



Editorial

Natural history of pulmonary atresia: The ACHD clinic perspective

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An increasing number of children born with congenital heart disease (CHD) survive into adulthood following single or multiple catheter based and/or surgical intervention [1–3]. Adult with congenital heart disease (ACHD) represents a very heterogeneous population of patients, with different anatomic, pathophysiological and electrophysiological problems after repair [3]. This is true even in patients classified as having the same cardiac defect, such as pulmonary atresia (PA). PA might present with various and complex anatomical features necessitating diverse surgical and interventional strategies [4–6].

Montanaro and coworkers [4] described clinical history and long term outcome of adults with PA, reviewing 15 years of data from their tertiary center. PA population was classified into 3 groups: PA-VSD, including patients within the spectrum of tetralogy of Fallot/double outlet right ventricle (PA-TOF and PA-DORV), with a complete repair or without repair (i.e. unoperated or with palliative arterial shunts); PA with intact ventricular septum: PA-IVS; and a miscellaneous group of PA, with different and complex associate anatomy. The conclusion was, nevertheless, surprising: morbidity and mortality were poorly correlated with the anatomic complexity and the consequent type of repair of the original PA defect. These findings are the result of a different point of view of the observer. The authors moved the “camera angle” from the CHD to the ACHD clinic, reviewing natural history of PA patients who reached the adulthood status. Their results are in contrast with a Finnish study [7], describing long term outcome of the PA + VSD population. They found that achievement of repair and initial size of true central

pulmonary arteries affect survival, irrespective of presence or absence of MAPCA. Differences between studies may be related to the different characteristics of the population studied, specific experience and resources of the 2 centers, and different periods of observation (longer in the Finnish study: 1970–2007; UK study: 2000–2015), reflecting different management strategies.

An interesting finding of the study by Montanaro et al. was that systemic ventricular dysfunction and resting oxygen saturations were predictive of all-cause mortality, due to heart failure and life-threatening arrhythmic events. This observation highlights the need of multi-imaging to better assess and monitor ventricular function in patients referred to ACHD clinics [8].

In conclusion, nowadays the majority of patients with PA surviving into adulthood carry residual lesions and sequelae putting them at risk of severe complications. Ventricular function and arrhythmic events should be regularly assessed and monitored in these patients. While cardiac surgery may improve anatomy and clinical status, morbidity and mortality are not really affected. In “end stage” patients, expert ACHD centers should offer advanced care planning and end-of-life issues [9].

Conflict of interest

The authors report no relationships that could be construed as a conflict of interest.

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