Cognitive and Psychologic Considerations in Pediatric Heart Failure

SETH A. HOLLANDER, MD,[1] AND EDWARD CALLUS, PhD[2]

Stanford, California and Milan, Italy

ABSTRACT

Because children with heart failure live longer both before and after cardiac transplantation, there is renewed focus on the quality and preservation of their intellectual functioning and psychosocial health. Children with chronic heart failure are at risk for delays in both cognitive development and psychologic functioning, though the extent and permanence of impairment is not well understood. Children with medically managed heart failure have been shown to be at increased risk for anxiety and depression, with a prevalence of emotional disorders similar to that of other children with congenital heart disease. The use of ventricular assist devices as a bridge to transplantation offers both risks and benefits for the preservation of intellectual and emotional function, with an increased risk for ischemic injury to the brain, but offers the advantage of allowing for cognitive stimulation and improved social interactions. A new population of children with heart failure, those outfitted with permanent ventricular assist devices in lieu of cardiac transplantation, may represent a particular risk group regarding social and cognitive function, but as of yet this is not well studied. Early intervention and school accommodations are recommended for those with cognitive, social, or emotional deficits, and brain imaging should be considered for those with persistent difficulties. Whenever possible, patients should be referred to psychologists and developmental specialists with experience in treating this patient population. (*J Cardiac Fail 2014;20:782−785*)

Key Words: Heart failure, cognitive, psychologic, ventricular assist, transplantation.

The impact of congestive heart failure (CHF), ventricular assist device (VAD) placement, and cardiac transplantation on cognitive and social function in adults is well described.[1−6] However, data regarding the effects of chronic ventricular dysfunction on the child's developing brain are limited. Because patients with heart failure live longer, both before and after cardiac transplantation, there is renewed focus on the quality and preservation of their intellectual functioning and psychosocial health. The following review summarizes our current understanding of the cognitive and psychologic issues surrounding children living with heart failure, including those bridged with VADs, for whom the implications of this technology on cognitive and social function remain to be fully understood.

Heart Failure That is Medically Managed

Disorders of Myocyte Function

Primary disorders of myocyte function are the most common cause of heart failure in children, with dilated cardiomyopathy (DCM) alone representing the most common indication for cardiac transplantation in children >10 years old and nearly one-half of transplantations in the infant population.[7,8] Cardiomyopathy is also the most common indication for VAD placement in children.[9] As clinical genetic testing technology improves, the association between cardiomyopathy and underlying metabolic or syndromic disorders is increasingly recognized in patients formerly thought to have idiopathic disease. Recent data suggest that 35% of pediatric patients with DCM or hypertrophic cardiomyopathy have an identifiable inborn error of metabolism, mitochondrial disease, or genetic syndrome.[10] In these patients, intellectual functioning may be impaired as a result of central

From the [1]Department of Pediatrics (Cardiology), Stanford University Medical Center, Stanford, California and [2]Pediatric and Congenital Heart Center, IRCCS Policlinico San Donato University Hospital, Milan, Italy.

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Reprint requests: Seth A. Hollander, MD, Clinical Assistant Professor, Department of Pediatrics (Cardiology), 750 Welch Road, Suite #321, Palo Alto, CA 94304. Tel: (650) 736-8600; Fax: (650) 725-8343. E-mail: seth.hollander@stanford.edu

See page 784 for disclosure information.

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nervous system comorbidities, independently from the severity of cardiac disease. For the remaining majority of patients with idiopathic or familial disease, it is assumed that normal social and cognitive development is achievable.

Because most children with end-stage cardiomyopathy progress to cardiac transplantation, long-term studies evaluating the effects of chronic heart failure on the brain independently from the superimposed neurologic sequelae associated with cardiopulmonary bypass and transplantation are limited. Cognitive impairment in adult patients with CHF, especially in the domains of vigilance and memory, is well described and appears to be reversible with cardiac transplantation.[11,12] Anatomic changes in brain gray matter volume in adults with chronic heart failure have also been reported.[6]

Data in children, however, are limited. Menteer et al reported that 4% of pediatric patients with severe DCM demonstrated cognitive deficits as evidenced by references to poor school performance, and this is likely to be an underestimation.[13] In the only series of its kind in the noncongenital heart disease CHF population, the gray matter atrophy seen in the brains of adults with chronic heart failure was also seen in a small series of 7 children with cardiomyopathy, despite the absence of ischemic and age-related degenerative changes. Although not correlated with clinical evidence of depression or cognitive abnormalities, the insulae, hippocampus, and anterior cingulate regions of the brain, which are responsible for controlling mood and memory, appeared to be disproportionally affected.[14] These findings have yet to be reproduced in a larger population, and an understanding of the relationship between these anatomic abnormalities and neuropsychologic impairment requires further study.

Patients with heart failure are also at increased risk for anxiety and depression, which, in adults, has been linked to an increased risk of hospitalization, arrhythmia, and death.[15−18] Some 44%−60% of children awaiting heart or heart-lung transplants show impaired psychosocial functioning, with 25%−35% of them meeting criteria for the diagnosis of an emotional disorder, a prevalence similar to children undergoing surgery for congenital heart disease (CHD).[19,20] Menteer et al recently reported that children with heart failure have a lower self-reported health-related quality of life related to their heart problems compared with children after heart transplantation; however, the prevalence of depression was lower than that reported for adult heart failure patients.[21] Improvements in psychologic adjustment by 1 year after transplantation also have been reported in the pediatric population, although 20%−24% of patients continue to demonstrate anxiety, depression, and behavior problems despite resolution of their heart failure.[20,22] In the first longitudinal study of its kind, Wray et al compared psychologic adjustment in 28 children and adolescents before heart or heart-lung transplantation and at 6, 12, and 24 months' follow-up.[23] Before transplantation, patients with heart failure demonstrated significantly higher behavioral and psychologic stress than healthy control subjects. By 1 year after transplantation, however, significant improvements in psychologic functioning resulted in no difference between the 2 groups, although a significant minority of children and adolescent still demonstrated psychologic difficulties. In a companion study by the same group, psychologic difficulties in those who did not improve after transplantation persisted up to 2 years after transplantation and appeared to be more prevalent in younger patients.[24] Conversely, Menteer et al studied 62 pediatric patients with DCM and severe heart failure, and although they were found to have a significantly lower health-related quality of life (Peds QL) scores than comparable patients who recently underwent cardiac transplantation, there was no difference in the incidence of depression between the 2 groups or when compared with a group of 24 healthy control subjects.[21]

Heart Failure in the Setting of Congenital Heart Lesions

The deleterious effects of CHD, cardiopulmonary bypass, and prolonged hospitalization following cardiac surgery on social and cognitive development are well described.[25−27] Although some patients with CHD also develop CHF, there are no studies that show that patients with CHD and CHF fare worse neuropsychologically than those with CHD alone. Given the increasing population of children and young adults with CHD proceeding to end-stage heart failure and cardiac transplantation, this subject requires further study.

Heart Failure Managed With a Ventricular Assist Device

VADs as a Bridge to Transplant

The use of VADs as a bridge to transplant is increasing. In 2010, 25% of pediatric heart transplant patients were supported with a mechanical support device.[28] Although the increased use of VADs as a bridge to transplant poses an increased risk of neurologic injury, it also has correlated with a dramatic reduction in extracorporeal membrane oxygenation (ECMO) use from 9.4% to 2.6% in recent years.[7] Despite the relatively high risk for stroke in pediatric VAD patients, including a 29% incidence of neurologic injury in children supported with the Excor Berlin Heart, the use of VADs rather than ECMO as the primary means of mechanical support likely reduces the likelihood of neurologic injury in long-term supported patients.[29] Moreover, the use of VADs in place of ECMO facilitates social and cognitive development in children awaiting transplant by allowing them to be awake, interactive, and participating in daily activities, including hospital school attendance.[30]

Most pediatric patients supported by a VAD proceed to cardiac transplantation, and the limited data available regarding the neurocognitive effects of VAD support often reflect the combined effect of both VAD support and transplantation. In 2012, Stein et al reported that 89% of patients bridged to transplant with a pulsatile VAD performed in the average range or above for general intellectual functioning, and slightly better than a control group of children receiving transplants without being bridged