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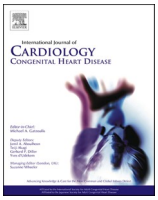

Screening and prevention in congenital heart disease: Whom, when, and how?

The treatment of congenital heart disease has been one of the triumphs of modern cardiology and cardiac surgery [1]. Most children born with congenital heart disease now survive to adulthood and this has created a new specialty with many challenges. Early treatment of congenital heart disease in childhood has often involved innovative surgery and interventional catheter approaches but, in many cases, patients are left with circulations that are very different from the 'normal' ones encountered by adult cardiologists which does affect outcomes [2].

For example, after the Mustard and Senning operations for transposition of the great arteries (TGA), the left and right ventricles end up 'the wrong way round' or indeed, there may be only one functioning ventricle after operations such as the Fontan procedure [3].

As a result, patients require long term 'secondary prevention' to manage a range of cardiac complications such as rhythm disturbances [4], ventricular dysfunction and valve failure [5]. They are also at risk of the consequences to other organs, such as the lungs, liver and brain from the disordered physiology that they had been left with, even after 'definitive repair'.

Despite the lack of rigorous testing with randomized clinical trials (with the exception of new therapies for pulmonary hypertension; [6]), huge progress has been made in both the prevention and care of such late complications which become progressively more common with longer follow up. Optimal results require specialists trained in adult congenital heart disease and as a result of improving clinical services, patients are now living for many decades even with the most complex congenital cardiac malformations [7].

1. The next challenge?

This remarkable success story has created the 'next challenge' as more and more patients are reaching the age at which acquired cardiovascular disease becomes manifest in the population [8]. The consequences of atherosclerosis, including myocardial infarction, stroke and heart failure are major causes of premature morbidity and mortality and become increasingly common after the age of 50 years. It is now clear, however, that the underlying pathological process of atherosclerosis begins decades earlier, in childhood [9], and progresses silently in response to risk factors, such as cholesterol, blood pressure, smoking and diabetes. As adults with congenital heart disease age, many are likely to be at least as vulnerable to the development and consequences of acquired cardiovascular disease on top of their original congenital malformation. Evidence suggests that cholesterol levels are similar to the general population. Furthermore, congenital heart defects may predispose to important vascular risk factors such as hypertension (for example, even after successful repair of coarctation of the aorta; [10]) and obesity (as a result of less healthy and less active lifestyles; [11]). Some of the circulations after repair of congenital heart disease maybe particularly vulnerable to acquired cardiovascular disease, such as those with impaired ventricular function or pulmonary hypertension. Patients after Fontan repair often have subclinical liver disease and depend on maintenance of low pulmonary vascular resistance and good function of their single ventricle. We still do not have sufficient information about the vulnerability of the coronary arteries to atherosclerosis and its complications after operations such as the arterial switch or aortic surgery. Thus, more than ever, 'primordial' or 'primary' preventative strategies for vascular disease are important in patients with congenital heart disease. More observational evidence will be required in larger populations with congenital heart disease on the development of known cardiovascular risk factors as well as the benefits of early lowering. Randomised evidence is still scarce for the value of different drugs, such as angiotensin converting enzyme inhibitors, betablockers and more recently mineralocorticoid antagonists and sodium glucose transport type-2 inhibitors (SGLT2-i) which have proved so effective in improving outcomes in non-congenital patients [12]. Nevertheless, we have good reasons to assume that blood pressure, lipids, weight management and diabetes prevention [13] will be equally if not more valuable in the adult population with congenital heart disease.

2. The current issue

This issue of *International Journal of Cardiology Congenital Heart Disease,* edited by Margarita Brida and Salvatore De Rosa, assembles a large number of excellent contributions on preventive measures and their effectiveness in patients with congenital heart disease. It starts with classic cardiovascular risk factors such as hypertension, reviewed by Giovanni Di Salvo, lipids and metabolic syndrome by Macjiej Banach, nutrition and dietary recommendations by Laura Dos Subira and exercise prescription and training by Werner Budts. But it goes far beyond primary prevention by discussing stroke and systemic embolism by Katja Prokselj, coronary artery disease by Salvatore De Rosa and atrial fibrillation by Berardo Sarubbi. Further it reviews liver disease, renal failure, pulmonary hypertension and sudden cardiac death – a true comprehensive compendium, which is a 'must' for everybody that is anybody in congenital heart disease and beyond.

The term 'prevention' is derived from the Latin word *praeventus*, ie to anticipate and hinder. To prevent, one has to foresee. The new strategies that are emerging to understand the development of acquired cardiovascular disease and to develop treatment plans which reduce cumulative exposure to modifiable risk factors, are likely to be the next 'big step forward' in the remarkable progress which has been made in lifetime

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management of patients with congenital heart disease.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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