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Editorial

Can patients be discharged after VSD closure?

Nashat H. [a], Montanaro C. [b,*]

[a] ACHD Centre Royal Brompton and Harfield Foundation Trust, London, UK; National Heart and Lung Institute, Imperial College London, UK
[b] GUCH Centre, St Bartholomew's Hospital, Barts Health NHS Trust, London, UK

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The evolution of congenital heart disease (CHD) globally rides on waves of success for many burdened. Advances in prenatal or early diagnosis, resulting in early corrective intervention has salvaged many young lives. Nevertheless, despite these respectable successes, interventions offer a range of complications, some unanticipated, thus necessitating lifelong follow up for a young population expected to survive as long as their 'healthy' peers [1].

Congenital heart disease, particularly defects precipitating abnormal pulmonary blood flow and pressure, predisposes individuals to pulmonary vascular remodeling thus potentiating pulmonary vascular resistance and disease. Guidelines introduced, encourage timely repair in those with a requisite panel of haemodynamics, thus protecting the delicate pulmonary vasculature from damage [2]. The prevalence of pulmonary arterial hypertension (PAH) is estimated at between 5 and 10% in this heterogeneous cohort. Modern medicine has interfered with the landscape of PAH-CHD, with notable reductions in patients with Eisenmenger syndrome and an emergence of PAH, often decades after, corrective repair. Furthermore, registry data has shown that this later group has a morbidity and mortality outcome similar to those with the aggressive idiopathic PAH. Therefore there is an urgent need to establish comprehensive screening methods and establish a risk stratification model in this expanding cohort, as we continue to explore optimal management at different stages of this disease.

In this edition article by Gabriels et al. '*Serial pulmonary vascular resistance assessment in patients late after ventricular septal defect repair*', the authors demonstrated that patients after ventricular septal defect (VSD) closure have altered pulmonary haemodynamics and right ventricle (RV) impairment when compared to 'healthy' controls [3]. Furthermore over a relatively short follow-up, patients had slight yet significant progression in pulmonary vascular resistance, albeit not clinically substantial. The most robust and gold standard method for assessing pulmonary haemodynamics, amongst all strata of PH, is the right heart catheterization. Nevertheless, methods to evaluate and validate pulmonary haemodynamics, using noninvasive techniques continue to evolve. This would allow a more feasible screening practice for patients at risk, a population that is set to grow [4]. There is mounting recognition of the clinical importance of pulmonary haemodynamics during exercise, particularly as a sign of early pulmonary vascular disease (PVD), left heart and lung disease. Importantly, there is growing evidence reported in the literature that these adverse exercise induced haemodynamics may unmask occult PVD [5]. However, this area remains challenged, more so for those with congenital heart disease, as a consensus agreement of resting pulmonary haemodynamics has been defined clearly, but as yet there is no clear widely accepted definition of 'exercise-induced' pulmonary hypertension [6]. As further studies of this kind continue, the body of evidence will allow for a consensus definition for this important disorder. Moreover, to date, there is no clear consensus on the use of advanced therapies in patients who show signs of exercise induced adverse pulmonary haemodynamics.

This notable work by Gabriels et al., serves many purposes. Firstly it supports the evidence that patients with congenital heart disease, regardless of defect complexity or 'complete' repair, necessitate lifelong follow up and screening for complications including PVD [7]. This is also supported by data from the Danish group who has demonstrated a number of subtle but important long-term abnormalities associated with "simple" lesions: adults with small, unrepaired atrial septal defect have been shown to have significantly impaired exercise capacity when compared to 'healthy' controls; the impairment was present even if, by the time of assessment, the defect had closed spontaneously [8]. Moreover, they demonstrated that, even if repaired in early childhood, adults after surgical VSD closure have altered pulmonary function [9], impaired heart rate variability and, particularly, those with complete right bundle brunch block have lower heart rate variability compared to controls [10]. The Danish group has also demonstrated that patients with small unrepaired VSD or VSD closed in early childhood had significantly higher RV end-diastolic volume index on cardiac magnetic resonance, and in particular those with small unrepaired VSD revealed higher RV stroke index compared to their controls; positive correlations

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* Corresponding author at: Grown Up Congenital Heart Disease Unit GUCH Barts Health NHS Trust, St Bartholomew's Hospital, West Smithfield, London EC1A 7BE, UK.
E-mail address: cm098cm098@gmail.com (C. Montanaro).

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were shown between RV end-diastolic volume indices and peak exercise capacity in patients [11].

Of note, the work by Gabriel et al. supports the validation of bicycle exercise stress echocardiography, a novel technique used to measure the relationship between the mean pulmonary arterial pressure to cardiac output (mPAP-CO) during supine incremental exercise. A disproportionate increase in the mPAP relative to CO during exercise reflects an increase in pulmonary vascular resistance, as evidenced in this paper. Furthermore, in this cohort, a lower exercise capacity correlated inversely with the mPAP-CO slope. This further supports the need for standard consensus definitions, of resting and exercise pulmonary haemodynamics, to help guide clinicians to make sensible evaluations and diagnoses. Importantly, this study shows potential methods to non-invasively assess patients and identify a potential risk stratification model for developing PVD in this cohort.

Additionally, as with many studies on individuals with congenital heart disease, the number of patients in this study was small. Patients were assessed at baseline and again at a median follow up of around one year. Further similarly designed studies with longer follow up periods would strengthen the deductions made in this paper. Unfortunately the techniques used were not suitable for all patients, with obvious limitations of inaccurate measurements and patients ability to comply with the exercise routine. Validations of the findings using non-invasive techniques with the gold standard method of invasive right heart catheterization would also strongly support the hypotheses.

In conclusion, despite the limitations in Gabriels et al. study, the data shown and methodology is original, paving the way for future studies for this and other cohorts of congenital heart disease. Non-invasive investigations such as echocardiography and exercise stress testing may help identify patients who may need closer surveillance and those who may benefit from further invasive investigations. This paper demonstrates conclusively that patients with “simple” lesions need a long-life surveillance in an expert centre.

Conflict of interest

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

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