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Performance Index for in Home Assessment of Motion Abilities in Ataxia Telangiectasia: A Pilot Study

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Abstract: *Background.* It has been shown in the very recent literature that human walking generates rhythmic motor patterns with hidden time harmonic structures that are represented (at the subject's comfortable speed) by the occurrence of the golden ratio as the the ratio of the durations of specific walking gait subphases. Such harmonic proportions may be affected—partially or even totally destroyed—by several neurological and/or systemic disorders, thus drastically reducing the smooth, graceful, and melodic flow of movements and altering gait self-similarities. *Aim.* In this paper we aim at, preliminarily, showing the reliability of a technologically assisted methodology—performed with an easy to use wearable motion capture system—for the evaluation of motion abilities in Ataxia-Telangiectasia (AT), a rare infantile onset neurodegenerative disorder, whose typical neurological manifestations include progressive gait unbalance and the disturbance of motor coordination. *Methods.* Such an experimental methodology relies, for the first time, on the most recent accurate and objective outcome measures of gait recursivity and harmonicity and symmetry and double support subphase consistency, applied to three AT patients with different ranges of AT severity. *Results.* The quantification of the level of the distortions of harmonic temporal proportions is shown to include the qualitative evaluations of the three AT patients provided by clinicians. *Conclusions.* Easy to use wearable motion capture systems might be used to evaluate AT motion abilities through recursivity and harmonicity and symmetry (quantitative) outcome measures.

Keywords: gait analysis; wearable sensors; rhythmic motor pattern; time harmonic structure; golden ratio; neurological disorders



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1. Introduction

Ataxia Telangiectasia (AT; MIM # 208900) is a rare genetic disorder characterized by early onset ataxia (lack of coordination of movements), neurodegeneration and multisystem involvement, including immunological defects, organ failure and predisposition to cancer [1,2]. The incidence worldwide is estimated to span from 1:40,000 to 1:100,000 individuals, and 1:300,000 in the West Midlands population. From a clinical point of view, AT is characterized by impaired coordination of the movement of voluntary muscles, with gait unbalance, oculomotor apraxia, altered speech fluency, anterior horn cell degeneration, peripheral neuropathy, and a cerebellar cognitive affective psychological profile. Despite the disease affecting, overall, the central and peripheral nervous system, the involvement