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Research Paper

## Charting the territory: Describing the functional abilities of children with progressive neurological conditions



Sandy K. Tatla<sup>a,\*</sup>, Liisa Holsti<sup>b</sup>, Gail Stephanie Andrews<sup>c</sup>, Leanne Feichtinger<sup>c</sup>,  
Rose Steele<sup>d</sup>, Harold Siden<sup>e</sup>

<sup>a</sup> BC Children's and Women's Health Centre, BC Children's Hospital Research Institute, 4500 Oak Street, Vancouver, BC, V6H 3N1, Canada

<sup>b</sup> Department of Occupational Science and Occupational Therapy, University of British Columbia and BC Children's Hospital Research Institute, Clinical Support Building, V3-327, 950 West 28th Avenue, Vancouver, BC, V5Z 4H4, Canada

<sup>c</sup> University of British Columbia, BC Children's Hospital Research Institute, F611-4480 Oak Street, Vancouver, BC, V6H 3V4, Canada

<sup>d</sup> School of Nursing, Faculty of Health, York University, Room 342 HNES Building, 4700 Keele Street, Toronto, ON, M3J 1P3, Canada

<sup>e</sup> Department of Pediatrics, University of British Columbia, BC Children's Hospital Research Institute, F612a-4480 Oak Street, Vancouver, BC, V6H 3V4, Canada

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### ABSTRACT

**Aims:** Little is known about the functional abilities of children with progressive genetic, metabolic, or neurological conditions (PNCs). In this study, children with PNCs were followed over a 2-year period to assess their functional abilities over time. Specific aims were to: 1) describe the changes in functional skills and the effects of age for children with PNCs, 2) assess changes in these children's need for caregiver assistance over time, and 3) examine relationships between these children's functional skills and need for caregiver assistance.

**Methods:** This study involved a longitudinal, descriptive design with three assessments occurring at Baseline, Year 1, Year 2. Functional skills and caregiver assistance were assessed by the Pediatric Evaluation of Disability Inventory (PEDI). The PEDI questionnaire was completed at baseline and then yearly by parents, along with the assistance of a trained research assistant (RA).

**Results:** The study was completed with 83 children (mean age at Baseline = 7.1 yrs,  $SD = 4.6$ ). Mean Functional skills scores were in the low ranges at Baseline and did not change significantly across time points ( $F(2, 71) = 0.437, p = 0.58$ ). Time point had no effect on caregiver assistance ratings ( $p < 0.2$ ); however, children required greater amounts of help with self-care at later time points than for other functional domains. Statistically significant correlations were found between PEDI-Functional skills and caregiver assistance ratings ( $r = 0.80-0.90, p < 0.01$ ).

**Conclusions:** Functional skills were low for these children overall, irrespective of age. In children with PNCs: 1) mean functional skills did not change significantly over time; 2) caregiver assistance scores remained stable and 3) functional skills and levels of caregiver assistance were strongly positively correlated. Further research to explore the long-term functional trajectory in children with a PNC is recommended.

\* Corresponding author.

E-mail addresses: [statla@cw.bc.ca](mailto:statla@cw.bc.ca) (S.K. Tatla), [liisa.holsti@ubc.ca](mailto:liisa.holsti@ubc.ca) (L. Holsti), [gandrews@cw.bc.ca](mailto:gandrews@cw.bc.ca) (G.S. Andrews), [feichtinger@cw.bc.ca](mailto:feichtinger@cw.bc.ca) (L. Feichtinger), [rsteale@yorku.ca](mailto:rsteale@yorku.ca) (R. Steele), [hsiden@cw.bc.ca](mailto:hsiden@cw.bc.ca) (H. Siden).

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## What this paper adds

This longitudinal study of children with progressive neurological conditions (PNCs) examines and describes their functional abilities over time using the PEDI. Functional skills such as communication, personal care, transfers and manipulation are involved in everyday activities of life. This work identifies the importance of assessing the functional abilities of these children on an ongoing basis to guide care. Though children's functional skills remained stable overall, the need for mobility modifications increased over time and children required significantly greater assistance in areas related to self-care compared to mobility and social function. The PEDI offers promise as a tool to assess and monitor functional changes in children with PNCs and the routine use of the PEDI in this population should be considered. Findings warrant further research to explore functional trajectories in this population.

## 1. Introduction

More than 50% of the children receiving pediatric palliative care in North America have a diagnosis of a progressive genetic, metabolic, or neurological condition (PNC) (Feudtner et al., 2011; Rogers et al., 2011; Siden et al., 2010). While a number of different terms have been proposed to describe this population, PNC in the field of pediatric palliative care is well understood by clinicians to mean children with severe life threatening, life limiting, and complex medical problems (Steele et al., 2014). The term 'genetic' is used in this context to represent both single and multiple gene conditions (including chromosome abnormalities and aneuploidy) whereby a resulting disruption in cellular function leads to deterioration and degeneration. These conditions are non-curable, so the current clinical emphasis is on treating the manifestations of the disease (e.g., seizures) until disease-modifying therapies or outright cures can be developed. These diseases correspond to Category 3 in the scheme developed by the charity "Together for Short Lives" (Chambers et al., 2009; Spicer, Macdonald, Davies, Vadeboncoeur, & Siden, 2015). Many of these conditions lead to early death before adulthood and in addition require ongoing care to achieve good quality of life. Though there are hundreds of individual diagnoses in this group, they collectively share a phenotype characterized by progressive constellation of impairments of the central nervous system and overlapping clinical manifestations.

Children with PNCs typically have impaired cognition, may or may not be able to communicate verbally, and have mobility that is often severely limited. Functional abilities may decline over the course of the disease, and due to their illness, they may require full time caregivers, wheelchairs and assistance with breathing or feeding tubes. Progression of the disease in children included in the cohort leads ultimately to an early death. However, the rate of disease progression and functional impact along the disease trajectory can be unpredictable. Their condition may be physiological or congenital, and onset occurs at differing rates, resulting in weeks or years of stable yet progressive decline (Steele, 2000).

Children with PNCs display impairments in cognition and in motor function and experience many symptoms such as pain and seizures (Steele et al., 2014). PNCs are all genetically-based with defects in protein assembly, function or clearance. These defects lead to impairment of one or more organs, but always affect the Central Nervous System. While the gene and protein defects may be distinct, there is seeming overlap in clinical phenotype; the focus of our program of research is in better delineating the clinical phenotype and trajectory of PNCs. Many of these children function at about a one-year old or younger level, without vocabulary, motor control over their limbs and trunk, or bladder control. They may require years of treatments and physical rehabilitation prior to succumbing to their life-threatening diseases. A paucity of information regarding the functional abilities of this population is available; little has been documented to describe the natural progression of this group over time (Siden et al., 2010; Steele et al., 2014).

Cerebral palsy is the leading cause of disability in children, and although children with CP were excluded from this study due to their injury resulting from static encephalopathies (e.g., hypoxic-ischemic encephalopathy), the functional limitations that they present with are similar to those of children with PNC. Much of the literature on children with severe disability comes from the CP population and is the most appropriate source of research results currently available (Ketelaar, Gorter, Westers, Hanna, & Verhoef, 2014).

Building on the focused studies on cerebral palsy, Steele, Siden, and colleagues evaluated symptoms in individuals with PNCs (Siden et al., 2010; Steele et al., 2014). They identified that these children experience multiple symptoms and that the presence of an enterostomy feeding tube was associated with a higher number of frequent and distressing symptoms, such as respiratory problems, pain, feeding difficulties and the need for extensive mobility modifications. Further, mobility loss and increased requirements for equipment modification have been reported as notable milestones of functional mobility for children in pediatric palliative care (Wood, Simpson, Barnes, & Hain, 2010).

Functional ability is a key indicator that can describe the degree to which an individual is engaging and participating in his or her daily life. Gaining a comprehensive understanding of the functional implications of living with a severe health condition is essential to delivering comprehensive and appropriate care that meets the needs of children with a PNC. Monitoring functional abilities may provide clinicians and parents with a standardized and objective approach to assess a child's performance in daily life, thereby supporting optimal care delivery (Worth, Darrah, Magill-Evans, Wiart, & Law, 2014). The Pediatric Evaluation of Disability Inventory (PEDI) (Haley, Coster, Ludlow, Haltiwanger, & Andrellos, 1992) is an instrument used internationally to assess and evaluate changes in function in children with disabilities (Worth et al., 2014), but it has not been used extensively in the PNC population though its use is increasing (Monteiro et al., 2014). Currently, very limited research is available to guide care for this population and the functional abilities of children with PNCs have not been reported. Consequently, the primary objectives of this longitudinal study were to: 1) describe the changes in functional skills and the effects of age for children with PNCs, 2) assess changes in these children's need for caregiver assistance and differences over time, and 3) examine relationships between these children's functional skills and need for

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