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EDITORIAL COMMENT

## Beyond the Short-Term in Neonatal Coarctation\*



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oarctation of the aorta occurs in 1 in 2,500 live births and in approximately 5% to 8% of patients with some form of congenital heart disease. It can be seen in isolation or in combination with more complex congenital heart diseases. This narrowing in the descending aorta can vary from mild to severe and when significant, represents a significant obstruction to flow to the lower body and a significant strain on the left ventricle (LV) (and right ventricle [RV] in fetal life) with subsequent left ventricular hypertrophy (LVH) in some patients. In neonatal life, the first symptom of significant coarctation of the aorta can be LV dysfunction or shock. The goal of therapy is to relieve the obstruction, prevent hypertension and its consequences, and subsequently, improve LV function in the short and long term. Timing for correction of the coarctation has evolved over time as surgery has improved and interventional tools have evolved. Corrective intervention often occurs in the neonatal period (at least 25% of the time), soon after the diagnosis is made. Balloon angioplasty and coarctation stenting, even in the neonatal period, is currently an accepted practice to rescue LV function prior to definitive surgical repair at a later date.

Echo derived strain analysis, both systolic and diastolic, has been used as an early detector of LV dysfunction in a number of myocardial diseases. It can also be used as a way to follow changes in myocardial function with intervention and a change in function over time. The advantages of these measures are that they are available, convenient, portable, computer generated, and reproducible.

In this issue of JACC: Advances, Hysko et al<sup>1</sup> used echo and strain analysis to evaluate biventricular function and immediate and short-term change in function, before and after catheter intervention or surgical intervention for patients with symptomatic neonatal coarctation. In their cohort of 26 patients who presented with LV dysfunction and heart failure, 10 patients underwent balloon angioplasty or stent and 16 patients had surgical repair. Ninety percent of the catheter intervention group had significant LV dysfunction with ejection fraction (EF) <50%; this was present in 38% of the surgical group. Despite many presenting in shock, all patients had successful intervention and had documented improved LV EF at the time of discharge. The authors further compared changes in echo-Doppler derived LV global longitudinal, radial, and circumferential strain and RV-free wall strain using 2-dimensional speckle tracking pre- and postintervention. In addition, the LV peak diastolic radial strain rate, peak diastolic circumferential strain rate, and peak diastolic radial velocity were evaluated pre and post intervention. They found that, in both catheter and surgical intervention groups, LV and RV systolic and diastolic strain indices improved and normalized predischarge. Thus, in the short-term, both interventions resulted in the return to normal of systolic and diastolic function indices in their population of neonates.

Coarctation of the aorta, however, is a complicated disease and presents between the neonatal period and adulthood, dependent on the severity of narrowing and associated symptoms. Once diagnosed, surgical- or catheter-based intervention is recommended. The current study, albeit a small population, suggests that LV dysfunction from isolated coarctation in the neonatal period can be reversed with

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prompt and proper relief of the obstruction. However, longer term follow-up is needed.

Despite adequate relief, it must be pointed out that there is a risk of recurrent coarctation after surgery. The literature has shown that recurrence is higher when patients are treated in the neonatal period compared to repair in childhood, adolescence, or adulthood. Hypertension followed by LVH and LV diastolic dysfunction are complications that can occur with persistent or recurrent coarctation. Recent studies have shown that systemic hypertension in adults causes a decline in global longitudinal strain even when there is normal LV EF. This is more prominent in patients with LVH, so adequate hypertension treatment is imperative to treat or prevent this dysfunction.<sup>2</sup>

With that said, it is important to diagnose coarctation early and treat early in any age group. But this is not enough in this disease. Careful follow up and prompt attention to any recurrent narrowing and hypertension is also important as this can lead to changes in LV systolic and diastolic dysfunction. This was illustrated in one study in adult patients following successful coarctation repair as an adolescent. They found that despite those patients having normal LV EF, measurements of LV global longitudinal and radial strain were found to be abnormal compared to controls. These reductions in strain parameters were again more pronounced in patients with LVH.<sup>3</sup> Even RV global longitudinal strain has been found to be abnormal in adult patients who have LV longitudinal strain abnormalities following coarctation repair.<sup>4</sup> The good news is that repair of recurrent coarctation can reverse these findings. In a group of patients repaired between 8 and 38 years, successful stent implantation improved LV global longitudinal strain in all patients in the short-term. Interestingly, patients who were younger at stent implant had persistent improvement in LV global longitudinal strain at 1 year follow-up. In addition, LV strain abnormalities associated with coarctation can improve over time, even in older patients who had catheter based or surgical repair.<sup>5</sup>

While this study by Hysko et al<sup>1</sup> is important as it shows the benefit of early and adequate intervention in neonates, it is not known whether this will translate into long term benefit. Overall, there is reason to be optimistic about LV function after coarctation or recoarctation repair. But what has not been studied in a systematic way is whether LV systolic and diastolic function can be salvaged in the long term with early intervention, be that in the neonatal period or later in life. In addition, large studies evaluating longitudinal LV systolic and diastolic function in patients who were repaired at various ages at diagnosis or recurrence are needed to guide therapy and prognosticate for patients as they age.

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