

# The Relationship Between Participant-Reported Energy Levels & Energy Expenditure & Sleep Among Individuals With Barth Syndrome

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Purpose Barth syndrome (BTHS) is a rare, X-linked genetic disorder characterized by skeletal myopathy, cardiomyopathy, and exercise intolerance due to early fatigue (Clarke et al., 2013). Fatigue in BTHS has proved difficult to quantify and has only been measured in terms of physiological fatigue excluding mental and emotional fatigue. The aim of this study was to understand how fatigue specifically impacts the lives of individuals with BTHS on a daily basis, and to be able to quantify fatigue in a way that can be objectively measured by examining the relationship between perceived ratings of fatigue and objective ratings of energy expenditure, examining the relationship between objective sleep data and perceived ratings of fatigue the following day, and examining age-related trends in the data. We hypothesized that TST, sleep efficiency, and number of awakenings would impact perceived fatigue the next day and that energy expended would correlate with perceived energy. Design This study used a descriptive study design. Participants were recruited with assistance from the Barth Syndrome Foundation. Sixteen participants with BTHS (6 adults, 5 adolescents, and 5 children) completed the study. Methods The participants wore an Actigraph GT9X Link activity tracker for 14 days to collect data on an individual's energy expended throughout the day and sleep information (sleep latency, sleep efficiency, total sleep time, and night time awakenings). Each participant also completed a sleep diary to validate these sleep periods as well as questions about perceived levels of fatigue on a phone app up to 6x/day. Statistical analysis was done through SPSS to compare perceived fatigue and objective energy expenditure data as well as sleep data at the following time periods: early morning (EM), late morning (LM), early afternoon (EA), late afternoon (LA), early evening (EEVE), and late evening (LEVE). Results Energy expended through each of the 6 time periods (EM, LM, EA, LA, EEVE, and LEVE) correlated with the perceived energy level ratings at these same times of day in adults ( $p < 0.001$ ), but not in children ( $p = 0.25$ ) or adolescents ( $p = 0.9$ ). Total Sleep Time (TST) the night before correlated with the average perceived energy ratings the next day in children ( $p = 0.027$ ) but not in adults ( $p = 0.8$ ) or adolescents ( $p = 0.106$ ). Sleep Efficiency the night before and number of awakenings in the night did not correlate with the average perceived energy ratings the next day in any of the age groups (Sleep efficiency: children  $p = 0.36$ , adolescents  $p = 0.186$ , adults  $p = 0.3$ ; Number of awakenings: children  $p = 0.5$ , adolescents  $p = 0.6$ , adults  $p = 0.07$ ). Conclusions In adults with BTHS, the measured energy expenditure highly correlated with perceived energy level ratings, but not in the children participants, indicating that the children and adolescents may not yet have the ability to accurately report or explain their fatigue. On the other hand, children's total sleep time significantly contributed to their average perceived energy level ratings the next day. This indicates that adequate sleep is particularly important for children with BTHS. There is a further need to study how to help children and adolescents explain their fatigue whether it be physical, mental, or emotional to help us best understand how their expended energy each day correlates with their perceived levels of fatigue. Impact Statement While this study focused on a specific rare genetic disorder, the research methodology used and knowledge gained from this study will further our understanding of fatigue and energy expenditure in disorders that involve skeletal myopathy and cardiomyopathy that could lead to exercise intolerance.

## References

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