

Editorial

Open Access



Preface to insights into congenital heart disease

P. Syamasundar Rao

Children's Heart Institute, University of Texas at Houston, McGovern Medical School and Children's Memorial Hermann Hospital, Houston, Texas 77030, USA.

Correspondence to: Prof. P. Syamasundar Rao, Children's Heart Institute, University of Texas at Houston, McGovern Medical School and Children's Memorial Hermann Hospital, 6410 Fannin Street, UTPB Suite # 425, Houston, Texas 77030, USA.
E-mail: P.Syamasundar.Rao@uth.tmc.edu

How to cite this article: Rao PS. Preface to insights into congenital heart disease. *Vessel Plus* 2022;6:35.
<https://dx.doi.org/10.20517/2574-1209.2021.109>

Received: 3 Aug 2021 **Accepted:** 10 Feb 2022 **Published:** 2 Jun 2022

Academic Editor: Alexander D. Verin **Copy Editor:** Xi-Jun Chen **Production Editor:** Xi-Jun Chen

Early detection of neonates with serious heart disease, including pulse oximetry screening and their rapid transport to tertiary care centers; availability of highly sensitive non-invasive diagnostic tools such as echo-Doppler studies, magnetic resonance imaging (MRI), computed tomography (CT); advances in transcatheter interventional procedures; improvements in pediatric cardiac anesthesia; and extension of complicated surgical procedures to treat children with simple and complex congenital heart defects (CHDs) have resulted in successful management of all children with heart disease. Consequently, almost all CHDs are diagnosed and "corrected". Defects that cannot be completely corrected can be effectively palliated. The last five decades have seen a great many advances as mentioned above, which resulted in the increased survival of children with CHD. The purpose of the Special Issue on "Insights into Congenital Heart Disease: Diagnosis and Management" is to present some of these advances in an easily readable format for the physicians interested in the care of infants, children, and adults with heart disease.

In the first paper^[1], I discussed issues related to the diagnosis of cardiac murmurs in children. Cardiac murmur is frequently heard on auscultation. Murmur is the frequent reason for the recognition of heart disease in children (with the exception of neonates). Mastery of skills of auscultation acquired by training and experience is important in diagnosing the causes of cardiac murmurs. Cardiac patient simulators and computer-assisted training methods have been used to educate students and residents; these methods should supplement bedside acquisition of auscultatory skills under the supervision of experienced clinicians and not become primary modes of training of our emerging physician pool. Murmurs are classified into



© The Author(s) 2022. **Open Access** This article is licensed under a Creative Commons Attribution 4.0 International License (<https://creativecommons.org/licenses/by/4.0/>), which permits unrestricted use, sharing, adaptation, distribution and reproduction in any medium or format, for any purpose, even commercially, as long as you give appropriate credit to the original author(s) and the source, provide a link to the Creative Commons license, and indicate if changes were made.



systolic, diastolic, and continuous types. The systolic murmurs are further divided into ejection systolic and holosystolic murmurs. The more common etiologies of ejection systolic murmurs are aortic stenosis, pulmonary stenosis, atrial septal defect (ASD), coarctation of the aorta, and functional heart murmurs. The causes of holosystolic murmurs are ventricular septal defect (VSD), mitral insufficiency (MI), and tricuspid insufficiency (TI). The diastolic murmurs are classified into early, mid and late (or presystolic) diastolic murmurs. The early diastolic murmurs are caused by aortic insufficiency (AI), pulmonary insufficiency, and pulmonary hypertension. Mid-diastolic murmurs are produced by increased flow across the mitral valve [secondary to large shunts across a VSD or patent ductus arteriosus (PDA) or moderate to severe MI] or increased flow across the tricuspid valve (due to ASD, partial or total anomalous pulmonary venous connection or moderate to severe TI). Other causes are Carey-Coombs murmur of rheumatic fever, Austin-Flint murmur of AI, and stenosis of the atrio-ventricular valves. The presystolic murmurs are produced by stenosis of the mitral or tricuspid valve and atrial myxoma. The continuous murmurs are more commonly produced by PDA, venous hum, or aorto-pulmonary shunt procedures. There are many other less common causes. Careful auscultation and other findings in history, physical examination, chest roentgenogram, and electrocardiogram will frequently help come up with an accurate diagnosis. Echo-Doppler studies are valuable and confirmatory in making the diagnosis, in quantifying the problem, and are very useful in directing the type of and timing of management.

In the next paper, Dr. Uppu^[2] from our institution described imaging of the defects in the atrial and ventricular septae. He stated that ASDs and VSDs account for the majority of the CHDs, and that these defects may be seen as isolated lesions or may occur in association with other defects. Clinical features are largely related to the size of the defect. ASDs produce right ventricular volume overload, while VSDs cause left heart volume overload. Initially, he reviewed the embryology of each defect separately, followed by a detailed anatomic description. Four types of ASDs, namely, ostium secundum, ostium primum, sinus venosus, and coronary sinus defects, were reviewed, presenting the location of the defect in beautifully drawn colorful heart models and then showed echocardiographic, MRI, and CT examples. Then, he reviewed all four types VSDs, i.e., perimembranous, inlet, muscular, and outlet VSDs demonstrating the echocardiographic features of each of these defects. He concluded that the understanding of embryology and anatomy is essential for correct diagnosis and planning for transcatheter and/or surgical interventions.

In the following paper, Ivy *et al.*^[3] from the University of the Incarnate Word School of Osteopathic Medicine, San Antonio, and the Children's Hospital of San Antonio, San Antonio, Texas, reviewed the role of three-dimensional (3D) visualization modalities [augmented reality (AR), virtual reality (VR) and 3D printing] for CHD surgery. They discussed the utility of 3D visualization modalities such as 3D printing, AR, and VR, and suggested that these techniques have changed the discipline of surgery in the last few years. These techniques have confirmed their value in planning preoperatively, in procedural training, and in guidance during surgery in many surgical spheres. They explored the existing applications of 3D visualization techniques in surgery and investigated their utility and potential in the sub-discipline of surgery for CHDs. The authors acknowledged limitations of these techniques, namely, high initial set-up costs, need for ready access to imaging technologies like CT or MRI, and availability of reconstruction tools. The authors concluded that all three modalities had been found to be useful in surgery in that they help the surgeons in pre-procedure planning, reduce postoperative complications, curtail the surgical duration, and advance the critical patient-physician relationship. Finally, they suggested that existing evidence indicates that these new imaging modalities are likely to result in better patient outcomes.

In the subsequent three papers, I presented an overview of echocardiography. In Part I^[4] of this review, I described principles of echocardiography and Doppler, outlined the technique of recording echo-Doppler

studies, presented methods of estimation of pulmonary artery pressure, reviewed methodology for evaluation of ventricular (left, right, and single) function, and demonstrated the usefulness of echo in evaluating multiple neonatal issues, including distressed neonate, infant of a diabetic mother, tracheo-esophageal fistula, Down syndrome, and cardiomegaly. Part II^[5] of this review described echocardiographic features of commonly encountered acyanotic CHDs. Echo-Doppler features of obstructive lesions, namely aortic stenosis, coarctation of the aorta, pulmonary stenosis, and branch pulmonary artery stenosis, were first presented, followed by a discussion of left-to-right shunt lesions, namely, ASD, patent foramen ovale, VSD, PDA, and atrio-ventricular septal defect. A special section dedicated to evaluating hemodynamically significant PDAs in premature infants was also included. Both congenital and acquired coronary artery anomalies were reviewed at the conclusion of the paper. Part III^[6] of this review described echocardiographic features of commonly encountered cyanotic CHDs. Echo-Doppler characteristics of more commonly seen defects such as tetralogy of Fallot, transposition of the great arteries, tricuspid atresia, total anomalous pulmonary venous connection, truncus arteriosus, and hypoplastic left heart syndrome were first reviewed. Then, less commonly observed lesions such as double-outlet right ventricle, double-inlet left ventricle, interrupted aortic arch, pulmonary atresia with intact ventricular septum, congenital corrected transposition of the great arteries, Ebstein's anomaly of the tricuspid valve, and mitral atresia with normal aortic root were reviewed.

In the next paper, Dr. Misra *et al.*^[7] of Children's Hospital of Michigan, Detroit, Michigan, detailed multimodality imaging of tetralogy of Fallot (TOF) with a focus on postoperative residua. They discussed the role of echocardiography, cardiac MRI, cardiac catheterization and cineangiography, and nuclear scan in this evaluation, along with the advantages and limitations of each technique. They suggested continued surveillance of post-operate TOF patients because of the existence of residual anatomic and hemodynamic abnormalities and the potential need for re-intervention, including percutaneous pulmonary valve replacement. They also stated that the afore-mentioned investigative methods are not exclusive of each other but are complementary to one another. They opined that echocardiography is useful in evaluating patients who are younger than 10 years of age. Beyond that age, the selection of the type of imaging modality used is largely dependent upon the clinical scenario, issues to be addressed, and institutional expertise while taking into consideration risks of the procedures such as radiation exposure and risk of cancer. The authors also provided several illustrations demonstrating features of residua following surgical correction of TOF. They concluded that the above investigative tools form the foundation of continuing care and surveillance of surgically repaired TOF patients.

In a subsequent paper, Dr. Misra *et al.*^[8] of Children's Hospital of Michigan, Detroit, Michigan, described cardiovascular involvement in multisystem inflammatory syndrome in children with COVID-19. The authors stated that SARS-CoV-2 infections in children usually have less severe COVID-19 infections than those seen in adults. Nevertheless, some children may exhibit severe multisystem inflammatory disease. The Center for Disease Control has named this entity a "Multisystem Inflammatory Syndrome in Children (MIS-C)". MIS-C results in important cardiovascular disease; the degree of cardiac involvement determines the clinical course and eventual outcome. The presenting findings of MIS-C are constant fever, gastrointestinal symptoms, polymorphic rash, conjunctivitis, and mucosal changes, somewhat similar to those seen in Kawasaki syndrome. The inflammatory markers are often elevated. A subset of MIS-C patients may present with hypotension and shock; this may be related to acute myocardial dysfunction or dilation of the systemic vasculature. The cardiac involvement includes myocarditis, pericarditis, valvulitis, coronary arteritis. Arrhythmias have also been documented. The authors reported the experience with MIS-C in 54 patients seen during the last year at their institution. They also reviewed cases reported in the recent literature, and the clinical protocol to be used in the diagnosis and management of MIS-C secondary to

COVID-19. The authors concluded that prompt identification and appropriate treatment might lead to satisfactory outcomes.

In the next paper, Drs. Saxena and Relan^[9] from All India Institute of Medical Sciences, New Delhi, India, reviewed issues related to pregnancy with CHD. Following the introduction of the subject, the authors described hemodynamic changes that occur during pregnancy as well as those that take place during delivery and the postpartum period. They indicated that the physiological changes that occur during pregnancy are additive to the already existing hemodynamic burden associated with CHD. They suggested that all women with CHD should be assessed for their risk of pregnancy and counseled prior to becoming pregnant; these should include risks both to the mother and fetus. They recommended using the modified World Health Organization's scoring system, the Cardiac Disease in Pregnancy (CARPREG) scores, for this assessment. They listed these scoring systems in nicely organized tables. This is followed by a discussion of the diagnosis and management of CHD during pregnancy. Discussion of management, including timing and mode of delivery, peripartum care, and surgical and transcatheter interventions for CHD during pregnancy. Then, they presented a review of the management of common complications seen in women with CHD, including heart failure, arrhythmias, bleeding/thrombosis (anticoagulation), and infective endocarditis. They addressed the management of specific CHDs, including Eisenmenger syndrome, CHD associated with pulmonary stenosis, left ventricular outflow tract obstruction, coarctation of aorta, residua following Fontan surgery, surgically repaired transposition of great arteries, and other congenital heart diseases. They concluded that advances in existing practices to assess and manage women with CHD might result in considerable improvement of outcomes both for the mother and fetus.

In the paper that follows, Drs. Sahulee and McKinstry^[10] from NYU Grossman School of Medicine, New York, NY, reviewed pharmacological management of low cardiac output syndrome (LCOS) after cardiac surgery in children. After defining the LCOS following cardiac surgery under cardiopulmonary bypass, the authors described its prevalence, pathophysiology, and available therapeutic options. Optimization of preload, maintenance of atrio-ventricular synchrony, methods to decrease afterload, and administration of pharmacologic agents to augment cardiac output are among the major therapeutic approaches to achieve a balance between tissue oxygen delivery and demand. Then, they presented a discussion of catecholaminergic inotropes (epinephrine, dopamine, and dobutamine), inodilators (milrinone and levosimendan), systemic vasodilators (sodium nitroprusside, nitroglycerine, nicardipine, nesiritide, phenoxybenzamine), and pulmonary vasodilators (inhaled nitric oxide, iloprost, citrulline) along with their relative merits in treating LCOS. Then, they reviewed adjunctive therapies using corticosteroids, thyroxine, vasopressin, and norepinephrine in the management of LCOS. Finally, they suggested that extracorporeal membrane oxygenation for patients who are refractory to medical management is detailed in the preceding sections. They concluded that there is no single pharmacologic agent that is useful in all patients to treat or prevent LCOS, that milrinone, epinephrine, and dopamine are the most frequently used pharmacologic agents at the present time, and meticulous multicenter clinical trials are required to determine the best option to manage LCOS.

In the final paper, Drs. Wang-Giuffre and Doshi^[11], also from our institution, discussed Cardiopulmonary Exercise Test (CPET) prognostic measures following Fontan operation. The authors initially reviewed the physiology of exercise in patients with Fontan circulation, stressing the importance of factors such as preload dependency, ventricular compliance, atrio-ventricular valve regurgitation, chronotropic competence, arrhythmias, peripheral vascular dysfunction, and abnormal ventilatory function. Then, they discussed cardiopulmonary exercise measures, namely, VO₂ peak (oxygen consumption at peak of exercise), submaximal measures (VO₂ at anaerobic threshold, O₂ uptake efficiency slope, and exercise

oscillatory ventilation), O₂ pulse, respiratory efficiency and spirometry, heart rate reserve, and chronotropic index. They suggested periodic evaluation of maximal metabolic measures, specifically, VO₂ peak, VO₂ at anaerobic threshold, oxygen pulse, oxygen uptake efficiency slope, and ventilatory efficiency, to identify subclinical alterations. The authors concluded that CPET is a key means for the surveillance of post-Fontan patients and that CPET, along with data from clinical, non-invasive imaging, and cardiac catheterization/angiographic information, are helpful in determining the need for and timing of re-intervention.

As one can see, there was a preponderance of papers from the University of Texas Health Science Center, my institution. It was not my intention to load up the manuscripts from my institution; the Journal's editorial staff and I have invited more than 100 physicians involved in the care of patients with CHD, and we received responses only from physicians currently contributing to this issue. It was not possible to include all the advances that occurred over the last few decades; only selected topics were included because of limitations of space. The authors of this Special Issue and I hope that we were able to pass on the knowledge of some of the diagnostic and therapeutic techniques to the interested reader and that these are helpful in caring for their patients with heart disease.

DECLARATIONS

Authors' contributions

The author contributed solely to the article.

Availability of data and materials

Not applicable.

Financial support and sponsorship

None.

Conflicts of interest

The author declared that there are no conflicts of interest.

Ethical approval and consent to participate

Not applicable.

Consent for publication

Not applicable.

Copyright

© The Author(s) 2022.

REFERENCES

1. Rao PS. Diagnosis of cardiac murmurs in children. *Vessel Plus* 2022;6:22. DOI
2. Uppu S. Imaging of atrial and ventricular septal defects. *Vessel Plus* 2022;6:21. DOI
3. Bui I, Bhattacharya A, Wong SH, Agarwal A. Role of advanced three-dimensional visualization modalities in congenital heart defect surgery. *Vessel Plus* 2022;6:31. DOI
4. Rao PS. Echocardiography: an overview - part I. *Vessel Plus* 2022;6:24. DOI
5. Rao PS. Echocardiography: an overview - part II. *Vessel Plus* 2022;6:25. DOI
6. Rao PS. Echocardiography: an overview - part III. *Vessel Plus* 2022;6:26. DOI
7. Misra A, Gupta P, Aggarwal S, Singh GK. Multimodality imaging for repaired tetralogy of Fallot. *Vessel Plus* 2022;6:32. DOI
8. Misra A, Safa R, Sanil Y, et al. Cardiovascular involvement in multisystem inflammatory syndrome in children with COVID-19. *Vessel Plus* 2022;6:14. DOI

9. Saxena A, Relan J. Pregnancy with congenital heart disease. *Vessel Plus* 2022;6:15. [DOI](#)
10. Sahulee R, McKinstry J. Pharmacologic therapies for the low cardiac output syndrome in children after cardiac surgery: evidence of their efficacy and trends in their use. *Vessel Plus* 2022;6:5. [DOI](#)
11. Wang-Giuffre EW, Doshi UH. Cardiopulmonary exercise test as a tool in surveillance after Fontan Operation. *Vessel Plus* 2022;6:29. [DOI](#)