

### Effect of age on exercise capacity and cardiac reserve in patients with pulmonary atresia with intact ventricular septum after biventricular repair.

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**Introduction:** In patients with pulmonary atresia with intact ventricular septum (PAIVS) biventricular repair appears to be the optimal treatment option in the absence of significant right ventricular (RV) hypoplasia. However, long term clinical outcome studies are limited. The purpose of this study was to evaluate exercise capacity and cardiac function during stress in children and young adults with PAIVS.

**Methods:** Ten PAIVS patients after biventricular repair, age  $20.4 \pm 3.7$  years, range nine to 43 years, underwent a cardiopulmonary exercise test, dobutamine stress magnetic resonance imaging (DS-MRI) and delayed contrast enhancement MRI. The response to physical and pharmacological stress and presence of myocardial fibrosis were evaluated and the correlation between patients' age and cardiac response to the physical and pharmacological stress was assessed.

**Results:** There was a strong negative correlation between the patients' age and physical exercise capacity (Figure 1-a, 1-b). RV E/A volume ratio at rest was negatively correlated with the patients' age indicating impaired RV diastolic function with age ( $r = -0.70$ ,  $p = 0.02$ ). The patients' age was negatively correlated with biventricular stroke volume (SV) response to pharmacological stress (Figure 1-c, 1-d).  $VO_2\max$  and  $O_2$ -pulse during physical stress were strongly correlated with biventricular SV response to pharmacological stress.  $\Delta RV$ -SV correlated with RV E/A volume ratio ( $r = 0.58$ ,  $p = 0.01$ ).

**Conclusion:** In PAIVS patients after biventricular repair exercise capacity and cardiac reserve decrease with age. This findings appears to be related to impaired diastolic RV function and decreased RV filling during stress, indicating that the function of the relatively small RV in PAIVS deteriorates in time.

**Figure 1:** Correlation between the patients' age and cardiac work indices in response to physical stress (a)  $VO_2\max$ , (b)  $O_2$ -pulse. Correlation between the patients' age and biventricular SV response to pharmacological stress; (c) LV-SV, (d) effective RV-SV.

