Received: 17 March 2022 | Accepted: 2 September 2022

DOI: 10.1111/dmcn.15420

ORIGINAL ARTICLE

Survival of individuals with cerebral palsy in Victoria, Australia: A longitudinal cohort study spanning four decades

Gina L. Hinwood[1,2] | **Hayley Loftus**[1] | **Daisy A. Shepherd**[1] | **Angela Guzys**[1] | **Dinah S. Reddihough**[1,3,4] | **Susan M. Reid**[1,3,4]

[1]Neurodisability and Rehabilitation, Murdoch Children's Research Institute, Parkville, Victoria, Australia

[2]Victorian Paediatric Rehabilitation Service, Monash Children's Hospital, Clayton, Victoria, Australia

[3]Department of Paediatrics, University of Melbourne, Parkville, Victoria, Australia

[4]Department of Neurodevelopment and Disability, The Royal Children's Hospital, Parkville, Victoria, Australia

Correspondence
Gina L Hinwood, Neurodisability and Rehabilitation, Murdoch Children's Research Institute, 50 Flemington Road, Parkville, Victoria, Australia.
Email: gina.hinwood@mcri.edu.au

Funding information
Victorian Government; Cerebral Palsy Alliance Research Foundation; Children's Hospital Foundation; Victorian Department of Health

Aim: To provide an updated description of the rates, trends, and predictors of mortality of individuals with cerebral palsy (CP), born in the Australian state of Victoria between 1970 and 2012.

Method: Data were extracted for 4807 individuals (2091 females; 2716 males). The probability of survival to 30th June 2017 was calculated using the Kaplan–Meier method. Mortality rates were calculated per 1000 person-years using age strata and compared with population mortality rates to produce mortality ratios. Cox proportional hazards regression was used to calculate hazard ratios for selected demographic and clinical characteristics and to estimate the effect of birth epoch on 15-year survival.

Results: There were 666 recorded deaths. Compared to the general population, mortality was higher for all persons with CP and highest for children aged 1 to 15 years (45–62 times). We observed 35% improvement in the probability of survival to 15 years for births in the 2000s relative to the 1970s (hazard ratio 0.65, 95% confidence interval [CI] 0.49, 0.86), but only 4% improvement for the subgroup with complex CP (hazard ratio 0.96, 95% CI 0.69, 1.33).

Interpretation: The observed improvements in survival for those born in the 2000s is likely related predominantly to a proportional reduction in complex CP within the cohort.

Cerebral palsy (CP) is a non-progressive, multifaceted condition that affects posture, limits movement, and sometimes coexists with other conditions such as epilepsy, intellectual disability, and sensory impairments. Worldwide, CP has been estimated to occur in 1.6 cases per 1000 live births in high-income countries,[1] but evidence suggests the rate is declining.[1–6] In Australia, the estimated birth cohort prevalence of CP for the birth years 2010 to 2012 was 1.4 cases per 1000 live births and in the Australian state of Victoria it was 1.2 per 1000 live births.[2]

Reported survival rates in CP are historically variable because of disparities between study samples that include different age groups and severity profiles. On the other hand, studies comparing survival rates in population cohorts in high-income countries, and when severity was taken into account, showed that rates were similar between geographic regions.[7–9] Because of the considerable variations in clinical presentations of CP, information on length of survival based on combinations of clinical characteristics is informative for people with CP and their families, clinicians, epidemiologists, and legal counsel in the event of litigation procedures. Moreover, life expectancy in CP is an important public health measure, informing service planning and public health policy.

As custodians of the Victorian CP Register, we have access to longitudinal, population-based data dating back to 1970.

DOI: 1111/dmcn.15346

In 2012, when we reported the survival of individuals with CP born in Victoria between 1970 and 2004, we observed no improvement in survival by birth decade.[10] These negative findings were unexpected given there had been advances in obstetric care and innovations in neonatal and paediatric treatment regimens that might have been expected to improve mortality. Lack of strong evidence in favour of improved survival was also reported from other groups with population registries, including another Australian study,[11] three studies from the UK,[12–14] and studies from Denmark[15] and Sweden.[16] In contrast, a Californian group showed modest improvement in survival over time among children who were aged 4 years and unable to lift their heads in prone, no improvement in survival for those aged over 15 years not fed via gastrostomy, and a possible weak trend for those with milder disability;[17] these findings have been supported by more recent research by the same group.[18]

CP is a lifelong condition, with little imminent possibility of cure. New or improved management practices are more likely to delay rather than prevent early death for those most severely affected. This effect has been demonstrated in two recent studies whereby early improvements in survival subsequently reverted to historical levels.[10,19] In contrast to the concept of delayed mortality due to improved management of complex CP, changes in the population distribution of CP phenotypes are likely to impact survival rates at all ages by changing the proportion most at risk. This has current relevance to survival trends because several research groups, including our own, have reported a reduction not only in birth prevalence of CP,[2–6,20] but also in motor severity and complex developmental disability profiles.[3,4,6,20] Our Victorian data showed that decreasing CP rates in the 2000s were associated with relatively greater decreases in the rates of Gross Motor Function Classification System (GMFCS) levels III to V, bilateral CP, epilepsy, and intellectual impairment.[4] Many factors have contributed to these temporal changes in clinical profiles of CP, including stabilization of neonatal death rates and improved neuroprotection in survivors of preterm birth and perinatal complications.

Considerable time has elapsed since we last reported survival for people with CP in Victoria, and it was deemed important to reassess whether survival has subsequently improved. The aim of the present study, therefore, was to (1) provide an updated description of the rates, trends, and predictors of mortality in individuals with CP who were born in the Australian state of Victoria between 1970 and 2012; and (2) examine if any recent improvements in survival in our most recent birth cohorts were related to changes in severity profiles of the population with CP.

What this paper adds

- Length of survival improved for Australians with cerebral palsy (CP) born this millennium.
- Improved survival was mainly because of a proportional reduction in complex CP.
- A small improvement in length of survival was seen for children with complex CP.

METHOD

Research setting and ethics

This population-based, observational study was performed at the Murdoch Children's Research Institute, Melbourne, Australia. Ongoing ethics approvals and monitoring of the activities of the Victorian CP Register is conducted by The Royal Children's Hospital Human Research Ethics Committee. For the data linkage, approval was obtained from the Australian Institute of Health and Welfare Ethics Committee.

Study population

This study assessed survival in individuals with CP who were born in Victoria between 1st January 1970 and 31st December 2012. The study population comprised participants in the Victorian CP Register, a long-standing, population-based research project that holds data on persons with CP born in Victoria, Australia since 1st January 1970. Data for the Victorian CP Register project are mainly obtained by searching medical records at Victoria's two tertiary paediatric hospitals. Confirmation of the CP diagnosis is made for each case at approximately 5 years of age based on accepted definitions and eligibility criteria.[21,22] Records are only removed if a case no longer meets the eligibility criteria.

Data collection, definitions, and data management

For this study, data extracted from the Register databank comprised date of birth, death status, date of death, sex, birth gestation, motor topography (unilateral [hemiplegia/monoplegia], bilateral [diplegia/triplegia/quadriplegia]), motor severity (mild, moderate, severe), and presence of epilepsy, blindness, bilateral deafness, lack of speech, and intellectual disability. Motor topography was classified according to the number of involved limbs and motor severity on grouped levels of the GMFCS.[23] Individuals who walked independently at the age of 5 years (GMFCS levels I and II) were categorized as having mild motor impairment. Individuals walking using a hand-held mobility device in most indoor environments were categorized as having moderate motor impairment (GMFCS level III), whereas those who were non-ambulant or only able to walk short distances with assistance or with the use of a body-support walker (GMFCS levels IV and V) were categorized as having severe motor impairment. An epilepsy diagnosis was determined on the background of

a history of at least two confirmed unprovoked seizures outside the neonatal period. The presence of blindness, deafness, lack of speech, and intellectual disability were, in the first instance, determined by formal testing and, if unavailable, by clinical interpretation.

Deaths were obtained via linkage with the Australian National Death Index, the Australian Institute of Health and Welfare's register of all deaths in Australia since 1980, and searches of the Victorian Death Index for deaths before 1980. Population age-specific mortality rates were derived from the Australian Bureau of Statistics.[24]

Statistical analysis

Lifetimes were calculated as the number of days from birth until death, or until the censor date of 30th June 2017, whichever came first. Survival data were summarized, and a Kaplan–Meier survival curve was constructed.

Frequency distributions of clinical characteristics of the population with CP were calculated. To estimate the mortality risk associated with each clinical characteristic, we used univariate Cox proportional hazards regression modelling to produce hazard ratios associated with each characteristic, together with the corresponding 95% confidence intervals (CI). Hazard ratios were estimated relative to the subgroup with the lowest mortality. We also performed multivariable Cox proportional hazards regression adjusting for (1) motor severity and (2) all selected clinical characteristics. Models were fitted under the assumption of proportional hazards. We assessed whether the assumption of proportional hazards was met for each model graphically and by performing a test based on Shoenfeld residuals. We considered violations in this assumption when interpreting our results.

After stratification by age, we calculated unadjusted mortality rates per 1000 person-years. The first age stratum was under 1 year, the second 1 year to 4 years; subsequent strata consisted of 5-year intervals, with a final age stratum of 45 years to 47 years. Person-years were computed for each age stratum by summing the total number of years that individuals were at risk of death. Mortality rates for CP were compared to population mortality rates to produce rate ratios. Comparison of survival to 10 years, 20 years, 30 years, and 40 years across four birth epochs was performed based on a combination of motor severity and number of additional impairments or conditions.

To assess temporal trends in survival, we subdivided the CP cohort into four epochs based on year of birth (1970–1979, 1980–1989, 1990–1999, 2000–2012) and used Cox proportional hazards regression to estimate the effect of birth epoch on 15-year survival. To evaluate whether any observed temporal change in mortality risk might be explained by changes over time in the population proportion of complex CP, we adjusted our regression model for complex CP and also stratified on complex CP. Categorization as medically complex CP was based on the presence of severe motor impairment and at least two additional impairments. To show temporal trends visually, we compared Kaplan–Meier curves across two birth epochs (1970–1999 and 2000–2012) for all CP and for CP stratified on complex developmental disability. All statistical analysis was conducted using Stata 16.0 (StatCorp 2019, College Station, TX, USA).

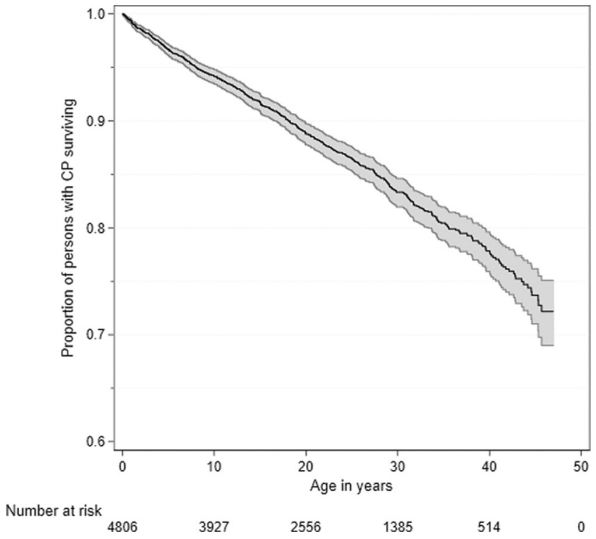

FIGURE 1 Kaplan–Meier graph showing survival estimates and 95% confidence intervals for persons with cerebral palsy (CP) born in Victoria, Australia, 1970–2012

RESULTS

The study cohort comprised 4807 individuals with CP, 2716 males (56.5%) and 2091 females (43.5%). Time in the study was a median of 21 years 4 months and a total of 108 156 person-years. There were 666 (13.9%) recorded deaths during the study period; 36 deaths occurred within the first year of life. The Kaplan–Meier survival curve for the population with CP is shown in Figure 1. Characteristics of the CP population related to clinical presentation of CP are summarized in Table 1.

Age-specific mortality is reported in Table 2. Unadjusted mortality rates were similar across age strata until age 40 years. When compared to the general population, mortality was higher for individuals with CP at every age. For those aged between 1 and 15 years, the mortality rate was 45 to 62 times the mortality rate in the general population, but was reduced to approximately 11 times the expected rate for deaths at 45 years to 47 years.

The probabilities of survival to 5, 15, 30, and 40 years for persons with CP were 96.7% (95% CI 96.14, 97.16), 91.8% (95% CI 90.89, 92.53), 83.3% (95% CI 81.94, 84.61), and 77.8% (95% CI 75.91, 79.61) respectively (Figure 1). Hazard ratios by motor topography, motor severity, and by the presence of epilepsy, lack of speech, intellectual disability, bilateral deafness, and blindness are presented in Table 1. Under the assumption of proportional hazards, the highest hazard ratios were seen for severe motor impairment relative to mild (hazard ratio 23.19, 95% CI 17.69, 30.41), lack of speech (hazard

TABLE 1 Participant characteristics

Characteristic	Total cohort *n* = 4807	Total deaths *n* = 666 (%)	Hazard ratio[a] (95% CI), unadjusted	Hazard ratio[a] (95% CI) adjusted for motor severity	Hazard ratio[a] (95% CI) adjusted for remaining variables
Sex					
Female	2091	288 (13.8)	1.00 (Reference)	1.00 (Reference)	1.00 (Reference)
Male	2716	378 (13.9)	1.05 (0.90, 1.23)	1.07 (0.92, 1.25)	1.14 (0.98, 1.33)
Motor topography					
Bilateral (diplegia/triplegia)	1311	32 (2.4)	1.00 (Reference)	1.00 (Reference)	1.00 (Reference)
Unilateral (monoplegia/hemiplegia)	1552	51 (3.3)	1.30 (0.84, 2.03)	2.15 (1.35, 3.44)	1.69 (1.06, 2.68)
Bilateral (quadriplegia)	1688	562 (33.3)	15.04 (10.53, 21.48)	3.30 (2.24, 4.87)	2.31 (1.55, 3.44)
Unknown	256	21 (8.2)	3.09 (1.78, 5.36)	2.60 (1.47, 4.59)	1.86 (1.05, 3.27)
Motor severity					
Mild	2780	59 (2.1)	1.00 (Reference)	-	1.00 (Reference)
Moderate	551	41 (7.4)	3.26 (2.18, 4.86)	-	2.22 (1.43, 3.46)
Severe	1400	548 (39.1)	23.19 (17.69, 30.41)	-	7.22 (4.90, 10.62)
Unknown	76	18 (23.7)	15.13 (8.92, 25.68)	-	3.01 (1.70, 5.34)
Epilepsy					
No	2891	171 (5.9)	1.00 (Reference)	1.00 (Reference)	1.00 (Reference)
Yes	1713	467 (27.3)	4.83 (4.05, 5.75)	2.22 (1.85, 2.65)	1.65 (1.37, 1.99)
Unknown	203	28 (13.8)	2.17 (1.45, 3.26)	2.36 (1.52, 3.66)	1.04 (0.66, 1.64)
Functional blindness					
No	4268	550 (12.9)	1.00 (Reference)	1.00 (Reference)	1.00 (Reference)
Yes	135	44 (32.6)	3.53 (2.59, 4.81)	1.39 (1.02, 1.89)	1.06 (0.78, 1.45)
Unknown	404	72 (17.8)	1.74 (1.36, 2.22)	1.96 (1.51, 2.55)	0.87 (0.64, 1.18)
Bilateral deafness					
No	4178	493 (11.8)	1.00 (Reference)	1.00 (Reference)	1.00 (Reference)
Yes	108	26 (24.1)	2.58 (1.74, 3.83)	1.76 (1.18, 2.61)	1.44 (0.97, 2.15)
Unknown	521	147 (28.2)	2.88 (2.40, 3.47)	2.95 (2.44, 3.56)	2.07 (1.64, 2.62)
Lack of speech					
No	3018	83 (2.75)	1.00 (Reference)	1.00 (Reference)	1.00 (Reference)
Yes	1286	464 (36.1)	15.72 (12.42, 19.88)	4.09 (3.16, 5.29)	2.79 (2.13, 3.64)
Unknown	503	119 (23.7)	10.83 (8.17, 14.35)	7.70 (5.75, 10.30)	4.69 (3.33, 6.61)
Intellectual disability					
No	1920	45 (2.3)	1.00 (Reference)	1.00 (Reference)	1.00 (Reference)
Yes	2487	557 (22.4)	10.16 (7.47, 13.81)	3.46 (2.52, 4.75)	1.77 (1.27, 2.48)
Unknown	400	64 (16.0)	9.40 (6.40, 13.80)	5.76 (3.86, 8.58)	1.70 (1.09, 2.66)

[a]Hazard ratios were estimated relative to the subgroup with the lowest mortality. -, not calculated; CI, confidence interval.

ratio 15.72, 95% CI 12.42, 19.88), and bilateral (quadriplegic) topography (hazard ratio 15.04, 95% CI 10.53, 21.48). The presence of epilepsy, intellectual disability, bilateral deafness, and blindness were also associated with higher risk of death at all ages (Table 1).

The probability of survival to 10, 20, 30, and 40 years by level of motor severity and number of additional impairments or conditions is shown in Table 3. The probability of survival to 40 years was 28.4% (95% CI 23.03, 33.94) for persons with severe limitations of gross motor function (predominantly non-ambulant) and at least three coimpairments or conditions. In comparison, the probability of survival to age 40 years for those with mild motor impairment and no additional impairments was 97.2% (95% CI 94.76, 98.50).

Comparatively better survival to 15 years was observed for persons born in the 2000s compared to earlier birth epochs (Table 4). Survival to age 15 years increased from 90.4% for persons born in the 1970s to 93.5% for those born between

TABLE 2 Age-specific mortality rates per 1000 person-years for CP and for the Victorian population and mortality ratios

Age in years	Number at risk	Person-years	CP deaths	Unadjusted CP mortality rate (95% CI)	Population mortality rate[a]	Mortality ratio (95% CI)
<1	4807	4789	36	7.5 (5.06, 9.97)	2.9	2.57 (1.75, 3.66)
1–4	4770	18 814	123	6.5 (5.38, 7.69)	0.1	45.90 (31.99, 67.05)
5–9	4580	21 332	113	5.3 (4.32, 6.27)	0.1	62.42 (41.20, 97.39)
10–14	3927	17 902	93	5.2 (4.14, 6.25)	0.1	59.40 (38.78, 93.50)
15–19	3225	14 435	95	6.6 (5.26, 7.90)	0.3	22.22 (16.67, 29.59)
20–24	2556	11 333	59	5.2 (3.88, 6.53)	0.3	16.63 (12.01, 22.79)
25–29	1987	8324	61	7.3 (5.49, 9.17)	0.4	17.29 (12.73, 23.21)
30–34	1385	5720	40	7.0 (4.83, 9.16)	0.5	14.39 (9.97, 20.29)
35–39	915	3564	23	6.5 (3.82, 9.09)	0.7	8.99 (5.61, 13.77)
40–44	514	1733	19	11.0 (6.03, 15.9)	1.1	10.27 (6.13, 16.24)
45–47	180	210	3	14.3 (0.00, 30.04)	1.2	11.82 (2.42, 34.92)

[a]Population mortality rate derived from the Australian Bureau of Statistics.[24] CI, confidence interval; CP, cerebral palsy.

TABLE 3 Percentage of survivors at 10, 20, 30, and 40 years by motor impairment severity

Motor severity + *n* additional impairments[a]	Start total	Total deaths	10-year survival (95% CI)	20-year survival (95% CI)	30-year survival (95% CI)	40-year survival (95% CI)
Mild motor impairment						
No impairment	1413	15	99.6 (99.15, 99.85)	99.4 (98.75, 99.73)	98.7 (97.51, 99.31)	97.2 (94.76, 98.50)
1 impairment	693	13	99.7 (98.80, 99.92)	99.1 (97.79, 99.62)	97.5 (95.28, 98.69)	95.6 (91.78, 97.61)
2 impairments	397	10	99.5 (97.98, 99.87)	99.1 (97.16, 99.72)	97.1 (93.86, 98.64)	94.1 (88.44, 97.02)
3 or more impairments	114	8	97.2 (91.49, 99.08)	96.2 (90.19, 98.56)	89.3 (79.07, 94.72)	89.3 (79.07, 94.72)
Moderate motor impairment						
No impairment	187	4	99.3 (96.27, 99.92)	99.5 (96.27, 99.92)	97.4 (91.65, 99.19)	95.8 (88.59, 98.51)
1 impairment	141	6	99.3 (95.07, 99.90)	98.2 (92.83, 99.56)	96.5 (88.75, 98.93)	89.7 (77.38, 95.52)
2 impairments	124	14	95.9 (90.41, 98.27)	92.1 (85.34, 95.83)	87.1 (78.51, 92.40)	81.3 (64.43, 90.68)
3 or more impairments	80	13	97.5 (90.31, 99.36)	90.5 (81.05, 95.37)	88.9 (78.91, 94.31)	69.4 (47.73, 83.49)
Severe motor impairment						
No impairment	83	3	100.0	98.4 (88.93, 99.77)	91.2 (73.85, 97.24)	91.2 (73.85, 97.24)
1 impairment	174	29	90.7 (85.20, 94.18)	83.1 (76.09, 88.27)	80.1 (71.72, 86.17)	77.1 (66.60, 84.66)
2 impairments	384	136	81.7 (77.46, 85.29)	70.9 (65.65, 75.45)	58.7 (52.33, 64.45)	49.2 (41.69, 56.32)
3 or more impairments	699	331	82.6 (79.55, 85.25)	60.1 (55.89, 63.96)	42.2 (37.40, 46.89)	28.4 (23.03, 33.94)

[a]Any intellectual impairment has replaced severe/profound intellectual impairment (in our 2012 paper[10]) as one of the five impairments/conditions. Number of additional impairments from a possible five, comprising epilepsy, intellectual impairment, blindness, deafness, and lack of speech. CI, confidence interval.

2000 and 2012 (Figure 2a). When we compared trends by birth epoch for complex CP only (Figure 2b), 15-year survival was 85.5% (95% CI 81.12, 88.88) for those born between 2000 and 2012 compared to 82.6% (95% CI 80.38, 84.51) for those born between 1970 and 1999.

Cox regression analysis showed 35% reduction in risk of death by 15 years for individuals born in the current millennium (hazard ratio 0.65, 95% CI 0.49, 0.86) relative to births in the 1970s (Table 4). Statistical adjustment for CP complexity in our Cox regression model indicated that the observed 35% improvement was reduced to 27% improvement in the adjusted model (adjusted hazard ratio 0.73, 95% CI 0.54, 0.99). When analysis was restricted to the complex subgroup, we saw only 4% reduction in risk between the two birth epochs (hazard ratio 0.96, 95% CI 0.69, 1.33).

DISCUSSION

Most children with CP will survive to adulthood, but it is clear from existing evidence that greater complex developmental disability is associated with increased risk of early death. Compared to the general population, mortality rates in the present study were higher for all age groups to 47 years. This finding is similar to that obtained from our previous study,[10] and to findings from other groups.[16] When compared to the general population, mortality ratios

TABLE 4 Birth decade comparisons of survival rates and mortality risk relative to the 1970 birth epoch

Birth epoch	5-year survival %	10-year survival %	15-year survival %	Risk of death	
				Unadjusted hazard ratio (95% CI)	Adjusted hazard ratio[a] (95% CI)
1970s	96.2	93.0	90.4	1.00 (Reference)	1.00 (Reference)
1980s	96.8	93.7	91.0	0.90 (0.73, 1.09)	0.98 (0.80, 1.21)
1990s	96.4	94.5	91.8	0.85 (0.68, 1.06)	1.02 (0.81, 1.29)
2000s	97.1	95.1	93.5	0.65 (0.49, 0.86)	0.73 (0.54, 0.99)

[a]Adjusted for cerebral palsy complexity. CI, confidence interval.

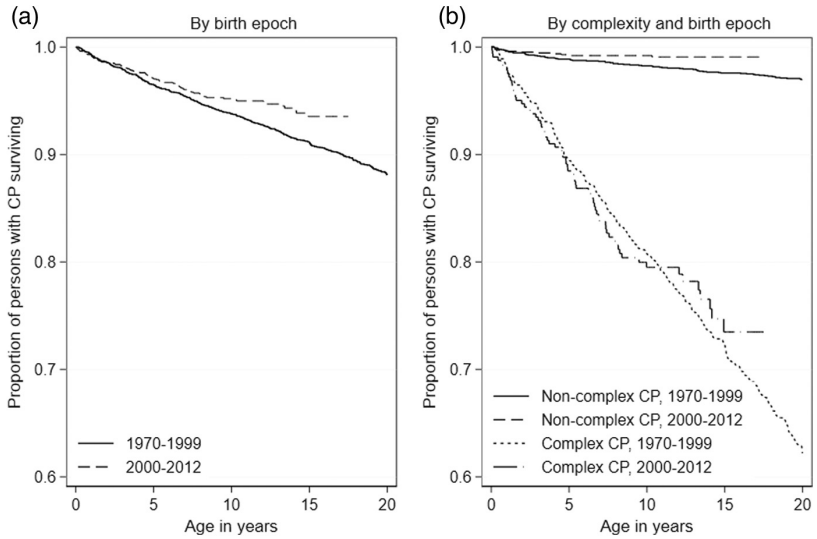

FIGURE 2 Kaplan–Meier graphs showing survival estimates by birth epoch for (a) all persons with cerebral palsy (CP) born in Victoria 1970–2012 and (b) the subgroup with complex CP

remained highest for those aged less than 15 years with the highest mortality ratio seen in the 5- to 9-year age group. More recently, other groups have reported a shift in relative mortality risk for populations with CP, from young children to adolescents and adults.[18,19]

Poor gross motor function, bilateral CP (quadriplegia), deafness, epilepsy, and lack of speech remained independent predictors for increased mortality risk. The present study supports previous findings that severe cognitive impairment in young people is associated with higher mortality risk.[10,11,16] In contrast to some previous studies, bilateral deafness was also associated with increased risk of death.[11,25] Although bilateral deafness is not common in CP, it is seen more frequently in individuals with poor gross motor function.[26]

Any temporal improvement in survival would indicate either better management of individuals at most risk of early death or improvement over time in overall severity of the CP population. Both outcomes would be valuable indicators of progress. There are comparatively few studies that have analysed survival among individuals with CP, and only a small number had the ability to analyse data across multiple decades.[7,10,16,18,19,27] Findings from other countries have been mixed, with most longitudinal population cohort studies reporting no improvement in survival for people with CP based on their time of birth.[11–16] Contrary to our previous study, we observed a trend towards improvement in survival to 15 years amongst all individuals with CP born between 2000 and 2012 compared to previous decades. At this stage, cases were still too young for us to fully ascertain the accuracy and longevity of this effect. It is possible that, with further data available, we may see a 'catch up' period, as previously described.[19]

Our study findings also include early indicators of a small improvement in survival within the complex CP group. Scoliosis surgery, improvements in nutritional management, and increases in the placement frequency and management of gastrostomy tubes (as described in Strauss et al.[17]), may be considered as reasons why survival has improved for complex CP cases.

Comparison between studies is difficult because of inherent differences in study samples across groups, classification criteria for CP, and differences in severity profiles. For example, some groups excluded children who died before 2 years,[16,28,29] or 1 year,[19] citing difficulties in assurance of an inarguable diagnosis of CP before this age and concern that infants, who may have received a CP diagnosis had they survived, are not included. Other studies only included children diagnosed with CP aged 4 years and over,[17,18] or only

analysed survival of adults with CP.[30] Levels of disability can vary widely between populations, and there are vastly different methods of data collection, variable classifications, analysis methods, and reporting of results. Variations in geographical location and socioeconomic status mean that comparisons with other studies, and generalizability of this study to other populations, must be made with caution.

Through statistical adjustment and adjustment by stratification, we were able to show that the substantially improved survival observed in persons born in the 2000 to 2012 birth epoch was largely a consequence of smaller proportions of cases with complex CP in the most recent birth epoch. To the censor date, the proportion of complex CP cases reduced from 26% in the 1970 to 1999 epoch to 20% in the 2000 to 2012 epoch. Some of the reduction in rates of severe or complex CP in Victoria has previously been shown to have occurred amongst children born very preterm, likely as a result of improved neonatal management.[31] Reduced rates of complex CP have also occurred in term livebirths,[10] but the reasons for this are not yet clear.

The Victorian CP Register project provided us with a very large, geographically defined, longitudinal cohort with nearly 50 years of follow-up and little risk of bias. As in previous studies,[7,17,18,32] our analysis took into account CP complexity, which added precision and interpretability to our results, improving generalizability and the potential for comparison with data from other countries. The same methodology for classifying the factors that are included in our rating of complexity has been used since the inception of the Victorian CP Register project, so changes over time in classification methods are unlikely to play a role in the findings.

Limitations

It is possible that a small number of persons with very mild CP (not requiring medical assistance or seeking services privately), were not included in this study. Some early deaths and deaths that occurred outside Australia may have been missed. Because of a lack of consistent data over the entire study period, we were unable to include some clinical factors that are known to affect survival, including tube feeding and nutritional status, severity of intellectual disability, and seizure control in epilepsy. In terms of the analysis, the assumption of proportional hazards may have been somewhat simplistic for this setting; although it was violated for some analyses, it did allow an easily interpretable estimate of the hazard ratio. We are also aware that inclusion in the study was conditional on survival to an age when CP could be diagnosed and the case registered, potentially increasing the risk of immortal time bias. However, we think we had reasonable success at ascertaining severe cases who died outside the neonatal period, so opted to accept a degree of bias instead of excluding ascertained cases that die early, thus potentially underestimating mortality or missing indicators of increased length of life.

Conclusions

This study demonstrates that individuals living with CP in the Australian state of Victoria continue to have reduced survival compared with the general population. What is encouraging is the observed improvements in survival for those born in the 2000s. Although some of this progress is likely related to the improvements in management of individuals with more complex disability, our study demonstrated that the recent improvement in survival could be largely attributed to a reduction in severity profiles. Ongoing research is required to determine if these trends continue, to clarify if there is indeed a 'catchup' period, and to better understand these findings.

ACKNOWLEDGMENTS

We wish to thank our participating families and the Australian Institute of Health and Welfare. The Victorian Cerebral Palsy Register received funding from the Lorenzo and Pamela Galli Research Trust, the Victorian Department of Health and Human Services, the Royal Children's Hospital Foundation, and the Cerebral Palsy Alliance Research Foundation; infrastructure support was provided by the Victorian Government's Operational Infrastructure Support Program.

DATA AVAILABILITY STATEMENT

The data that support the findings of this study are available on request from the corresponding author. The data are not publicly available due to privacy or ethical restrictions.

ORCID

Gina L. Hinwood https://orcid.org/0000-0002-2618-4798
Dinah S. Reddihough https://orcid.org/0000-0003-3634-7906
Susan M. Reid https://orcid.org/0000-0001-9415-5246

REFERENCES

1. McIntyre S, Goldsmith S, Webb A, et al. Global prevalence of cerebral palsy: A systematic analysis. Dev Med Child Neurol 2022. https://onlinelibrary.wiley.com/doi/10.1111/dmcn.15346
2. ACPR group. Australian Cerebral Palsy Register bulletin, birth years 1995–2014. Sydney: Cerebral Palsy Alliance Research Institute, 2020.
3. Hollung S, Vik T, Lydersen S, Bakken I, Andersen G. Decreasing prevalence and severity of cerebral palsy in Norway among children born 1999 to 2010 concomitant with improvements in perinatal health. Eur J Paediatr Neurol 2018; 22:814–21.
4. Reid SM, Meehan E, McIntyre S, et al. Temporal trends in cerebral palsy by impairment severity and birth gestation. Dev Med Child Neurol 2016; 58:25–35.
5. Sellier E, Platt MJ, Andersen GL, et al. Decreasing prevalence in cerebral palsy: a multi-site European population-based study, 1980 to 2003. Dev Med Child Neurol 2016; 58:85–92.
6. Sigurdardottir S, Thorkelsson T, Vik T. Trends in prevalence and characteristics of cerebral palsy among Icelandic children born 1991 to 2010. Dev Med Child Neurol 2016; 58:45.
7. Brooks J, Shavelle R, Strauss D. Survival in children with severe cerebral palsy: A further international comparison. Dev Med Child Neurol 2012; 54:383–84.

8. Hemming K, Hutton JL, Colver A, Platt M-J. Regional variation in survival of people with cerebral palsy in the United Kingdom. Pediatrics 2005; 116:1383–90.
9. Shavelle RM, Straus DJ, Day SM. Comparison of survival in cerebral palsy between countries. Dev Med Child Neurol 2001; 43:574.
10. 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; 54:353–60.
11. Blair E, Watson L, Badawi N, Stanley FJ. Life expectancy among people with cerebral palsy in Western Australia. Dev Med Child Neurol 2001; 43:508–15.
12. Hutton JL, Colver AF, Mackie PC. Effect of severity of disability on survival in a north-east England cerebral palsy cohort. Arch Dis Child 2000; 83:468–74.
13. Hutton JL, Cooke T, Pharoah POD. Life expectancy in children with cerebral palsy. BMJ 1994; 309:431–5.
14. Hutton JL, Pharoah PO. Effects of cognitive, motor, and sensory disabilities on survival in cerebral palsy. Arch Dis Child 2002; 86:84–9.
15. Nielsen J, Uldall PV, Rasmussen S, Topp M. Life expectancy in children with cerebral palsy in Eastern Denmark. Ugeskr Laeger 2002; 164:5640–43.
16. Himmelmann K, Sundh V. Survival with cerebral palsy over five decades in western Sweden. Dev Med Child Neurol 2015; 57:762–7.
17. Strauss D, Shavelle R, Reynolds R, Rosenbloom L, Day S. Survival in cerebral palsy in the last 20 years: signs of improvement? Dev Med Child Neurol 2007; 49:86–92.
18. Brooks J, Strauss D, Shavelle R, et al. Recent trends in cerebral palsy survival. Part II: Individual survival prognosis. Dev Med Child Neurol 2014; 56:1065–71.
19. Blair E, Langdon K, McIntyre S, Lawrence D, Watson L. Survival and mortality in cerebral palsy: observations to the sixth decade from a data linkage study of a total population register and National Death Index. BMC Neurol 2019; 19:111.
20. Galea C, McIntyre S, Smithers-Sheedy H, et al. Cerebral palsy trends in Australia (1995–2009): a population-based observational study. Dev Med Child Neurol 2019; 61:186–93.
21. Rosenbaum P, Paneth N, Leviton A, Goldstein M, Bax M. A report: the definition and classification of cerebral palsy. Dev Med Child Neurol 2007; 49 Suppl:8–14.
22. Smithers-Sheedy H, Badawi N, Blair E, et al. What constitutes cerebral palsy in the twenty-first century? Dev Med Child Neurol 2014; 56:323–28.
23. Palisano R, Rosenbaum P, Walter S, et al. Development and reliability of a system to classify gross motor function in children with cerebral palsy. Dev Med Child Neurol 1997; 39:214–23.
24. Australian Bureau of Statistics. Dataset: Deaths, year of registration, age at death, age-specific death rates, sex, states, territories and Australia (Victoria), 2012 Available from: https://stat.data.abs.gov.au/Index.aspx?DataSetCode=DEATHS_AGESPECIFIC_REGISTRATIONYEAR.
25. Baird G, Allen E, Scrutton D, et al. Mortality from 1 to 16-18 years in bilateral cerebral palsy. Arch Dis Child 2011; 96:1077–81.
26. deLacy M, Reid SM, ACPR group. Profile of associated impairments at age five years in Australia by CP subtype and GMFCS level for birth years 1996–2005. Dev Med Child Neurol 2016; 58 Suppl 2:50–56.
27. Hemming K, Hutton JL, Bonellie S, Kurinczuk JJ. Intrauterine growth and survival in cerebral palsy. Arch Dis Child Fetal Neonatal Ed 2008; 93:F121-6.
28. Touyama M, Touyama J, Ochiai Y, Toyokawa S, Kobayashi Y. Long-term survival of children with cerebral palsy in Okinawa, Japan. Dev Med Child Neurol 2013; 55:459–63.
29. 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; 53:808–14.
30. Hemming K, Hutton JL, Pharoah POD. Long-term survival for a cohort of adults with cerebral palsy. Dev Med Child Neurol 2006; 48:90–5.
31. Doyle LW, Roberts G, Anderson PJ, Victorian Infant Collaborative Study Group. Outcomes at age 2 years of infants <28 weeks' gestational age born in Victoria in 2005. J Pediatr 2010; 156:49–53.
32. Strauss D, Brooks J, Rosenbloom L, Shavelle R. Life expectancy in cerebral palsy: an update. Dev Med Child Neurol 2008; 50:487–93.

How to cite this article: Hinwood GL, Loftus H, Shepherd DA, Guzys A, Reddihough DS, Reid SM. Survival of individuals with cerebral palsy in Victoria, Australia: A longitudinal cohort study spanning four decades. Dev Med Child Neurol. 2022;00:1–8. https://doi.org/10.1111/dmcn.15420