40 Years of Pediatric Innovations: Congenital Heart Disease

Here is another in our series of discussion and debate over advances in pediatrics over the past 40 years.
~ Stanford T. Shulman, MD

Welton M. Gersony, MD: The innovations in congenital heart in the last 40 years that have had the most positive effect on patient outcome are the emergence of successful neonatal cardiac surgery; the remarkable development of the field of interventional of cardiac catheterization, including arrhythmia management; and most recently, nonsurgical valve replacement. Imaging, drug development, and genetics are other areas that have seen great progress. Let’s review a few highlights.

The heart-lung machine has allowed cardiac surgery for adults and adolescents since the 1950s, but for many years, the technology could not be successfully miniaturized for infants. The indication for open heart surgery in a child was that he/she must be over 35 lb. If the patient weighed less than 35 lb, it couldn’t be done.

As technology improved, the era of neonatal and infant surgery began. The most impressive success story is that of Transposition of the Great Arteries (TGA), which, before the era of catheter and surgical interventions, resulted in 98% mortality in the first days of life. Now, with neonatal surgical intervention, 70% to 80% of patients with HLHS and other forms of single ventricle survive past infancy.

Unlike in TGA, these children continue to face cardiac issues as they grow older. It was not long ago that 90% of cardiac catheterizations were diagnostic studies. This has totally reversed; 90% are interventional procedures. With modern imaging techniques (echocardiography, MRI), diagnostic catheterization in pediatric cardiology has become virtually extinct. New catheter interventional techniques have replaced open heart surgery for many children with congenital heart disease. Holes are closed, channels are opened, and valves are replaced. Ablation of abnormal electrical pathways and insertion of pacemakers and defibrillators have revolutionized arrhythmia management.

The accuracy of fetal echocardiography has led to great strides in prenatal identification of cardiac lesions. Currently, 70% to 80% of neonates evaluated at our medical center already have a cardiac diagnosis.

The effect of early diagnosis on survival is not totally documented, but there is a distinct advantage for a cardiac specialty unit being forewarned so obstetrical management of the mother and urgent care of the neonate can be optimal.

The drugs that have had the most impact include the newer inotropes that are especially useful in pediatric cardiac ICUs. Modern diuretics are powerful and effective and, when used properly, are rarely associated with clinical side effects. Treatment indications with beta-blockers have changed significantly in recent years, as has been true for adults.

In the neonate, the development of prostaglandin as a drug to keep the ductus arteriosus open after birth is an important factor in the success of surgery. In patients with pulmonary outflow obstruction, pulmonary blood flow is maintained in the critical hours before surgery via a patent ductus. In neonates with obstructed aortic blood flow (HLHS, aor-
tic coarctation, critical aortic stenosis), blood flow from the pulmonary artery to the aorta through the ductus allows adequate systemic blood flow immediately after birth until surgery can be carried out. Prostaglandin administration stabilizes the infant’s condition and increases the chances for effective surgery.

Drug therapy for arrhythmias has not advanced as much as one might have hoped, but interventional treatment with ablations, pacemakers, and defibrillators have had a huge effect on management of cardiac arrhythmias.

Perhaps the most important innovations in congenital heart disease are emerging in genetics. Understanding the genomic basis of some patients with cardiac syndromes has defined therapeutic and preventive management; however, the effects of these advances are just beginning. Much more investigation is needed before the laboratory-to-bedside component of genetic research becomes part of day-to-day management.

Dr. Shulman: With the recognition over the last 40 or 50 years of the relationship between strep and rheumatic fever has been the near disappearance of acute rheumatic fever in the US, although it thrives in some underdeveloped countries of the world. Now, there is recognition of another acquired heart disease. I just recently received two calls about children, and we see other children, with serious refractory Kawasaki disease (KD) with resulting coronary artery abnormalities.

Dr. Gersony: I’m struck by the improvement in early diagnosis. One of the interesting frontiers you now see explored in cardiology is understanding how cardiac disease affects brain development, even in utero and before surgery. We used to think that if there was a developmental delay, it had to do with the difficult surgeries. Now we understand that babies with a hypoplastic left heart have altered circulation in utero and have brain architecture that lags by several weeks.

One of the really exciting frontiers is the increased partnership among the cardiologists, cardiac intensivists, neonatologists, and developmental specialists in taking a population that now, as you said, routinely survives and helping these children live normal, satisfying lives.

Dr. Gersony: This is an important point. Abnormalities in fetal cerebral blood flow may account for the coexistent developmental disabilities associated with congenital heart defects such as HLHS. This may be even more important than some causative determinants related to hypoxemia, catheterization complications, and other environmental factors.

Stan L. Block, MD, FAAP: I saw a 22-year-old mother who, at age 14, had two major episodes of syncope. She subsequently had normal EGG and ECG, normal everything. Because of the severity of her symptoms, I was on heightened alert, so I sent her to the cardiologist. He put her on the treadmill and she developed V tach; he thought the V tach was epinephrine-induced, initially. Several months later, a friend of mine and the cardiologist told me about the normal ECG-variant of long QT syndrome (LQTS) — with genetic testing only available then at the University of Pittsburgh. We later found out there’s about 10 in the whole family who have this LQTS, including babies, sisters, mothers, and fathers, up the entire family tree.

It brings us back to genetics and the issue of sudden death syndrome in athletes. What are we going to do about this in the future? How are we going to diagnose it? There’s discussion of doing ECGs on every teen and what that means to us in private practice. It’s a huge burden of time, energy, and money, and a huge issue in practice for us. I’d say this is one of those beckoning problems for the cardiologist.

Dr. Gersony: LQTS is complicated. There are various types, each with different degrees of concern in terms of prognosis. There is no data to indicate that mass ECG screening would be effective in preventing sudden death. In many cases, a diagnosis of LQTS is far from a simple “yes or no” based on a screening ECG.

It is important for the pediatrician to recognize the difference between neurologic and cardiac syncope, and make an immediate referral to an appropriate specialist. In evaluating such patients, a careful family history must be obtained, and this may lead to an early diagnosis. Once the presence of LQTS has been established, preventive therapy is effective.

Dr. Block: Then there’s the multiple causes of sudden death of the athlete.

Dr. Gersony: In Italy, screening electrocardiography is standard for all athletes. There is an ongoing debate as to whether to institute such a program in the United States to diagnose cardiomyopathy and protect athletes from sudden death. The ECG has many variations within the normal range. Without absolute proof of efficacy, the screening process could negatively affect the quality of life of countless young men and women, based on inaccurate interpretation of ECG as abnormal.

Dr. Block: The cost and time to do all these athletes could be cost-prohibitive.

Dr. Gersony: Based on both risk/benefit and cost/benefit analyses, universal ECG screening in the US is not warranted at this time.

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