

**Patient/Family Lay Summary:** FON Case Review Conference July 2023

**Coordinated by:** Children’s Wisconsin and Children’s Hospital Colorado

**Topic:** Physical Activity Programs for Individuals with Fontan Circulation

**The Problem:** Multiple research studies have demonstrated that exercise capacity, or the maximal physical ability that a person is able to sustain, declines over time in individuals with Fontan circulation. Exercise capacity, in turn, is an important outcome that is associated with overall physical health, psychosocial wellness, and quality of life. Multiple factors likely account for this decline in exercise capacity after the Fontan operation, including the unique Fontan physiology, decreased muscle mass, abnormal lung function, and perceptions about the benefits and safety of exercise among providers, patients, and caregivers. Recent studies have shown that physical activity and exercise training can improve exercise capacity in children, adolescents, and adults with Fontan circulation. However, despite these benefits, most patients may not receive formal advice on physical activity, sports, and exercise training during their routine clinic visits.

**Clinical Course of the Cases Presented:**

Case 1 was a 9 year old girl with hypoplastic left heart syndrome (HLHS) and Fontan circulation who presented during a routine clinic visit with symptoms of shortness of breath with activities and feeling that she “can’t breathe in air fast enough” when exercising. A formal cardiopulmonary exercise test (a.k.a. “stress test”) confirmed that she had reduced exercise capacity. Therefore, she was referred to the Children’s Wisconsin “Steppin’ It Up” Physical Activity Program. The goal of this program is to encourage patients to be more physically active with the goal of improving exercise tolerance. During the program, our patient had 4 multidisciplinary in-person clinic visits over 12 months. At these visits, she met with a cardiologist and an exercise physiologist to provide an individualized age-appropriate activity prescription, a psychologist to discuss any psychosocial barriers limiting physical activity, physical and occupational therapists to evaluate and treat any physical limitations to activity, and a dietician. Following the program, she reported marked improvement in her symptoms with exercise and physical activity. During high school, she went on to participate competitively on the diving and rock climbing team. Repeat exercise testing after graduating high school showed that her exercise capacity was maintained and did not decline over time. During the case conference interview, she reported the role that physical activity and sports had on her overall psychosocial health and development.

Case 2 was a 14 year old boy with pulmonary atresia with intact ventricular septum and Fontan circulation who expressed desire to improve his physical fitness and “get faster and stronger.” He was referred to the Children’s Hospital of Colorado “Heart Chargers” Physical Activity Program. This program is a 12-month, home-based, personalized exercise program for patients with congenital heart disease with goal of encouraging physical activity and improving fitness, as a lifestyle change. The program was initially designed for patients with Fontan circulation and more recently has expanded to patients with cardiomyopathy and other forms of congenital heart disease. Unlike the Wisconsin program, the Colorado program utilizes telemedicine visits and measures physical activity remotely with a fitness tracker. During the visits, patients are provided personalized exercise prescriptions that include 30-45 minutes of aerobic, strength-training, and respiratory muscle strengthening exercises, as well as the option to meet with a dietician. Following completion of the 12-month program, our patient demonstrated marked improvement on his cardiopulmonary exercise stress test and participates competitively on his high school Cross Country team.

**Important Points, Lessons Learned, and Potential Solutions:**

1. Multiple factors contribute to the risk for reduced and declining exercise capacity in individuals with Fontan circulation.
2. There is growing evidence that exercise training is safe and effective in improving exercise capacity in people with Fontan circulation.
3. We demonstrated 2 examples of dedicated multidisciplinary physical activity programs within cardiac care centers that have been successful in encouraging patients to be more physical activity by providing individualized exercise prescriptions and monitoring safety of exercise.
4. Challenges experienced by both programs included patients/families not being able to commit to the 12-month long programs, including loss of social support and catchment area, and difficulties with maintaining the program due to difficulties with provider availability and insurance reimbursement.