Outcome in cerebral palsy: life-expectancy

Jane L Hutton

Abstract

The life-expectancy of people with perinatally acquired cerebral palsy (CP) can be similar to that of the general population, or substantially reduced. The most important factors associated with reduced survival are disabilities of motor, cognitive or visual functions. Prematurity and low birth weight are associated with lower rates of disability and better survival. In contrast, those born light for gestational age have higher rates of severe disability than those of normal birth weight. Very severely disabled children who were born very heavy for gestational age have the worst survival. In recent years, survival has improved for children with severe disabilities and adults with gastrostomies, but not for the majority of people with CP. A 2 year old with severe CP has about a 40% chance of living to age 20, in contrast to a child with mild CP, for whom the chance is 99%. CP, respiratory diseases, epilepsy and congenital malformation are the most commonly recorded causes of early death.

Keywords birth weight; cerebral palsy; cognitive impairment; disability; gestational age; life-expectancy; psychomotor impairment; sensory impairment; survival

Introduction

Cerebral palsy (CP) has a prevalence of about 2 per 1000 live births, with considerable variation by birth weight, but a consistent excess of males. The rate of CP per 1000 neonatal survivors of normal birth weight is 1.1–1.4, but for very low birth weight infants (less than 1000 g) it is 57–78, with similar rates for gestational age. CP is more prevalent in infants small for gestational age than those slightly heavy for gestational age.

Several definitions of CP have been proposed. The crucial components of all the definitions are that the disability: is of the brain; produces disorders of movement or posture; occurs pre- or peri-natally; and is non-progressive; although non-progressive, the symptoms and signs may vary as the child matures.

Families of people with CP of perinatal origin usually wish to know how long their child is likely to live. Health and social care providers, whether insurance companies or state authorities, need information on life-expectancy in order to plan for the medical, educational and social needs of these children and their families. If medical liability is admitted, information on life-expectancy is an essential component in deciding the quantum of a settlement. In the last two decades, reliable estimates of life-expectancy have become available.

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Life-expectancy

The 'life-expectancy' is the arithmetic mean of all the lifetimes of the relevant population. Care is needed in reading government actuarial tables of life-expectancy, as they usually give the 'residual life-expectancy for age X', i.e. the average additional number of years that a person aged X will live. For example, an English woman aged 20 years has a life-expectancy 57.3 and is expected to live to age 77.3 years. In the UK, civil courts wish to know the age at which 50 of a notional cohort of 100 people like the plaintiff will have died, which is the median life. The median expected life for a 20-year-old English woman is 79.6 years (59.6 additional years).

Clinical experience alone is insufficient for reliable estimation of the probability of survival in CP. In order to obtain a good estimate of the survival experience of a population of people with CP, we need to have a representative group of such people, with information on factors which affect life-expectancy, and to know precisely when they died, or how old they were when last known to be alive. All syndromes that have progressive cerebral disability must be excluded from survival estimates, and early diagnoses should be verified as children grow. Some countries provide official statistical data on dates and causes of death, or dates of emigration, to researchers.

The most reliable source of information on survival is a precisely defined geographical cohort with known dates of birth, sex, birth weight, gestational age and severity of functional impairments, and reliable notification of fact and date of death. Registers or databases which focus on people who receive treatment or services related to CP can also be informative. However, it is essential to assess the completeness of registers. If cases are collected from a service which focuses on provision of education for those with cognitive deficits, reliable information on people with normal or good cognitive ability will not be available. A region with an excellent health service might attract people from other regions, so that the service disease register does not represent a geographical cohort.

In order to estimate the effects on survival of the severity of functional disability, each severity category must have a specific definition. The categories must be mutually exclusive. A wide range of survival estimates can be found by varying the combination and levels of disability. In any large clinical dataset, it is unusual for every variable to have a complete set of values. The reasons why some values are missing may severely bias a survival estimate. For example, when the intelligence quotient (IQ) is used as a measure of cognitive ability, a missing value for IQ could arise if the child had limited communication because of other severe impediments. Also, children who die young are more likely to have information missing. For the general UK CP cohort, the survival to 10 years for children with at least one disability not recorded is as poor as that of those with three severe disabilities.

Demographic factors

More males than females have CP, with the ratio varying between 1.2 and 1.6.[1–20] Females have slightly better overall survival than males: a combined estimate of UK CP survival from age 2 years

shows that 86% of women live to age 30 years, compared with 84% of men (Table 1). This difference is minimal relative to the effect of disability on life-expectancy. Survival from age 2 years is considered to allow for verification of early diagnoses.[6]

Survival is strongly associated with birth weight and gestational age, but not as might be expected. Low birth weight (less than 2500 g) and premature (less than 37 weeks) infants have better survival than term infants.[6,8–10] The UK CP 30-year survival from age 2 years is 92% for very low birth weight and premature infants, compared to 81% for term and normal weight births (Table 1). As low birth weight infants might be twins, or premature or growth restricted, results for birth weight standardized for gestational age, sex, parity and multiplicity (StBW) are given.[5] Life-expectancy is similar for children born with light (less than 10th centile), typical (10–90th centile) or heavy (more than 90th centile) for their StBW: the 30-year survival rates are 82%, 85% and 83%, respectively (Table 1). Part of this difference in survival arises from the higher proportions of severe disabilities among term or normal birth weight infants. For example, in two English cohorts (Mersey and North of England), 36% and 34% of normal birth weight infants had severe mental disability, compared to 26% and 23% of those weighing less than 1500 g at birth.[8,10] Forty per cent of infants light for gestational age (less than 3rd centile) had severe mental disability, compared with 22% of those between the 26th and 75th centiles.

Two databases permit long-term follow-up of people with CP.[7,14,16] Excess risks of death compared to the general population generally decreased with age.[17] For UK women, but not men, an increase in relative risk of death after age 50 years was found.[7] The Californian study reported a decline in ambulatory function over the age of 60 years, along with a poorer life-expectancy for those individuals who had lost mobility.[14]

UK 10-, 20-, 30- and 40-year survival estimates

Characteristic		% living from age 2 to:			
		10	20	30	40
Sex	Female	96	89	86	83
	Male	95	88	84	79
Birth weight (g)	<1500	97	94	92	92
	1500–2499	96	91	87	82
	≥2500	94	87	82	78
Gestational age (weeks)	<32	97	94	91	91
	32–36	97	92	88	86
	≥37	94	87	82	76
Standardized birth weight (centiles)	<10th	95	87	82	78
	10–90th	95	90	85	82
	<90th	95	88	83	77
Severe disabilities	None severe	100	99	98	97
	Only Ment	98	95	93	92
	Amb or Mand	97	93	89	87
	Amb, Mand, Ment	86	60	53	46
	Amb, Mand, Ment, Vis	70	41	31	26

Amb, ambulation; Mand, manual dexterity; Ment, mental ability; Vis, vision

Table 1

Disabilities

Life-expectancy is significantly poorer in those with severe disability.[1,2,4,6–10,16,18,20] The clinical type of CP is associated with survival, in that quadriplegia is associated with a higher risk of death,[9] but the particular impairments are more relevant to survival.

In the UK and Europe, common definitions of severe functional disabilities are used.[4,6,19,20] Severe manual disability is defined as the child being unable to feed or dress themselves; the Californian definition is similar.[16,18] A severe intellectual disability is defined as an IQ less than 50. Severe ambulatory disability indicates that the child is unable to walk even with aids, and requires a wheelchair. A severe visual impairment is defined as vision in the better eye less than 6/60. A severe hearing impairment is defined as a hearing loss in the better ear more than 70 dB averaged across frequencies 0.5–4 kHz.

Estimates for a general UK CP cohort, for children who lived to at least age 2 years, with each disability considered separately, show that 95–99% of children with at most moderate disability live to adulthood (age 20 years; Figure 1). Two-thirds of those who require a wheelchair, or have severe intellectual disability, live to about age 20 years. Survival to age 20 is slightly lower for those with severe manual disability (59%) and severe visual disability (55%).

Californian Mental Retardation Database records whether, when lying, a person can lift their head or roll, has a record of epilepsy, and if tube feeding (gastrostomy) is used.[16,18] The less mobility a person has, the poorer is their life-expectancy. A child who cannot lift their head when lying on their stomach has a mortality rate 14 times higher than a child who can roll freely.[18] A record of epilepsy and tube feeding are associated with increased risks of death. The frequency and type of epileptic seizures and the level of control of seizures are usually not known, so the effect on survival cannot be accurately assessed.

The Californian database has relatively high levels of severe ambulatory impairments (73% in children, 35% in adults),[16,18] compared with 26–36% in young people in the UK,[8] Canada[2] and Australia,[8] and 17% in a UK adult study.[7] Canada and Australia record slightly lower rates of severe cognitive disability (16% and 24%, respectively) than the other countries (21–44%). Manual disability is severe in 20–25% across these countries. Blindness is recorded in 6–11% of children. Although the prevalence of severe disabilities varied between the five UK regions, there were no differences in survival experience once disabilities were taken into account. Children whose ambulation is severely impaired are likely to have severe manual disabilities (72%) in contrast to less than 5% of those who have some mobility.

Combined effects of risk factors

As children with CP can have a wide range of levels of disabilities, it is sensible to consider the combined effects of multiple disabilities. Statistical models which allow the joint effects of several variables to be assessed show that severe cognitive, motor (manual and ambulatory) and visual disabilities all jointly