Advanced Circulatory Support Workshop

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Introduction

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The past decade, and especially the past five years has witnessed an evolving understanding of heart failure in pediatric and adult patients. Most therapies were originally directed at improving cardiac output and reducing fluid overload. However, no adult or pediatric studies have shown long-term benefit from positive iontropic agents, and therapies today are directed at reducing ventricular remodeling. The new adult model of heart failure is a progressive disease that evolves where after an index event which damages the heart muscle and disrupts the heart muscle's ability to generate force. Compensatory mechanisms ensue: sympathetic activation, activation of salt and water retaining mechanisms and activation of vasodilatory molecules, initially keeping the patient from becoming symptomatic. However, patients ultimately transition to overtly symptomatic heart failure as these mechanisms, once considered compensatory, ultimately become toxic. Neurohormonal and cytokine activation lead to ventricular remodeling with alterations in myocyte molecular biology and cellular composition, as well as changes in left ventricular chamber geometry, all of which contribute to the deterioration in cardiac function. This has proven to be an excellent working model for adult patients. The first class of drugs shown to improve symptoms, improve long-term survival and reduce ventricular remodeling in adults was angiotensin converting enzyme (ACE) inhibitors. Subsequent multicenter studies have shown similar benefits from the use of beta blockers, spinolocatone, and angiotensin II blockers. Adult studies have also shown no benefit, and even reduced survival from higher serum levels of Digoxin. It remains unclear whether pediatric patients with heart failure will benefit from these same therapies. There is a wide spectrum of heart failure etiologies in children. Most children have structural abnormalities that may be surgically corrected or palliated. And there are increasing numbers of single ventricle patients, many of whom have a single right ventricle, who progress to heart failure. It is clear that both genomic and endocrine markers will be used in the future to more accurately define patients with heart failure. Brain naturetic peptide (BNP) is a useful plasma marker of heart failure in adults, and has recently been found to be elevated in children with heart failure, even without evidence of systolic dysfunction. Stem cell injections may improve reduce or reverse myocardial remodeling. Recently published studies of adult patients receiving stem cell injections into the myocardium produced improved revascularization. Today, there is a better understanding of receptor physiology and the potential harmful effect of chronic or high-dose catecholamine stimulation. Finally, there is a great need for continued refinement in the treatment of heart failure because of the relatively poor outcomes after heart transplantation in children. The Pediatric Heart Transplant Study Group recently reported that almost one-third of pediatric heart transplant recipients experience graft failure within 8 years of transplantation.

During this workshop we will focus on new modalities for assessing myocardial performance and on new therapies for advanced circulatory support. We will mostly focus on therapies for acute treatment of circulatory failure like mechanical support of the circulation, new pharmacologic agents, and new pacing modalities.

New NONINVASIVE Modalities for Assessing MYOCARDIAL PERFORMANCE

Wanda C. Miller-Hance, MD

Educational Objectives:

This section of the workshop will focus on:

1. Demonstration of how the new echocardiographic indices of myocardial function are derived.
2. Discussion of the clinical applications of echo-Doppler modalities and magnetic resonance imaging in the functional assessment of the failing heart in the pediatric age group.

Echocardiography:

Myocardial Performance Index

The myocardial performance index (MPI or Tei Index) is a useful, non-invasive, Doppler-derived tool that can be used to assess global left ventricular function. This parameter incorporates aspects of both systolic and diastolic function. The performance index is the ratio of the sum of iso-volumetric contraction and relaxation times over the ejection time.

MPI= (ICT + IRT)/ET

ICT, isovolumic contraction time

ET, ejection time

IRT, isovolumic relaxation time

This Doppler-derived index is considered to be an accurate parameter in the echocardiographic assessment of left ventricular contractility. A benefit of this measurement is that it is not affected by loading conditions. Prolongation of the myocardial performance index has been shown to correlate with global impairment of left ventricular function.

Doppler Tissue Imaging and Strain Rate Imaging

Several emerging echocardiographic techniques offer the potential for an enhanced quantitative assessment of myocardial performance. New methods that provide for measurements of global and regional left and right ventricular function include Doppler tissue imaging and strain rate imaging.

Doppler tissue imaging (DTI) enables quantitative assessment of myocardial tissue velocities. This technique relies on the direct interrogation of specific regions of the myocardium by pulse wave Doppler thus allowing for evaluation of tissue motion throughout the various phases of the cardiac cycle.

Strain relates to the deformation of a myocardial tissue segment over the cardiac cycle whereas strain rate measures the rate of deformation.

Magnetic Resonance Imaging:

Magnetic resonance imaging provides accurate and reproducible images with high spatial and temporal resolution of cardiovascular structures. These capabilities have stimulated the enthusiasm in the applications of this modality to the evaluation of cardiac function.

The ability of this technique to acquire images in any desired plane offer huge advantages over standard imaging approaches and allows for direct visualization of global and regional function. The images generated not only provide for a rapid qualitative assessment but permit an accurate and reproducible quantitative evaluation of myocardial performance.

New Pharmacologic Therapies

Avinash Shukla, MD

1. Vasopressin
2. Prostacyclin and Sildenafil
3. T3
4. Nesiritide (BNP)
5. Levosimendan
6. Fenoldapam

New Electrophysiologic modalities for managing Cardiac dysFunction

Wanda C. Miller-Hance, MD

Educational Objectives:

This section of the workshop will address the following topics:

1. Rationale for cardiac resynchronization therapy.
2. Review of clinical trials.
3. Application of this therapeutic modality to pediatric heart failure.

A relatively common feature in chronic heart failure is abnomal ventricular electrical activation. Alterations in the conduction pathways result in delay in the onset of right or left ventricular systole. The ventricular dyssynchrony is manifest on the electrocardiogram as prolongation of the QRS duration. It has been suggested that the intraventricular conduction delay may further impair the ability of the failing heart to generate an adequate stroke volume and at the same time may increase the severity of mitral regurgitation. The finding of an intraventricular conduction delay has been associated with increasing degrees of cardiac dysfunction and an increased risk of death in patients with advanced heart failure.

Cardiac resynchronization therapy (CRT) consists of a pacing modality in which a left ventricular pacing lead placed via the coronary sinus is used in conjunction with a right ventricular lead to "resynchronize" the heart. The goal of this approach is to ensure stimulation of the left ventricular myocardium at or near the time of right ventricular depolarization. Benefits of CRT include improved contractile function, reduced myocardial oxygen consumption and reversal of deleterious remodeling associated with chronic heart failure. A number of clinical trials have documented improvements in functional class and quality of life in patients receiving CRT.

Bridging to recovery or transplant: Mechanical support of the circulation in infants and children

Laura K. Diaz, MD

A. ECMO and pediatric heart disease: Indications and outcomes

B. Ventricular assist devices: Indications and outcomes

   1. Centrifugal pumps
   2. Pulsatile pumps
      - Thoratec
      - Berlin Heart
      - MEDOS HIA-VAD
   3. Axial pumps

C. Future trends in mechanical support

D. Recovery from myocarditis: dystrophin studies

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