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Editorial

Pulmonary function in repaired congenital heart disease: Our attention must reach beyond the mended heart

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During the recent decades of great surgical and medical achievements in the treatment of congenital heart disease (CHD), research focus has shifted from just survival to exploration how the heart condition and its treatment is affecting the patients' health in multiple ways.

The great age of milestones in thoracic surgical treatment of CHD lasted from the 1950s to the late 1980s [1,2]. It began with early attempts of extra-corporal circulation enabling open heart surgery. More and more congenital heart defects became repairable, more and more lives were saved. Finally, during the 1980s, the development of the Norwood procedure in hypoplastic left heart syndrome, and the Jatene procedure of arterial switch in transposition of the great arteries were the latest game changing surgical innovations in CHD.

Improved survival created a novel field of competency in cardiology: the care for the adult patient with congenital heart disease. The adult patient population with CHD has outnumbered the pediatric CHD population in developed countries. However, not only do these adult patients face the need of redo-interventions and surgery. The burden of aging and life-style related cardiovascular morbidity also hits them harder than other groups.

With increasing survival and an aging patient population, knowledge of the human cost of repaired congenital heart disease has evolved. A broad spectrum of late consequences and comorbidities in repaired CHD has been studied. Neuro-developmental outcomes in children with congenital heart disease have been a concern for both professionals and caregivers for a long time [3]. Other organ systems have lately received attention of the researchers. Renal dysfunction [4], impaired bone health [5], protein-losing enteropathy [6], lymphatic abnormalities [7], and chronic liver disease [8] are just a few examples of such comorbidities. In particular, the long-term challenges in patients with a Fontan-circulation have given rise to the question, how many problems are solved and how many are created with the Fontan-type palliation of univentricular heart defects.

Impaired pulmonary function in a large cohort of CHD patients has previously been demonstrated by Alonso-Gonzalez et al. [9]. In their study, pulmonary dysfunction was correlated to complexity of the underlying CHD. In this issue, J. Heiberg and colleagues present a Danish study showing altered pulmonary function in adults after early repair of an isolated ventricular septal defect, commonly considered as a simple form of CHD [10]. This publication contributes with interesting data to the field of adult congenital heart disease. The authors have done commendable work with a double-blinded study design.

It is methodologically difficult to analyze which factors are the most important contributing to pulmonary dysfunction in CHD: is it the early surgical trauma from opening an infant's chest? Is it genetics leading to developmental alterations of both cardiac and pulmonary structures? Or is it a chronic detrimental process in the lungs triggered by a significant left-to-right shunt and increased shear stress in the pulmonary vessels early in life?

Based on results from studies like this, it should be remembered that also the repaired simple cardiac lesions might have long-term implications. Follow-up is often terminated though after few years when simple lesions have been repaired without residual pathology. Long-term comorbidities in adult CHD patients with simple lesions should make us consider seeing even the “simple” CHD patients from time to time during adult life.

Conflict of interest

No conflicts of interest.

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