him?

3.

NICU

Stephanie is a 38 week gestational age baby whose mother had no pre-natal care. Stephanie had anomalies and small size noted at birth and was intubated for ineffective respiratory effort and transferred to our NICU. Her exam suggested trisomy 13, confirmed by karyotype. Cardiac echo showed a large VSD. She is now 5 days old on minimal ventilator settings and oxygen, but shows no signs of being able to breathe on her own. She is minimally responsive on no sedatives, but does grimace weakly with painful stimuli. Parents are young and overwhelmed. The attending has recommended extubation and focus on comfort with the expectation of a fairly prompt death.

Questions:

1. How do we help these young parents?
 2. What options or strategies for end-of-life care planning are there?
 3. In what ways may Stephanie be suffering (or might at withdrawal)?
 4. Where might any withdrawal of support happen?
 5. What else needs to happen before withdrawal?
- 4.

Issues:

1. Pros/cons of setting (i.e., hospital, residential hospice, home hospice)
2. Stopping dialysis may result in an unpredictable course—acute hyperkalemia/arrhythmia, marked uremia, fluid overload, acidosis.
3. DNar/DNI order should be discussed, obtained if death from renal failure is expected.
4. Artificial fluids may have a role in alleviating thirst, but exacerbate fluid overload (pulmonary edema, secretions), especially in anuria.
5. Blood product choice involves benefits vs. burdens.
6. With a wide range of terminal manifestations, thorough anticipatory guidance should be provided to the family.

Words that might work:

Stopping some of these treatments doesn't mean that you or we are giving up, we are having a different focus. We will continue to care for her.

Our decision to stop this isn't what will cause her to die, it is her underlying disease.

Children who are this sick are not going to feel hungry and may not be thirsty. Forcing fluids into them will likely cause more discomfort.

She's not dying because she's not eating, she's not eating because she is dying.

2.

Issues:

1. Prognostic and diagnostic certainty may allow for more specific and confident recommendations.
2. Family/community support (spiritual rites, cultural issues, grandparents).
3. Location of withdrawal should consider parental supports/preferences.
4. Compassionate extubation and “de-medicalizing” care
5. Prepare to treat symptoms in the baby (e.g., dyspnea, secretions).

Words that might work:

We will do everything we can to make sure she is comfortable.

What can we do to help you remember your baby and to make your time left with her as special as possible for you?

I'm glad you both had a chance to hold her.

You've both been really good parents to her.

4.

Issues:

1. Non-escalation of therapy (e.g., higher pressors, starting dialysis)
2. Non-initiation of therapies (e.g., CPR, dialysis)
3. Withdrawal of care (and optimal sequence)
4. Ownership of actions: physician recommendations vs parental choices
5. Other family/community support (including religious leader)

Words that might work:

What might Jason want to say if he could talk?

What is most important to you that we stay focused on with Jason's care?

I know you are wishing and hoping for a miracle and recovery. If that does not turn out to be possible, what else are you hoping for?

There is no more I/we can do medically to help him recover. We can keep his heart beating with our current support. I plan to continue the level of care we are providing, but if his body can't keep going with all of this support, he has no chance of making it. We will know more in a few hours or days, as his body tells us whether or not it can be healed.

1.

Issues:

1. Prognosticating pediatric brain injury is often difficult.
2. Different parents define ‘normal’ very differently (i.e., be specific).
3. Decreasing inotropic support usually results in rapid loss of blood pressure and consciousness, providing non-pharmacologic sedation.
4. Sequence of withdrawal may affect the realization of goals. (e.g. If pressors are stopped first, there might not be much time for holding once he is extubated.)
5. Stop paralytics before discontinuing ventilatory support.

Words that might work:

What are you afraid of as we anticipate that he will die?

What would be most important to Josh?

What is important to you?

Who is important to be close by? In your family what else is important?

We will make sure he is comfortable.

3.

PICU Cardiac ICU NICU

Roscoe is 9 months old and has only been out of the hospital for 3 weeks of his life. His congenital heart disease was complicated by the development of progressive pulmonary hypertension that is no longer responding to medical or surgical therapy. He has had 6 trips to the operating room and now has a persistent chylothorax, and a poorly healing sternotomy wound. Parents are used to him being intubated, as he is currently. They are very used to life in the ICU. The team, including his cardiac surgeons, feel they are out of curative or palliative options that will enable Roscoe to leave the hospital alive. Even with maximum therapy for his pulmonary hypertension, a life-ending crisis is inevitable and could happen within days to weeks. Roscoe appears uncomfortable to the staff whenever awake, so he remains deeply sedated most of the time.

Questions:

1. When is it appropriate to initiate goals of care conversations?
 2. What decisions are likely to arise in the near and distant future?
 3. How do you respond to, "Oh, that Roscoe! He's been fooling you guys since day one. He's proven you wrong so many times. He's a fighter!"
 4. How might we recognize the suffering of the staff caring for Roscoe?
 5. If the family were to accept the inevitability of Roscoe's death, what type of decisions would be available?
- 5.

NICU

Deborah is a 26 week preemie, now 13 weeks old who has been receiving treatment for gram negative and fungal sepsis, grade 4 IVH, and persistent moderate ventilator requirement. At a team and family meeting, tracheostomy was declined by the parents, and a DNAR/DNI was agreed to. The baby was extubated, with a decision not to re-intubate, nor to escalate supportive care. She stabilized again over 6 hours. Now, with scheduled morphine and lorazepam, she has looked comfortable. She remains on OG tube feeds, diuretics and antibiotics. This morning however her secretions, tachypnea, and work of breathing are all increasing. She has no fever.

Questions:

1. What intended life prolonging therapies may cause discomfort?
 2. What happens when the child who was expected to die stays alive?
 3. How is parental trust affected when predictions do not come to be?
 4. When is a 'Menu of Care' appropriate (what we will and won't do)?
- 7.

PICU

Maria is a 14 year old girl with static encephalopathy, and the developmental age of an 18 month old. She was admitted for recurrent pneumonia, the fourth admission in a year. Before her current illness she was able to walk and play. With this admission, she has been intubated for 4 weeks and two extubation attempts have failed in the last 5 days. Both times she endured less than 12 hours, and failed due to hypercarbia, hypoxia, and a weak cough with poor oromotor secretion control. Her devoted mother requests 'focus on comfort' since Maria would "never be able to tolerate a tracheotomy."

Questions:

1. What will happen if Maria is extubated again?
 2. What strategies might be used in withdrawing ventilatory support?
 3. Should antibiotics be continued?
 4. What orders should accompany a DNI in this case?
 5. What does the other parent think? Other family members?
 6. What symptoms should we be ready to treat?
- 6.

PICU Cardiac ICU NICU

Eva is expected to die within hours to days from her incurable underlying disease. Since extubation 6 hours ago and discontinuation of tube feedings, she has been comfortable in appearance and seems to be in a peaceful sleep. She is getting 0.2 mg/kg of morphine every 3 hours, and 0.1 mg/kg of lorazepam every 3 hours p.r.n. agitation. In the last 15 minutes the pulse has increased by about 25% above the previous baseline. Her last dose of morphine was 2.5 hours ago, the last dose of lorazepam 1 hour ago.

Questions:

1. How does one best assess pain or dyspnea?
 2. What is the difference between pain, dyspnea and agitation?
 3. If this were assessed as increasing pain or dyspnea, what change in morphine and/or lorazepam administration would you try?
 4. How soon before you re-assess?
 5. What are the limits to opiate use?
- 8.

Issues:

1. Prognostic uncertainty in serial failed extubations.
2. Validate the mother's application of best judgment.
3. DNI/DNAR is appropriate in the face of expected respiratory failure.
4. Distinguish noxious/non-noxious therapies to continue or forego
5. Pros/cons of withdrawing ventilatory support quickly vs. gradually: compassionate/prompt extubation vs. terminal wean.
6. Anticipate air hunger and secretions in either scenario.

Words that might work:

We agree that a trache in a child like Maria will likely cause more discomfort and decreased quality of life. She would very likely never get used to being suctioned.

You are really tuned in to your daughter's needs and what she would consider a good quality of life.

We promise to do everything we can to keep her comfortable when we take out the tube. We will be watching carefully to see how she responds.

She may surprise us when we take out the tube. We should be ready for anything. We will let her guide us as to how best to make sure she is comfortable.

6.

Issues:

1. Use guidelines for assessing and treating dyspnea, pain and agitation (e.g., W.H.O. pain ladder)
2. Document both your dosing and your assessment of the reason you are increasing.
3. Comfort medication needs may escalate drastically at end of life.
4. Principle of double effect: we can accept a side effect (respiratory depression) if we are using a drug for the purpose of, and with doses intended to address a treatable symptom.

Words that might work:

We will continue to watch for signs of discomfort. Tell us if you are worried.

We are using doses of medicine that are enough to help.

She is dying from her underlying disease. We are fortunate to be able to help relieve her pain.

While it is possible that the medicine may slow her breathing a little, it is our desire to use just enough medicine to keep her comfortable.

When we treat pain and shortness of breath with these medicines, People who are older and can talk often wake up once their distress is relieved and say 'thank you, I feel better.'

Issues:

1. There are triggers and opportunities throughout the course of advanced disease for having ongoing goals of care and end-of-life conversations.
2. It is important to recognize signs of suffering when parents and staff are 'desensitized'. Morally distressed staff can feed into parental suffering.
3. Parental understanding of illness may underappreciate medical reality
4. Pulmonary hypertension is one of the diagnoses that may not show visible signs of lethality.
5. When death appears inevitable, it is appropriate for medical teams to suggest/recommend/ask about DNR status.

Words that might work:

With severe pulmonary hypertension, CPR will not be able to bring him back if his heart stops.

He sure has been an amazing fighter. In our experience, some children, even babies, may need a loving parent to say, "you can stop fighting....we will be ok"

Sometimes we have to reconsider what we are fighting for. Do you see anything different about the way he is reacting to the things we are doing to him?

When a child is close to death, I have a harder time accepting that any suffering is OK.

5.

Issues:

1. Set expectations, but allow for uncertainty when possible
2. Readdressing goals of care as the situation develops.
3. Distinguish between life prolonging care vs. death-prolonging care
4. Benefits vs. burdens of any therapy: feeds, IVF, antibiotics, etc
5. Diuretics can be palliative medicines

Words that might work:

Now that we all agree that our goal is to continue only the things that will keep her comfortable, we may have some other decisions to make about how best to do that.

How does she seem to be handling her feedings?

What is important to you now?

What are you most worried about?

Many loving parents in this situation would like to spend whatever time is left being able to treat her like a baby, and not like a patient. If any of the tubes seem to bother her, they don't have to stay.

We have medicines that can be given in other ways (skin, nasal spray, drops under the tongue)

7.

PICU

Jacob is a 4 year old boy from a very religious family who 2 days ago suffered a prolonged warm water near drowning and very prolonged resuscitation. He is on high vent settings, paralytics, as well as continuous infusions of morphine and midazolam. His brain imaging shows neurologically devastating ischemic damage and evidence of intracranial hypertension. The religious leader of the community has arrived and counseled the parents that stopping life support would be acceptable.

Questions:

1. How does being “very religious” affect parental decision-making?
2. What role should the religious leader play?
3. In what order should therapies be withdrawn?

9.

PICU

Hector is a 12 year old previously healthy child who presented with a brain abscess and meningoenitis secondary to sinusitis. Despite aggressive care, including debridement, decompressive craniectomies, external ventricular drains, and broad spectrum antibiotics, his ICP has been difficult to control and his neurologic status has deteriorated over the last 3 weeks due to edema and hemorrhage.

This morning, he stopped breathing over the ventilator, became bradycardic, and was noted to have anisocoria. ICP therapies were escalated. En route to the CT scanner, his EVD subsequently returned very sanguinous drainage, and the CT confirmed a new large intracranial hemorrhage. After returning to the PICU and consulting with neurosurgery, Hector’s pupils remained fixed and dilated and he was unresponsive. The family is updated and believes that their son is “gone,” and want to end his suffering now.

Questions

1. How does one assess for brain death?
2. Can a family request stopping therapy before brain death is ascertained?
3. What are the options for withdrawing care?
4. What feelings might the parents be experiencing?

11.

PICU

Sarah is a 19 year old young woman with cystic fibrosis complicated by end-stage lung disease, home oxygen dependence, and bipap. Prior to this hospitalization, she indicated in advanced directives that she wanted initial attempts at aggressive life support, including intubation and CPR. She had conveyed to her mother (DPOAHC) that if her course was deemed irreversible, she wanted to be made comfortable and allowed to die.

Sarah presented to the ER 2 weeks prior with multi-drug resistant polymicrobial pneumonia, was intubated and admitted to the ICU where she developed septic shock, severe ARDS, and pulmonary hemorrhage. She has recovered hemodynamically and is off pressors, but remains on high ventilator settings and an FiO2 of >60% with saturations in the mid 80’s.

On appropriate sedatives, her mental status waxes and wanes, she intermittently follows commands but is frequently confused and isn’t able to maintain focus for conversations. The ICU team and pulmonologists believe her recent illness has irreversibly damaged Sarah’s lungs and that she will not likely ever be free of the ventilator.

The mother is struggling with the decision to “quit” on her daughter, even though she knows and believes that Sarah wouldn’t want to go on like this. She is afraid of extubating Sarah, because she does not want her last memory of her daughter to be watching her struggle to breathe.

Questions:

1. How can Sarah’s own wishes and needs be respected?
2. Who is the ultimate decision maker?
3. What are the dilemmas inherent to parents as surrogate decision-makers?
4. What are the options for withdrawal?

10.

PICU

William is a 14 year old boy with cerebral palsy, mild developmental delay, and seizures. His parents found him unresponsive in the morning, dusky in color, breathing faintly, with no palpable pulse. They called 911 and paramedics intubated him in the field, after which his PEA resolved. He was admitted to the PICU and stabilized from a cardiopulmonary perspective, but was found to have severe hypoxic ischemic brain injury believed secondary to status epilepticus. On day 5 of his hospital course, his routine bedside clinical exam is suggestive of brain death (not breathing over ventilator, pupils fixed and dilated, unresponsive). The family indicates that they are ready to withdraw life support, but would also like to donate their son’s organs.

Questions:

1. What are the requirements for declaring brain death?
2. Does a family’s insistence on continuing life-support in a brain dead child have to be respected?
3. Who needs to be involved in the discussion of organ donation?
4. What are the ethical and legal issues that should be considered?

12.

Issues:

1. Pros/cons to different withdrawal strategies:
Terminal ventilator wean (gradual titration of comfort medications)
vs. prompt extubation (ready to control acute symptoms)
vs stopping antibiotics, fluids (allow disease to fulminate)
2. Burden of decision-making on parents.
3. Establish the legality of decision-making (varies by state law).

Words that may work:

We will do our very best to make sure that Sarah is comfortable.

If we slowly decrease the ventilator support, she will fall into a deeper coma.

You have been a great mother and support for Sarah for all of these years.

How can we help you?

We are all fortunate in that Sarah has given us the gift of letting us know her wishes. The damage to her lungs is too severe for her to survive and it is clear that she wanted to be allowed to die if this was the case.

It is most likely that Sarah knows that you are here and that she can hear you. Please feel free to remind her how much you love her, let her know that you will miss her and remember her always, and tell her that it is OK for her to go.

10.

Issues:

1. The pediatric brain death criteria of 1987 were recently updated in 2011, and aspects remain controversial and non-uniform. State law and hospital policy vary somewhat in the application of the criteria.
2. Brain death is legally dead. Comfort medications are not considered indicated, as brain death requires no demonstrable responsiveness to the environment (i.e. brain function is essential for somatic suffering).
3. Brain death usually affords the most options for organ donation.
4. Medical staff need to know the process of organ donation, including what happens to the body after organ and tissues are harvested.

Words that might work:

I am/we are so sorry. I / we wish things had gone differently.

It is a very gracious gift that you and William are offering.

What can we do to help your family?

What is important to you and your family at this time?

12.

Issues:

1. Religiosity is highly individual in application (miracle seeking versus acceptance of tragedy)
2. Stopping (or reversing) paralytics / allowing them to wear off is necessary to allow assessment of comfort (prevent distress)
AND to avoid the appearance of euthanasia.
3. Prompt extubation vs. slower weaning of support will depend on the patient responsiveness (e.g., deeply comatose patients do not particularly benefit from terminal weans).
4. Discontinuation of monitors as support is being withdrawn can help family focus on the child, not the monitors.

Words that might work:

His inability to respond or move indicates a deep level of coma. It is unlikely that he is in pain.

I would talk to him and touch him. At some level he knows you are here with him.

When we remove the breathing tube, I'm not sure if he will take some breaths. We will be ready to give him medicine so that he does not feel uncomfortable.

9.

Issues

1. Withdrawal is ok in this circumstance even without brain death.
2. Discontinuing monitors, ICP control meds, followed by empiric comfort medications and prompt compassionate extubation would be very reasonable.
3. Be ready to treat patient distress
4. Be wary of attempting medications to control unpreventable terminal manifestations (e.g., obstructed breathing, death rattle, etc), for which anticipatory guidance is often more helpful.
5. Parents may second-guess their own behaviors/actions and/or those of others in response to the sinusitis that led to the abscess.

Words that may work:

We are/ I am so sorry that we could not save him.

What is important for you, for your family?

Would you want to hold him, lie with him when we take out the breathing tube?

He might gasp or have some noisy breathing.....I will be right here to make sure we treat any discomfort.

Together we have given Hector every possible chance to survive, but it is not possible for him to overcome this latest bleed in his brain.

11

Issues

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13.

Cardiac ICU

Hector is a 12 year old with palliated heart disease who presented with a brain abscess and meningoenephalitis secondary his persistent right to left shunt. Despite aggressive care, including debridement, decompressive craniectomies, external ventricular drains, and broad spectrum antibiotics, his ICP has been difficult to control and his neurologic status has deteriorated over the last 3 weeks due to edema and hemorrhage.

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Questions

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2. Can a family request stopping therapy before brain death is ascertained?
3. What are the options for withdrawing care?
4. What feelings might the parents be experiencing?

13.