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HMS 2013
Castle Society

Neil Samuel Ghiso Foundation - Fellowship Report

Parental Perspectives on Quality of Life of Children with Advanced Heart Disease: Implications for Alleviation of Lifetime Burden of Disease

My summer as a Ghiso fellow has been truly a fabulous learning experience. I was able to gain experience in palliative care research and exposure to clinical care of chronically and terminally ill children. The knowledge and skills I have acquired have strengthened my commitment to pursuing a medical career in which I practice compassionate medical care of children coping with chronic illness. I am incredibly grateful for the support of the Ghiso Foundation as well as my mentors, Dr. Betsy Blume, Medical Director of the Heart Failure/Transplant Program, and Dr. Joanne Wolfe, Director of Pediatric Palliative Care at CHB. In this final report, I summarize the results and status of my research, as well as my experience in the clinical setting.

RESEARCH

I am very pleased with my Summer 2010 progress and productivity. As a result of this extremely positive experience, I intend to continue working closely with my research mentor, Dr. Blume, throughout my second year and beyond in order to see our research project to completion.

In 2009, Dr. Blume received a grant from CHB to conduct a two-fold study: 1) a retrospective medical chart review of children who passed away as a result of advanced heart disease over a three year period at CHB, and 2) an extensive survey of these patients' parents designed to better understand patient suffering. The overall purpose of the study is to assess the physician, patient, and parental factors that predict ease of suffering at the end-of-life in pediatric patients with advanced heart disease. The IRB proposal for the chart review portion was approved this past spring. While I had expected the IRB for the survey portion of the study to be approved in a similar time period, it unfortunately underwent a significant hold-up by the Cardiology Department's Scientific Review Committee prior to submission to the IRB this summer. We addressed the Committee's concerns regarding length and phasing of survey, and then submitted our full proposal for review by the IRB this past July. We recently received comments from the IRB and have already addressed their concerns and resubmitted the proposal for consideration.

The resistance from the Cardiology Department and the IRB to approve the study has been frustrating, as this same type of study with parents has been carried out numerous times in the pediatric oncology population. Thus, there seems to be some degree of resistance purely because of the new setting, in pediatric cardiology patients, where end of life care is not typically addressed in the same manner. What makes this even more frustrating, and provides even greater purpose for our study, is the obvious immense need for palliative care research relating to children with heart disease. I am confident that we will obtain IRB approval in our latest submission, and I have worked to get all aspects of the survey implementation in order so we can begin right away. We plan to do a pilot study first with four families – two from the pediatric oncology population and two from the pediatric cardiology population. We hope this will help us to make final adjustments to the survey prior to carrying out its formal implementation.

Despite these challenges in getting the survey portion of the study approved, I have made enormous headway on the chart review. I started with the patients in the January 2007-July 2009 Cardiology Bereavement Database (which contains any child who died in that time who had ever been seen for a cardiology consultation). I reviewed 468 records for inclusion, and ultimately included 111 patients. Exclusion criteria were: 1) patient passed away outside the hospital; 2) patient was older than 21 years of age at time of death; 3) patient's primary diagnosis was prematurity, active cancer, sepsis, congenital diaphragmatic hernia, or sudden infant death syndrome. All patients with a primary diagnosis of pulmonary hypertension were included.

I helped to develop a medical record data extraction tool composed of 45 indicators based on the study's purpose as well as variables relevant to end-of-life experience reported in the literature. The tool

was used to capture demographics, lifetime burden of cardiac disease, types of support within 24 hours of death, and modes of death.

Several variables were designed to assess lifetime burden of disease. All procedures billed as surgeries were reviewed and categorized as definitive surgeries (conducted in an Operating Room and treatment-directed) or bedside surgeries (such as mediastinal washouts, thoracotomies, and ECMO cannulation). In addition to the standard treatment-directed surgeries, the following procedures were accounted for as 'definitive' if conducted in an Operating Room: pacemaker insertion, diaphragm plication, and shunt revision. Total numbers of catheterizations were also recorded. To assess the intensity and density of interventions over the patients' lifetimes, total number or definitive surgeries and catheterizations were each divided by length of life. Tracheostomy, gastrostomy, and peritoneal drain placements were also recorded. ECMO cannulation dates were recorded and total days on ECMO over patient lifetimes were calculated.

End-of-life care indicators are described as follows. Location of death, length of last hospital stay, documentation of end-of-life discussion with family, and presence of family at bedside at time of death were obtained from final discharge notes. Types of mechanical ventilation, medications, feeding, mental status and mechanical cardiac support (ECMO, ventricular assist device, cardiopulmonary bypass) were recorded from notes within 24 hours prior to death. Types of end organ failure were extracted by reading numerous inpatient records and consult notes. The extraction tool contains seven sub-questions to assist in categorizing the type of death as: during resuscitation, intervention withdrawal, or comfort care. Primary and secondary causes of death were coded based on organ systems affected as well as such clinical conditions as sepsis, capillary leak syndrome, and graft rejection. Patient or family resources accessed, including pastoral care, social work, pediatric advanced care team, child life, and psychology, were noted as well. Finally, documentation of autopsy was noted.

Midway through the summer, I had reached about 65% of total data abstraction goals. However, research abstract deadlines for the American Heart Association and American Association for Hospice and Palliative Medicine (AAHPM) were fast approaching. Dr. Blume encouraged me to submit abstracts based on my preliminary data. I have included the abstract here:

Title: Patterns of end of life care in children with advanced heart disease

Background: Despite significant advances in the treatment of children with advanced heart disease, a small percentage of children die, and little is known about their end-of-life experience. This study describes patterns of care of patients with advanced heart disease who experience in-hospital deaths.

Methods: 79 patients <21 yrs who suffered in-hospital deaths at Children's Hospital Boston from January 2007-July 2009 were included. Medical records review included patient demographics, disease characteristics, types of support and modes of death.

Results: Median age at death was 6mo (1 d-20 yrs), while median length of last hospital stay was 22 days (1-193). Seventy-four percent of the patients had complex congenital heart disease and 20% had known genetic diagnoses. Nearly 40% of patients were from out of state and 4% were international. The median lifetime number of catheterizations and surgeries were 2 (0-59) and 4 (0-18), respectively. At end-of-life, children had tracheostomies (2), gastrostomy tubes (13), and peritoneal drains (16). Within the last 24 hours of life, 85% of patients were mechanically ventilated, 86% were on inotropic support and nearly 30% were on mechanical circulatory support. Just over 60% of patients had evidence of end organ failure at time of death (42% neurological, 44% renal, 50% pulmonary, 10% hepatic); 50% of cases involved multi-organ failure. Over half (58%) of patients were completely sedated and most (97%) received analgesics. Regarding mode of death, 22 patients (28%) died during resuscitation, 42 (53%) had interventions withdrawn and 5 (6%) patients died receiving comfort care.

Conclusion: While overall survival of children with heart disease continues to improve, children who die experience a high burden of care including chronic hospitalization, repeated interventions and frequent use of advanced supportive technologies. Further research from the perspective of children and their families would help to better understand their end-of-life care needs.

For the AAPHM conference, I was instructed to include measurable behavioral objectives that would be given to attendees if my abstract was selected for a presentation: 1) Increase awareness of the importance of end-of-life care specifically for the pediatric cardiac disease population; 2) Increase

utilization of end-of-life care practices for children with advanced heart disease; 3) Extract baseline characteristics of children dying of advanced heart disease in order to carry out prospective studies and interventions at home institutions.

I am currently working to analyze the full chart review database with a biostatistician in the Cardiology Department and intend to submit a manuscript with those results to a journal in the coming months. I believe that the results of this chart review alone will provide critical information for practicing clinicians and researchers in the fields of pediatric cardiology and palliative care. Literally no other studies have ever looked at end of life care issues relevant to children with heart disease, and I sincerely hope that this initial study will provide impetus for further research that will identify ways in which we can change clinical practice to reduce lifetime burden of disease and relieve suffering. I hope that our survey of parents may help us to identify some of those changes that we need to make.

As outlined in my initial proposal, I still intend to test my own hypothesis from the survey portion of the study. I will assess whether or not the degree to which parents perceive their child's level of suffering depends on the lifetime burden of their child's disease. This is intended to provide a broader picture of quality of life for chronically ill patients and the potential for adjustments to standards of patient care to alleviate suffering at all points of illness, not just in the acute end-of-life period. The preliminary results from the chart review have certainly shown that lifetime burden of disease tends to be high for many of these children. I feel it is critical to understand how this may relate to degree of suffering.

CLINICAL EXPERIENCE

Through shadowing Dr. Blume and intensivists in the Cardiac Intensive Care Unit (CICU), I was able to gain significant clinical exposure to chronically ill children in the hospital setting. One particularly vivid moment happened on the second full day of working in the hospital. On rounds my first day with Dr. Blume in the CICU, I had the opportunity to see a number of patients, including Jose, a 5-year old boy born with a single ventricle who had been staying in the CICU for the past 5 months. He had undergone several 'palliated' surgeries early in life to give him an opportunty for survival. At a young age following those interventions, Jose's family spent a few years away from medical care and perhaps missed early signs of developing heart failure. At approximately age four, Jose developed renal, liver, and respiratory failure as a result of poor perfusion, and at this point, cardiac transplant was no longer an option. Jose's parents were adamant about continuing to provide all possible forms of treatment-directed interventions, so Jose was maintained on the maximal pressor support as well as 'treatments' for each of his organ failures. They were not willing to hear about any of the 'comfort care' options. On my second day in the hospital, Dr. Blume received a page telling her that Jose was 'coding.' We rushed up the CICU. I had no idea what I was getting myself into. I had never seen a code, except on medical dramas on TV. I had never seen a patient pass away. And I had never imagined that a death could seem so traumatic and chaotic. The family could not believe that their son was dead, and kept shouting – 'he's going to wake up, I know he's there…" over and over. The code team continued with chest compressions long after they knew that Jose was dead, just to ensure the family that all measures had been done. I was so emotionally upset by what I had witnessed. I felt helpless in the situation and so distraught by the family's pain. This experience showed me first-hand what a 'sudden,' and yet not-so unexpected death could be like, and gave me strengthened resolve to better understand how to best care for children and their families in the period of time during which the child no longer has realistic chance of survival, and at the end of life itself.

I also had the wonderful opportunity to spend several days this summer with the PACT team. I observed all aspects of their involvement in care – from intake, to family meetings, to examinations of patients, to discussions of hospice care, to discharge of patients on various forms of home support. I was able to see how absolutely essential the role of the PACT team is in coordinating care for patients and families dealing with difficult chronic illness. I also saw the importance of PACT team involvement not just at the final end of life period, but throughout various phases of illness.

EVALUATION - Successes and Challenges

Lastly I would like to address the points I outlined in my Evaluation and Potential Obstacles sections of my initial protocol. I include here the measures I set out for my own evaluation of success:

1) Extent to which the general survey is effectively modified with additional questions for my secondary hypothesis. I successfully added questions to the survey to reflect lifetime burden of disease as well as degree of suffering at end of life.

2) Completion of the survey portion of the study from time of parent enrollment to comparison of data collected with physician and patient factors. I was not able to even begin this portion of the study, unfortunately. However, I plan to be heavily involved in the entire process once we begin implementation.

3) Completion of medical chart abstraction from patient files. I successfully completed this objective.

4) *Completion of data analysis*. I was able to conduct preliminary data analysis of the chart review for submission in two conference abstracts. Final chart review analysis is currently underway.

5) Survey results showing appropriate statistical power (for an estimated 80% reporting of "suffering" with 80 parents, estimate proportion of suffering within $\pm$ 8%). Not yet completed.

6) Extent of perceived benefit of research for future medical practices and development of educational tools. Not yet able to assess.

7) Sufficient breadth of opportunities to engage with chronically ill patients; and 8) routine observation of palliative care team in hospital and related specialists involved in coordinating patient care. I am grateful that I had numerous opportunities to observe patient and family interactions.

Thus far, I am only able to evaluate one of the potential obstacles listed in my initial proposal - that extensively complicated medical charts would make consistent data abstraction difficult. While learning how to read, code, and analyze patient charts took a good deal of time and teaching from Dr. Blume as well as from a Cardiology fellow, I eventually felt confident in conducting the chart abstraction on my own. For a few complex indicators, such as "cause of death" or "type of death," my codes were reviewed by at least one other member of the study staff for accuracy and consistency.

FINAL THOUGHTS

My experience as a Ghiso fellow this Summer 2010 has strengthened my passion for compassionate medical care and my commitment to pursuing further training in palliative medicine. Whatever medical discipline I choose to enter, I know that I will make patient and family hopes, values, and goals a priority in coordination of care.

Dr. Blume continues to be a fabulous mentor. She has consistently made time to work with me to develop the research methods as well as take me on Rounds in the hospital. She is an excellent role model of someone who has made a successful career in a pediatric discipline, demonstrating in both her clinical practice and academic research her commitment to compassionate care for children nearing end of life and with terminal diagnoses.

Dr. Wolfe, a Co-Investigator on the survey portion of the study, has also been a great mentor. I have had the opportunity to shadow her in the hospital as she leads the PACT team on Rounds and in her interactions with patients. In addition, I have learned a great deal from her past research through both reading and active discussions concerning the care for children with end-stage cancer.

I would like to sincerely thank the Ghiso Foundation for their support of my work with Dr. Blume and Dr. Wolfe.