

## *Adolescent with end-stage cystic fibrosis - Suggested issues and responses*

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 (dad is completely out of the picture), and care at another CF center. In the last 3 years she has been a patient here but has 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.

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”

|   | <b>Patient issues</b>   | <b>Family issues</b>  | <b>MD issues</b>  | <b>Nurse/RT/etc. issues</b>  |
|---|---|---|---|--|
| <b>I Anticipating the end</b>               | Will I be in pain?<br>What will it feel like to die a respiratory death?<br>Will I be alone?<br>Who will be with me?<br>Will I be awake?<br>What happens when I die?  | Was this my fault?<br>Can I stay? Should I stay?<br>How do I deal with my guilt?<br>What do I/we do next?<br>Can we really do what she wants us to do?<br>When she gets close to the end, will we be able to cope?<br>How will we live without her SSI?<br>Can we afford a funeral? | Can I control her symptoms without contributing to an earlier death?<br>What do I do for dyspnea?<br>Are there limits to drug use?<br>Can I hold myself together emotionally?<br>How often do I come in and check on her? | Does the doctor know what to do?<br>Are we going to stop the fluids and the monitors?<br>What is my role in support?<br>Will the palliative care team be here or near? |
| Useful questions and phrases for the family | <i>The goal is to make sure you are comfortable. What is most important for you tonight? What are you most nervous or worried about? What else might help? Who do you want to be with you? Social Work / Spiritual care can be helpful in addressing some of your concerns.</i> |   |   |  |

|  | <b>Patient issues</b>  | <b>Family issues</b>                                | <b>MD issues</b>   | <b>Nurse/RT/etc. issues</b>  |
|--|--|---|--|--|
| <b>2. Pain, discomfort, dyspnea</b>  | Should I tolerate discomfort for the sake of my family?<br>Will the medicines make me so sleepy that I won't be able to talk with my family?   | Will the medicines make her stop breathing?         | For dyspnea:<br>Morphine 0.05 to 0.1mg per kg<br>or<br>Hydromorphone 0.01 mg/kg<br><br>Morphine or hydromorphone PCA<br><br>Inhaled opiates: there is anecdotal, but no controlled randomized studies that support their use, but a trial may be useful. | Nasal cannula may make talking easier than the mask.<br>Turn of the monitors.<br>Positioning options for increased comfort.<br>Fan to relieve dyspnea. |
| Useful questions and phrases for the patient and the family  | <i>We want to focus on you (your daughter, your sister) and not on the monitor. If more oxygen makes you feel better, we will turn it up. We will use medicine in doses that will lessen that feeling that it is hard to breathe. At large doses, these medicines can slow down someone's breathing. We do not anticipate using high doses, but if we do our intention is to use just enough to relieve discomfort.. Remember our goal now is to keep her comfortable.</i> |   |  |  |
|  | <b>Patient issues</b>  | <b>Family issues</b>                                | <b>MD issues</b>   | <b>Nurse/RT/etc. issues</b>  |
| <b>3. I's &amp; O's of dying</b><br><br><b>May include:</b><br><br><b>Secretions, noisy breathing</b><br><br><b>Nausea, vomiting (not likely)</b><br><b>Bleeding (not likely here)</b> | Will it hurt me to eat or drink?   | Noisy, raspy breathing is distressing to listen to. | Turn of IV fluids and tube feeding if secretions are a problem.<br><br>Robinul (glycopyrrolate)<br>Atropine<br>Scopolamine patch (may take hours to have an effect. Sedating)  |  |
| Useful questions and phrases for the family  | <i>Remember that our main objective is your comfort. If you feel hungry and want to eat or drink something, you can certainly do so. We would recommend something light and easy to digest—perhaps starting with apple juice or ice chips. If that goes well we can see about trying other liquids or even solids.</i>   |   |  |  |

|   | <b>Patient issues</b>  | <b>Family issues</b>   | <b>MD issues</b>  | <b>Nurse/RT/etc. issues</b>  |
|---|--|--|---|--|
| <b>4. Seizures, agitation, myoclonus</b>    | I'm scared, I'm nervous.   | I can't stand to see her so scared!  | Treat pain and dypnea more aggressively. Benzodiazepines at small doses is needed (lorazepam 0.01mg/kg)<br>Myoclonus may be from high d           | Low lighting, comfortable chairs for the family. "De-Medicalize" as much as possible.  |
| Useful questions and phrases for the family | <i>Don't be afraid to reminisce about some of your best times together.<br/>Is there anything in particular that is important for you to say to each other right now?</i>  |  |   |  |
|   | <b>Patient issues</b>  | <b>Family issues</b>   | <b>MD issues</b>  | <b>Nurse/RT/etc. issues</b>  |
| <b>5. Last breaths</b>                      | How is my family going to do after I'm gone?   | Am I going to be able to hold it together?<br>What will it look like?<br>Will she be scared? | Time to death is unpredictable. Breathing will become irregular. The 'last gasp' is often not the last breath.<br><br>What should I be ready for? | Will I be able to control my emotions?<br>What should I say?<br>Am I going to give her that last dose of morphine that 'tips her over the edge'? |
| Useful questions and phrases for the family | <i>Don't be afraid to tell her that you are going to miss her, that you are going to be OK, and that it is OK for her to let go.<br/>One thing that you can let her know: You promise that someone will be with her, that you won't leave her alone.<br/>Let us know how much or how little you would like me to be present in the room with you. Different families have different needs and I would like to meet your expectations to the best of my ability.<br/>We may know the end is near when her breathing becomes irregular and has pauses and some gasps followed by pauses. It won't surprise us if you hear a large breath or gasp followed by a pause that you think is her final breath....and then she takes another. Let us know if she looks uncomfortable or scared.</i> |  |   |  |
|   | <b>Patient issues</b>  | <b>Family issues</b>   | <b>MD issues</b>  | <b>Nurse/RT/etc. issues</b>  |
| <b>6. After the death</b>                   |  | What do we do now?   | Pronouncing death:<br>Advise family that you will be listening  | Time for family to be with the body.<br>What tubes or IV's can and can't be removed?   |

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|   |  |  | <p>for 2 minutes. Check heart, pulses, breathing, at least one reflex.</p> <p>Note the time of death as the time of your exam, not the report of the nurse or family.</p> <p>Paperwork, death note, death forms.<br/>Gift of life, autopsy consent or decline.</p> | Are there religious or cultural norms? |
| Useful questions and phrases for the family | <p><i>Erica has stopped breathing and her heart is no longer beating. She is gone...I am going to record the time of death as ____.</i></p> <p><i>I am so sorry for your loss.</i></p> <p><i>You can spend as much or as little time as you would like with Erica's body.</i></p> <p><i>Would you like to take part in preparing her body?</i></p> <p><i>I need to ask you again: have you thought whether having her body examined in an autopsy would be of some value?</i></p> <p><i>When you and your family decide it is appropriate, her body will be removed to a private part of the hospital where the funeral home can pick her up. Other family members could visit her body there.</i></p> |  |  |  |

### Learning Objectives

|                                   | <b>Trigger Question(s)</b>                                  | <b>Learning objectives</b>  |
|-----------------------------------|---|---|
| 1 Anticipating the end            | "Am I going to make it till the morning?"                   | <ol style="list-style-type: none"> <li>1. Identify the sources of distress in the patient, the family and the support staff</li> <li>2. Practice using language that can comfort and reassure patients and families.</li> <li>3. Recognize the need for autonomy in the ill and dying adolescent</li> </ol> |
| 2. Pain, discomfort, dyspnea      | "What can you give her that won't make her stop breathing?" | <ol style="list-style-type: none"> <li>4. Assess dyspnea and pain</li> <li>5. Know doses and routes of at least one opioid</li> <li>6. Learn phrases that can reassure the family that using an opioid is safe and appropriate.</li> </ol>  |
| 3. I's & O's of dying             |   |   |
| 4. Seizures, agitation, myoclonus | "Why is she so agitated? Can you help her?"                 | <ol style="list-style-type: none"> <li>7. Distinguish delirium from agitation near end of life</li> <li>8. Identify sources of agitation in a dying patient</li> <li>9. Describe at least one pharmacologic and one non-pharmacologic method to treat agitation near the end of life.</li> </ol>            |
| 5. Before the last breaths        | "What should we be ready for?"                              | <ol style="list-style-type: none"> <li>10. Explain the process for approaching patients and families regarding autopsy and gift of life.</li> </ol>   |
| 6. After the death                | "Now what?"   | <ol style="list-style-type: none"> <li>11. Review the 'check list' of post-mortem care items.</li> <li>12. Learn phrases that may comfort family members.</li> </ol>  |

