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Integrating Upper- and Lower-Limb Exercises in Virtual Reality Video Games: A Novel Approach in Pediatric Clinical Exercise Rehabilitation for Duchenne Muscular Dystrophy

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Abstract
In clinical exercise rehabilitation, training of the lower extremities is recognized as a key contributor in delaying the decline in functional status and the onset of disability in children with Duchenne muscular dystrophy. Recent evidence revealing the importance of upper-limb exercise as an adjunct intervention has led to the evaluation of clinical trials regarding the integration of exercise modalities involving the upper and lower extremities. One method to integrate both modalities of exercise is through the use of interactive video game (exergaming) technology, which is currently an active area of research to improve neurorehabilitation outcomes in children with developmental disabilities. We propose that a novel approach involving clinical exercises of the upper and lower extremities in virtual reality video games can lead to marked enhancements in the functional status of children with Duchenne muscular dystrophy. **Health & Fitness Journal of Canada 2016;9(4):22-28.**

Keywords: Duchenne Muscular Dystrophy, Video Game-Based Therapy, Exergaming, Clinical Exercise Rehabilitation, Physical Activity, Health Promotion

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Introduction

Duchenne muscular dystrophy (DMD) is an inherited X chromosome-linked recessive neuromuscular disease that affects 1 in 3600-6000 live male births (Emery, 1991). The pathogenesis of DMD is characterized by the absence of dystrophin (a protein that maintains muscle integrity) leading to progressive degeneration of muscle function, respiratory failure and subsequent premature death. In skeletal musculature, continual muscular disuse leads to wheelchair dependency, reduction in the capacity to perform activities of daily living, and a decline in health-related quality of life (Kanagawa and Toda, 2006). With increasing life expectancy as a result of advancements in disease-retarding treatments, improving the quality of life has become increasingly important in palliative management for DMD (Jansen et al., 2015; Kohler et al., 2009). Because ambulatory status is associated with quality of life, delaying the loss of functional abilities in the lower extremities has been a clinical focus in pediatric rehabilitation for DMD.

To preserve strength in the lower extremities in children with DMD, assisted cycling has been the conventional focus in clinical exercise rehabilitation. However, there is growing support for the use of upper-limb exercise as an adjunct intervention because ambulatory status is strongly correlated with the functionality of upper limbs in children with DMD (Alemdaroğlu et al., 2015). The evidence suggests that an integration of upper- and lower-limb exercises can lead to greater preservation of skeletal muscle endurance and delay in the loss of ambulation.

One method to integrate upper- and lower-limb exercises is through the use of video game technology. The integration of video games in physical rehabilitation programs has been shown to achieve similar health-benefit results in comparison to conventional therapy alone (Bonnechère et al., 2016). A novel strategy in pediatric rehabilitation for DMD is the use of virtual reality video games as a platform to integrate clinical exercises that involve the upper and lower extremities.

The primary purpose of this narrative review and commentary is to evaluate contemporary evidence regarding the use of upper- and lower-limb exercise as a rehabilitation strategy for children with DMD, and to provide an evidence-based recommendation on how the integration of these exercises in virtual reality video games can elicit marked enhancements in their functional statuses.

Key Findings

Recent advancements in clinical research have generated insight regarding the efficacy of bicycle training programs on the functional status in children with dystrophinopathies (Huijben et al., 2015; Jansen et al., 2013; Sveen et al., 2008). Consistent with current international guidelines that recommend submaximal-intensity exercises in DMD management (Bushby et al., 2010), the literature suggests that low-intensity bicycle training is a safe, feasible and beneficial protocol to preserve skeletal muscle endurance, ambulatory status and functional abilities in children with DMD.

Jansen et al. (2010) demonstrated that an assisted, low-intensity cycling program can preserve skeletal muscle endurance and delay the loss of functional abilities in boys with DMD. In this randomized controlled study, 30 ambulant or wheelchair-dependent boys with DMD were assigned to one of two groups: intervention (24-week cycling program) or control (24-week cycling program following a 24-week waiting period). The exercise protocol consisted of 15 min cycling sessions 5 times per week. Participants were instructed to use both arms and legs to cycle at a constant speed of 65 rpm, assisted by electrical motor support on the mobility trainer. The Motor Function Measure and the Assisted 6-Minute Cycling Test were the outcome measures in this study. The Motor Function Measure score (100%) measured the following functional abilities: 1) standing and transfers; 2) axial and proximal motor functions; and 3) distal motor functions (Bérard et al., 2005). The Assisted 6-Minute Cycling Test is an endurance test of the arms and legs, in which participants were instructed to complete as many revolutions as possible in 6 min (Jansen et al., 2012). Outcome measures were assessed before, during and after the cycling task.

Three key findings emerged from the study by Jansen et al. (2013). First, it was observed that Motor Function Measure scores were stable in the intervention group, but significantly decreased in the control group, suggesting that a low-intensity cycling program is sufficient to prevent functional deterioration in children with DMD. In fact, further analysis revealed that 6 months of assisted bicycle training prevented a 6.3% reduction in functional deterioration from the total Motor Function Measure score during the intervention period as observed in the control group (Jansen et al., 2013). Second, no significant group differences were observed for the Assisted 6-minute Cycling Test. One interpretation of this finding is that low-intensity cycling does not impose the muscular and aerobic demands to physical activity as observed in moderate-to-vigorous intensity exercises. Third, no adverse events were observed during the study. Collectively, the findings suggest that a low-intensity protocol may be a safe, feasible and beneficial intervention for children with DMD irrespective of their ambulatory status (Jansen et al., 2013). Similar findings were observed in a follow-up study when an assisted, low-intensity bicycle training program was individualized for a 9-yr-old ambulatory girl symptomatic of an intermediate form of DMD (Huijben et al., 2015).

Collectively, the evidence suggests that a low-intensity, arm and leg bicycle-training program is sufficient to preserve skeletal muscle endurance in children with DMD. In particular, the evidence highlights the clinical importance of upper-limb exercises, that improving functional ability of the upper extremities in the early stages of DMD can accrue marked benefits in preserving functional status in the later stages of the disease. The findings by Alemdaroğlu et al. (2015) provide additional support for this claim.

In the study by Alemdaroğlu et al. (2015), 24 ambulatory children diagnosed with DMD and living with preserved upper extremity function were randomly assigned to one of two groups: intervention (arm ergometry cycling: 40 min sessions, 3 times weekly) or control (upper-extremity range of motion exercises: 40 min sessions, 5 times weekly). Exercises were performed over 8 weeks, and outcome measures were assessed pre- and post-training. The major finding was that both forms of exercise positively impacted arm function, endurance and ambulation at different levels (Alemdaroğlu et al., 2015). Not surprisingly, most gains were accrued from arm cycling, which elicited significant positive effects on muscular endurance, performance of daily activities, arm function and ambulation status, but not muscle strength; in contrast, range of motion exercises improved grip strength and muscular endurance (Alemdaroğlu et al., 2015).

The observations made by Alemdaroğlu et al. (2015) are clinically relevant to contemporary DMD management strategies. While upper-extremity range of motion exercises only preserved strength in distal upper extremity muscles, arm cycling was observed to protect the strength of proximal muscles, which are significant contributors to upper extremity function (Alemdaroğlu et al., 2015). However, since functional abilities are lost in a proximal-to-distal manner, preserving proximal function via arm cycle exercises can delay the loss of distal functions (e.g., grip strength) that are also integral to performance in activities of daily living. Thus, by coupling arm cycling and range of motion training, the benefits accrued from the isolated strengthening exercises may contribute significantly to the preservation of functional abilities in the later stages of DMD. The findings and implications made by Alemdaroğlu et al. (2015) suggest that integrating upper-limb exercises with traditional lower-limb interventions can markedly improve functional status in children with DMD.

One method to integrate both upper- and lower-limb exercises is through the use of video game technology. There is growing evidence in support of using interactive video games as a supplementary to conventional strategies. For instance, a systematic review of 126 investigations revealed that in majority of the cases, physical rehabilitation programs that integrated video game-based therapy conferred similar health-benefit results as observed in conventional therapy alone (Bonnechère et al., 2016). This systematic review suggests that non-clinical video game-based therapy can be beneficial in training programs for children with DMD. To build and extend upon this analysis, we postulate that video games with a clinical focus can lead to even greater neurorehabilitation outcomes. Virtual reality video games offer this possibility.

A novel approach in pediatric rehabilitation for DMD is the use of virtual reality video games that integrate clinical exercises involving the upper and lower extremities. The exercises described by Jansen et al. (2013) and Alemdaroğlu et al. (2015) can be adapted in virtual reality settings. For instance, while low-intensity stationary cycling of the arms and legs can be used to simulate ambulation in a virtual environment, range of motion exercises allow participants to practice their reach for, and transfer of, virtual objects. Applying virtual reality games to incorporate upper-limb range of motion exercises is the novel factor. Several lines of evidence provide support for the use of interactive video games to improve upper-limb movements in children with developmental disabilities.

Range of motion exercises can be integrated into virtual reality settings via motion capture devices, which have been used in conjunction with commercially-available consoles. For instance, Microsoft's Xbox™ Kinect motion capture system (Luna-Oliva et al., 2013; Zoccolillo et al., 2015), Nintendo's Wii™ Fit software (AlSaif and Alsenany, 2015; Tarakci et al., 2013), and Sony's PlayStation™ EyeToy device (Jannink et al., 2008; Sandlund et al., 2011) are actively being investigated for their efficacy as an adjunct therapy to improve upper limb movements in children with cerebral palsy (a non-progressive, developmental movement and postural disorder). Other innovative approaches including Scratch 2.0, an augmented-reality program that creates a real world and virtual reality interface for physical interaction, have also been used to improve neurorehabilitation outcomes in children with developmental disabilities (Lin and Chang, 2015). The combined use of a stationary bike and a virtual-reality video game is a novel, evidence-based approach that integrates clinical exercises involving the upper and lower extremities to improve functional status in children with DMD.

Integrating upper- and lower-limb exercises in virtual reality video games confers several advantages: 1) arm and leg cycling activities retain clinical requirements; 2) different upper-limb physical activities (e.g., range of motion exercises) can be integrated with cycling tasks; 3) virtual reality video games can be adapted (e.g., selection of difficulty level) to individualize exercise prescriptions (Zoccolillo et al., 2015); 4) the cost of commercial consoles is low; and 5) the appeal in video games can optimize therapy compliance, since adherence to therapy has been identified to be a barrier for children, particularly in home settings (Harris and Reid, 2005).

This novel approach in clinical exercise rehabilitation builds and extends upon recent advancements in research regarding the use of video game technology to improve neurorehabilitation outcomes in children with developmental disabilities (Knights et al., 2016). The application of upper- and lower-limb exercises as described in this review has been shown to be safe (e.g., a very low risk for exercise-related adverse events), feasible, and beneficial interventions to preserve skeletal muscle endurance, ambulatory status, and functional abilities in children with DMD (Alemdaroğlu et al., 2015; Jansen et al., 2015). The efficacy of this approach can be tested using the Motor Function Measure and the Assisted 6-Minute Cycling Test, which are safe, feasible and easily reproducible outcome measures to assess functional abilities and skeletal muscle endurance, respectively (Bérard et al., 2005; Jansen et al., 2012).

Conclusion

In conventional practice, upper-limb exercises are often introduced into rehabilitation when functional ability of the upper extremities is already compromised (often in the later stages of the disease) (Alemdaroğlu et al., 2015). In the literature, several lines of evidence support the application of upper-limb exercises in conjunction with lower-limb interventions, because preserving the functionality of the upper extremity in the early stages of DMD can assist in delaying the loss of functional abilities to perform activities of daily living with the onset of disease progression (Alemdaroğlu et al., 2015; Jansen et al., 2015).

The novel approach of integrating clinical exercises involving the upper and lower extremities in virtual reality video games has the potential to improve the functional status in children with DMD. The efficacy of this evidence-based recommendation can benefit from future research; the justification for further research stems from the relatively limited volume and quality of evidence in support of a blueprint for best practice in clinical exercise rehabilitation for children with DMD.

Authors' Qualifications

The authors' qualifications are as follows: Henry Lai, BSc, BKIN; Darren Warburton, MSc, PhD, HFFC-CEP.

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