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Editorial

Pulmonary artery dilatation in congenital heart disease: Size doesn't matter

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A 76-year-old patient was referred for consideration of surgical repair of aneurysmal dilatation of the pulmonary artery, which was found incidentally on chest X-ray. On computed tomography and angiography the maximal diameter of the main pulmonary artery was measured 66 mm. Pulmonary artery pressures were within normal range and echocardiography revealed mild pulmonary valve regurgitation. No compression of bronchi or coronary arteries was detected. The patient was asymptomatic. What should we advise?

This real-life case vignette highlights a typical scenario in our day-to-day clinical practice in the care for patients with congenital heart disease. On numerous occasions we are confronted with rare conditions, for which the evidence to guide our decision-making regarding prophylactic interventions is sparse or nil. Thus, in such cases, decision-making is often guided by extrapolation from other disease conditions or anecdotal experience.

In the paper published in this issue of the International Journal of Cardiology, Gallego and colleagues present the results of a carefully executed review of pulmonary artery dilatation and its associated complications in patients followed in their program for adults with congenital heart disease [1].

Although transthoracic echocardiography often allows assessment of pulmonary artery dilatation, in case of suboptimal echocardiographic windows, a simple plain chest X-ray, as used in the present study, may serve as a valid screening tool for detection of significant pulmonary artery dilatation but requires confirmation by either computed tomography or magnetic resonance imaging (see Fig. 1).

Gallego and colleagues found a high prevalence of pulmonary artery dilatation, affecting every fifth patient in their cohort. Pulmonary artery dilatation was most common in patients with pulmonary valve stenosis and shunt lesions, particularly in those with concomitant pulmonary arterial hypertension, while largest pulmonary artery dimensions were found in patients with tetralogy of Fallot with the absent pulmonary valve syndrome variant.

During follow-up, progressive dilatation and complications were rare and occurred predominantly in patients with pulmonary hypertension. While the extent of pulmonary artery pressure was found to be associated with complications, sheer pulmonary artery diameter was not.

Although it may be tempting to use fixed diameters for prophylactic interventions in patients with dilated pulmonary arteries, as commonly executed in patients with aortic dilatation, the results of this study discourage the use of any fixed diameter thresholds for prophylactic intervention in patients with pulmonary artery dilatation. In this regard, this study highlights that extrapolation on the base of vague similarities between different disease entities may be misleading and inherit the risk of overtreatment of affected patients. A typical example of this phenomenon is the change in treatment algorithms for ascending aortic dilatation in patients with bicuspid aortic valves. In the past, based on similarities on histological appearance of dilated aortas in patients with bicuspid aortic valves and patients with connective tissue disorders (such as the Marfan-syndrome), an aggressive approach for prophylactic aortic replacement had been strongly advocated for patients with bicuspid aortic valves and dilated aortas, based on diameter-criteria derived from cohorts with connective tissue disease [2]. However – despite these histological similarities of the aortic wall – the natural history of ascending aortic dilatation and its risk of complications is fundamentally different in patients with bicuspid aortic valves compared to patients with connective tissue disease [3]. Therefore recommendations had to be revised in more recent recommendations [4]. In the meanwhile, many patients with bicuspid aortic valves and moderate aortic dilatation had undergone prophylactic major aortic surgery often with concomitant valve replacement, exposing them to the increased long-term risks of prosthetic heart valves (e.g. increased risk of endocarditis and long-term risks of oral anticoagulation in case of mechanical valve prosthesis).

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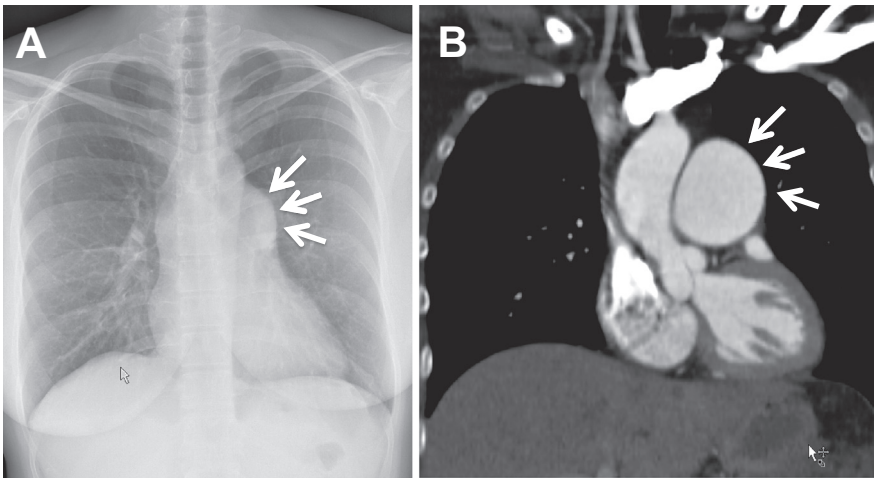

Fig. 1. Chest X-ray (Panel A) of a patient with dilated main pulmonary artery (arrows) and corresponding chest computed tomography scan (Panel B).

Although randomized controlled trials are considered the gold standard to gain evidence in clinical medicine, many important clinical questions are not amenable to randomized clinical trials. As congenital heart disease is a life-long disease encompassing many rare conditions, careful observational studies of long-term outcomes often provide important insights in risk factors for adverse events. This evidence allows a more rational counseling and decision-making for our patients.

We thus must appreciate the work of Gallego and colleagues provided in this current study and we should encourage all efforts for ongoing observational studies and carefully conducted registries, as these studies and registries will importantly increase our knowledge on long-term outcomes in adults with congenital heart disease and will eventually improve the care for our patients.

Conflict of interest

No conflict of interest.

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