JIM Introduction

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Introduction The 99th Berzelius symposium: the Cardiac Patient from Birth to Adulthood

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Content List: – Read more articles from the symposium: "The 99th Berzelius symposium: The Cardiac Patient from Birth to Adulthood".

Approximately one child out of 100 is born with a congenital heart defect, and a major part of them need an intervention. Thanks to the success in paediatric heart care, most children born with congenital heart disease are now expected to reach adulthood. This has consequently led to a growing population of adults with repaired congenital heart disease. In fact, the number of adults with complex congenital heart disease has since long outnumbered the children [1]. However, many heart lesions must be regarded as repaired and not cured. As a consequence, long-term complications pose a potential threat as the patients grow older. It is intuitive to expect that most of the late complications are directly related to the heart lesion which also is true for many patients. With the steadily increasing number of survivors, other types of complications such as neurocognitive disorders including dementia [2], cancer [3] and acquired cardiovascular disease have emerged as associated with congenital heart disease.

In the 99th Berzelius symposium: The Cardiac Patient from Birth to Adulthood – February 2019, a number of sessions covered the late sequels in adults with congenital heart disease and three reviews based on this symposium are published in the Journal of Internal Medicine [4–6]. Arianne Marelli describes the dramatic changes in epidemiology, with success in early survival and an increasing number of adults living with congenital heart disease, as a paradigm shift and that congenital heart disease should be considered a lifespan condition [5]. She also introduces the concept of disease trajectories, complex constructs to describe the patients' healthcare journey. These trajectories move beyond the mortality outcome and are hypothetically described for some of the complex congenital heart lesions. The lifetime perspective is stressed with the goal of optimal healthcare delivery throughout life.

As patients with congenital heart disease grow older, they are also exposed to the risk of acquired cardiovascular disease. In fact, the risk for coronary artery disease may be higher in this population [7]. Added coronary artery disease on pre-existing congenital heart disease may have severe consequences. This problem is reviewed by Eero Jokinen [4], with a focus on underlying causes and primary prevention, a field where there is a paucity of studies and evidence. Patients with coronary artery disease may have a slightly different risk-factor profile, and there are also specific factors that may be of importance. Here, both denervation of the coronary arteries by previous heart surgery and previous cyanosis are mentioned. An active and early primary prevention is advocated as atherosclerosis is known to start early.

The most common structural congenital heart defect, *that is* persistent foramen ovale (PFO), is actually not considered as a congenital heart lesion but rather a normal variant that exists in up to one third of adults. Often, there is a valve-like function that only permits right-to-left shunting when the right atrial pressure transiently exceeds the left atrial pressure, *for example* during the Valsalva manoeuvre. PFO has since long been considered to be involved in cryptogenic stroke, albeit without clear evidence. After a cryptogenic stroke, the shunt was frequently closed, if other causes were excluded. However, in 2013 three negative trials were published that changed the practice to a more conservative approach. Intervention with closure of the shunt was only performed in highly selected cases such as after repeated strokes where no other causes were shown. In new trials, with an adjusted design, a clear positive effect of closure of PFO after cryptogenic stroke was shown [8–10]. Again, the practice was changed but now towards an increased use of catheter closure of PFO. Lars Søndergaard, the first author of one of the trials

[10], describes the state of the art of PFO closure including review of the previous three trials, cost-effectiveness and prognosis.

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