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GUEST EDITOR'S PAGE

Adult Congenital Heart Disease

The Quest to Address the Simple and the Complex

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Congenital heart disease (CHD) is the most common inborn defect affecting ~0.8% of newborns. Thanks to advances in pediatric cardiology and innovations in surgical and interventional procedures, there has been a dramatic improvement in long-term survival of children born with CHD. As a result, and for more than a decade now, the number of adult patients with CHD exceeds the number of children, and this number continues to grow exponentially encompassing progressively elderly patients. Furthermore, increasing number of patients with complex CHD is now surviving to adulthood, posing new challenges and opportunities to health care providers.

It is a fact that CHD represents one of the major successes of modern medicine (1). However, as we see in this special issue of *JACC: Case Reports*, the majority of adult patients born with CHD are not cured by their pediatric cardiac operation or intervention, and many face challenges and complications of living with the consequences of their underlying cardiac defect (2). The road to successfully addressing these emerging complications is a long and arduous one. Professor Jane Somerville, back in 1988, as the John Keith Lecturer, rightly entitled her talk "Out of the blue and into the pink. Is it so rosy for the cardiologist?" referring to the cyanosed female patient with Ebstein anomaly who had had reparative surgery and would have had challenges later on in life. We are faced with a very heterogeneous population with such a diverse spectrum of anatomy, physiology, and a variable adaptation to residual hemodynamic lesions, often having multiorgan involvement and further complications by acquired heart disease and noncardiac comorbidities, all these contributing to quality of life and late outcomes. Indeed, our adult patients with CHD continue to be afflicted by higher mortality rates compared with the general population, with the highest mortality observed among patients with complex CHD, such as patients with the Fontan operation or the Eisenmenger syndrome. The leading cause of death in adults with CHD is now heart failure. Therefore, it is appreciated that adult patients with CHD have special needs and require lifelong surveillance and highly specialized health care, which has led to the development of tertiary adult CHD centers all over the world (2). It is paramount that patients with moderate-to-complex CHD receive such expert care and support for life.

Ongoing research and growing understanding of the late pathophysiology of CHD are continuously refining all aspects of care for these patients. Putting the patient in the very center of these efforts further advances the existing model of care (3). A personalized patient path should be the way forward, starting from prenatal diagnosis and planning, through childhood, and with a seamless transition to adult care with a lifelong trajectory (**Figure 1**). We have proposed that patient-centered adult CHD care should include a single-day assessment at the CHD center for all patients, including those transitioned from pediatric care, where a comprehensive and thorough assessment takes place, including imaging, cardiopulmonary exercise testing (or 6-min walk distance), biomarkers, genetics, and consultation

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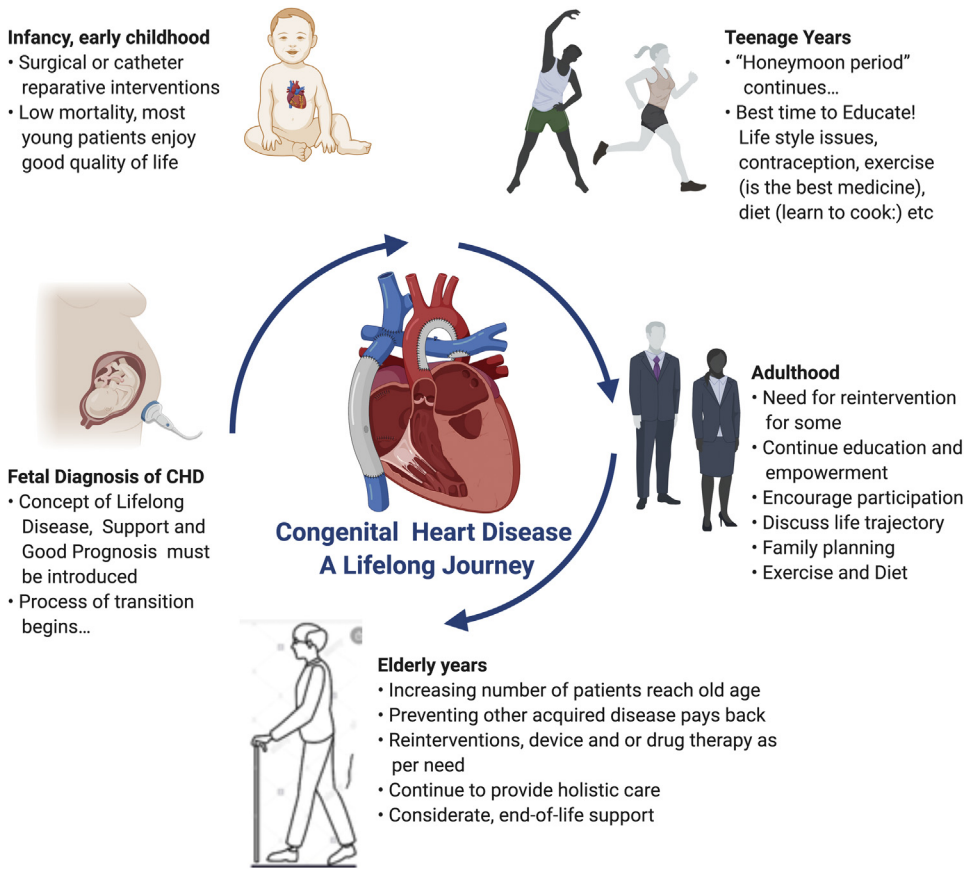
FIGURE 1 Congenital Heart Disease: A Lifelong Disease



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with the adult CHD Consultant and Clinical Nurse Specialist (1,3). At this crucial visit, patients should be risk-stratified and receive all necessary information about their condition, prognosis, lifestyle, family planning and contraception, what to do with emergencies, etc. All this information should be shared in an accessible way, such as in a form of a mobile application—currently piloted by our group—so that patients have their files with them and can share with other health care professionals as per need. This, we submit, may be a more effective way of reaching out to other disciplines and providers that CHD patients need input and care from (who are often lost in the "abyss world of CHD").

Subsequent follow-up must be personalized. Patients with simple CHD defect and at the good end of the spectrum may be managed in "nonspecialist" clinics with access to specialist care, if required. Conversely, patients with moderate and complex CHD will require closer surveillance at tertiary settings, albeit technology may assist here with remote monitoring, thus potentially reducing frequency of visits to the CHD Hub. As the world moves into the digital era, medicine follows, and CHD should not be an exemption to this. The advanced model of CHD care, discussed herewith, should utilize and further develop state-of-the-art breakthroughs, namely:

- Novel genomic and molecular testing techniques
- Artificial intelligence for "online" risk stratification of CHD patients and imaging analysis, including volumetric data
- Refinement of cardiac surgical and catheter interventions and innovation (such as bio-resorbable devices for atrial septal defect or patent foramen ovale closure or new, durable, biological prosthetic valves)
- Multimodality imaging and techniques of 3-dimensional printing, computational modeling, and computer-generated real-time digital

holograms depicting the depth and spatial relationships of the heart, great vessels, and the surrounding structures. These models can be used to study CHD anatomy, plan surgical procedures, and teach trainees and patients

- Fast access to CHD databases with automatic notifications regarding specific actions required

Heart failure advanced therapies, including cardiac resynchronization therapy, mechanical support, and transplantation are still underutilized in CHD compared with non-CHD population (4). Mechanical pumps are not designed to accommodate specific CHD needs such as supporting the systemic right ventricle or the Fontan circulation. Funding bodies and industry need to be enticed to invest in this growing population, and a stronger and clearer voice from the adult CHD community, including patients, may facilitate this. Until then, outcomes of complex CHD patients may remain suboptimal. We must do all that we can to mount these long-due investments.

Last but not least, personalized CHD care includes patient education and patient empowerment (5). Thorough understanding of the patient's condition is essential. Patients must, consequently, assume responsibility for their life and care, including healthy lifestyle choices such as daily exercise and healthy diet. Moreover, educated patients should be an integral part of the decision-making process when it comes to major interventions.

In conclusion, we have come a long way with the endless pursuit of excellence and innovation on the bumpy and rewarding road of CHD care, but there is still much to do. With the CHD patient at the center of our pursuits, we form a strong basis for future growth, needs, and developments, and for a better patient health care experience and life journey, irrespective of complexity of CHD disease, ethnicity, or geography.

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