Author/s:
Vemuri, S;Baker, L;Williams, K;Hynson, J

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Title

The last two years of life for children with severe physical disability: observations from a tertiary paediatric centre.

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Authors

Dr Sidharth Vemuri [a,b]

Dr Louise Baker [b,c]

Professor Katrina Williams [b,d,e]

Associate Professor Jenny Hynson [a,d]

[a] Victorian Paediatric Palliative Care Program

[b] Department of Neurodevelopment and Disability, The Royal Children's Hospital, Melbourne

[c] Victorian Paediatric Rehabilitation Service

[d] Department of Paediatrics, The University of Melbourne

[e] Murdoch Children's Research Institute

Address of the institution at which the work was carried out together

The Royal Children's Hospital, Melbourne

50 Flemington Road, Parkville VIC 3052

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Corresponding author

Dr Sidharth Vemuri

Victorian Paediatric Palliative Care Program

3 West Clinical Offices, The Royal Children's Hospital, Melbourne

50 Flemington Road, Parkville VIC 3052

Email: sidharth.vemuri@rch.org.au

Phone: 03 9345 5374

ABSTRACT

Aims: To describe the clinical course of children with severe physical disability (SPD) in the two years prior to their death and identify whether these children had palliative care involvement and advance care planning prior to death. To investigate whether there is a difference between children with progressive (PSPD) and non-progressive (NPSPD) aetiologies of SPD.

Method: A retrospective cohort analysis of 48 children with SPD, who died between 1 January 2013 and 1 January 2015 at The Royal Children's Hospital, Melbourne. Clinical charts were reviewed to collect data about type of SPD, frequency and duration of hospital admissions, duration of palliative care involvement (if any), and presence of an advance care plan.

Results: The majority of children were admitted in the six months before their death, and over a third were admitted to the intensive care unit (ICU). There was a significant increase in the frequency of hospital admissions as the study cohort approached death (p=0.003). The majority of children with SPD were offered a referral to a palliative care service, with referrals more likely in children with PSPD (90%) compared to children with NPSPD (57%). While approximately 60% of children

in each cohort had an advance care plan, there was a trend towards this being formalised earlier in children with PSPD (p=0.09).

Conclusion: The increase in hospital admissions prior to death in children with SPD suggests an opportunity for greater consistency in offering advanced care planning and palliative care, especially to those with NPSPD.

What is already known on this topic?

- The 20-year mortality rates for children with SPD are 40-60%.
- In many cases, there is a steady period of physical decline prior to death.
- There is variation in the approach to end of life management.

What this paper adds?

- This cohort of children with SPD had an increasing frequency of hospital admissions as they approached death.
- There are opportunities for greater consistency with referral to a palliative care service and advance care planning in children with SPD, regardless of aetiology.
- There may be key features in the clinical course of children with SPD that may assist in predicting death.

Keywords

Death

Severe physical disability

Paediatric palliative care

Advance care planning

INTRODUCTION

Much of the research conducted in paediatric palliative care has focused on children with malignant conditions. Although children with severe physical disabilities (SPD) are known to have a high 20-year mortality rate (40-60%)[1-4], relatively little is known about their experience of care. Cerebral palsy is the most common form of SPD[3], and the degree of physical disability is related to the risk of death.[1, 2, 5, 6] There are other pathologies that produce similar clinical phenotypes, however, the absolute number of these is small, and there is limited literature on disease-specific mortality rates. It is widely accepted that, regardless of aetiology, children with SPD will have a shortened life. It may be expected that children with aetiologies of SPD that cause ongoing degeneration in the central nervous system (progressive SPD) have a more predictable pathway towards death, compared to children with cerebral palsy (non-progressive SPD).[7] However, there are no published studies describing either path.

There is increasing consensus that timely referral to palliative care and opportunity to participate in advance care planning can enhance the overall experience and satisfaction of patients and families of children with life-limiting conditions. Potential benefits include improved control of complex symptomatology and greater opportunity for families to discuss goals of care and make choices regarding interventions,[8, 9] reduction in suffering and improvement in quality of life.[10] This may reduce trauma to families, as they are better prepared for the terminal phase, and create opportunities for consideration of non-acute settings for care.[8] Palliative care can be integrated with, and sit alongside, ongoing efforts to prolong life and this approach may be appropriate for many years for children with SPD.

The uncertainty in diagnosing dying in children with SPD can make appropriate timing of advance care planning and introduction to palliative care challenging.

Current clinical practice depends on the physician's experience and individual preference as to the timing of such discussions. Commonly, these issues are raised in the intensive care unit (ICU), at the time of an acute deterioration where decisions are made to withhold or withdraw life-prolonging therapies.[7]

The aim of this study was to describe the clinical course of children with SPD in the two years prior to their death. We also aimed to identify the extent to which children with SPD had palliative care involvement and advance care planning in the two years prior to death and investigate whether practices differ according to whether the child has a progressive (PSPD) or non-progressive (NPSPD) aetiology of SPD.

METHODS

This retrospective cohort analysis was conducted at the Royal Children's Hospital, Melbourne (RCH), the largest tertiary paediatric referral centre serving families living in Victoria, Tasmania, and regional New South Wales. Ethics approval for the project was granted by the hospital's Human Research Ethics Committee.

Patient selection

Using the hospital's patient administration system, we identified all children who died between 1 January 2013 and 1 January 2015, and were either managed by the Developmental Medicine, General Medicine, Neurology or Metabolic teams, or had relevant diagnosis codes from 'The International Statistical Classification of Diseases and Related Health Problems, 10th Revision, Australian Modification (ICD-10-AM)'[11] including E70-80, E88, E83.0, E90, F82, F84.2, G00-26, G30-32, G35-37, G46, G92-94, G98-99, P00-04, P05, P07, P15.9, P20-21, P35-37, P50-61, P70.4, and P90-96, assigned in previous hospital admissions from July 1998 to December 2014.

Eligibility criteria

Of the children identified, we included those aged between one and 18 completed years at the time of death who had SPD. Death may have occurred in an inpatient, respite or outpatient setting. SPD was defined as impairments in all areas of motor function, with physical impairments that restricted voluntary control of movement, and the inability to independently mobilise, even with adaptive equipment.[12] Children were excluded if their SPD was a result of congenital skeletal anomalies. Eligible children needed to have been reviewed at least once by a hospital-based medical professional, in an inpatient or outpatient setting, in the two years prior to death.

Patient grouping

Children were grouped as either NPSPD or PSPD. PSPD were defined as SPD due to disease processes that caused ongoing degeneration in the central nervous system, with the clinical phenotype of developmental regression. This included conditions such as Rett syndrome, Batten disease and mitochondrial disorders. NPSPD were defined as SPD resulting from a single, static insult to the central nervous system (e.g. cerebral palsy).

Data collection

Individual hospital records were accessed to extract data related to clinical features of SPD, age at death, number and length of unplanned overnight hospital admissions, including admission to ICU, between 18-24 months (T24), 12-18 months (T18), 6-12 months (T12) and 0-6 months (T6) prior to death. Palliative care involvement and the presence of an advance care plan were recorded. During the study period, the only option for structured documentation of advance care planning atRCH was the "Treatment plan for a child with a life-limiting condition" document.

This document aimed to define preferences and goals for future medical care, including communicating resuscitation plans to staff who may not know the child well. Study data were collected and managed using REDCap (Research Electronic Data Capture) tools hosted at the Murdoch Children's Research Institute, which is partnered with the RCH.[13] REDCap is a secure, web-based application designed to support data capture for research studies.

Data analysis

Descriptive analyses were performed using Stata 13.1 (StataCorp. 2013. *Stata Statistical Software: Release 13*. College Station, TX: StataCorp LP). The Mann-Whitney test was used to identify any statistical significance of differences in gender and circumstances of death, between children with PSPD and NPSPD. Hospital lengths of stay, including admission to ICU, were compared between the four time periods before death, using absolute values, and median values with interquartile ranges (IQR) to identify any potential trends. Analysis of the incidence rate ratio (IRR) of overnight hospital admissions, and summary frequencies of overnight hospital admissions for each of the four six-monthly time periods before death were compared using Poisson regression. To assess differences between timing of palliative care referral, advance care planning, and death, log transformation of the dataset and Student's T-test analysis was used.

RESULTS

There were 48 children eligible for inclusion in this study, with an equal proportion of males and females. Twenty (42%) children had a PSPD. There was no evidence of gender differences among children with PSPD and NPSPD (p=0.56). The causes of PSPD included Rett syndrome (n=2), mitochondrial disorders (n=2), Batten disease (n=3), Aicardi syndrome (n=2), leukodystrophy (n=2), GM1 gangliosidosis (n=1),

infantile neuroaxonal dystrophy (*n*=2) and Cockayne syndrome (*n*=1). There were five children who had no causal diagnosis for their PSPD.

Circumstances of death

The median age of death was 5.9 years (IQR 2.0 – 7.9 years) in children with PSPD and 8.8 years (IQR 5.0 - 14.3 years) for children with NPSPD. In this relatively small sample, there was a non-significant trend for children with PSPD to die earlier, with median age of death 2.9 years earlier (95% CI 0.0, 6.2 years; *p*=0.06).

Hospital admissions

Most children were admitted to hospital and over one third were admitted to ICU in the last six months of their life (Table 1). As a group, in their last six months of life, 15 children with PSPD accounted for 38 admissions to RCH, and 18 children with NPSPD accounted for 25 admissions to RCH. Each child admitted to ICU was also admitted to the ward and was only admitted to ICU once during their last six months.

Table 2 lists the number and median duration of unplanned overnight hospital admissions for children with NPSPD and PSPD. There was an increase in the frequency of admissions in both groups as children approached death. For the whole cohort, there was evidence of a 32% increase in the number of overnight hospital admissions for each six-monthly time period as death approached. This result suggests that the expected increase in overnight admissions in the four six-monthly time periods approaching death could be between 10% to 58% (*p*=0.003). There was no evidence to suggest longer admissions for the study cohort, or a difference between children with PSPD and NPSPD, as they progressed towards death.

The number of ICU admissions increased as the whole cohort progressed towards death, but this did not reach statistical significance. Children with PSPD only used ICU within the last six months of life, while children with NPSPD had ICU admissions in all four six-monthly time periods. There was a non-significant trend toward children with NPSPD having longer lengths of stay in ICU in the last 12 months of life.

Palliative care referral

There were 34 (71%) children with SPD who had been referred to the RCH palliative care service, in the two years prior to death. Eighteen (90%) children with PSPD were referred, with a median time of referral before death of 44.5 months (IQR 7.7 - 62.5). Of the 15 children with PSPD who were admitted in their last six months of life, three (20%) were referred to palliative care in the last two months of their life. Sixteen (57%) children with NPSPD were referred to palliative care, with a median time between palliative care referral and death of 20.1 months (IQR 0.6 - 121.2). For the 18 children with NPSPD who were admitted in their last six months of life, three (17%) were referred on the day of their death, and two (11%) were referred in the last two months of life. While a greater proportion of children with PSPD were referred to palliative care, there was no statistical association between the timing of this referral and aetiology of SPD.

Advance care planning

An advance care plan documented in the two years prior to death was found in the records of 28 (58%) children. Twelve (60%) children with a PSPD had an advance care plan created a median of 8.5 months (IQR 2.3 - 44.6) before death. Of the 15 children with PSPD who were admitted in their last six months of life, five (33%) had an advance care plan developed in their last month of life. Sixteen (57%) children with NPSPD had a documented advance care plan, created a median of 1.7 months (IQR 0.4 - 20.6) before death. Of the 18 children with NPSPD who were admitted in

their last six months of life, seven (39%) had an advance care plan developed in the last week of their life. There was a trend suggesting that advance care plans were created earlier in children with PSPD (p=0.09).

Place of death

There were 23 (48%) children who died in hospital, 10 (21%) children who died at home, and six (12%) children who died in a respite facility. Location of death was not documented in nine (19%) children. These findings were similar for both groups, with nine (45%) children with PSPD, and 14 (50%) children with NPSPD dying in hospital. There was no evidence to suggest an association between location of death and palliative care involvement.

DISCUSSION

There is a paucity of literature describing palliative care referral patterns and advance care planning practices in children with SPD. This study supports the findings that children with PSPD were referred to palliative care more frequently when compared to children with NPSPD.[7] While there was no evidence to suggest children with PSPD were referred to palliative care earlier, there was a trend toward children with PSPD having advance care plans documented earlier. This may reflect greater clinician comfort in raising issues about end-of-life care in children with PSPD compared to NPSPD, despite a significant mortality rate in the latter group.[1-4]

Similar to recent published data[7], most of our patient cohort died in hospital than home or hospice, even with palliative care involvement. In the paediatric oncology literature[9, 14], it is suggested that many parents would prefer to care for their dying child at home. While examination of parental preference for end of life care was

outside the scope of this study, the available findings may suggest this might not be the case in the setting of SPD. One possible explanation for the predominance of deaths in hospital is prognostic uncertainty and the challenge of trying to differentiate reversible causes of deterioration, compared to one related to end of life progression. Another explanation may be that families find comfort in the familiarity of hospital surroundings, particularly if there has been a high rate of hospital utilisation during their child's life.

There was a trend towards an increased number of unplanned overnight admissions in the two years prior to death, regardless of aetiology of SPD. There are no directly comparable published data. The clinical significance of this finding should not be over-interpreted as a predictor of impending death, as other studies[15-17] demonstrate that children with NPSPD experience a greater frequency of admissions than those with milder physical disabilities.

Duration of admissions did not increase in this cohort of children with SPD as death approached. However, children with NPSPD had longer admissions in the 12 months before their death when cautiously compared to other published data.[15, 17]

Our data suggest an increased need for ICU in the last six months of life in children with SPD, however there are no comparable published data. This may reflect greater frailty as these children approach death.

Study limitations

This is a small cohort analysis with considerable inter-patient variation. In an attempt to identify trends, and to take into account variability, we employed non-parametric tests of the data, but larger studies will be required to decide if observed trends are statistically significant.

This study examined the medical records from a single tertiary institution, which may mean that the most severe forms of both physical disability and adverse outcomes are over-represented. This data source may not accurately reflect health service provision, as it does not include data about medical presentations and management at other hospitals. As such the findings of this study should not be generalised to non-tertiary paediatric settings.

Being a retrospective study, we relied on clear documentation within medical records, especially in identifying the timing of referral to palliative care. The presence of a completed "Treatment plan for a child with a life-limiting condition" document was used as the indicator of advance care planning. However, this may under-represent the frequency of such discussions, as it may not reflect those that are documented within patient letters or discharge summaries. Furthermore, it would not capture similar forms that were created for the same child at other institutions, as these would be filed as external correspondence within the individual hospital record. There was a large number of records where location of death was not documented, highlighting the importance of clear documentation to assist in data collection in this area of research.

CONCLUSION

Although the majority of children with SPD have advance care planning and are referred to palliative care in the two years prior to death, practice is not consistent. We identified a small number of children with NPSPD who were referred to palliative care on the day of their death and our findings suggest that clinicians may be more likely to address end-of-life issues in children with PSPD. This may result from the expected and perhaps more predictable, clinical degeneration in this patient group.

Despite a significant mortality rate among children with NPSPD, we question whether there is a greater hesitancy to refer to palliative care and enter into advance care planning. A postulated explanation for this hesitancy is the difficulty in diagnosing dying.

We have demonstrated that regardless of the aetiology of SPD, there is a greater frequency of hospital admissions as these children approach death. Additionally, there was also a trend for greater utilisation of ICU in the last six months of life, illustrating a need to collaborate with staff in the ICU to ensure holistic care is provided during these admissions. The next challenge is to explore this trend further in a larger patient cohort and to compare it with the hospital utilisation requirements of a matched cohort of surviving children with SPD. Findings from such studies may assist with predicting death, leading to greater consistency in the provision of important elements of care, such as advance care planning and referrals to sources of support.

REFERENCES

1. Baird G, Allen E, Scrutton D, Knight A, McNee A, Will E, et al. Mortality from 1 to 16-18 years in bilateral cerebral palsy. *Arch Dis Child*. 2011;96(11):1077-1081.
2. Brooks JC, Strauss DJ, Shavelle RM, Tran LM, Rosenbloom L, Wu YW. Recent trends in cerebral palsy survival. Part II: individual survival prognosis. *Dev Med Child Neurol*. 2014;56(11):1065-1071.
3. Hutton JL, Pharoah PO. Life expectancy in severe cerebral palsy. *Arch Dis Child*. 2006;91(3):254-258.
4. Westbom L, Bergstrand L, Wagner P, Nordmark E. Survival at 19 years of age in a total population of children and young people with cerebral palsy. *Dev Med Child Neurol*. 2011;53(9):808-814.
5. Reid SM, Carlin JB, Reddihough DS. Survival of individuals with cerebral palsy born in Victoria, Australia, between 1970 and 2004. *Dev Med Child Neurol*. 2012;54(4):353-360.
6. Reddihough DS, Baike G, Walstab JE. Cerebral Palsy in Victoria, Australia: Mortality and causes of death. *Journal of paediatrics and child health*. 2001;37:183-186.
7. DeCourcey DD, Silverman M, Oladunjoye A, Balkin EM, Wolfe J. Patterns of Care at the End of Life for Children and Young Adults with Life-Threatening Complex Chronic Conditions. *J Pediatr*. 2017.
8. Fowler K, Poehling K, Billheimer D, Hamilton R, Wu H, Mulder J, et al. Hospice referral practices for children with cancer: a survey of pediatric oncologists. *J Clin Oncol*. 2006;24(7):1099-1104.
9. Vickers J, Thompson A, Collins GS, Childs M, Hain R. Place and provision of palliative care for children with progressive cancer: a study by the Paediatric Oncology Nurses' Forum/United Kingdom Children's Cancer Study Group Palliative Care Working Group. *J Clin Oncol*. 2007;25(28):4472-4476.
10. Hammes BJ, Klevan J, Kempf M, Williams MS. Pediatric Advance Care Planning. *J Palliat Med*. 2005;8(4):766-772.
11. Organisation WH. ICD-10 International Statistical Classification of Diseases and Related Health Problems 10th Revision. Geneva2011.
12. Palisano R, Rosenbaum P, Walter S, Russell D, Wood E, Galuppi B. Development and reliability of a system to classify gross motor function in children with cerebral palsy. *Dev Med Child Neurol*. 1997;39:214-223.
13. Harris PA, Taylor R, Thielke R, Payne J, Gonzalez N, Conde JG. Research electronic data capture (REDCap)--a metadata-driven methodology and workflow process for providing translational research informatics support. *J Biomed Inform*. 2009;42(2):377-381.

14. Cohn RJ, Goodenough B. Health professionals' attitudes to videoconferencing in paediatric health-care. *J Telemed Telecare*. 2002;8(5):274-282.
15. Meehan E, Freed GL, Reid SM, Williams K, Sewell JR, Rawicki B, et al. Tertiary paediatric hospital admissions in children and young people with cerebral palsy. *Child Care Health Dev*. 2015;41(6):928-937.
16. Berry JG, Hall DE, Kuo DZ, Cohen E, Agrawal R, Feudtner C, et al. Hospital Utilization and Characteristics of Patients Experiencing Recurrent Readmissions Within Children's Hospitals. *JAMA*. 2011;307(7):682-690.
17. Young NL, McCormick AM, Gilbert T, Ayling-Campos A, Burke T, Fehlings D, et al. Reasons for hospital admissions among youth and young adults with cerebral palsy. *Arch Phys Med Rehabil*. 2011;92(1):46-50.

TABLES

Table 1. *Number and proportion of children with overnight admissions to the ward or ICU in the four six-monthly time periods (T6-T24) before death*

	PSPD (n=20)		NPSPD (n=28)		TOTAL (n=48)	
	ICU	Ward	ICU	Ward	ICU	Ward
T24	0	6 (30%)	3 (11%)	11 (39%)	3 (6%)	17 (35%)
T18	0	11 (55%)	4 (14%)	14 (50%)	4 (8%)	25 (52%)
T12	0	12 (60%)	6 (21%)	14 (50%)	6 (13%)	26 (54%)
T6	7 (35%)	15 (75%)	10 (36%)	18 (64%)	17 (35%)	33 (69%)

All children admitted to ICU were also admitted to the ward. ICU, intensive care unit; NPSPD, non-progressive severe physical disability; PSPD, progressive severe physical disability.

Table 2. *Number and duration of hospital admissions for children with SPD in the four six-monthly time periods (T6-T24) before death*

	PSPD	NPSPD
Number of unplanned overnight admissions		
T24	10	10
T18	12	11
T12	23	24
T6	38	25
Number of ICU admissions		
T24	0	3
T18	0	4
T12	0	6
T6	7	10
Median length of stay in hospital, in days (IQR)		
T24	13 (3.5-26.25)	5 (2-16)
T18	9 (4-15)	5.5 (2.25-11.5)
T12	7.5 (4.75-11.25)	19 (3.25-35.25)
T6	16 (9.5-45.5)	7 (3.25-34)
Median length of stay in ICU, in days (IQR)		
T24	0	8 (4.5-20)
T18	0	3 (1.75-4.25)
T12	0	12 (7.5-20.25)
T6	7 (5.5-16)	11 (2.25-23.75)