

# Charting the Territory: Children and families living with progressive life-threatening conditions

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**OBJECTIVES:** To increase awareness of the topic of paediatric palliative care among practicing physicians in Canada by exploring the impact of a child's neurological or rare genetic life-threatening condition on the affected child and his/her parents.

**METHODS:** Cross-sectional, baseline results from an observational, longitudinal study, Charting the Territory, which followed 275 children and 390 parents from 258 families. Parents completed multiple surveys, for themselves and their child.

**RESULTS:** These children had a high symptom burden. The three most common symptoms were pain, sleep problems and feeding difficulties; on average, they had 3.2 symptoms of concern. Despite analgesic use, the frequency of pain episodes and distress were invariant over time, suggesting that treatments were not successful. Parents experienced anxiety, depression and burden; at the same time they also reported positive life change and a high degree of spirituality. The child's condition resulted in parental changes in living arrangements, work status and hours devoted to caregiving. Nearly two-thirds of families were involved with a palliative care team; the size of the community in which a family resided did not make a significant difference in such involvement.

**CONCLUSIONS:** These families experience many challenges, for the patient, other individual members and the family as a whole. At least some of these challenges may be alleviated by early and organized palliative care. Effective interventions are needed to enhance symptom management for the ill child and to alleviate the various negative impacts on the family.

**Key Words:** Genetic conditions; Metabolic diseases; Neurology; Palliative care

## Explorer de nouveaux territoires : les enfants et les familles qui vivent avec une maladie évolutive au potentiel fatal

**OBJECTIFS :** Mieux faire connaître les soins palliatifs aux médecins en exercice du Canada en explorant les effets de maladies neurologiques ou génétiques rares au potentiel fatal sur l'enfant atteint et ses parents.

**MÉTHODOLOGIE :** Les auteurs publient les résultats transversaux et initiaux d'une étude d'observation longitudinale, Explorer de nouveaux territoires, auprès de 275 enfants et de 390 parents de 258 familles. Les parents ont rempli de multiples sondages en leur nom et au nom de leur enfant.

**RÉSULTATS :** Les enfants présentaient un lourd fardeau de symptômes. Les trois plus fréquents étaient la douleur, les troubles du sommeil et les problèmes d'alimentation. En moyenne, ils avaient 3,2 symptômes inquiétants. Malgré l'utilisation d'analgésiques, la fréquence d'épisodes de douleur et de détresse ne changeait pas au fil du temps, ce qui laisse supposer l'échec des traitements. Les parents ressentaient de l'anxiété, de la dépression et un fardeau, mais signalaient également un changement de vie positif et une spiritualité marquée. En raison de l'état de l'enfant, les parents modifiaient leur mode de vie, leur statut professionnel et les heures consacrées aux soins. Près des deux tiers des familles recouraient à une équipe de soins palliatifs, sans que la dimension de leur communauté y ait une incidence significative.

**CONCLUSIONS :** Ces familles éprouvaient de nombreux problèmes, tant pour les patients et les autres membres de la famille que pour l'ensemble de la famille. Au moins certains problèmes peuvent être atténués par des soins palliatifs instaurés rapidement et organisés. Des interventions efficaces s'imposent pour améliorer la prise en charge des symptômes de l'enfant et en réduire les divers effets négatifs pour la famille.

### CASE PRESENTATION

PD is a seven-year-old girl with severe neurological impairment due to mitochondrial encephalomyopathy. She is intellectually disabled, nonverbal, visually impaired and nonambulatory. She experiences seizures several times a month. She is fed through a gastrostomy tube (G-tube), has a chronic cough and has been hospitalized at least annually with aspiration pneumonia. She also has hypertonic cerebral palsy. PD uses a specially modified wheelchair and is followed by orthopedic surgery for contractures, hip dysplasia and scoliosis.

The pregnancy and delivery were uneventful, but in infancy she was delayed in achieving milestones. At one year of age, her parents were referred to a paediatrician. An initial workup found

no immediate cause for delay and a referral was made to a neurologist, physiotherapist and speech-language pathologist. While awaiting the neurology appointment, PD was hospitalized for a seizure; this accelerated the workup, but it was not until a muscle biopsy at 20 months of age that a diagnosis of mitochondrial disease was made.

PD attends school for 3 h each day in grade 2; she tires easily if she attends for longer. An educational assistant in the classroom helps her with a program based on an individual education plan. A community nurse sees PD at school once a month and as needed. This nurse also coordinates the care plan for PD's respite. PD's family has access to 20 h of respite care per week in their home, provided by staff who are registered nurses

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or licensed practical nurses. The family uses 4 h respite slots to take care of their other children. The respite nurse provides the gastrostomy feeds, scheduled medications and physiotherapy; otherwise, the parents do this work. One weekend each month, PD receives respite care at a children's hospice 90 min away.

PD lives with her parents and two siblings (an 11-year-old sister and a two-year-old brother) in a community of 90,000, 2 h from a major metropolitan area. Her father is a civil engineer who has recently accepted a new position in a rural area. Her mother is an accountant, but has not worked since PD was first hospitalized at 14 months of age. PD's father grew up on military bases. He graduated from high school in Quebec but identifies himself as being "from all over". PD's mother immigrated to Canada from Hong Kong at two years of age and lived in Alberta. The maternal grandparents reside nearby and see the family frequently. The family identify as Catholic and say that religion is "moderately important" to them. PD's sister is devoted to her and knows how to manage tasks such as starting a G-tube feed or positioning her in the wheelchair. Her sister plays hockey and, recently, much of her time is spent at tournaments. One parent tries to be with the sister at practices or games. The two-year-old brother is healthy and very active. During that pregnancy, the mother underwent prenatal testing; results were negative.

Children with progressive metabolic, neurological or chromosomal conditions constitute an important group within the broad spectrum of children with complex, chronic conditions. Specific conditions are uncommon, but can be broadly categorized as a single group because of the problem that unifies them: they are progressive and involve impairment/injury of the central nervous system (CNS) (1,2). While it is difficult to determine precisely how many children have severe life-threatening conditions due to these progressive disorders, estimates based on data from one Canadian program suggest that they occur, at minimum, at a rate of eight per 10,000 population (3).

What is known is that approximately 50% of the annual 2500 childhood nontraumatic deaths in Canada occur because of such disorders (4,5). Across North America, these diagnoses account for more than one-half of the children who receive paediatric palliative care (4-6). Families of children with progressive, noncurable conditions anticipate an unknown lifespan; the children often endure uncomfortable symptoms; and families must cope with emotional and spiritual challenges as the condition progresses along an uncertain trajectory toward death.

For these children, for whom there is neither a cure nor a specific life-prolonging treatment, the focus is on optimal symptom management, quality of life and family support. Paediatric palliative care, therefore, should begin at diagnosis and focus on anticipating and relieving the child's symptoms while supporting the family to ensure the best possible outcomes in the face of early death. Unfortunately, there is a lack of research evidence on which to base such care (1,2,6-8). The terminal outcome of the disease may be known, but the onset, timing, patterns and severity of symptoms are not; consequently, best practices and strategies for symptom relief are often unclear.

Furthermore, little is understood about the biopsychosocial/spiritual impact on these families and how to support them. There is a critical need to develop a solid understanding of disease progression, and the experiences of children and families over time. The goal of our research program is to close knowledge gaps and provide new information about the best care for these vulnerable children and their families. To meet this goal, and to provide better information to clinicians regarding the clinical

trajectory of affected children (AC) and the psychosocial experiences of their families, we conducted a five-year longitudinal study, 'Charting the Territory' (CTT) (CIHR MOP-89984) to track children diagnosed with progressive metabolic, neurological or chromosomal conditions and their families to determine and document the clinical progression of the condition and the associated biopsychosocial/spiritual experiences of the family. The present study provides a summary of recent findings from the CTT study; detailed analysis is available elsewhere and more will be forthcoming (9).

## METHODS

The present descriptive, correlational study enrolled children zero to 19 years of age and their families (parents, and siblings seven to 18 years of age). Recruitment occurred at seven Canadian and two American centres. Patients were recruited via clinician letters sent from programs in neurology, genetics, metabolic diseases, complex care, palliative care and general paediatrics. Study information was also disseminated via advocacy groups and social media. Children eligible to enroll had to have a progressive neurological, metabolic or chromosomally based condition with impairment of the CNS. Other organs may be affected as well. Children whose CNS impairment was due to a hypoxic-ischemic injury were excluded. Baseline data collected from parent reports and chart reviews included family demographic information; health and function information regarding the AC; coping and health assessments of eligible, consenting siblings; and biopsychosocial/spiritual assessments of at least one (ideally both) parents. If there was >1 AC in a family, then data were collected about each child.

Data collection included monthly parental assessment of seven symptoms chosen from the literature, which showed these symptoms were highly prevalent in this population and of significant concern to families (pain, dyspnea, feeding intolerance, dysomnia, constipation, seizures and arousal) (10,11). An annual functional assessment of the child was completed with a standardized tool (12). One 'designated parent' for each family completed the monthly, semiannual and annual reports on behalf of the AC and any siblings in the study; older siblings also completed self-assessments. In two-parent families, the 'nondesignated' parent completed semiannual survey tools about their own health and psychosocial well-being. Instruments assessed family functioning, marital satisfaction, health status, anxiety, depression, stress, burden, grief, spirituality and growth; and assessment by parents of siblings' health every six months; siblings who were old enough assessed their own health, and all participating siblings completed a coping measure. The impact on parents participating in the present study was assessed after one year and at the end of the study. If the child died during the study, family data collection continued after a waiting period of at least six months if the family agreed to continue. Chart reviews were conducted at enrollment and at the conclusion of the study or at the time of the child's death.

## RESULTS

Data were collected between July 2009 and December 2013. A total of 358 families were contacted, 258 of whom enrolled (275 children, 70 siblings, 390 parents), from nine sites. There were 93 specific diagnoses for 227 of these children, while 48 did not have a diagnosis but otherwise had characteristics of progressive, noncurable conditions affecting the CNS. The diagnoses could be characterized into 12 broad condition groups. Selected details regarding these children are shown in Table 1.

**TABLE 1**  
**Selected characteristics at baseline for ill children (n=275)**

Demographic characteristic	Ill children
Sex	
Female	139 (50.5)
Male	136 (49.5)
Top concerns that brought parent to doctor*	
Not meeting developmental milestones	54 (19.6)
Perinatal: diagnosis or problem noted	51 (18.5)
Seizures	47 (17.1)
Change in muscle tone	34 (12.4)
Prenatal: diagnosis or problem noted	33 (12)
Feeding difficulties	30 (10.9)
Breathing difficulties	21 (7.6)
Losing developmental milestones	21 (7.6)
Age at entry to study, years	6.6±5.2 (0–19) <sup>†</sup>
Age at initiation of diagnostic evaluation, months (n=261)	12.1±25.5 (–5.8–183.5) <sup>†‡</sup>
Age at diagnosis, years (n=203)	2.2±3 (–0.4–15.7) <sup>†‡</sup>
Time since parent first sought medical treatment, weeks (n=261)	315.2±248.9 (7.9–980) <sup>†</sup>
Time since child's diagnosis, weeks (n=203)	269.6±231.3 (0–903.7) <sup>†</sup>
Time from initial diagnostic evaluation to confirmation of diagnosis, months (n=197)	11.9±23.6 (0–177.9) <sup>†</sup>
Primary condition	
Multiorgan congenital abnormalities	54 (19.6)
Severe neurological impairment – not yet diagnosed	45 (16.4)
Lysosomal/peroxisomal leukodystrophy	44 (16)
Mitochondrial encephalopathy/myopathy	31 (11.3)
Neurodegenerative disease	23 (8.4)
Structural central nervous system abnormalities	22 (8)
Epileptic encephalopathy	19 (6.9)
Small molecules diseases	13 (4.7)
Neuromuscular diseases	10 (3.6)
Other metabolic diseases	3 (1.1)
Congenital disorders of glycosylation	7 (2.5)
Other conditions not otherwise specified	4 (1.5)

Data presented as n (%) unless otherwise indicated. \*Some responses are not reported in the table; overall, numbers totaled more than sample size because some parents provided multiple responses; <sup>†</sup>Data presented as mean ± SD (minimum – maximum); <sup>‡</sup>Negative values reflect prenatal evaluations and diagnoses

Children in the cohort had a high symptom burden according to parent report (9). At the time of enrollment, pain/irritability had been experienced by 55.2% of the children; 50.2% had sleep problems, while problems with feeding including nausea, vomiting and/or intolerance had affected 48%. Other important symptoms included constipation (47%), respiratory difficulties (46.5%), seizures (40.2%), and problems with alertness and interaction (34.7%). Parents reported an average of 3.2 symptoms per child at baseline. The less ambulatory a child was, the more likely he/she was to experience a greater number of symptoms ( $r=-0.297$ ,  $n=271$ ,  $P\leq 0.001$ ) (9). The presence of breathing difficulties ( $\chi^2[1, 271]=17.49$ ,  $P<0.001$ ), pain ( $\chi^2[1, 270]=7.26$ ,  $P=0.01$ ) feeding difficulties ( $\chi^2[1, 271]=7.15$ ,  $P=0.01$ ) and constipation ( $\chi^2[1, 270]=6.38$ ,  $P<0.05$ ) significantly differed by the presence of a G or J tube. A tube-fed child was more likely to have these symptoms. Tube-fed children also had a higher total number of symptoms than orally fed children (mean ± SD 3.7±1.7 versus 2.6±2.0); children with G or J tubes experienced more symptoms

overall ( $t[269]=-4.36$ ,  $P\leq 0.001$ , difference  $-0.99$  [95% CI  $-1.4$  to  $-0.5$ ]). On average, children were taking 3.5±2.3 medications to address symptoms (9).

During the course of the study, 54 (19.6%) of the children died. Thirty-three of these children had advance directive orders specifying no attempted resuscitation (DNAR), three did not and the status of 15 was unknown. A total of 170 families (65.9%) had involvement with a palliative care team. DNAR presence was strongly correlated at baseline with the involvement of a palliative care team ( $P<0.001$ ).

PD has many symptoms and ancillary problems found in children with progressive conditions in the CTT study. She has seizures and respiratory difficulty, requires artificial enteral nutrition and mobility aids. It is important to inquire about the other symptoms she may be experiencing, such as pain or sleep disturbance. It is also important to remember that symptoms fluctuate over time, even monthly. While parents may bring distressing symptoms to a clinician's attention, having a mental 'checklist' of the common ones may be of assistance.

There were 390 participating parents within the 258 families in the study. A total of 258 designated parents and 132 nondesignated parents participated. Information regarding these parents is shown in Table 2. Parents were, for the most part, born in their current country of residence (Canada or the United States, respectively). If they were immigrants to Canada, they had been in the country for quite a long time (17.2±12.3 years). Forty percent had at least some university education. In addition, religion was important to them. Table 2 provides a picture of the types of internal support and resources individual parents may report.

Parents reported experiencing challenges as families. Slightly more than one-half ( $n=202$ ) noted that their occupation or job status changed as a result of the child's illness (Table 2). On average, annual income was below that of the Canadian median based on the 2012 census (13), but above the American median (14). Most families found it difficult to meet all their needs at better than an 'adequate' level and many had difficulty meeting the additional costs associated with an ill child (Table 3). There was a variety of alterations in living arrangements attributed to the child's condition: eg, 10.1% moved to another city, 3.1% moved in with relatives, 14% moved into a different style of home and 28.7% renovated.

On average, families devoted 85.9±58.5 h per week caring for the affected children in the family (Table 2). Therefore, an important source of support to families is provided through the respite care that they can access, sometimes through formal programs and sometimes via unpaid family or friends. More than one-half ( $n=138$ ) of the study families relied on unpaid caregiving and had, on average, 3±2.6 such caregivers who provided almost 14 h/week of aid. Concurrently, families received care through programs such as home nursing, hospices or other support. Families identified that 46.5% of them received respite care once per week or more and an additional 8% received it two or three times per month. However, 45.6% of families received minimal respite ranging from bimonthly to none at all.

Some parents experienced anxiety, depression and burden. Clinical levels of anxiety were reported by 25.5% of parents and 61.2% reported at least moderate to severe burden. Approximately one-third ( $n=127$ ) reported much higher than average stress. Mild depression was present in 25.2% of parents and an additional 12.5% scored in the major depression range. At the same time,

many parents also reported positive life changes (45.2%) and high degrees of spirituality (94.7%).

PD's parents face the challenges of raising a young family with the additional work and costs associated with caring for a child with a severe progressive condition. Their focus is on her quality of life, maintaining her health and integrating her into the community. At the same time, they have had to make choices; her mother is staying home, and the family relies on paid and unpaid caregiving for support. Clinicians caring for children such as PD need to be aware of the 'patchwork quilt' that many families arrange for care. They should not only inquire about enrollment in government or charitable programs, but also determine the level of care provided informally. They should also ask about how parents are coping – for many parents, it is a complicated experience with inherent contradictions of both burden and growth. The assumption that the experience is only one of ongoing depression or grief should not be made. In fact, recent literature on resilience indicates that clinicians should identify a family's strengths and encourage the family to build on those strengths (15).

In the CTT study cohort, 53.4% of families lived in either metropolitan areas or large cities according to Statistics Canada definitions, and 25.3% were in medium/small cities with populations of 10,000 to 99,999 (Table 3). Access to some services, such as respite care, did not appear to be dependent on community size; of the families who accessed formal respite programs, both home and inpatient based, there was no difference regardless of community size. Moreover, access to palliative care team involvement did not appear to differ. A large proportion of children included in the study were involved with a palliative care team (65.9%), across all community sizes. Even in communities with populations <10,000, 78.4% of study patients identified as being involved with a palliative care team. In addition, families were equally likely to have or not have an advance directive regardless of community size.

The CTT study underscored that families are resourceful and develop supports wherever they are. PD's family, however, may face a challenge if they move because of her father's job. They will lose the caregiving support of the nearby grandparents and will be farther from the paediatric hospice where they currently receive respite. They may continue to have a connection to a paediatric palliative care team situated at a tertiary care centre. This support will be important to them as they move into a rural area where clinicians have limited exposure to children with progressive, complex health conditions.

## CONCLUSION

Most paediatricians know children similar to 'PD', and have clinical experience with the myriad symptoms. There is little evidence, however, to augment that clinical experience. Addressing the challenging symptoms affecting children with progressive metabolic, neurological or chromosomal conditions has received insufficient attention in the literature, which is mostly focused on cellular mechanisms of disease or development of treatments. Similarly, there is scant literature on the impact of these conditions on other family members. In the CTT study, we elected to follow children, their parents and siblings prospectively to create a detailed 'map' of symptom trajectory and of the impact on family

**TABLE 2**  
Selected characteristics of parents at baseline (n=390)

Demographic characteristic	Both parents
Sex	
Female	249 (63.9)
Male	141 (36.1)
Role in family	
Mother	248 (63.6)
Father	141 (36.2)
Grandmother	1 (0.3)
Relationship to ill child	
Biological parent	371 (95.1)
Adoptive parent	12 (3.1)
Step parent	6 (1.5)
Legal guardian	1 (0.3)
Age, years, mean $\pm$ SD	38.4 $\pm$ 8.2
Female	37.2 $\pm$ 8
Male	40.5 $\pm$ 8
Country of birth – Canadian cohort	
Canada	234 (71.1)
Elsewhere	95 (28.9)
Length of time in Canada if immigrant, years, mean $\pm$ SD	17.2 $\pm$ 12.3
Country of birth – United States cohort	
United States	58 (95.1)
Elsewhere	3 (4.9)
Highest level of education completed	
< At least some university	233 (59.7)
At least some university	157 (40.3)
Importance of religion <sup>†</sup>	
Very important	188 (48.6)
Of medium importance	108 (27.9)
Of little importance	52 (13.4)
Not at all important	39 (10.1)
If working, current job allows parent enough time to care for child <sup>†</sup>	
Yes, completely	130 (52)
Only partially	94 (37.6)
No	26 (10.4)
Change to job status as result of child's illness <sup>†</sup>	
No	187 (48.1)
Yes	202 (51.9)
Hours/week giving care to child in past month (total for both children if 2 AC <sup>†</sup> ), mean $\pm$ SD (n=377)	85.9 $\pm$ 58.5

<sup>†</sup>Numbers do not total to sample size because of missing data

members. The present article highlights some of the early findings from our work; deeper analysis is underway. In our next steps, we will describe the longitudinal nature of children's symptoms and function, longitudinal data on parent well-being and details regarding physical decline, advance care planning and interventions at death; we also will examine differences between experiences in Canada and the United States.

A concern when undertaking studies of this nature is whether families will participate. To recruit families, the site investigators worked closely with subspecialist colleagues. Many of these clinicians, as well as research ethics boards, were concerned about contacting families in which a child has a life-threatening condition. We used surveys to study the impact of the research on families. Not a single parent respondent regretted participation, 96% believed the study had value and 97% supported conducting research in paediatric palliative care (16). Families' strong support for research highlights the potential

**TABLE 3**  
**Family variables (n=258)**

Family variable	Designated parent
Size of community*	
Metropolitan area (≥1 million)	78 (30.4)
Large city (100,000–999,999)	59 (23)
Medium/small city (10,000–99,999)	65 (25.3)
Town (1,000–9,999)	38 (14.8)
Village (300–999)	13 (5.1)
Other	4 (1.6)
Average household income, \$*	
<40,000	73 (29)
40,000–<80,000	92 (36.5)
80,000–<120,000	57 (22.6)
≥120,000	30 (11.9)
Current income meets needs*	
Completely	20 (7.8)
Very well	23 (9)
Adequately	87 (34.1)
With some difficulty	83 (32.5)
Not very well	34 (13.3)
Totally inadequate	8 (3.1)
Family income changed as result of child's illness*	
No	72 (28.1)
Yes†	184 (71.9)
Parent stopped working/unable to work	92 (50)
Parent went to part-time	43 (23.4)
Income decreased	42 (22.8)
Parent needs to work flexible hours	22 (12)
Increased medical costs	19 (10.3)
Other	12 (6.5)
Income decreased drastically	11 (6)
Income improved	7 (3.8)
Parent went to full-time	2 (1.1)
Parent stopped being a student	2 (1.1)
Responsible for some costs resulting from child's illness*	
No	49 (19.4)
Yes	204 (80.6)
If 'yes', difficulty in managing these costs (1–10), mean ± SD (n=200)	5.8±2.5

Data presented as n (%) unless otherwise indicated. \*Numbers do not total to sample size because of missing data; †Numbers do not total to sample size because multiple responses could be reported

impact new evidence-based knowledge may have for them, and was encouraging to us.

Our work had limitations. In the present study, we were able to follow children and families for three years at most. These conditions evolve slowly; thus, more time is needed. Second, this was not an inception cohort; children were recruited at varying stages of disease. Third, it was not a registry-based study, so we did not follow every child with a relevant condition. While recruitment was not directly through the palliative care teams, most of the families were involved with palliative care. This may be one reason why there was so much penetration of service even into smaller communities. Families whose children have rare conditions are likely to be followed at tertiary centres and more likely to encounter palliative care teams. This may not hold true for all children who need such care, eg, children with hypoxic-ischemic injury who were not part of the present study or those with noncurable malignancies who wish to be at home.

The goal of the present summary article was to highlight areas of importance to generalist and subspecialist paediatricians. Clinicians should be aware of emerging information on symptom patterns. They should also focus attention on the well-being of siblings and parents, identifying both areas of concern and strengths. Children live in the context of the family and their quality of life impacts, and is impacted by, their family.

We plan to continue our analysis and knowledge dissemination activities, and are seeking funding to extend the time-frame of the observational cohort to better understand their clinical trajectories and family experiences. Our hope is to develop the evidence base to target support and interventions that will benefit children with these progressive conditions and their families.

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