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Treatment of Epstein's Anomaly Related Exercise-Induced Pulmonary Hypertension - A Case Report

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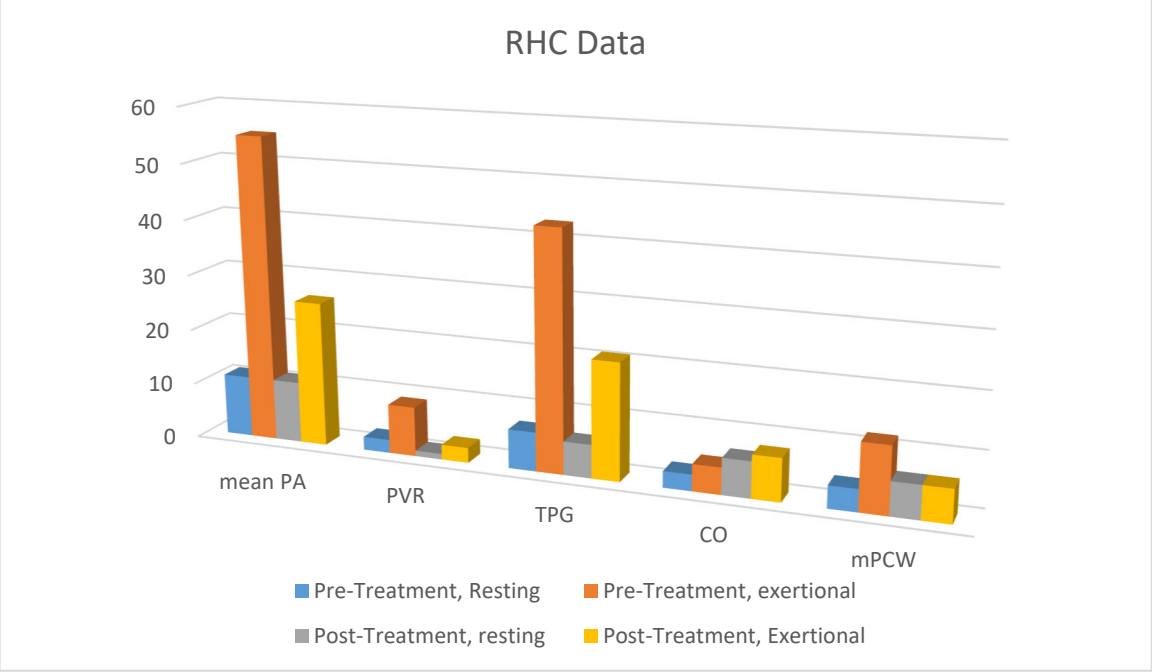
Background: Prevalence of Epstein's anomaly is 1 in 20,000 births in US. When identified as an adult, the treatment is primarily surgical for those with heart failure symptoms. Medical management of heart failure and arrhythmias is recommended for temporary management of symptoms prior to surgical intervention. Literature review reveals lack of data regarding treatment for exertional pulmonary hypertension in symptomatic patients with Epstein's Anomaly.

Methods: 62 year old male with recurrent exertional syncope, severely dilated RV-RA, with clinical RV failure and volume overload. After diuresis, right heart catheterization (RHC) confirmed exertional PAH. Patient was treated with sildenafil 20 mg TID and macitentan 10 mg daily for a year. RHC with resting and exertional data, echocardiogram, six minute walks were performed prior and post medical therapy.

Results: PA mean pressures were normal at rest, however, with minimal exercise, increased to 55 mmHg, without change in bilateral filling pressures with exercise. After 6 months of PDE 5 + ERA dual therapy, patient's syncope resolved, exercise mean PA pressure improved to 26 from 53 mmHg, PVR reduced to 2.7 from 8.9 woods units. Six minute walk distances improved from 197 to 325 meters. WHO functional class improved from IV to II without any subsequent hospitalizations or surgery for treatment of Epstein's Anomaly.

Conclusions: Treatment of adult patients with exertional pulmonary hypertension associated with Epstein's Anomaly with standard PAH therapy may provide significant improvement in functional capacity, resting and exercise hemodynamics, which may in turn reduce right ventricular failure related hospitalizations and potentially lower the risk of surgical complications at time of valve surgery.

Figure 1. RHC Data



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