
Becker’s muscular dystrophy (BMD) is associated with abnormal cardiac findings in 75% of cases; up to one third will develop ventricular dilatation leading to congestive heart failure, at times necessitating cardiac transplant. Candidates are selected from a base of heart failure patients who are usually New York Heart Association (NYHA) class III or IV. Treatment in a phase II cardiac rehabilitation program after transplantation is associated with functional improvement in patients without BMD, but there are no reports of patients with this disorder. We present the case of a 38-year-old man diagnosed with BMD with associated dilated cardiomyopathy. The patient was a NYHA class IIIa and underwent orthotopic cardiac transplantation for intractable heart failure followed by treatment in a phase II cardiac rehabilitation program. At the end of cardiac rehabilitation, his 12-minute walking distance had improved from 716.28 to 929.64m (30% improvement), he had increased his conditioning metabolic equivalent level from 3.5 to 5.5 (55% improvement), he had a weight loss from 81.65 to 78.93kg, and his body mass index changed from 23 to 22kg/m². The patient now has returned to work, is using a stationary bicycle once a day for 30 minutes, and is walking 1 hour a day. This suggests that treatment in a cardiac rehabilitation program is effective in patients with BMD after cardiac transplant.

Key Words: Case report; Muscular dystrophy, Duchenne’s; Rehabilitation.

© 2005 by the American Congress of Rehabilitation Medicine and the American Academy of Physical Medicine and Rehabilitation

Becker’s muscular dystrophy (BMD) belongs to a small group of disorders known as the dystrophinopathies. Dystrophin is a large structural protein (3685 amino acids, 427kd) that is present in all types of muscle, as well as the brain. Dystrophin seems to play a role in membrane structure and stability. Because of the large size of the gene, it is prone to mutation. Disorders of dystrophin primarily affect males because the gene is on the X chromosome. There is a spectrum of disease, with 3 clinical entities. In Duchenne’s muscular dystrophy (DMD), there is a frameshift mutation that results in a complete lack of dystrophin. This causes a rapidly progressive degeneration of skeletal muscle, usually presenting in the first 5 years of life, with death occurring in late adolescence or early young adulthood, commonly from pulmonary complications. Although there is often some cardiac involvement, it is generally much less progressive than the skeletal muscle involvement and is, in general, not clinically significant. At the other end of the spectrum is X-linked dilative cardiomyopathy (XLDCM). In this disorder, mutation results in a truncated dystrophin protein that is still functional. In this disorder, the heart is much more involved, leading to a rapidly progressive dilative cardiomyopathy in adolescence, with little skeletal muscle involvement. The grey area zone between these 2 extremes is BMD.

BMD was first described by Becker in 1955 as a disorder similar to DMD, but with much slower progression.1 As with XLDCM, a truncated dystrophin protein is present. Many mutations that result in a dysfunctional dystrophin are possible, leading to a wide phenotypic variation depending on the function of the abnormal protein. BMD may be recognized in adolescence with mild weakness and pseudohypertrophy,2 or much later in life with primarily cardiac involvement.3 This variation is simply a result of the large number of possible mutations in the dystrophin gene and how dysfunctional the dystrophin protein is to skeletal and cardiac muscle. A wide range of cardiac manifestations have been documented in BMD, from very subtle signs of cardiac involvement to severe myocardial disease leading to death or to refractory heart failure treated by heart transplantation. BMD is associated with abnormal cardiac features in 75% of patients; up to one third will develop ventricular dilatation leading to congestive heart failure. A number of authors have reported cases of cardiac transplantation in patients with BMD,4,5 but have not discussed the rehabilitation aspects. The benefits and risks of exercise in patients with neuromuscular disorders (NMDs) are controversial. We present objective findings of the benefits of cardiac rehabilitation following orthotopic heart transplant (OHT) in a patient with BMD.

CASE DESCRIPTION

JD is a 38-year-old man who was diagnosed with BMD during adolescence. He developed congestive heart failure (CHF) during his fourth decade as a result of dilated cardiomyopathy. Before developing CHF, JD was quite functional, independent with ambulation and self-care, and employed full-time. Pharmacologic treatment was instituted, eventually progressing to home dobutamine infusion for approximately 4 months. JD was classified as a New York Heart Association (NYHA) class IIIa (table 1). JD was placed on the cardiac transplant list, and eventually received an OHT. His hospitalization was uncomplicated and his CHF resolved before discharge. After completing phase I cardiac rehabilitation, JD was classified as NYHA class II and was discharged to his home, with follow-up in an outpatient phase II cardiac rehabilitation program.

Before admission into the phase II cardiac rehabilitation program, JD underwent cardiopulmonary exercise testing to
evaluate his functional capacity and to determine a baseline exercise prescription. He exercised on an electronically braked cycle ergometer using a ramping protocol increasing 5W/min. His resting seated heart rate and blood pressure were 107 beats per minute (bpm) and 130/80mmHg, respectively. The test was stopped due to general fatigue. He achieved a peak workload of 15W, a peak heart rate of 115bpm, and a peak blood pressure of 152/80mmHg. His peak exercise electrocardiogram (ECG) revealed a sinus tachycardia with no arrhythmias or ischemic ECG changes. He achieved a peak oxygen uptake of 11.4mL.kg⁻¹.min⁻¹ or a peak level of 3.3 metabolic equivalents (METS).

JD underwent treatment in the phase II cardiac rehabilitation program for 3 times 1 week for 12 weeks. His adherence and compliance to the prescribed program were excellent. JD’s therapy was carried out on a Schwinn Airdyne ergometer, a NuStep recumbent arm and leg ergometer, and a recumbent cycle ergometer. JD’s exercise prescription was established at a target heart rate range of 110 to 132bpm, a rating of perceived exertion (RPE) of 11 to 15 (on a Borg scale of 6–20), or a target MET level of 3.0 to 5.0 METS. The target heart rate was based on recommendations for cardiac transplant patients, and not the preparticipation cardiopulmonary exercise, as that was limited by general fatigue and not a true maximal exercise test. Table 2 summarizes JD’s progress from the beginning to the end of the program. All of JD’s ECG rhythm strips were within normative limits without any indications of heart block and/or arrhythmias.

JD achieved significant progress on 3 successive 12-minute walks. He covered a distance of 716m during his third cardiac therapy session, 899m during his 18th cardiac therapy session, and 929m during his 36th cardiac therapy session. This represents a 30% improvement in his walking distance for 12 minutes over 12 weeks. In addition, JD’s weight decreased from 81.6 to 78.9kg and his body mass index improved from 23 to 22kg/m².

After completion of his phase II program, JD was transitioned to a home exercise program. This consisted of exercising for 15 minutes on the Schwinn Airdyne ergometer and walking for a minimum of 60 minutes. He has also returned to work. JD had regular follow-up examinations by his cardiologist, and there was no evidence of rejection.

**DISCUSSION**

Cardiac rehabilitation after OHT offers unique challenges in the patient with BMD. First, all patients with OHT have a new resting cardiovascular physiology as well as response to exercise. The major change is the heart has been denervated. This results in a higher resting heart rate and a much lower maximal exercise heart rate. Increased cardiac output with exercise is more dependent on intrinsic cardiac controls (ie, Frank-Starling mechanism). Resting blood pressures are in the normative range. The pretransplant changes in peripheral physiology remains the same in patients with OHT for a prolonged period of time after heart transplantation. Skeletal muscle biopsies 6 weeks after OHT are grossly abnormal and include intracellular lipid and glycogen accumulations, increased pinocytosis, and markedly thickened capillary basement membranes. A number of reviews are available on post-OHT physiology. Specific to cardiac rehabilitation after OHT in patients with BMD are the concerns about exercise in NMDs. Bennett and Knowlton raised concern regarding “overwork” in diseases with partial innervation, what they felt was the possibility of “continued voluntary activity to overuse a muscle to a point where long-lasting impairment of that muscle results.” Clinical experience was cited, and 5 cases were discussed (4 postpolio, 1 cervical spinal cord injury). There are a number of reports of improved or maintained muscular strength and endurance in various NMD in prospective studies.

**CONCLUSIONS**

We present the first objective data that cardiac rehabilitation is beneficial after OHT in patients with BMD. Because of the rarity of this condition, it is likely impossible to perform well-controlled studies to examine the risks and benefits of exercise in these patients. Cases and case series should continue to be reported to provide more information about the care of these patients.

**References**


Suppliers
a. Nautilus Inc, 16400 SE Nautilus Dr, Vancouver, WA 98683.
b. NuStep Inc, 5111 Venture Dr, Ann Arbor, MI 48108.