

### Exercise capacity and cardiac reserve in children with corrected pulmonary atresia with intact ventricular septum after univentricular and biventricular repair.

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**Introduction:** Management of pulmonary atresia with intact ventricular septum (PAIVS) is challenging and is determined by the degree of the right ventricular hypoplasia after birth. Clinical outcomes of biventricular repair appear favorable to a Fontan operation; however, data on superiority of biventricular repair regarding exercise performance are conflicting. We investigated the response to physical and pharmacological stress in surgically corrected PAIVS patients, with assessment of the presence of myocardial fibrosis.

**Methods:** Sixteen pediatric patients (7 patients after univentricular repair, age  $11.8 \pm 2.6$  years, 7 patients, age and sex matched, after biventricular repair, age  $13.7 \pm 4.5$  years, and 2 patients after one and a half (1.5) ventricular repair underwent a cardiopulmonary exercise test, dobutamine stress magnetic resonance imaging (MRI) and delayed contrast enhanced MRI. The presence of myocardial fibrosis was assessed and the differences between both groups in response to physical and pharmacological stress were compared between the uni- and biventricular group.

**Results:** The univentricular repair group showed impaired exercise capacity in contrast to normal exercise capacity in the biventricular repair group. At rest, heart rate and left ventricle (LV) functional parameters were not different between both groups. With dobutamine, LV ejection fraction increased in both groups. However, LV stroke volume (SV) increased only in patients after biventricular repair ( $+11.3 \pm 5.0$  ml,  $p < .001$ ), but not in patients after univentricular repair ( $-0.04 \pm 3.6$  ml,  $p = .9$ ), HR increase was inadequate in the univentricular repair group. Maximum oxygen consumption ( $VO_2\max$ ) and oxygen-pulse ( $O_2$ -pulse) were strongly correlated with LV-SV during dobutamine (Figure 1) but not to LV-SV at rest. In both groups, no myocardial fibrosis was detected. The results of the 2 patients after 1.5 ventricular repair were comparable to the univentricular repair group.

**Conclusion:** Impaired exercise capacity in children with PAIVS after univentricular repair is related to decreased cardiac reserve due to impaired LV filling during stress, and inadequate chronotropic response. In contrast, pediatric PAIVS patients after biventricular repair show normal exercise capacity and cardiac reserve. These findings support the superiority of biventricular correction of PAIVS over uni- and 1.5 repair during mid-term follow-up.

