



Case 2: Adolescent with end-stage cystic fibrosis

Background: Erica is a 17 year old high school senior with advanced, end-stage cystic fibrosis. During the early years of her life compliance with therapy had been episodic, complicated by multiple family moves, divorce of parents, and care at another CF center. In the last 3 years that she has been a patient here she has developed progressive cor pulmonale, and is felt to be very high risk for heart-lung transplant.

Between her twice yearly hospitalizations for exacerbations of her CF, she has been active. She has many friends at school, and has always told her mother and brother that she is ready for a short life as long as it can be ‘as normal as possible’. She is admitted this week with an aggressive lung infection which despite broad spectrum antibiotics and aggressive non-invasive support, is not responding. A trial of bi-pap was ‘very uncomfortable’. Erica told her mother, “I am not afraid to die.”

At a family meeting yesterday, the team of specialists spoke with Erica, her 38 year old mother, and her 21 year old brother. The decision was made to enter an “Allow Natural Death (do not intubate, do not resuscitate) Order.” Erica spoke with her brother and several of her friends earlier today. She has directed her brother as to which of her belongings will go to which of her friends and relatives. Hospice and home hospice were offered, but Erica prefers to remain in the hospital, acknowledging that she will not survive for long.

This evening she has been more short of breath than earlier. She is on 4 liters of nasal cannula oxygen with a sat of 85%. RR is 40, Pulse 120, BP 110/65. She will intermittently cough up some green sputum.

You are on call this evening, with the understanding that “care is to be directed at comfort---she may not survive till the morning”

	Questions	What are we worried about?	What do we say?	What do we do?
I Anticipating the end	“Am I going to make it till the morning?”			

Author(s): CS Mott Children's Hospital Pediatric Palliative Care Team, 2010-2011.

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2. Pain, discomfort, dyspnea “What can you give her that won’t make her stop breathing?”

3. I’s & O’s of dying (unlikely in this case)

What are we worried about?

What do we say?

What do we do?

4. Seizures, agitation, myoclonus (not likely for this case) “Why is she so agitated? Can you help her?”

5. Before the last breaths “What should we be ready for?”

6. After the death “Now what?”

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