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Pediatric Cardiomyopathy as a Chronic Disease: A Perspective on Comprehensive Care Programs

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Abstract

Substantial numbers of children with cardiomyopathy are now surviving into adulthood, making it essentially a chronic disease. As a chronic condition, it may be best treated through comprehensive, multidisciplinary treatment programs. Such programs have improved health outcomes and reduced costs in managing other pediatric chronic diseases and heart failure in adults, but the treatment and cost implications of programs for managing pediatric cardiomyopathy are unknown. We investigated the treatment and cost implications of establishing such programs by reviewing cost-effectiveness studies of similar programs, estimating the current inpatient costs of this diagnosis, and interviewing experts in the field about the need and desirability of these programs. According to our findings, comprehensive pediatric heart failure programs do exist, but they have not been evaluated or even described in the literature. Consensus among experts in the field is that such programs are highly desirable, and similar programs have reported tremendous cost savings through early and intensive management: the return on investment has been as high as 22 to 1. Another study reported that mean length of stay decreased from 83.9 to 10.6 days, mean annual admissions decreased from 2,796 to 1,622, and median hospital charges decreased from $26.1 million to $14.6 million. In conclusion, limited experience and strong circumstantial evidence suggest that, despite substantial costs, comprehensive multidisciplinary pediatric heart failure programs would be highly cost-effective and beneficial to patients, families, and institutions alike.

Keywords

Pediatric cardiomyopathy; pediatric heart failure; comprehensive programs; multidisciplinary programs

INTRODUCTION

With an estimated incidence of 1.13 cases per 100,000 children, and with 40% of children dying or requiring heart transplant within 2 years after diagnosis, pediatric cardiomyopathy is a rare, but serious condition in children.[1] However, a substantial proportion of these children undergo heart transplantation and survive, and others remain event-free for years.

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The Pediatric Cardiomyopathy Registry (PCMR) reports survival rates after a diagnosis of dilated cardiomyopathy[2] with freedom from death or transplantation ranging from 69% at 1 year to 46% at 10 years and for idiopathic hypertrophic cardiomyopathy[3] 94% at 1 year to 85% at 10 years (Table 1).

Cardiomyopathy is the leading cause of transplantation in children older than 1 year of age, and the percentage of children with cardiomyopathy who receive heart transplants has not declined over the past 10 years.[4] In 1993, the American Heart Association (AHA) reported that pediatric cardiovascular disease cost more than $8 billion per year in medical expenses and lost contributions to the US gross national product.[5] In comparison, the AHA estimates the direct and indirect costs for adult heart failure at $33.2 billion in 2007, out of a total of $431.8 billion for all cardiovascular diseases in the United States.[6] There has not been an estimate published for children with heart failure or cardiomyopathy to date.

Consequently, the majority of children diagnosed with cardiomyopathy are now living longer and many more are surviving into adulthood. The case can be made that pediatric cardiomyopathy has now become a chronic disease, complete with high associated costs.[2, 3]

In other chronic pediatric diseases, as well as in adult heart failure, various comprehensive treatment programs have improved health outcomes and reduced costs. On the basis of the above facts, we suggest that pediatric cardiomyopathy, as a chronic disease, would benefit from adopting the approaches taken to other pediatric chronic diseases.

In this article, we review the current literature and thinking in this field and make a case for establishing multidisciplinary pediatric comprehensive heart failure management programs.

METHODS

Literature Review on Multidisciplinary Treatment Programs

To identify comprehensive heart failure programs and their economic impact, we searched PubMed for articles published in English using the following keywords: pediatric heart failure, economics, cost-effectiveness, and comprehensive health programs. No restrictions were placed on country of origin, publication dates, or type of economic valuation, and no quality measures were used to exclude otherwise suitable topics.

Inpatient Cost Estimate of Pediatric Cardiomyopathy

The Healthcare Cost and Utilization Project (HCUP) is a family of health care databases and related software tools and products developed through a Federal-State-Industry partnership and sponsored by the Agency for Healthcare Research and Quality (AHRQ).[7] The database includes the largest collection of longitudinal hospital care data in the United States, with all-payer, encounter-level information beginning in 1988. We queried the database to assess the inpatient costs incurred for children with a principal diagnosis of cardiomyopathy.

Interviews with Pediatric Health Professionals and Advocates

We interviewed health professionals specializing in pediatric cardiomyopathy to identify how they care for their patients with this diagnosis and to determine what changes they believed were needed to improve this care. We also interviewed the founder and Executive Director of the Children's Cardiomyopathy Foundation (CCF) and a Department of Pediatrics administrator.

RESULTS

Literature Review on the Costs of Cardiac Treatment

We identified 23 unique citations to multidisciplinary cardiac treatment programs, only one of which was targeted to children. Other relevant studies are also described below.

The costs of pediatric cardiac treatment

In a cost-effectiveness study of pediatric heart transplantation,[8] data from 95 children undergoing transplantation at one medical center between 1997 and 2004 were reviewed to determine the cost of transplantation, including pre-transplant care, organ procurement, initial hospitalization, and follow-up care. The reported costs for pediatric heart transplant and follow-up services were estimated at $465,494 for the initial year.[8] The costs of primary pediatric heart transplantation relative to the benefits, expressed as quality-adjusted year of life (QALY) gained, was reported as $49,679 per QALY gained, which was close to the commonly accepted benchmark of a cost-effective therapy of $50,000 per QALY. The cost of re-transplantation, however, was barely within the outer limit, which is considered to be $100,000 per QALY. The base case estimate for re-transplantation was $87,883 per QALY, and sensitivity analysis identified the range from $70,834 to $103,661 per QALY. The authors suggest that with the high costs of transplantation and the increased frequency of re-transplantation in treating graft failure, which accounts for 6% of heart transplants, improvement in survival after re-transplantation is needed to bring its cost–effectiveness in line with acceptable standards. This would be addressed by comprehensive heart failure programs in attempting to delay the use of this therapy or to select better candidates who may need less intensive and less costly care with more benefit derived when transplantation is utilized.

The costs of treating other chronic pediatric diseases

The influence of chronic disease on resource use in common acute pediatric conditions was estimated in a retrospective study of 30,379 admissions of patients with and without chronic diseases for common pediatric conditions, such as concussion, appendicitis, and viral illness.[9] Of the 30,379 admissions, 4,737 patients had at least 1 of the selected chronic conditions. Children without chronic disease had shorter mean length of stay than did patients with at least 1 chronic disease (2.53 vs. 3.05 days, $P<0.001$). In addition, mean total hospital charge for patients without chronic diseases was lower ($2,614 vs. $3,663, $P<0.001$) than for patients with at least 1 chronic disease.

The resource use and cost of care for children with chronic conditions was also investigated in a comprehensive care program at the Children's Hospital at Strong, University of Rochester Medical Center.[10] To expand ambulatory care coordination and comprehensive services to children with chronic conditions, the hospital began hiring additional personnel, including nurses, social workers, and psychologist in 1985. Funding was provided by a regional insurance company in 1989. All children with chronic conditions, independent of their insurance status, had access to the services. Data were collected from 1984 to 1995 and the following results were reported. Mean length of stay decreased from 83.9 to 10.6 days ($P<.001$), and mean annual admissions decreased from 2,796 to 1,622($P<.001$). Median hospital inpatient charges, adjusted for cost of leaving, decreased from $26.1 million to $14.6 million. The return on the $3.6 million investment by the insurance company was a total savings on inpatient care of $77.7 million, an impressive return-on-investment ratio of 22 to 1.

A novel approach for reimbursing intensive case management in children with diabetes also resulted in substantial savings.[11] Three intensive case management services—specialty education, round-the-clock telephone access to an educator, and quarterly educator assessments of self-management skills—were reimbursed through a contract with a third-party

payer. Data were collected and analyzed for 15 months after contract initiation. There were 22 hospital admissions for diabetic ketoacidosis (DKA) for 16 different patients within the first 15 months after the contract was signed. Ten out of 16 patients chose to participate in the intensive case management program described above. The results showed that the program had a significant effect on frequency of hospitalization and emergency department visits: there was only one subsequent emergency department visit or hospital admission for DKA among the participants, whereas 5 non-participants were rehospitalized for DKA (P=.039). In addition, the program made a significant impact on the total cost, which included education, hospitalization, and emergency department visits. The total cost of nonparticipant was 125% greater than that of a participant: $2,396 vs. $1,063 (P=.008). Considering the success of the program above, a similar approach could be applied to managing other costly chronic diseases.

The costs associated with adult comprehensive heart failure programs

As described above, studies focusing on the successful management of pediatric chronic diseases, other than cardiomyopathy, have had impressive results. In the adult cardiology literature, specifically adult heart failure, many reports describe the effectiveness of comprehensive heart failure programs. Several such programs are briefly summarized below.

A randomized trial investigated the effect of a nurse-directed, multidisciplinary intervention in high-risk patients with congestive heart failure on the rates of readmission within 90 days of hospital discharge, quality of life, and cost of care.[12] The intervention included comprehensive education for the patients and families, a prescribed diet, social-service consultation, and early discharge planning, as well as a review of medications and intensive follow-up. Readmissions for heart failure decreased by 56.2% (54 in the treatment group vs. 24 in the control group, P=.04), and readmissions for other causes decreased by 28.5% (40 vs. 29, P not significant). In addition, the quality-of-life scores at 90 days for patients in the treatment group were significantly higher than those in the control group. Finally, the overall cost of care was $460 less per patient in the treatment group as a result of the reduction in hospital admissions.

Over a 3-year period, Fonarow et al.[13] enrolled 214 adults at a single center with heart failure (New York Heart Association functional class III or IV), who were accepted for heart transplantation, and who were then discharged to await elective transplantation. Before discharge, patients were thoroughly evaluated, their medical therapy was adjusted, and they received intensive education on diet, exercise, and overall awareness of their condition. Patients were followed closely after their discharge. Death and transplantation were also tracked as end points. Functional status, hospital readmission rate, and estimated hospital costs were compared for 6 months before and after referral. Hospital readmissions were reduced by 85%, functional status improved (as indicated by congestive heart failure functional class and aerobic capacity), and the estimated savings in hospital readmission costs were $9,800 per patient. A follow-up report by Fonarow et al.[14] using the OPTIMIZE-HF trial data showed increased use of evidence-based measures and shorter length of stay across 259 hospitals in the United States.

Comprehensive inpatient education and discharge planning combined with outpatient support have been beneficial in elderly patients with congestive heart failure.[15] An experienced cardiac nurse educator coordinated a targeted inpatient congestive heart failure education program linked with comprehensive discharge planning and immediate outpatient reinforcement through a home health care program. As a result, 6-month readmission rates fell from 44.2% to 11.4%. In addition, the decreased use of skilled nursing services and home health care during outpatient follow-up saved an average of $1,541 per patient in the interventional group.

The Hackensack University Medical Center Heart Failure Program developed a multidisciplinary team approach to heart failure that focused on case management and the

continuum of care.[16] The Program emphasized the importance of continuous care in the patient's home and provided a comprehensive set of discharge instructions. An integral part of the Program was the discharge education program, which included dietary guidelines, instructions on proper medication intake, as well as smoking cessation and exercise. In addition, the staff received education and training on the pathophysiology, treatment, and physical assessment of the disease, among other areas of care. Finally, various support programs, such as an inpatient heart failure unit, an outpatient center, and a heart failure homecare team were established. The Program performed well on the Joint Commission on Accreditation of Healthcare Organizations and the Centers for Medicare and Medicaid Services core measures. For example, 100% of program participants received angiotensin converting enzyme inhibitors at the time of discharge, and all received smoking cessation counseling. Moreover, length of stay declined on average from 7 to 5 days.

A meta-analysis assessing the effectiveness of multidisciplinary heart failure management programs on hospital admission rates analyzed 8 randomized controlled studies that compared usual care with a nonpharmacologic patient education interventions targeted at increasing patient knowledge of heart failure.[17] Some intervention components included nurse, dietitian, and geriatric cardiologist inpatient visits, educational videos and booklets, as well as counseling, and nurse home visits and telephone calls. The pooled relative risk reduction in hospital readmissions was 21%, in favor of the intervention group with a number needed to treat of 9.

During the American College of Cardiology 2007 meeting, two disparate abstracts were presented. The multi-center COACH trial from the Netherlands enrolled adults with heart failure into one of three counseling programs. The results showed an improvement in mortality reduction (15%), but the primary combined endpoint was not significant, as rehospitalization rates were not decreased. The smaller REMADHE trial from Brazil showed improvement in mortality, re-hospitalization, and other cost measures. The lead author of the COACH trial, Dr. Jaarsma, commented that, "close intensive nurse-led advising and counseling in chronic heart failure patients might decrease mortality at the 'cost' of more-shorter-hospitalizations."[18] Also noted was the older population in COACH (mean 71 versus 51 years) which may have affected the outcomes.

The programs described above are among many successful examples of comprehensive adult heart failure programs improved health outcomes, reduced length of stay, and decreased number of hospital readmissions, which, in turn, led to substantial cost savings. The American Heart Association established the "Get with the Guidelines" initiative advocating that hospitals measure and report performance and quality measures in an effort to improve care at the institution level.[19] The AHA reports improved outcomes and reduced events at participating centers. Such programs have set a precedent for establishing pediatric comprehensive heart failure programs in the near future.

Estimated Cost of Pediatric Cardiomyopathy

We found no published estimate on the cost of cardiomyopathy, only estimates of related disease and procedures. In our own cost-of-illness analysis of The Healthcare Cost and Utilization Project's data, estimated total costs were almost $120 million (Table 2). This estimate is only a fraction of the cost because it includes only inpatient treatment costs. Initial, diagnostic and outpatient treatment costs add substantially to this total. Another source of cost associated with cardiomyopathy is heart transplant, which we reported above, as $465,494 for the initial year of surgery and follow-up care per child.

Interviews with Clinicians

We interviewed six pediatric cardiologists who practice in some of the major academic medical centers across the United States (personal communication with Charles Canter, MD, Professor of Pediatrics, St. Louis Children's Hospital, Washington University School of Medicine; Steven Colan, MD, Professor of Pediatrics, Children's Hospital Boston, Harvard Medical School; Daphne T. Hsu, MD, Professor of Clinical Pediatrics, Morgan Stanley Children's Hospital of New York-Presbyterian College of Physicians & Surgeons, Columbia University; Paolo Rusconi, MD, Associate Professor of Pediatrics, Holtz Children's Hospital, Leonard M. Miller School of Medicine, University of Miami; Jeffrey A. Towbin, MD, Professor of Pediatrics, Texas Children's Hospital, Baylor College of Medicine, Steven A. Webber, MBChB, Professor of Pediatrics, Children's Hospital of Pittsburgh, University of Pittsburgh School of Medicine. September– October 2007). All interviewees reported having pediatric comprehensive heart failure programs at their institutions. These programs had been operating for 3 to 17 years. Several are financed by the patient's health insurance, and some draw their resources from the sponsoring institution. Additional resources came from non-profit organizations for one program. The components of these programs varied in terms of team composition, but they all included pediatric heart failure and heart transplant specialists. Some of the personnel were not full-time in the programs. Four of the programs routinely measured and recorded health outcomes of their patients. Only one program measured the associated costs of treatment. Although all programs published their clinical research findings, none had described the nature of their program or the overall efficacy. Three programs collected feedback from patients and families, as well as from associated medical personnel. All interviewees identified at least several important elements missing from their programs. Interview highlights are summarized in Table 3.

Local and National Support Groups

To obtain the perspectives of patient advocates and parents, we interviewed the founder and Executive Director of the Children's Cardiomyopathy Foundation (CCF). After experiencing the deaths of her two children from cardiomyopathy, she established the CCF in 2002 to promote research, education, advocacy, awareness, and support for families of children with this disease.[20] Currently, some 580 families are registered with CCF, both in the United States and internationally.

Based on CCF's experience, generally physicians provide excellent care when treating a child with cardiomyopathy, but often the families' need for basic information and practical guidelines for living with a chronic disease is not met. As a result, the family is unsure how to deal with the typical challenges that the disease presents as the child grows older; for example, whether they should speak to a geneticist, nutritionist, or access ancillary services. In the founder's opinion, physicians do not always have the time to discuss these topics of non-urgent care. Consequently, families often go to nurses for this kind of information and support resources, who in turn go to CCF.

Educating families at the beginning of their child's care, when they are the most overwhelmed, should be part of the overall management of the disease. Comprehensive care centers would greatly facilitate this form of multidisciplinary treatment, much like models of care for pediatric cancer and cystic fibrosis. The Cystic Fibrosis Foundation is a good example of how a foundation established a network of care centers that it accredits and funds to help develop these programs.[21] The lack of any support mechanism at the hospital was the driving force for to the establishment of CCF, and the creation of these groups is one of CCF's main activities. At the time of the interview, CCF had facilitated the establishment of support groups in Atlanta, Detroit, Durham, and Boston. The groups are led by CCF-registered families who work closely with their local hospital and appoint a medical advisor and meeting facilitator.

Since its establishment, CCF has recognized the value of collaborating with other disease-specific foundations. When children present with cardiomyopathy as the initial symptom of some underlying, but unknown genetic disorder, CCF tries to get families to the appropriate specialty center to determine the cause of the disease. Once a definitive diagnosis has been made, families may shift to working with other more specialized organizations, such as the Noonan Syndrome Support Group, Barth Foundation, or Parent Project Muscular Dystrophy. However, with only one-third of children with cardiomyopathy receiving a diagnosis, the majority with idiopathic disease remain involved with CCF.[22] (Personal communication with Lisa Yue, the founder and Executive Director of CCF. October 2007.)

To receive additional insight from the nursing perspective into parental support groups, we interviewed a nurse practitioner, who is a strong proponent of such groups (personal communication with Kathleen McGrath, RN, MS, CPNP, Advanced Practice Nurse, Pediatric Cardiology, Golisano Children's Hospital, Assistant Professor of Nursing, University of Rochester. October 2007). As part of a task force for the American Heart Association's Council on Cardiovascular Nursing, she co-authored "Guidelines for Parent Support Groups."[23] Although the guidelines were written focusing on the parents of children with congenital heart diseases, the principles can be applied to other pediatric heart conditions. The document recommends the assessment of parental needs, discusses the types of information and resources these families could find useful, as well as describes how to establish a successful parental support group. When setting up a support program, importance is placed on including physicians, nurses, and social service personnel, in addition to parents and other family members. This brings an important clinical perspective and greatly simplifies any issues that come up in dealing with access to care and therapy options. Once the support group is established it is crucial to develop clear goals specific to the needs of the children and their families, such as emotional support goals, resource information goals, and educational goals. The next step is determining the group's structure and organization, i.e. leadership and the roles of all participants. Finally, evaluation of the group's progress via participant feedback and resolution of relevant issues both greatly improve the effectiveness of the group.

DISCUSSION

The Nature and Costs of Chronic Diseases

Chronic diseases, according to the Centers for Disease Control and Prevention, contribute most heavily to death, illness, and disability.[24] The Institute of Medicine has also commented on the deficiencies in treating patients with chronic disease.[25] These issues have brought about suggestions from researchers on the best method to provide comprehensive care to those suffering from chronic diseases.

Wagner et al.[26] summarized the current concepts in the Chronic Care Model (CCM). The authors argued that the primary reason for the current problems in treating chronic diseases is the mismatch between the patients' needs and care delivery systems that are largely designed for acute illnesses. The CCM integrates all the medical, as well as auxiliary services, necessary for the effective treatment of chronic disease. The CDC focuses on preventable chronic diseases of adulthood, also echoed in the CCM, but do not directly address pediatric chronic diseases. This omission has been attended to by the field of pediatric oncology. Survivors of pediatric cancer have been written about extensively with regard to the long-term effects of initial treatment.[27, 28] The cumulative incidence of chronic health conditions were estimated to be 73% at 30 years of follow-up.[29] Improvements to initial treatment, as well as longer and more comprehensive follow-up management,[30] have been stressed in a disease process with substantial sequelae. A systematic review commissioned by the National Health Service on cost-effective approaches to cardioprotection during chemotherapy[31] identified modifications to administration of anthracyclines and addition of cardioprotectants during therapy[32, 33] as

strategies. Another study[34] showed that cardiac function of childhood cancer survivors, which was depressed directly following doxorubicin therapy, appeared to rebound during the first 6 years of follow up only to decline again with longer follow-up. This result proves that an early recovery is not necessarily a long term recovery and continuing follow up in comprehensive heart failure programs would be worth considering. On a comprehensive level, the Children's Oncology Group collaboratively established guidelines for long-term care published on the internet for easy access.[35] The issues surrounding the development of a care program and the barriers to quality care have been addressed as well.[36, 37]

The Sources of Costs for Pediatric Cardiomyopathy

In a series of manuscripts related to the National Heart Transplant Study,[38–40] Evans catalogued the sources of direct and indirect costs for adult heart transplantation when there was debate about its utilization and management. In a similar fashion, we provide the reader with a review of sources of costs related to pediatric cardiomyopathy.

Most children present with acute congestive heart failure, often to the emergency department, and their families will incur the costs of hospitalization as well as the emergency department visit. The length of stay and the intensity of the diagnostic work up will be the major determinants of cost, and most children will undergo a panel of laboratory tests, electrocardiography, echocardiography, and chest radiography. At certain institutions, variable involvement of non-cardiology consult services and the associated tests add to the costs.

Relative to symptoms and their severity, any of several medications and therapies may be prescribed. After stabilization and discharge, patients complete a series of follow-up visits at the pediatric cardiology clinic, in addition to routine visits to their primary care pediatrician. The diagnostic work-up may continue in outpatient testing centers, in cooperation with medical geneticists. Case-findings in immediate family members may appear on screening echocardiograms for latent disease. The PCMR reported that only one-third of all cases in the Registry had a causal diagnosis for cardiomyopathy.[22] Delayed diagnosis can involve repeated interactions with different medical providers, thus increasing the cost of care.

Some children will regain normal heart function, and their cardiomyopathy may resolve altogether, as seen in a moderate proportion of myocarditis-induced dilated cardiomyopathy cases. The remaining children continue with office visits and intermittent hospitalizations. Their physical functioning may be impaired, further reducing their overall quality of life. Certain children may experience sudden death, and others may deteriorate over various lengths of time. Children who deteriorate slowly enough are often evaluated for heart transplant. The most serious cases may utilize bridge therapies, such as ventricular assist devices or extracorporeal membrane oxygenation. Additionally, families may relocate from great distances to be close to tertiary care centers.

The costs attributed to heart transplantation include those of the hospital stay and the costs of specialists, including transplant surgical teams on both the donor and recipient ends. The follow-up costs of anti-rejection medications and specialist visits, in addition to those with pediatric cardiologists continue, often until death or a second re-transplant is necessary.

The indirect costs of death due to pediatric cardiomyopathy are just as important as those billed for direct medical services. The premature loss of life affects society by losing a potentially productive member, as well as its effect on the child's parents who suffer this loss. Even if the child does not die prematurely, the indirect costs to the children and their families are felt all through the disease course. For the children, the impact on their social and psychological wellbeing is just as important as their physical functioning. These children have their lives disrupted by the same medical interventions which keep them alive. For the family, the loss of

productivity is felt while family members must attend to the frequent outpatient and potentially long inpatient visits. The stress of these events can initiate schisms or further disrupt fragile relationships leading to family strife or divorce. Preliminary investigation of the effect of cardiomyopathy on functional status has shown impaired physical and psycho-social functioning in a substantial proportion of children with cardiomyopathy across five clinical centers.[41] Current work is underway to evaluate this domain in a larger cohort across 10 clinical centers from the PCMR Study Group.

Comprehensive Program Composition

Given the paucity of published reports, there is also uncertainty as to whether these programs should focus solely on cardiomyopathy or if they should be structured as part of a comprehensive program for pediatric heart failure, transplant, or cardiac disease, among several options. This decision will ultimately come down to priorities and resources at the institutional level, as well as the size of the pediatric cardiomyopathy population seen at a given center. For example, in a rural setting where very few children with cardiomyopathy live, there may not be a critical mass of families for a face-to-face support group solely for cardiomyopathy, but there may be enough children dealing with other cardiac diseases or even chronic diseases in general that may benefit the families equally well. There also exists family support groups not specific to any pediatric disease in particular, such as Parent to Parent-USA and Family Voices.[42, 43] These groups may be able to fill this void at the local level, although parents can still affiliate with CCF on the national level. Even in larger centers, we have seen programs caring for children with cardiomyopathy which are integrated into transplant care. The financial realities in a given center may force the sharing of resources, but the goal of improving the care for children and their family through a multidisciplinary approach remains at the core. Nevertheless, we do not know if there is any significant difference in outcomes or family satisfaction between generalized versus specific care program models. These questions should be addressed in future research.

The areas of nutrition[44] and exercise rehabilitation[45] have been addressed in the previous and current issues of Progress in Pediatric Cardiology. They echo the need for research into the efficacy of interventions in children with cardiomyopathy.

Initiating or Formalizing Pediatric Comprehensive Heart Failure Programs

Medical professionals who wish to initiate a comprehensive heart failure program or to formalize an existing program should include input from the hospital administration in this process. In an interview with experienced pediatric administrator the following points were identified. For a new initiative, it is helpful to submit a formal proposal, whose complexity should be commensurate with requested funding. The components may include an executive summary, a description of the initiative, an explanation of its importance, a budget, and a relevant market analysis demonstrating patient source and a comparison to other programs in the area. If applicable, one could use a strengths, weaknesses, opportunities, and threats (SWOT) analysis, a strategic planning tool used for specifying the objective of the project and evaluating the external and internal factors that help or impede achieving the objective. In addition, it is important to address how the initiative could affect health outcomes, improve the efficiency of service delivery, contribute to the mission of the establishment, and fit in with its overall structure.

It is particularly important to define the audience for the proposal. The difference between hospital and academic pediatric departments affect the weight that economic arguments hold. Potential research opportunities and fund utilization should be emphasized when presenting the initiative to academic department administration, while the return on investment should be the focus of discussion when presenting to hospital administration. In both cases, emphasis on

patient care and outcomes is vital. Regardless, administrative assistance is often available in preparation of necessary documents to get the project off the ground in either scenario or when presenting to both. (Personal communication with Dennis L. Harris, Assistant Chairman of Administration, Department of Pediatrics, Leonard M. Miller School of Medicine, University of Miami. September 2007.)

Potential Barriers to Program Standardization

At the 2007 American Heart Association's Scientific Sessions, the informal "Ask the Experts" panel discussion on care of children with heart failure revealed an array of different practice patterns among the approximately 30 attendees. In an informal discussion with some participants, opinions were put forward that training experiences influence future independent patient care in issues that included administration of intravenous immunoglobulin or performance of diagnostic and follow-up biopsies. When discussion of trial data began, the issues similar to those addressed by Gidding in his editorial[46] regarding the Carvedilol Trial[47] reemerged, namely that there is not the same quantity and quality of randomized control trials in pediatric cardiology as compared to adults. One concern is that potential sample size for studies in children is relatively small, however, biases in what is standard of care also act as barriers for multi-site studies. Medical centers find difficultly in agreeing on what constitutes the control treatment. The ethical dilemma arises because optimal outcomes are desired, but equipoise is not viewed in the same context by virtue of training and institutional habits. This is not to say that all care is divorced from evidence, just that the acceptance of certain nuances is not universal. A set of diagnostic algorithms were proposed to assist in identifying cardiomyopathies of genetic origin in a multi-disciplinary effort.[48] Additionally, the International Society of Heart and Lung Transplantation issued the first practice guidelines for pediatric heart failure management based on existing studies and in context of adult guidelines.[49] Focusing on pharmacotherapeutics and devices, the document did not address the ancillary services we have highlighted in this article. Furthermore, it is unclear how extensively either the algorithm for diagnosis or the practice guidelines have been adopted for routine use.

Limitations of the Study

Our review of the literature and cost estimates is clearly not definitive, only suggestive. Ideally, a full economic evaluation of an existing multidisciplinary program would be conducted. Additionally, consensus on composition of comprehensive programs would be established.

Conclusions

Pediatric cardiomyopathy is a chronic disease with substantial negative impacts on children and their families, as well as significant healthcare costs. Advances in medicine and heart transplantation have moved the focus of care from the acute stage of disease to a chronic disease care model. Chronic diseases in children and adult chronic heart failure have been well studied, and their impact on physical and psychosocial health is better understood. Despite years of research and previous calls to action in the pages of this journal,[50, 51] there is limited data for comprehensively managing pediatric cardiomyopathy, and much of the treatment approach is based on experience with adult cardiomyopathy or professional consensus. Given the direct and indirect costs of pediatric cardiomyopathy, the comprehensive multidisciplinary care exemplified in other pediatric chronic diseases and adult heart failure should be extended to pediatric cardiomyopathy.

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Table 1

Survival rates from the Pediatric Cardiomyopathy Registry Data

	DCM			HCM
Survival from	Death	Transplant	Death or Transplant	Death or Transplant
1 year	87%	79%	69%	94%
5 years	83%	70%	54%	90%
10 years	77%	66%	46%	85%

DCM=Dilated cardiomyopathy, HCM=Hypertrophic cardiomyopathy Data from Towbin et al., 2006; Colan et al., 2007.

Table 2

Estimated Inpatient Costs of Pediatric Cardiomyopathy as the Principal Diagnosis*, 2005†

Age range	Number of Discharges		Mean Charges		Aggregate Charges	
years	N	SE	S	SE	S	SE
			Hypertrophic Obstructive Cardiomyopathy (ICD-9 Code 425.1)			
<1	unreliable	unreliable	unreliable	unreliable	unreliable	unreliable
1–17	193	46	88,673	13,581	17,092,916	4,942,355
			Other Primary Cardiomyopathies (ICD-9 Code 425.4)			
<1	205	54	75,557	16,659	18,269,797	6,488,741
1–17	621	147	130,330	20,348	84,202,762	26,732,894
Total	1,019				119,565,475	

SE=Standard Error

*Estimates for all other categories of cardiomyopathy (ICD-9 425) were unreliable.

†Weighted national estimates from the HCUP Nationwide Inpatient Sample (NIS), 2005, Agency for Healthcare Research and Quality (AHRQ), based on data collected by states. Total number of weighted discharges in the US is based on 39,163,834.

Table 3

Interview Highlights with Clinicians on Pediatric Comprehensive Heart Failure Programs

Program components:

- Heart failure specialists
- Heart transplant specialists
- Geneticists
- Specialists in neuromuscular disease
- Social workers
- Financial coordinators
- Pharmacists
- Dietitians
- Psychologists
- Physical and occupational therapists
- Electrophysiologists

Measured and reported health outcomes:

- Transplant success rate
- Number of referrals, hospitalizations, and deaths
- Success of genetic testing in establishing a cause
- Success of enrollment in clinical trials
- Success of different therapies

Missing program components:

- Additional cardiologists, nurse practitioners, psychologists, research coordinators, financial advisors, nutritionists, social workers, research nurses, and data analysts
- Exercise and rehabilitation programs
- Database developed specifically for cardiomyopathy patients at their medical center for local quality assessment and research
- Round-the-clock telephone access
- Community organizations
- Support groups
- Parental-child education on nutrition, exercise, and medication adherence