Wearable Devices and Mobile Health – an Opportunity for Innovative Remote Patient Monitoring in Niemann-Pick Type C

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INTRODUCTION AND OBJECTIVES

NIEMANN-PICK C (NPC) is a rare inherited neurodegenerative autosomal recessive disorder primarily diagnosed in infancy. Disease symptoms include frequent falls, ataxia and behavioral problems. There is a lack of validated, age-related, disease-specific and points available to screen gait and ataxia long-term in NPC. Doctors often assess NPC impacting the Pirlo Disability Scale (PDS). Other methods employed to assess ataxia and gait ambulation can include GAITRite assessment and 5-Minute Walk Test (5MWT).

METHODS

Data collection

NPC patients were recruited from the UK Specialist Centre. Doctors assessed each patient with PDS, 5MWT and GAITRite. Parents completed PROs about their child’s disability, answering questions regarding Disease Impact Severity, Family Impact and Wider Impact developed by the International Niemann-Pick Disease Association and pushed at regular intervals to them via a mobile app. Parents were identified in collaboration with the patient group. Participants were also given a wearable device (a 3D accelerometer) which calculates step data in 38-minute epochs and transmits it via Bluetooth to a paired smartphone app downloaded to patient’s personal (or care givers) phone. No restriction was made on the wear time of the device.

Data summaries

Adherence: Zero steps recorded over a 24-hour daily cycle was interpreted to mean that a patient did not wear the device for that day and therefore was considered to be “non adherent”. Wearable data summaries are based on the adherent days only.

Peaks: We defined peaks of activity as a local maximum value. To identify local maximum points we used the function findpeaks from the peakfinder package in R. Since this function might identify relatively lower values which nevertheless satisfy the criteria of having locally increasing and decreasing values of activity around them, we looked at the peaks exceeding a determined quintile of the distribution of peak values for steps. We found that the third quintile of the overall distribution, including zero count, provided a robust definition of peak threshold. The number of peaks was then standardized by dividing it by the number of valid days in which the device was worn.

RESULTS (1)

• We collected data from 4 pediatric NPC patients at various stages of the disease according to PDS, PRO and 5MWT assessments. Patient specific baseline characteristics are displayed in Table 1.

• The data cut-off differ for different patients, ranging from 16 days to 95 days. High adherent has been achieved in all scenarios. On average, patients adhered on 70% of the days when asked to wear the device continuously (Table 2).

• Patients 4 and Patient 3 seemed to be the most and least active, with an average of 11254 and 7006 daily steps, respectively (Table 3).

• Patients 4 and Patient 1 seemed to have the most and least intense activity with the average daily peak count of 4 and 2, respectively (Table 3).

• The 24-hour cycles for average steps per epoch for each patient are displayed in Figure 1. Although each one is slightly different, there are general similarities according to the time that activity for each patient starts in the day (e.g. patients wake up) and ends. There is also a peak of activity seen around lunchtimes, with a large dip just after that and with a general reduction in activity then before bedtime.

• Different activity patterns between the patients were noticed when comparing weekday vs. weekend (Figure 2). Patient 1 generally is more active in a weekday only up to the lunchtime, and after that activity seemed to be very similar during the whole week. Patient 2 seemed to be more active over the weekend. Patient 3 has very similar activity patterns during the week day and weekend while Patient 4 is more variable with an extremely high activity noticed during the weekend.

• Table 5 shows that daily steps and daily peaks were perfectly correlated, but that neither daily steps nor daily peaks correlated with the 5MWT, PDS or PRO. PDS and PRO was negatively correlated with the 5MWT, suggesting that the more severe disease score the less patient was able to walk during the 5MWT.

• Table 5 reports a correlation coefficient, an absolute value > 0.5 is required for significance at 10% for n = 4.

DISCUSSION AND CONCLUSIONS

Through this study it has been demonstrated that it’s possible to achieve remote, continuous, long time series monitoring of patients’ PA. In selecting the technical capabilities of the device used there was a tradeoff between battery life and the 30 minute epoch resolution. However, even this resolution was great enough to detect individual patient’s patterns and changes in PA.

Further work is required to understand why the wearable device data did not correlate with the currently used clinical data. This may be achieved by conducting a larger scale study with a higher number of patients. Data analysis demonstrated that each individual patient has physical activity patterns that are personalized to them, in terms of timing of day and of the week. It is crucial to be able to capture and define such personalisation in order to identify subtle changes in disease improvement or progression.

REFERENCES


2 [2] Patients were recruited at Great Ormond Street Children’s Hospital and Manchester’s Children Hospital in the UK between February and July 2016.

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