The CVDY Council once again had a spectacular program at Scientific Sessions 2009 in Orlando, Fla. The sessions began with the annual Taussig Lecture presented by Dr. Andrew Reddington, followed by the presentation of the Outstanding Research Awards in Pediatric Cardiology. These went to the top three abstracts that were received for this year’s conference. It is significant to note that all three winning abstracts were the result of work performed by large multiinstitutional, collaborative groups and two of them involved randomized, prospective clinical trials. In addition, at this year’s Scientific Sessions, Dr. Jane Newburger was honored with both the AHA’s Clinical Research Prize and the Cardiovascular Disease in the Young Meritorious Achievement Award (presented by the Council). In addition, Dr. Gail Pearson, who has been instrumental in supporting clinical investigation relevant to congenital heart disease at the NIH, received the Cardiovascular Disease in the Young Distinguished Achievement Award.

As always, I invite input from you, the membership, to let us know what issues we should be addressing in our Council. If you are interested in becoming more involved in CVDY and its committees, please let me know. As always, I encourage those of you who have been longtime AHA members and volunteers to consider applying to become a Fellow of the American Heart Association.

The Congenital Cardiac Defects Committee and Adult Congenital Heart Disease Committee have been very active in several areas of patient/parent education and peer-reviewed scientific statements. Our most significant publications include the print and Web version of “If Your Child Has a Congenital Heart Defect” and the Web version of “Adults with Congenital Heart Disease.” These booklets are updated and have wonderful new pictures which will serve as important resources for patients and families. There are several ways to get to this information including heart.org/chd, heart.org/chdbooklet, and heart.org/achd. In the coming months we will be updating and expanding the Web site for fetal cardiac care, translating our new adult and pediatric content into Spanish, and reorganizing and updating the “FAQs from the Pediatric Cardiologist” section.

The AHOY Committee has historically been one of the most active committees of the CVDY Council. In the last year the committee has published three scientific statements and three more are in progress (one is in the process of writing and two others are awaiting MOC commissioning). At Scientific Sessions 2009, the committee discussed the possibility of developing a physician education program in preventive cardiology for children, using as a model the Adult Preventive Program. It also began efforts to respond to a request of the NHLBI Working Group on Obesity and Other CV Risk Factors in Congenital Heart Disease to produce guidelines for promotion of physical activity for children with congenital heart disease.

The committee also suggested programs to be submitted for Scientific Sessions 2010. Additional discussion addressed the collaboration of AHOY with the Healthcare Pillar of the Alliance for a Healthier Generation (a program of the Clinton Foundation and Robert Wood Johnson Foundation for prevention and treatment of childhood obesity). The Alliance Healthcare Initiative, which receives input from AHOY, has made major progress in ensuring that children get the insurance coverage they need to fight obesity. The committee members were energized by this progress and will continue advocacy efforts both regionally and nationally.

My colleagues serving on the CVDY Membership/Communications/Newsletter Committee, Dr. Arthur Pickoff (arthur.pickoff@wright.edu) and Dr. Ashwin Prakash (ashwin.prakash@cardio.chboston.org) and I look forward to receiving input and newsletter content from CVDY Council members. We ask for your suggestions. Please send requests for future content and any advice for improving communication and enhancing CVDY membership. We ask also that you give serious consideration to becoming an author of future content. Feel free to contact me if you have any questions or comments.
Committee on Rheumatic Fever, Endocarditis, and Kawasaki Disease (RFEKD)

Recent and ongoing activities of the committee address a variety of topics. A scientific statement was recently completed and will be published in Circulation in January 2010 that addresses cardiovascular implantable electronic device infections. This work is cosponsored by the Heart Rhythm Society and is the first iteration of the document.

The committee has developed an outline for a paper that examines data that purportedly links periodontal disease to the development of coronary artery disease. A systematic review is underway and the subsequent writing of a manuscript is underway with a writing group defined and tasks assigned.

Subsequent discussions within the group will review what role echocardiography has in the Jones criteria for the diagnosis of rheumatic fever and the need for updating related statements from the AHA.

Program Committee

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Seema Mital, MD, FAHA, FRCP(C)

Scientific Sessions 2009 in Orlando, Fla. was an exciting and well-attended meeting with a new format that was very well received. The attendees had an opportunity to hear exciting new results from leading researchers, including updates on basic, clinical and translational research in heart disease in the young as well as adults with congenital heart disease. The International Congress on Genetics and Genomics of Cardiovascular Disease intersected well with the other core sessions and was very well attended.

Dr. Andrew Redington from the Hospital for Sick Children gave the Helen B. Taussig Memorial Lecture: “Taussig, Tetralogy and Turmoil: From Childhood to Adulthood and Back.” At the CVDY Council dinner, Dr. Perri Klass, a pediatrician, journalist and writer from New York, gave the William J. Rashkind Memorial Lecture, entitled “A World Full of Stories: Doctors as Readers, Writers and Characters.” Dr. Jane Newburger was awarded the AHA Clinical Research Prize and the Meritorious Achievement Award and Dr. Gail Pearson received the Distinguished Scientist Award.

The Outstanding Research Awards session showcased the results of two major clinical trials performed by the Pediatric Heart Network as well as a multiinstitutional study in adults with congenital heart disease. The recipients were:

- Richard Ohye, Ann Arbor, MI — Outcomes of the Norwood Operation in Infants Randomized to a Modified Blalock-Taussig versus Right Ventricle-to-Pulmonary Artery Shunt: The Pediatric Heart Network Single Ventricle Reconstruction Trial
- George Lui, New York, NY — Heart Rate Response as a Predictor of Pregnancy Outcome in Women with Congenital Heart Disease
- Daphne Hsu, Montefiore Children’s Hospital, NY — Enalapril Does Not Improve Growth or Ventricular Function in Infants with Single Ventricle: A Multicenter Clinical Trial

The Program Committee for 2009–10 includes Seema Mital (Chair), Wolfgang Radtke (Vice Chair), William Mahle, Andrew Atz, Charles Canter, Macdonald Dick and Jonathan Chen. The committee requests that members submit suggestions for topics and speakers for Scientific Sessions 2010 to be held in Chicago. We look forward to another exciting program next year.

Additional evaluation is planned to determine if revised follow-up recommendations are needed for patients with Kawasaki disease.

The committee plans additional discussion regarding the development of a statement that reviews peripheral vascular infections. In addition, an update of the endocarditis management document which was last published in 2005 is warranted.

Science Recap

William Mahle, MD, FAHA

Sessions 2009 provided the venue for the most recent and important clinical, translational and basic research in pediatric cardiology and congenital heart disease.

The scientific program saw the first presentation of a number of randomized trials in children that will no doubt be seminal events.

Results of the Single Ventricle Reconstruction (SVR) Trial, sponsored by the Pediatric Heart Network, were presented by Dr. Rick Ohye from C.S. Mott Children’s Hospital. This randomized trial of 555 children compared two types of shunts as part of the Norwood operation for hypoplastic left heart syndrome. The primary outcome measure, freedom from death or transplantation at 12 months of age, was 68.7 percent. Children who received a right ventricular-to-pulmonary artery shunt had a significantly greater freedom from death or transplantation, 73.6 percent vs. 63.3 percent in the children who received a modified Blalock-Taussig shunt.

The differences between the two groups became less prominent with longer follow-up, suggesting some
intermediate-term survival benefit with the modified Blalock-Taussig shunt. The study did not identify any difference in length of stay or other markers of morbidity at the time of the Stage I operation. There was a higher rate of unintended cardiovascular procedures in children who underwent right ventricle-to-pulmonary artery shunt. At 14 months, there was no difference between the groups in right ventricular function or dimension as assessed by echocardiography.

This first-ever randomized trial of congenital heart surgery demonstrated an early benefit in children undergoing a right ventricle-to-pulmonary artery conduit. Intermediate-term follow-up of this cohort should provide important insights into the outcome of hypoplastic left heart syndrome patients.

The Infant with Single Ventricle (ISV) Trial, also supported by the Pediatric Heart Network, was presented by Dr. Daphne Hsu from Montefiore Medical Center. This study compared administration of enalapril to placebo in infants with single ventricle to determine whether ACE inhibitor therapy might improve growth, measures of ventricular function and geometry, or neurodevelopmental status over a 14-month period. Of 230 children randomized, 182 completed the analysis. There was no difference with respect to weight for age, left ventricular mass or volume, or other measures of cardiovascular status including BNP levels and Ross heart failure scores. The entire cohort had lower developmental test scores, but again, there was no difference between the treatment and placebo groups.

This study is important given previous reports that up to 60 percent of patients with functional single ventricle receive ACE inhibitors. It may be that there are other populations for whom ACE therapy is beneficial, but at this point, the strength of a randomized trial will help guide therapy in the near future.

Multicenter adult congenital consortium data presented by Dr. George Lui from Columbia-Presbyterian Medical Center examined exercise testing and outcomes of pregnancy in women with congenital heart disease. Data from eight adult congenital heart centers in North America demonstrated that adverse cardiac events during pregnancy were common, occurring in 18 percent. The chronotropic index was an important predictor of outcomes: patients with a higher index had fewer adverse events during pregnancy. While the mechanism is not entirely understood, it underscores the importance of risk-stratification for young women with congenital heart disease considering pregnancy.

Sessions focused on a number of key areas in clinical and translational medicine.

There was keen interest in the potential role of glucose management around the time of cardiac surgery.

A large study, of over 700 infants from a single institution, presented by Dr. Li and colleagues from Beijing, China, reported the impact of hypo- and hyperglycemia on infants undergoing congenital heart surgery. Low serum glucose following open-heart surgery was associated with higher risk of complications after surgery. There seemed to be a U-shaped relationship between serum glucose levels in the postoperative period and morbidity. Very high serum glucose levels, >250 mg/dL, were associated with increased risk of longer intensive care unit stays.

A study from the Hospital for Sick Children in Toronto presented by Dr. Moga and colleagues similarly suggested that hyperglycemia was an important factor in adverse events following congenital heart surgery. When paired with residual lesions, patients were at greatest risk. Moderate hyperglycemia (>180 mg/dL) was associated with the highest risk for adverse outcome.

One intriguing area of interest in congenital heart care is the impact of gestational age on outcomes. A provocative study by Costello et al. suggested an adverse impact to children born at less than 39 weeks compared to those who are born in the range of 39 to 41 weeks gestation. Children born before 39 weeks gestation tended to have at least a two-fold increase in adjusted mortality, as well as longer duration of mechanical ventilation. Gestational age had a U-shaped relationship with mortality, as children born beyond 41 weeks also had increased risk. This seminal study should help guide plans for perinatal care. It will be important to avoid early delivery for convenience’ sake, and to emphasize the benefits of term delivery when possible.

Several studies focused on the aortopulmonary collaterals in the setting of functional single ventricle.

Using MRI, Whitehead and colleagues found that patients after a bidirectional Glenn procedure continue to have a large proportion of pulmonary blood flow (>50 percent) supplied by aortopulmonary collaterals and suggest that the so-called “ventricular volume unloading” following a Glenn may be misleading. Once the Fontan operation has occurred, some aortopulmonary collaterals resolve, so perhaps there may be a benefit to completing the Fontan at a relatively early age.

There has been interest in coil embolization of aortopulmonary collaterals in patients with single ventricle. Using data from a previous Pediatric Heart Network study, Banka et al. failed to demonstrate an association between the practice of embolization of aortopulmonary collaterals and later functional outcomes.

Thacker and colleagues presented the use of Budesonide in the treatment of protein-losing enteropathy. Administering open-label oral Budesonide to children and adolescents improved serum albumin and reduced symptoms. While it has been known for years that systemic corticosteroids can improve symptoms, side effects have generally precluded long-term therapy. This novel approach, using an oral corticosteroid which is 90 percent metabolized in the liver upon first pass, holds promise.

Dr. Zeisberg from Children’s Hospital, Boston presented elegant data regarding the development of endocardial fibroelastosis (EFE). Using a mouse model, as well as analysis of resected tissue from children with EFE, they demonstrated that endothelial to mesenchymal transition, a mechanism by which endothelial cells can acquire a mesenchymal phenotype, is a critical component in the development of EFE.