Cardiology in the Young

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"Failing Fontans:" an emerging challenge in paediatrics

Tracy Hills[1], Erika Mejia[2] and Rachna May[3]

[1]Department of Pediatrics, Vanderbilt University Medical Center, Nashville, TN, USA; [2]Department of Pediatrics, Northwestern University, Chicago, IL, USA and [3]Department of Pediatrics, University of Oklahoma Health Sciences Center, Oklahoma, OK, USA

Case Report

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Corresponding author:
Rachna May; Email: rachna-may@ouhsc.edu

Abstract

As staged palliative surgery for patients with hypoplastic left heart syndrome ages to greater than 50 years in practice, providers are increasingly faced with the challenges of managing patients with "failed Fontan" physiology. In this brief case report, we discuss different ways paediatric palliative care can support this population.

Introduction

Hypoplastic left heart syndrome is nearly universally fatal if not managed with staged surgical palliation. Although surgical and medical advances have improved outcomes for this patient cohort, 12-year transplant-free survival is only 60%.[1] The third of these staged procedures, the Fontan, was developed in the late 1960s, and now there is an increasing number of patients with hypoplastic left heart syndrome reaching adulthood.

Medical comorbidities seen in these patients include heart failure, plastic bronchitis, protein losing enteropathy, cirrhosis, renal insufficiency, and thromboembolic disease.[2] With the accrual of morbidities, patients experience a range of symptoms and an expanse of difficult decisions regarding medical management, surgical or catheter-based interventions, and advance care planning.[3]

This patient population faces overlapping challenges related to their chronic disease while confronting legal and social obstacles found in adulthood. As patients age, their medical understanding, their values, and decision-making preferences become increasingly more complex, and the transition from youth to adult care can be particularly tumultuous for patients and families. Early involvement of paediatric palliative care helps ease both medical and psychosocial burdens for these patients.[4]

Case presentation

RC is a 19-year-old male with hypoplastic left heart syndrome with left pulmonary artery atresia status who underwent Fontan palliation at the age of 3 years old and subsequently developed failed Fontan physiology characterised by elevated venous pressures (25–30 mmHg), biventricular heart failure, cirrhosis, and multifocal hepatocellular carcinoma.

Paediatric palliative care first met RC, then 15 years old, in a care conference with gastroenterology, oncology, and cardiology. His family was informed there were no further disease-modifying medical therapies or surgical interventions available. Due to anatomical challenges and significantly elevated human leukocyte antigen sensitivity, multiple centres declined him for combined heart/liver transplant. Furthermore, oncology did not have chemotherapy to offer. A year after this meeting, RC's family contacted paediatric palliative care with concerns about worsening symptoms, including fatigue, poor appetite, nausea, and abdominal pain. Hospice was discussed, but his family was not amenable to hospice enrollment and was still seeking second opinions.

Six months later, RC was admitted with abdominal pain and ascites and was found to have progression of his hepatocellular carcinoma. RC declined an offer to trial sorafenib as he and his family felt his quality of life was acceptable and the burdens of sorafenib outweighed the benefits. Over the next two years, RC had multiple admissions for diuresis and experienced progression of symptoms including fatigue, shortness of breath, and pedal and scrotal oedema.

As RC approached the age of majority, paediatric palliative care discussed health care proxy with RC. He shared he deferred medical decision-making to his parents and felt comfortable with their decisions should he not be able to speak for himself. RC stated his goal was to "live another twenty years" and that his focus was on the positive. He wished to be at home over the hospital and requested outpatient diuresis versus short stay admissions for intravenous diuretics.

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Despite efforts to remain home, RC was readmitted with significant bilateral lower extremity oedema and fatigue. A trial of milrinone was initiated for quality-of-life benefits. RC did not want an indwelling line and requested milrinone discontinuation after a few days. However, once milrinone was discontinued, RC developed significant acute renal failure and became obtunded. After multiple discussions about end-of-life and goals of care with his family and paediatric palliative care, his parents declined dialysis and requested to place a "Do Not Resuscitate" order. Ultimately, his acute renal failure self-resolved, and his parents reverted code status back to full resuscitation. Once RC was able to communicate again, he opted to stay on milrinone, despite his initial hesitations.

RC was discharged after 38 days on milrinone, bumetanide, metolazone, methadone, oxycodone, ketamine, and diazepam for symptom management. His primary goal remained maximising time at home, and he also wished to proceed with non-invasive treatments to prolong life and be pain-free. He and his parents did not wish to have an advanced directive in place, given the unlikely turnaround post renal failure. Based on RC's goals, hospice care was readdressed, and RC agreed to enroll.

Once home, RC was followed monthly in clinic jointly by cardiology and paediatric palliative care. He had ongoing issues with fatigue, low appetite, and worsening oedema. Four days after his last clinic visit and four months after hospital discharge, RC collapsed at home. His mother called emergency medical services; resuscitation efforts were discontinued by his mother on arrival to the local emergency room. Palliative care continued to follow RC's family for one-year postmortem to provide bereavement support.

Discussion

Paediatric palliative care best practices include partnering with the patient and family to provide holistic, longitudinal, and goal concordant care throughout the course of a chronic, life-limiting illness in partnership with the medical care team.[5] Paediatric palliative care consultation in the care of adolescents and young adults patients or with end stage heart failure is not well studied.[6] Few available studies show that paediatric palliative care is underutilised in the adolescents and young adults population, but well accepted.[7] When paediatric palliative care is involved, conversations about goals of care, advanced care planning, and identification of a health care proxy occur earlier in the course of illness.[4]

Paediatric palliative care followed RC for five years prior to his death advocating for goal concordant care and partnering with the cardiology team in symptom management. RC continued to live his life as he wished, graduating from high school, moving out of his parent's home, and occasionally racing cars. The complexity of decision making—desiring a "palliative" plan, while refusing to address code status—is seen in this case. The focus on living well over advanced care planning is common in adolescents and young adults in other disease states.[8] This paradigm can be challenging for medical teams to navigate, further lending importance to the collaboration of paediatric palliative care and cardiology at this stage of illness.[3,7]

Another unique aspect to this case is the challenges of hospice enrollment on home milrinone. Home milrinone is safe to use in the paediatric hospice population, and patients often demonstrate a reduction in symptoms including fatigue, decreased appetite, and respiratory discomfort.[9] However, due to cost, patients on home milrinone may not qualify for hospice enrollment, dependent on state laws and payor. Concurrent care, or the ability for patients to receive life prolonging therapies in addition to hospice care, is available to patients under the age of 21 years enrolled in Medicaid, under the Affordable Care Act.[3,10] RC was enrolled into hospice under concurrent care, allowing him to continuing receiving milrinone during that time.

Conclusion

Palliative care consultation is important for end-stage Fontan patients, to address medical and quality of life goals, advanced care planning, and emotional and spiritual distress. Early integration of paediatric palliative care into the multidisciplinary team can significantly impact patient and family outcomes, ensuring that they receive holistic and goal concordant care through the final parts of their illness.

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