CE Symptom Management Series
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Communication, Comfort, and Closure for the Patient With Cystic Fibrosis at the End of Life

The Role of the Bedside Nurse

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Cystic fibrosis is a life-threatening genetic disease that causes persistent lung infections and progressively limits the ability to breathe. The median predicted survival age of the patient with cystic fibrosis is 40 years. In the terminal care of the patient with cystic fibrosis, the role of the bedside nurse is critical in providing seamless, interdisciplinary care in order to promote a ''good death'' and ensure that the patient's and family's wishes are respected at the end of life (EOL). Key components of the bedside nurse's role in EOL care for the cystic fibrosis patient include the facilitation of interdisciplinary patient/family–centered communication, the provision of comfort to ease suffering from breathlessness and pain, and the promotion of patient and family closure within the family and among caregivers. This case study depicts the critical role of the bedside nurse in a young female patient with cystic fibrosis at the EOL to honor patient and family wishes and promote a peaceful, dignified death.

KEY WORDS

closure, comfort, communication, cystic fibrosis, end of life, nursing role

Cystic fibrosis is a life-threatening, genetic disease that causes persistent lung infections and progressively limits the ability to breathe.[1] Approximately 30 000 people in the United States are living with cystic fibrosis (70 000 worldwide), and nearly half are aged 18 years or older. Ongoing research has identified breakthrough treatments, which have added years to the lives of people with cystic fibrosis, such as lung transplantation as a viable option.[2,3] Today the median predicted survival age is close to 40 years, but although there has been significant progress in treating this disease, there is still no cure.[1] Advanced care planning should occur during the process of care, with the goal of ensuring that the patient's and family's wishes will be respected at the end of life (EOL).[4] The role of the bedside nurse is critical in providing and coordinating the interdisciplinary delivery of patient/family–centered terminal care to the patient/family, which honors their wishes and facilitates a peaceful, dignified death. The following case study will illustrate key components of the role of the bedside nurse, including communication, comfort, and closure in providing EOL care to cystic fibrosis patients who choose to terminate treatment.

CASE STUDY

TS (anonymous patient initials), a 29-year-old married woman with 2 children younger than 5 years, was admitted to a progressive care unit because of respiratory failure and complications from cystic fibrosis. She had become dependent on a mechanical ventilator during this hospitalization, and when TS was informed that transplantation was not an option, she and her family decided together to withdraw ventilator support. TS was able to clearly indicate understanding of her situation and make her wishes known to the medical team; therefore, it was not necessary to consult psychiatry in this situation. When Cindy, the bedside registered nurse, began caring for TS, she was not aware of the terminal wean that was being considered. This decision was made within several hours of her first day caring for TS. During subsequent shifts (four of the following five 12-hour day shifts) in which Cindy remained the primary bedside nurse, further plans were made with various details considered in collaboration with the primary provider and palliative care consultation services. TS was given the chance to determine the day of her death. This allowed for time to say good-bye to her terminal husband, her 2 young sons, her parents, and her sister. Initially, she had requested that her terminal wean be completed at night, after she read a bedtime story to her children. Because of hospital staffing including consideration of nursing, social work, and respiratory therapy availability, it was determined that her wean should occur on a

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weekday during daytime hours. TS agreed with this. During the weekend, she was able to spend time with her family. Social work had arranged for child-life services to be available to support her sons during this time. Child-life services consist of experts who work with children of adults who have a chronic/acute debilitating illness and assist them in coping and dealing with their parent's illness and death.

On the day of her death, TS was visited by multiple hospital staff whom she had contacted to inform them about her terminal wean, including both nurses and physicians of her outpatient care team from the cystic fibrosis clinic. Other hospital nurses with whom she had developed friendships with also came to visit. If she was sleeping, staff was encouraged by Cindy to leave a note for her. Most of these visits occurred before her family arrived, with the exception of her husband. After her family arrived, including her pastor from home, the pulmonary team providers completed their morning rounds. It was confirmed that she wanted to complete her terminal wean that morning, and she was given the choice of the nurse practitioner or physician assistant who would be involved with her care for the day. After this, her children came in to visit, and she reassured them that they could always talk to her even when they would no longer be able to see her. Her children left the room and were cared for by extended family in the family waiting area. This space had been marked for their specific use, and other families were encouraged to use a second waiting area or space on adjoining units.

At this time, a morphine drip and intravenous lorazepam had been ordered and obtained. TS was visibly fatigued and asked to take a nap. Her husband shared the hospital bed with her and held her while she did this. Her family left to get food and returned approximately an hour later, at which time she was awake, but still sleepy. Ventilator support was withdrawn at this time, at TS's request. The morphine drip was titrated, and the family was encouraged to let nursing staff know if they noticed any signs of discomfort. The respiratory therapist remained near the room as well and also monitored the patient for discomfort. After approximately half an hour, TS's parents requested that Cindy let them know when she thought their daughter was near death so that they could allow TS time alone with her husband. Cindy encouraged them to leave about half an hour afterward, and TS died as her husband held her approximately 15 minutes later. In the meantime, TS's parents were being supported by their home pastor, and other family was present to watch her sons. After TS passed, her husband went to the waiting room, and her parents and sister returned to the bedside. They were allowed time with one another and with her body without hospital staff present. As her father left, he came to Cindy and thanked all of the staff involved with her care. He described that he had worked as a paramedic and that he had seen many deaths, but none that had gone as well as his daughter's.

In hindsight, this was a "good death." It was made possible by many contributing factors, including that goals of care preferences, and decisions were conveyed by TS and her family over the course of several days. Although not all of her wishes could be accommodated, including a nighttime death, efforts were made to honor wishes that could be met to support the patient and her family. The clinical course of her death was eased by the ready availability of interdisciplinary staff. Nurses helped assist with Cindy's other assigned patients so that she could remain available to TS and her family during the morning of the terminal wean. A respiratory therapist remained nearby to manage ventilator settings, and the physician assistant was readily available, which allowed for medical orders to be adjusted and for the morphine infusion dosage to be increased quickly when requested. The social and spiritual portion of her death was eased by support from child-life services, social work, and their home pastor. Continuity of care was also critical during this period. TS and her family knew the staff who was involved in her care and had worked with them for several days before her death. While not all of these factors are possible to ensure, they were beneficial to TS's and her family's comfort during her transition to death.

Key Component of Care: Communication

TS and her family decided to terminate mechanical ventilation treatment when informed of her deteriorating clinical condition and prognosis. Communication was key in honoring the patient's and family's wishes in this situation (Table). TS had a tracheostomy, and the bedside nurse was able to establish communication methods with TS through lip reading and writing on a notepad. Her family also readily interpreted TS's communications to the nurse as needed.

Interdisciplinary communication was imperative in streamlining the patient and family's wishes and goals of care. Cindy coordinated communication among the healthcare team including primary service providers, the palliative care consultation service, respiratory therapy, social work, and other nursing staff.[5,6] The time and place for the terminal wean were mutually agreed on with the expertise and assistance of the palliative care team, and comfort measures were provided to prevent suffering. Flexibility was imperative because TS's initial choice was to be discharged and terminate the ventilator in the home setting. This was not an option because she lived in a rural area not served by hospice. An alternative arrangement was mutually decided on to turn TS's hospital room into a home-like environment and honor her wishes there. Once the plan was decided by the family, the staff implemented it supportively and professionally, with dignity and privacy.[4] Integral to the success of the implementation of family's wishes was having consistency in nursing

TABLE Key Components of Nursing Care in EOL for the Cystic Fibrosis Patient

Key Components of Care	Nursing Interventions
Communication	1. Facilitation of interdisciplinary communication and collaboration between nursing staff, primary service providers, palliative care specialists, social workers, respiratory therapists, physical therapists, chaplains, and child-life services
	2. Maintenance of consistent primary bedside nurse caregivers to facilitate clear communication between shift caregivers, which conveys clear patient and family information and preferences for care
Comfort	1. Physical discomfort: opioids, morphine
	2. Dyspnea: Opioids, fan
	3. Mucus clearance: coughalator bid, prn suctioning
	4. Anxiety: lorazepam
	5. Ventilator setting management
Closure	1. Legacy formation with patient
	2. Memory formation with immediate family and children
	3. Facilitate timing of good-byes from health caregivers, friends, and family
	4. Reminiscence

Abbreviations: EOL, end of life; prn, pro re nata.

care staff provided over the final 3 days on both the day and night shifts. This allowed for enhanced communication with the disciplines and clear handoffs to caregivers on off shifts that included patient and family preferences. In addition, having nurses familiar with TS allowed that minimal report time be spent on the patient's clinical background and more time be focused on psychosocial aspects of care.[7] Most importantly, by having consistent nurse caregivers on both shifts, therapeutic relationships were developed between the patient, family, and staff. This allowed the nurses to implicitly know the patient's and family's wishes, dynamics, and the goals of care without reiteration, thus creating less anxiety in the situation for the patient and family.

Key Component of Care: Comfort

The primary symptoms of discomfort in patients with terminal cystic fibrosis include breathlessness, difficulty expectorating sputum, and chest pain.[8] Breathlessness is often treated with a combination of drug therapies such as midazolam and opiates, nondrug therapies such as noninvasive ventilation, cognitive behavioral therapy, and general measures such as oxygen, sitting in an upright position, use of a cool air fan, breathing control methods, and reassurance.[8-11] TS was on a ventilator at the EOL. The respiratory therapist managed the ventilator to promote maximum comfort for the patient. Symptoms of breathlessness were addressed by having a fan lightly blowing on the patient and the administration of low doses of morphine and lorazepam while on the mechanical ventilator.[8-11] Tracheostomy suctioning was intermittently done as needed and per the patient request, combined with the coughalator vest twice a day to assist with mucus clearance. These nursing interventions were focused on providing comfort and ease of breathing. During the ventilator withdrawal process, the patient was made comfortable with a titrated morphine infusion and lorazepam administration, and rapid withdrawal occurred.[9] The nurse was a key in coaching the family through this process.

In addition, breathlessness can be linked with a vicious cycle of anxiety and fear, which can be treated with cognitive behavioral therapy.[8,12] This involves recognition that it is not just the effect of a physical symptom but the patient's perception and response to the symptom that may be important.[8] In TS's case, the palliative care physician, physician assistant, and social worker assisted the patient and family with clear identification of values and wishes in goals of care. The family strongly supported the patient in her desire to terminate care and withdraw the ventilator, and a plan was developed. Knowing that their plan was supported and facilitated by all members of the interdisciplinary team assisted to some extent in relieving the patient and family members' anxiety and fears related to suffering and the unknown.

Chest discomfort in the patient with cystic fibrosis may be related to frequent coughing episodes resulting in stress on the chest wall.[12] Other sources of pain may include vertebral or rib fractures secondary to osteoporosis or headache from hypoxia.[12] The nurse must have adequate knowledge of symptom etiology and management to provide appropriate relief of discomfort at EOL.[11,13]

Key Component of Care: Closure

As a young adult, closure for TS was primarily important with her family members, including her husband, children, and parents. Legacy formation is important to address in young adults facing their own mortality. Those with children may focus on what their children will remember and how not to be forgotten in their eyes.[14] Offering anticipatory guidance, labeling grief, and providing empathy to encourage self-reflection about developmentally appropriate losses helps the patient to feel understood and respected.[14] Reminiscence was effectively used as the family viewed picture albums and told stories of happy occasions and trips. TS valued reading bedtime stories to her children, and the staff facilitated this with her as a way to allow them to remember her last days in a positive way. She also made memories with her children by taking her picture with them and making linked fingerprints together in clay. The nurse encouraged TS and her mother to clip locks of TS's hair for her children and sign cards that would be delivered to them on their subsequent birthdays. Support for the children also came from child-life experts who guided the children through the experience and assisted with interpreting the situation for them. Spending her last moments with only her husband present also honored TS's wishes to acknowledge her role as a wife. TS's parents were present during the last 3 days and helped to communicate these wishes to the nursing staff so that they could be honored. Pastoral support and presence were invaluable to the family during this time and was even more meaningful because this was provided by their home pastor who knew TS and her family.

It was also important for TS to have closure with interdisciplinary health care providers including her primary physician and out patient nurses who had treated her for several years. Many came to her hospital bedside and she was able to thank them for their support and concern, and achieve satisfaction through saying good-byes. Notably, many of the care providers told the family that it had been an honor and privilege caring for TS over the years, and expressed appreciation for the therapeutic relationship which had developed. Timing and coordination of these health care professionals was assisted by the bedside nurse. Final satisfaction for TS and her family occurred through the decision to donate her body to medical science to learn about the pathophysiological impact caused by the cystic fibrosis disease and treatment.

Barriers to Care

Although many aspects of this case study made it an exemplar for a "good death," there were opportunities to improve the situation. The nurse, Cindy, was not aware of TS's situation when she chose her patient assignment. The terminal wean for TS had been in consideration by the patient, family, and medical team for a few days, but this information was not conveyed during change of shift report, and Cindy did not know that she was signing up for this grave situation and potential terminal wean. She believed that this knowledge would have been beneficial in determining her readiness to be able to deliver the best quality of patient and family-centered care for TS and her family. In addition, when the decision was made to move forward with the terminal wean, staffing workload assignments could not be adjusted to compensate Cindy for the increased time needed in supporting the patient and family, coordinating the interdisciplinary team and terminal wean process, and so on. A nursing colleague with nearby assigned patients, implicitly understood the dynamics of the situation and provided immeasurable support with whatever Cindy needed for TS and her other patients. Cindy was extremely grateful that her colleague recognized ways to help with this workload burden.

Technological challenges occurred when the low-pressure ventilator alarms were not shut off by respiratory therapist during the terminal wean. The respiratory therapist had to stand at the ventilator to continually silence them, which was somewhat distracting to family members in the room. Other technological challenges included the beside computer documentation that needed to be done to verify medication administration and other charting. The family had expressed wishes that the computer be turned off, as well as the cardiac monitor. Cindy was able to have a portable computer brought outside the patient's room to perform necessary documentation. The telemetry was turned off in the room and able to be visualized at the nursing station to monitor the patient's status. It is challenging to minimize technology to promote a peaceful death in the hospital environment.

Finally, Cindy recognized that after caring for TS during the 3 days leading up to the terminal wean she felt emotionally exhausted from the situation.[15] Although Cindy was understanding and supportive of TS's decision to withdraw care, the family dynamics of having a young wife with 2 very young children compounded the sadness of the situation for both family and caregivers. It was difficult for Cindy to bounce back and return to "work as normal" immediately after the patient died. Cindy found that support from nursing management to remove her from staffing for a few minutes before returning to the care of her other patients for the rest of her shift would have been extremely helpful. In addition, Cindy believed that providing for staffing flexibility by allowing her to take the following day off from work would have contributed to her resilience in subsequently returning to patient care. Other supportive measures to ensure nursing resiliency could include daily multidisciplinary debriefings,[16] education about holistic practices such as mindfulness and meditation,[17] as well as dedicated unit spaces for nurses to relax and restore.[17]

Conclusion

The role of the bedside nurse is critical in the delivery of patient/family–centered care for the cystic fibrosis patient at the EOL. In this case, the same nurse cared for the patient during the last 3 days of her life. Having consistent nursing staff enhanced knowledge of the patient's and family's needs and preferences for care, thereby decreasing their frustration and anxiety when interacting with new staff. The bedside nurse also played a key role in the collaboration and communication with interdisciplinary team members' delivery of the care so that goals of care were known and facilitated. Delivery of terminal care was seamless and within a supportive environment. Patient comfort for the patient with terminal cystic fibrosis was also enhanced immensely by the consistent bedside nurses who understood patient comfort and airway management needs as well as values and goals of care. In summary, the bedside nurses in this case study were able to assist the interdisciplinary team in facilitating TS and her family in having a peaceful, comfortable death, which honored their wishes and provided a dignified closure.

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