EDITORIAL

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The growing importance of adult congenital heart disease

The phenomenal success of paediatric cardiology and cardiac surgery over the last 50 years has brought with it a new and daunting responsibility for the adult cardiologist, and that is coping with the growing numbers and complexity of children surviving into adult life, with palliated or repaired congenital heart disease.

From the 1960s onwards, complex congenital heart lesions that were almost uniformly fatal in infancy have now been transformed to conditions where long-term survival is the expectation, but where few of the surviving patients are left with "normal hearts". Rather, a variety of haemodynamic, electrophysiological and even psychosocial consequences accompany these children on their journey into adult life. Thus, for example, where transposition of the great arteries was usually fatal in the first year of life before the era of open-heart surgery, survival rates of well over 90% into adult life are now expected, but often with residual volume loads, pressure loads, re-entry circuits and/or complex management issues related to genetic counselling, pregnancy management, etc.

The burden of adult congenital heart disease, in fact, has now grown to the extent that by the year 2000, there were nearly equal numbers of adults and children with severe congenital heart disease (Marelli et al Circulation 2007;115:163-172). The implication of this observation would be that we need almost as many specialists in adult congenital heart disease as there are in paediatric cardiology, with important resource and training implications.

Nevertheless, the specialty of "adult congenital heart disease" is in its infancy and hardly exists at all in many countries where there are such adults who require specialised care. Most adult cardiology training programs focus appropriately on the management of coronary artery disease, valvular disease, heart failure and arrhythmia, with very little if any training given in congenital heart diseases. It is exceptional for an adult cardiology trainee to spend any time in a Paediatric Cardiology Unit learning about the diagnosis and management of congenital heart diseases, but it is equally rare for paediatric cardiologists to wish to continue the care of children with heart disease beyond teenage years. Furthermore, children's hospitals in general discourage the continued care of patients beyond the paediatric age range, for a variety of reasons.

Thus adults with congenital heart disease, many of whom have complicated medical and non-medical issues, often "fall between two stools": too old for the paediatric cardiologist who really understands the anatomy and physiology of their lesions and too complex or different for the adult cardiologist whose area of expertise does not usually encompass repaired congenital heart diseases.

This special issue of SA Heart is thus dedicated to a giving a glimpse into the world of adult congenital heart disease. Whereas the management of complex post-operative lesions such as transposition of the great arteries, pulmonary atresia, tetralogy of Fallot and single ventricle circulations is probably the province of the specifically trained cardiologist in adult congenital heart disease, many other lesions will present to the adult cardiologist for diagnosis and care. We have selected the topics for this special issue with a view to providing important background information about these relatively less complicated aspects.

The issue opens with papers covering two of the commonest of the so-called "simple" congenital heart lesions that present in adult practice: mitral valve prolapse and regurgitation, and sequelae from coarctation of the aorta. Three more general papers follow, including a spectacular view into the imaging advances in congenital heart disease provided by Magnetic Resonance and CT scanning, an excellent review of the variety of important arrhythmias that might be encountered in the adult with congenital heart disease and finally a primer into the issues of pregnancy and genetic risk in adult survivors with CHD.

Each country in which paediatric cardiology and cardiac surgery have been successfully practiced for 20 years or more is now obliged to consider the adequate and coordinated provision of services for young and older adults with congenital heart disease. Often this involves specific training of selected specialists in overseas centres where such coordinated adult congenital heart programs already exist, such as in the USA or the UK. Such trainees are then well placed to replicate models of coordinated care in their countries of origin and this must be prospectively planned. The successes of Paediatric Cardiology of yesterday must be cared for in an environment that will maximise their chances of continued high-quality and long-quantity life, where their cardiac condition permits. To provide anything less than optimal adult congenital cardiac care would be to fail our mission of consolidating the fabulous successes of paediatric cardiac medicine and surgery over the last 50 years.