ABSTRACT

MUSCLE CONTRIBUTIONS TO CARDIAC FUNCTION AND EXERCISE ABILITIES IN PATIENTS WITH FONTAN

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Single ventricle anatomy is the most severe of the congenital heart defects (CHD) requiring a palliative process resulting in complete cavopulmonary flow and a single designated ventricular pumping chamber. The final step of palliation, the Fontan, has become the namesake for this population of CHD patients. Despite improved survival, individuals with Fontan experience long-term sequelae that includes decreased exercise capacity.

Inadequate venous return and diminished cardiac output are primary causes of exercise intolerance in patients with Fontan. The peripheral muscle pump's ability to augment venous return is not fully understood. Impaired Fontan circulation may result in muscle hypoperfusion atrophy, yet the contributions of peripheral muscle to exercise performance requires further investigation. The purpose of this dissertation was to establish the muscle contributions to cardiac function and exercise abilities in patients with Fontan.

A review of cardiopulmonary exercise tests (CPET) to create reference values for cardiac performance was conducted. Regional blood flow to the peripheral muscle during exercise was assessed via near infrared spectroscopy in children and adolescents matched by age and dominant ventricle. Finally, primary and secondary measures of muscle function were assessed in children and adolescents as predictors of aerobic exercise performance (CPET).

Declines in exercise tolerance occur in preadolescence and accelerate throughout adolescence and into adulthood. Deficits in primary measures of muscle including strength, size, and estimated glycogen were seen in patients with Fontan. Secondary measures of a 30-second sit to stand, 6-minute-walk test, and balance also decreased. Predictors of exercise capacity include the 6-minute-walk test, 30-second sit-to-stand, physical activity assessment, and balance scores. Females with Fontan experience more aggressive declines in fitness that is accompanied by significant differences in peripheral muscle perfusion and peripheral muscle strength.

Exercise capacity can be predicted by measures of muscle function. The findings of regional blood flow abnormalities in combination with decreased muscle size, strength, and glycogen content provide evidence that deficits in the peripheral muscle pump exist and likely contribute to the reduced exercise capacity seen in children and adolescents with Fontan. While muscle deficits are apparent, the contributions of physiologic mechanisms versus barriers to physical activity requires investigation.