

Title

Rett syndrome: establishing a novel outcome measure for walking activity in an era of clinical trials for rare disorders

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Keywords

Rett syndrome, physical activity, accuracy, repeatability

Abstract

Background: Rett syndrome is a pervasive neurological disorder with impaired gait as one criterion. This study investigated the capacity of three accelerometer-type devices to measure walking activity in Rett syndrome.

Methods: Twenty-six participants (mean 18 years, SD 8) wore an Actigraph, ActivPAL and StepWatch Activity Monitor (SAM) during a video-taped session of activities. Agreement was determined between step-counts derived from each accelerometer and observation. Repeatability of SAM-derived step counts was determined using pairs of one-minute epochs

during which the same participant was observed to walk with the same cadence.

Results: The mean difference (limit of agreement) for the Actigraph, ActivPAL and SAM were 41 (SD 33), -16 (SD 21) and -1 (SD 16) steps/min, respectively. Agreement was influenced by a device/cadence interaction (p < 0.001) with greater under-recording at higher cadences. For SAM data, repeatability of step-count pairs was excellent (intraclass correlation coefficient 0.91, 95% CI 0.79-0.96). The standard error of measurement was 6 steps/min and we would be 95% confident that a change ≥17 steps/min would be greater than within-subject measurement error.

Conclusions: The capacity of the SAM to measure physical activity in Rett syndrome allows focus on participation-based activities in clinical practice and clinical trials.

Introduction

Rett syndrome is a rare neurological disorder usually caused by a mutation in the X-linked *MECP2* gene [1]. Following largely normal early development, there is period of developmental regression with loss of hand and communication skills and development of hand stereotypies and impaired gait [2]. The resultant disability is severe with dependence for most activities of daily living [3] and co-morbidities such as epilepsy [4], poor growth [5] and scoliosis [6] may ensue. Nevertheless, using video data from individuals in the Australian Rett Syndrome Database (n = 99) we found that approximately 43% of girls and women were able to walk without assistance with a further 27% able to walk short distances with some assistance [7]. The ability to walk is in part related to the specific *MECP2* mutation [8] and may or may not decline with age [9].

Beyond the measurement of gross motor skill levels [7], measures of functional performance, such as step counts, can assist an understanding of the extent gross motor skills are used during daily life. There is growing knowledge and understanding of the biological basis for neurological disorders such as Rett syndrome [10] and clinical trials are planned to test therapeutics that hope to reverse some signs [11]. Therefore validated measures of physical activity, such as step counts, are important not only to monitor function

during daily life but also to use as outcome measures in these new trials of pharmaceutical agents.

Accelerometers measure the rate and magnitude of body movements and are able to calculate the intensity, frequency and duration of physical activity [12]. Models with variable levels of sophistication are available and there is potential to measure physical activity in persons with atypical walking patterns. For example, there is some validation of the Actigraph in children with cerebral palsy [13] and in those with acquired head injury [14]; the ActivPAL has been used to characterize activity in patients with Parkinson Disease [15]; and the StepWatch Activity Monitor[™] (SAM) has produced accurate and reliable measures in the elderly [16], those with respiratory conditions who walk very slowly [17] and Alzheimer's disease [18]. Each is worn at a different body location, steps are counted using different algorithms and their comparative application in Rett syndrome is not known. We were the first to collect initial data on the accuracy of the SAM in 12 girls and women with Rett syndrome [19]. Although the sample size was small and the SAM under-reported steps where gait was extremely slow [19], accuracy was promising in this sample which comprised participants with a range of gross motor skill levels.

The population-based Australian Rett Syndrome Database (ARSD) was established in 1993 and collects data longitudinally from families with a daughter affected by Rett syndrome [20]. Recruiting families from the ARSD, this study sought to expand our previous investigation and examine the capacity of three different accelerometer-type devices (Actigraph, ActivPAL and SAM) to measure step counts accurately and consistently.

Methods

Participants

Families from the ARSD [20] who lived in either Western Australia or Victoria in Australia were recruited if their daughter with Rett syndrome was able to walk independently or with

assistance. Diagnosis of Rett syndrome was confirmed using diagnostic criteria [2] or the presence of a pathogenic *MECP2* mutation. Families were visited at home or school for data collection.

Devices

Three different accelerometer-type devices were used. The Actigraph[™] GTX3 (ActiGraph, Pensacola, FL, USA) is a triaxial accelerometer with inclinometer that attaches to the waist using an elasticized belt and measures activity counts, vector magnitude and steps taken. The ActivPAL[™] (PAL Technologies, Glasgow, UK) is a uniaxial accelerometer that attaches to the thigh using an adhesive pad. It has dual sensors including an inclinometer. This device detects thigh inclination and limb movement and provides information pertaining to the number of steps taken. It also separates time spent in supine and sitting from time in standing and walking. The SAM (OrthoCare Innovations, WA, USA) is an accelerometertype device that attaches to the ankle using a Velcro strap and responds to acceleration, position and timing. It measures the number of steps taken.

Procedures and measures

Each device was programmed using proprietary instructions. Thereafter each participant was fitted with all three devices and encouraged to undertake physical activities for 20 to 30 minutes with supervision and assistance as necessary. Activities, which included walking at different speeds and on different terrains and inclines, were video-taped by an investigator. For the video-taped observations, a step was counted when the foot cleared the ground and moved in either a forward, sideways or backward direction. Two trained observers counted steps during each video session and mean values were used for analyses.

General and complex gross motor skills were measured with the recently developed Gross Motor Scale for Rett syndrome [7]. The general gross motor skill subscale comprised 10 items pertaining to tasks such as sitting, standing and walking as well as standing up from sitting. The maximum score is 40 with higher scores representing better general gross motor skills. The complex gross motor skills subscale comprised five items that were more complex in nature and include walking on a slope, stepping over an obstacle, bending down to touch the floor, standing up from the floor and running. The maximum score is 20 with higher scores representing better complex gross motor skills [7].

Ethics approval for this study was provided by the Human Research Ethics Committee at Curtin University, Western Australia and families provided written informed consent for their daughter to participate in the study.

Analyses

Agreement

The numbers of steps observed on the video-tapes was the criterion measure. For each device, Bland-Altman analyses [21] were conducted to determine agreement (i.e. accuracy) between the average step count each minute derived from the video-tape and the average step count each minute, recorded by the device. Hierarchical random effects modelling was used to assess the influence of device type and cadence on agreement. The model used agreement in step count (device – video-tape) as the dependent variable with minute (within participant) as random effects and device type and cadence as fixed effect predictors. In addition, a model incorporating an interaction effect of accelerometer type with cadence was assessed to determine whether accuracy varied with cadence.

Repeatability

For the best performing device, repeatability was assessed using step counts recorded by the device over two separate one-minute epochs that were characterized by an identical number of video-taped steps. If there was more than one pair of minutes that met this criterion, the first pair was analyzed. Using these data, an intraclass correlation coefficient (ICC) was calculated. The standard error of measurement, defined as the square root of the mean square error term using repeated measures analysis of variance, was determined and then used to calculate the minimal detectable difference (SEM x $\sqrt{2}$ x 1.96) [22].

Data are expressed as mean (SD) unless otherwise specified. p < 0.05 was used to denote statistical significance.

Results

In December 2012, the ARSD included 392 girls and women born since 1976 of whom 64 (16.3%) had died since its inception. Twenty-nine families were contacted regarding participation in the study and 28 provided consent giving a recruitment fraction of 96%. Two girls were unwell at the time of assessment and therefore 26 girls and women (mean age of 18 years, SD 8) participated. Most of the common mutation categories were represented (C-terminal [n=4], p.R294X [n=4], p.R133C [n=3], p.R270X [n=3], p.R168X [n=2], large deletion [n=2], p.R306C [n=1], p.T158M [n=1], p.R255X [n=1], p.R133H [n=1], other [n=1]). Three did not have a pathogenic mutation. Twenty-one (80.8%) were able to walk independently and five (19.2%) needed assistance. Scores on the general motor scale ranged from 15 to 40 and on the complex motor scale ranged from 5 to 20. The mean duration of the video-taped session of activities was 12.1 minutes (SD 6.5) in duration and a total of 313 minutes of activity were available for analysis. All participants appeared comfortable wearing the three devices and no participant jogged or ran during the testing session.

Agreement

Bland-Altman plots showing agreement between the average steps/minute for each participant as observed in the video-tape with that derived from each of the devices are presented in figures 1, 2 and 3. The SAM was inadvertently not activated for one girl giving SAM data for 25 of the 26 girls and women. The mean difference (limit of agreement) for the Actigraph, ActivPAL and SAM were -41 (SD 33) steps/minute, -15 (SD 21) steps/minute and -1 (SD 16) steps/minute, respectively.

Insert figure 1, 2 and 3 about here

The mean cadence for all video-taped minutes was 60 (SD 24) steps/minute. The random effects model confirmed that step counts derived using the SAM had the best agreement with the steps counted during the video-taped activities. Agreement varied by device (p < 0.001) and compared to the SAM, on average the Actigraph under-recorded by 41 steps/minute (95% confidence interval [CI], 39 to 43) and the ActivPAL under-recorded by 15 steps/minute (95% CI, 13 to 17). Agreement was also influenced by cadence (p < 0.001) and an interaction effect between device and cadence (p < 0.001). All devices showed a tendency to under-record at high cadences with the Actigraph and ActivPAL under-recording also at slow cadences (figure 4).

Insert figure 4 about here

Repeatability

The video-tape analysis revealed that 20 participants took the same number of steps in at least two different one-minute epochs. Reliability of the step count pairs as measured by the SAM was strong (ICC 0.92, 95% CI 0.82 to 0.97). The standard error of measurement was 6 steps/minute and the minimal detectable difference was 17 steps/minute, indicating that an observed difference using the SAM on the same individual of at least this magnitude would be necessary to be 95% confident that the difference was greater than measurement error.

Discussion

We have extended our previous assessment of the measurement properties of the SAM in Rett syndrome [19] to include a larger sample size and the testing of additional accelerometer-type devices. We chose to investigate these particular devices as preliminary data supporting their validity had been previously reported in persons with other neurological disorders [13-16,18]. Compared with steps counted during a video-tape of the assessment session, superior agreement was demonstrated between the steps derived using the SAM compared with the ActiGraph and the ActivPAL. This was true on average across the sample and over most cadences. Step counts measured via the SAM also demonstrated good repeatability in this population.

The Actigraph showed the greatest differences from our observed video steps, particularly at higher cadences. Using earlier models of this device, moderately strong relationships between activity counts and observed activity were observed in children with cerebral palsy [13] and strong correlations between activity counts and self-reported physical activity were found in adults with multiple sclerosis [23]. Actigraph data correctly classified bouts of low and moderate physical activity but underestimated bouts of vigorous activity in adults with traumatic brain injury [14]. However, each of these studies examined similarities in measures using different devices rather than agreement with a gold standard measure which examines differences. More recently in the general population, the Actigraph GTX3 which was used in the current study underestimated steps particularly during slower cadences (< 67m/min) compared with directly observed steps [24]. Our data also underestimated steps, in part due to slow cadences but also likely due to the abnormal gait patterns in our sample. Specifically, normal walking is characterized by a co-ordinated pattern of movement of the centre of gravity in horizontal, lateral and vertical planes [25]. The triaxial Actigraph records accelerations in all three planes and uses algorithms to then calculate step counts from the recorded acceleration episodes. Inspecting the raw data collected in our sample, we noted very few vertical displacements, indicating that vertical movements of the pelvis generally did not exceed the threshold for inclusion in the algorithm for identifying steps. These data nevertheless provide additional understanding of pelvic movement during gait in Rett syndrome and perhaps the Actigraph GTX3 provides a simple method of basic gait analysis in a clinical group for whom laboratory assessment is practically very difficult. Whilst the Actigraph is widely used in physical activity research in

populations with largely normal gait patterns [26], at present, its utility in populations with a neurological disorder including Rett syndrome may be limited.

We found that the ActivPAL also under-reported steps in Rett syndrome. Studies of healthy young participants have reported accurate measures of step counts during comfortable walking speeds across land [27] and during treadmill walking [28]. Nevertheless, accuracy has been compromised during free-living walking sessions [28] or at speeds slower than 40 meters/minute [24]. In our sample, the ActivPAL under-recorded to a lesser extent than the Actigraph but agreement between step counts recorded using this device with the video-tape analysis was also compromised.

Compared with the ActivPAL, the SAM has demonstrated similar [27] or superior [24] accuracy. In those with a chronic respiratory condition, the SAM was superior to the ActivPAL in identifying steps [17]. For those with gait affected by neurological impairments, the SAM had good accuracy in those with incomplete spinal injury [29] and Duchenne Muscular Dystrophy [30], and superior accuracy when compared with a Caltrac accelerometer [31] in patients with stroke. In our sample, the SAM recorded a similar step count to the observed during the video-analysis during the most commonly used cadences. Therefore, our data together with this earlier work suggests the SAM is currently the superior choice to measure walking activity in persons affected by a neurologic disorder.

We also identified pairs of minutes during which the observed step count was identical and found the repeatability of the SAM step counts to be excellent. We calculated the SEM independently from ICC calculations [22] and for any individual, we are 95% confident that a change \geq 17 steps/minute would be greater than measurement error. This threshold allows for the recording of additional fidgety or rocking movements that may not be steps but which would reflect natural test-to-test variability. During a bout of physical activity, an increase of at least 17 steps in a minute could be seen as important to identifying individual

improvement and a feasible target when aiming to improve walking activity levels in Rett syndrome.

Rett syndrome is a rare disorder with an incidence of 1 in 9,000 live female births [32]. The ARSD recruits from multiple sources throughout Australia and as such is population-based. We recruited families from two states of Australia to enable testing of an adequate sample size. Our participation fraction for the current study was high giving confidence that our sample of girls and women with walking skills was representative of the clinical population. Further, our sample demonstrated clinical variability in terms of age [33], mutation type [8] and gross motor skills [20] and therefore the results of this study pertain to the range of clinical characteristics seen in ambulant girls and women with Rett syndrome.

Observation of videoed steps was our criterion measure but we acknowledge the potential for human error when identifying each step on the video. Nevertheless, we developed an operational definition for a step prior to the analysis of the video tapes and two observers independently counted the steps. We defined cadence as the number of steps in a minute but we also acknowledge that each minute likely included episodes at different speeds. The number of steps in a minute was our best estimate of cadence since those with Rett syndrome cannot walk at predetermined cadences because of cognitive impairment and dyspraxia [34]. For similar reasons, our study comprised assessment in natural "free-living" conditions rather than laboratory-based testing with a strict testing protocol, and therefore we cannot identify cut-points in constant cadences below or above which agreement would be unsatisfactory. However, "free-living" assessment enabled estimates of device accuracy in the conditions of use. In the general population, measurement properties of the Actigraph and ActivPAL have both been shown to record fewer steps than the SAM in "free-living" conditions [24]. We also acknowledge that the development of condition-specific algorithms for devices such as the Actigraph and ActivPAL may improve their application in persons with neurological impairments.

In conclusion, our novel field observations enabled pragmatic assessment of how well three accelerometer-type devices could detect steps in Rett syndrome. Our data demonstrated that the SAM was the most accurate device and provides a platform for future analyses of whole day activity and sedentary behaviors. Repeatability of step counts recorded using the SAM was good suggesting that a small difference would be necessary to be confident of a change that exceeded measurement error. Over time, there have been shifting paradigms in the assessment and management of disability issues particularly in association with the introduction of the International Classification of Functioning Disability and Health [35]. That is, there is reduced focus on quantifying capacity or impairments and increased interest in quantifying participation in activities of daily life. Participation-based activities such as walking have potential to be critical influences on wellbeing and quality of life [36] and our study puts the spotlight firmly on walking-based activities and promotion of active lifestyles for this group. The capacity to collect accurate measures of physical activity in Rett syndrome enables additional monitoring of clinical progress and can contribute to rigorous evaluation of interventions such as cell-based molecular therapies in our new current era of clinical trials for rare disorders.

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Declaration of interest statement

The authors declare that there is no conflict of interest.

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Figure 1



Average steps/min (device and video analysis)





Average steps/min (device and video analysis)

Figure 3



Average steps/min (device and video analysis)





Implications for Rehabilitation

- Many girls and women with Rett syndrome are able to walk on their own or with assistance but with altered movement patterns.
- Validated measures of physical activity, such as step counts, have potential to monitor function during daily life.
- Compared with other forms of accelerometer-type devices such as ActiGraph and ActivPAL, the StepWatch Activity Monitor (SAM) measured step counts with good accuracy and repeatability.
- The capacity of the SAM to measure physical activity in Rett syndrome allows focus on participation-based activities in clinical practice and clinical trials.