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Gait pattern in Duchenne muscular dystrophy

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ABSTRACT

We investigated the gait pattern of 21 patients with Duchenne muscular dystrophy (DMD), compared to 10 healthy controls through 3D Gait Analysis.

An overall observation of gait pattern in our DMD patients when compared to controls confirmed the data previously reported for small dystrophic groups. An excessive anterior tilt of pelvis and abnormal knee pattern in loading response phase were found. Since during the swing phase the DMD foot is too plantarflexed, patients adopt a higher flexion and abduction of the hip in order to advance the swinging limb. Velocity and cadence of DMD patients resulted similar to those calculated for healthy subjects, whereas stride length was reduced and step width was increased.

We then divided the DMD patients in to two subgroups (treated with steroids and untreated), and we observed that the only statistically significant differences between the two groups in Gait Analysis parameters were found for the maximum of ankle power.

3D Gait Analysis gives objective and quantitative information about the gait pattern and the deviations due to muscular situation of DMD subjects; being our study a single moment evaluation, it is otherwise unable to unravel changes only detectable through serial analysis during the time course of the disease and, if any, due to the treatment.

1. Introduction

Duchenne muscular dystrophy (DMD) is the commonest childhood muscular dystrophy with a worldwide incidence of 1 in 3500 male live births. It is an X-linked recessive disorder, due to mutations in the dystrophin gene. Untreated, boys with DMD become progressively weak during their childhood and stop ambulation at a mean age of 9 years. Confinement to a wheelchair is followed by the development of scoliosis, respiratory failure and cardiomyopathy. Without intervention, the mean age at death is 19 years [1,2]; no curative treatment is known.

Some studies have reported substantial long-term functional benefits from the steroid therapy: prolonged ambulation into the mid-teens, reduced severity of scoliosis, major preservation of respiratory and cardiac function [3–18] reflected by changes in weakness, which affects the proximal muscle groups first and the distal ones later, compromises the child's posture and gait. A dynamic quantitative assessment of gait in DMD patients would be important in evaluating the progression of the disease and the benefits of any therapy [19]. Few studies focused on gait of DMD children [20–23], sample sizes were small and/or not homogenous.

The aim of the present study was the characterization of the gait pattern of a large group of DMD children at a single time point of their clinical history compared to healthy age and sex matched controls using three-dimensional Gait Analysis.

2. Materials and methods

2.1. Patients

Twenty one DMD children were involved in this study; the diagnosis of DMD was established according to internationally accepted criteria [24]: progressive muscular weakness, increased muscle plasma enzymes, muscle biopsy identifying fiber degeneration and absence of the dystrophin protein, alterations in the dystrophin gene (deletions or other genetic alteration).

Exclusion criteria included absence of additional conditions such as cerebral palsy, behavioural and/or psychiatric disturbances, acquired brain or spinal injuries, deafness, severe visual impairment, epilepsy and the ability to walk independently, without aids.

Ten healthy children, age and sex matched, were recruited (all boys, mean age: 88.7 ± 14.0 months). Patients and controls did not differ significantly in age, body weight and height (Table 1).

Ten out of 21 DMD children began steroids treatment in other hospitals before coming to ours. Four children were under prednisone (0.75 mg/kg day) and six under deflazacort treatment (0.9 mg/kg day) [5,11,15]. Treatment was started at a

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