Unraveling the mysteries of cardiac contraction has been a major preoccupation of the cardiac investigators. More recently, the right ventricle is in focus, especially the right ventricle in the systemic position. The differences between the right and the left ventricles in myocardial architecture, blood supply, metabolism, response to loading conditions, and embryological origin have been well documented (1). However, why the right ventricle in systemic position is compatible with long life only in some patients has not been clearly understood, except perhaps teleologically. Evaluating the right ventricle has been difficult due to its complex geometric shape and position. With advances in imaging modalities such as tissue Doppler imaging, speckle tracking imaging (STI), cardiac magnetic resonance (CMR), and 3-dimensional echocardiography, the global and regional functions of the right ventricle can be better assessed and could potentially improve our understanding of the systemic right ventricular function.

In this issue of iJACC, Khoo et al. (2) report on the systemic right ventricle function in patients of hypoplastic left heart syndrome (HLHS). The investigators cleverly used the “unnatural experiment” of staged correction of HLHS and the available time window to gain insights into the right ventricle function and adaptation in this scenario. They studied 20 patients before and a few months after the Norwood operation and analyzed several echocardiographic parameters using conventional imaging and STI. The patients were studied at the mean age of 4 days and 150 days, respectively. The conventional indicators of global right ventricular function such as right ventricular fractional area change and myocardial performance index did not change, but the right ventricle had become more spherical. The right ventricle contractility, as measured by isovolumic acceleration, strain, and strain rate, had decreased. The longitudinal strain decreased and the circumferential strain did not change, hence the ratio of longitudinal/circumferential strain decreased. They also studied mechanical dyssynchrony by STI and found that mechanical dyssynchrony was evident at both stages, and with time, the mechanical dyssynchrony indexes improved somewhat. The echocardiographic strain parameters correlated with CMR-derived right ventricle mass, ejection fraction, and volumes obtained 0 to 80 days after the echocardiographic studies in 12 patients. Overall, the investigators conclude these findings as evidence of “left ventricle-like” adaptation of the right ventricle facing a systemic load. These novel observations may have important prognostic and therapeutic implications.

Because STI is a non-Doppler method, it is not dependent on angle of interrogation and is being increasingly used to study right ventricular function (3). However, the need for an excellent 2-dimensional image and manual tracking of borders, the issue of appropriate frame rates, the variability in the results from different algorithms, and other technical issues remain (4). Normative data of right ventricle function in children using STI and information on right ventricle function in other congenital heart disease is gradually accumulating (3). The study by Khoo et al. (2) is important in this regard, as it adds to such data. Low intraobserver and interobserver variability and
The normal right ventricle contracts anomalously, the right ventricular function in patients with systemic right ventricle may be more common than anticipated and may not be the result of coronary abnormalities but may occur due to demand-supply mismatch. Patchy fibrosis, in the adults with systemic right ventricle, has been reported on gadolinium CMR. This could possibly result from ischemia. Thus, the finding of post-systolic strain index warrants further exploration.

Despite tremendous progress in the operative management of HLHS, there is a significant mortality in the interstage period (10). As pointed out by Atallah et al. (10), the established independent risk factors for interstage mortality in post-Norwood palliation include tricuspid valve regurgitation and right ventricular dysfunction. The focus of the study by Khoo et al. (2) is on the systemic right ventricle function, but the variations of the left ventricle in HLHS may also have clinical influence, albeit less significant (11). The right ventricular function in patients with systemic right ventricle after Fontan operation has also been studied with echocardiographic parameters as in the study by Chow et al. (12), and RV dysfunction in these patients remains a clinical concern at follow-up.

Mechanical dyssynchrony in the right ventricle is complex. The normal right ventricle contracts most peristaltically like a bellow and the normal infundibulum is activated 25 to 50 ms later than the inlet of the ventricle (1). Both the ventricles synchronously deliver the same output into 2 circuits with vastly different loads. However, when faced with increased load, this pattern of right ventricle contraction is altered. The free right ventricle wall in the patients with idiopathic pulmonary hypertension contracts well into the ventricle diastole, and mechanical dyssynchrony is the rule (13). The present study also showed mechanical dyssynchrony in the basal state as well as later, but it improved with time despite worsening in some parameters of longitudinal contractile function. Mechanical dyssynchrony in the right ventricle does not correlate with QRS duration, and whether it can be favorably altered by resynchronization therapy is not clear. One study (14) showed improvement in mechanical function of right ventricle. But the observed benefits were smaller in another study (15) in the group with systemic right ventricle or single ventricle. Further studies are required to identify the potential benefits of resynchronization therapy in the failing right ventricle.

Right ventricular mass/volume relationship in congenital heart disease can now be reliably evaluated with CMR and 3-dimensional echocardiography. Although, right ventricular mass correlated with basal mechanical dyssynchrony indexes in this study (2), numerous correlations in a small number of patients are a potential limitation of the study. The increase in ventricular mass can be explained by the changes in myocardial work associated with the left ventricle-like contractions of the systemic right ventricle. Indeed, these findings are consistent with observations of Pettersen et al. (16), who studied 14 patients of transposition of great arteries following Senning surgery and compared them to healthy controls using tissue Doppler imaging and tagged CMR. In patients with transposition of great arteries, the circumferential strain was higher than the longitudinal strain in the right ventricular free wall, which is akin to findings in normal left ventricle, and the opposite was true in the normal right ventricle. The strain measured at the interventricular septum was not different, and the ventricular torsion was absent in the systemic right ventricle. Similarly, increased circumferential contractility of the right ventricle has also been described in pulmonary artery hypertension (17). Whether it is a reflection of right ventricle dysfunction or adaptation is not very clear. In the normal right ventricle, the middle layer circular fibers are not as prominent.
as in the left ventricle. With hypertrophy, circumferential contraction may have mechanical advantage. Whether the right ventricle hypertrophy actually results from the circular middle layer fibers, however, remains unclear. Similarly, the role of right ventricular twisting will require further investigations. Recent CMR-based studies have shown a dramatic decrease in apical rotation and circumferential strain in the single ventricle of left ventricular morphology compared with the control group (18). Yet in a systemic right ventricle, significant twisting may not be seen (16); the mechanism underlying these disparate observations needs to be clarified. Increasing application of STI, 3-dimensional STI, and CMR will undoubtedly further refine the nuances of right ventricle function in the future. The “systemic right ventricle question” continues to intrigue investigators.

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