# Long-term survival for a cohort of adults with cerebral palsy

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**The aim of this study was to investigate long-term survival and examine causes of death in adult patients with cerebral palsy (CP). A 1940–1950 birth cohort based on paediatric case referral allows for long-term survival follow-up. Survival is analyzed by birth characteristics and severity of disability from age 20 years (and age 2y for a subset of the data). Survival outcome compared with that expected in the general population based on English life tables. The main cohort consisted of 341 individuals, with 193 males and 148 females. Conditional on surviving to age 20 years, almost 85% of the cohort survived to age 50 years (a comparable estimate for the general population is 96%). Very few deaths were attributed to CP for those people dying over 20 years of age. Females survived better than males. However, females faced a greater increase in risk relative to the general population than did males. We conclude that survival outlook is good though lower than in the general population. The relative risk of death compared with the UK population decreases with age, although it shows some indication of rising again after age 50 years. Many more deaths were caused by diseases of the respiratory system among those dying in their 20s and 30s than would be expected in the general population. Many fewer deaths than expected in this age group are caused by injuries and accidents. For those people who die in their 40s and 50s, an increase in deaths due to diseases of the circulatory system and neoplasms is observed. More deaths than expected in this age group are due to diseases of the nervous system.**

The survival patterns of people with cerebral palsy (CP) have been described previously, usually considering the effect of the severity of physical, cognitive, and sensory disabilities (Evans et al. 1985; Hutton et al. 1994, 2000; Hutton and Pharoah 2002). It is has been widely reported that those with less severe disabilities, and even some children with severe disabilities, may survive well into adulthood (Evans et al. 1985, Blair et al. 2001). Indeed, over 80% of infants with early impairment CP will survive beyond their 30th birthday, rising to over 95% for those without learning disability* (Hutton and Pharoah 2002). However, most studies, principally those based in the UK, have been limited by short follow-up times and unable to consider long-term survival and, therefore, focus on childhood and early adulthood survival patterns. One study from the USA that has focused on adult survival (Strauss and Shavelle 1998a) found that individuals, aged 15 years and over, lacking key functional skills had additional life expectancies of 11 years, whereas those high-functioning adults had life expectancies close to the general population. Further work found that excess mortality risks compared with the general population decreased with increasing age (Strauss and Shavelle 1998b).

Furthermore, for adults over 60 years a decline in motor function along with poorer survival was observed (Strauss et al. 2004). Long-term survival outcomes would enable appropriate planning in terms of funding and services, would be of value to patients and families, and might help in structuring medicolegal settlements.

We evaluated long-term survival patterns based on a cohort of individuals with CP followed for up to 60 years and made comparisons with the general population. The way in which the data were collected restricted the amount of information available on childhood survival, and our main focus was on survival in adulthood (after age 20y).

We also considered, to a lesser extent, survival patterns from childhood. Changes in patterns of underlying causes of death, by age at death, were also examined and again compared with that expected in the general population.

## Method

Children and young adults with CP in the Bristol area of the UK in the 1950s were identified from the records of a paediatrician (Woods 1957, 1963). From 1951 to 1964, information on all those attending a special clinic for children with CP at the Bristol Children's Hospital was recorded on punchcards. The patients comprised those referred by paediatricians, public health and school workers, and others. Although it is not possible to verify, all children with CP from Bristol and its surrounds were reported to have been included in these records: 'Since 1951, I have recorded all the cases under my care on professionally designed punchcards, which I filled in immediately after seeing the child involved … In this way, we saw all the cerebral palsied children in Bristol, and many from the Somerset and Gloucestershire areas' (Woods 1994).

The cohort consisted of children who were born over four decades (1930s–1960s).

### IDENTIFICATION OF THE CP COHORT

Patient information on punchcards was transcribed to a computer database. In assigning a patient as having CP we were dependent on the information given on the punchcard. We

*US usage: mental retardation.

use the definition of CP as used by the Surveillance of Cerebral Palsy in Europe (2000) collaboration. Inevitably, some patients were not included because inadequate data did not allow for identification of CP, and few cases may have been included that did not fully meet this formal definition of CP. Only patients labelled as having quadriplegia, paraplegia (at the time the term paraplegia was consonant with diplegia as currently used), monoplegia, athetosis, mixed spasticity, and ataxia were assumed to meet the criteria of having CP. Any patients so labelled as having paraplegia but where there was an indication of a spinal injury were excluded, as were any other patients where there was an indication that the paralysis was not of cerebral origin. Several records specified that the CP was a result of a cerebral insult in infancy or childhood (e.g. meningitis, encephalitis). If there was mention of a postnatal insult, after age 28 days, the child was classified as having late impairment CP and excluded from subsequent analysis. All others were assumed to be early impairment CP. The patients so diagnosed were included, provided subsequent notes did not indicate a progressive disorder. Some patients were included even though data describing the severity of their functional disabilities were missing. As a comparator, we also considered a cohort of CP cases from the Mersey Cerebral Palsy register (Hutton and Pharoah, 2002) born between 1966 and 1969 (the closest available birth years to this Bristol cohort).

Ethical permission was obtained from the Local Research Ethics Committee to have the cases flagged at the National Health Service Central Register (NHSCR) of the Office for National Statistics. The UK system of death registration, in which by law all deaths must be registered, allows studies such as this to trace facts and causes of death with NHSCR. Using a flagging system, NHSCR provides notification of dates of death for all people in the study, whether residing in the Bristol region, or elsewhere in the UK. Dates of emigration are given when individuals leave the UK permanently. Individuals are traced on the system through a variety of sources, including name, date of birth, and NHS number. Formal name changes, for instance due to marriage, are accommodated within the system (although adoptions would not be unless the adopted name were known). This ensures that the fact of death or immigration is recorded for virtually all people in this study.

### IDENTIFICATION OF REPRESENTATIVE COHORTS

The original survey was performed between 1951 and 1964 (inclusive). Allowing for a notification lag of up to 5 years, we assumed completeness up to the birth cohort year 1959. The survey identified both prevalent and incident cases, as referral cases consisted of both young infants and older children. A few individuals (29) were identified from the 1930s birth cohort, but are not considered here due to a clear under-ascertainment. Our analysis, therefore, focuses on two decades of birth, the 1940s and the 1950s. Due to the retrospective nature of the data collection, the cohort is not representative of childhood survival patterns, as the severest cases are likely to have died before ascertainment. Two representative subcohorts are, therefore, selected. The first of these representative subcohorts is referred to as the adult subcohort. This subset includes only those individuals who survived into adulthood (here defined as age 20 years), as analysis showed all participants to be ascertained by their 20th birthday. The second representative subcohort is referred to as the incident cohort. This subset, which may be used to predict childhood survival patterns, includes only those birth years that were ascertained prospectively. For this we consider the period 1954–1959, conditional on survival to age 2 years and similarly allowing for a 5-year notification lag.

### AVAILABLE COVARIATES

Gestational age, birthweight, sex, date first assessed, and year of birth was recorded. The level of cognitive disability was measured by IQ, grouped according to severe disability (IQ$<$50) and not severe disability (IQ$\geq$50). It is not known what test procedure was used to determine the IQ. The records had a statement of the child's ambulatory and manual ability. We classified children based on whether their disability was severe or not. For ambulation, a child was considered severely disabled if dependent on a wheelchair. A child was classified as having a severe manual disability if unable to self-feed or dress. In addition, the punchcards also recorded whether visual disability was present. We classified a child as having a severe visual disability if the child was registered as blind or attended a school for the partially sighted. Severity of disability was classified as unknown where it was not possible to determine the level of disability from the punchcard records. Similar classifications were used for the Mersey cohort (Hutton and Pharoah 2002). For infants born in the 1950s, these severity levels referred to disability as assessed in infancy. For infants born in the 1940s the assessment referred to disabilities at an older age.

### STATISTICAL ANALYSIS

Lifespans from birth until death were calculated for all individuals who were notified as having died. For those individuals still alive, lifespans were defined as times from birth until the censoring date, March 2004, which allowed for a 6-month notification lag for deaths. Kaplan–Meier survival estimates, conditional on survival to age 20 years, were computed (using the adult subcohorts) at ages 30, 40, 50, and 60 years (where follow-up times allowed). Differences between groups were evaluated using the log-rank test (Cox and Oakes 1984). Survival estimates by birth characteristics and level of intellectual disability are presented (for other disabilities the amount of missing data suggest very little information for the severely disabled). Some comparative survival estimates are presented for the Mersey cohort. Estimates of length of life, from age 2 years, are provided for the incident cohort and stratifications are provided by some birth characteristics (restrictions here are due to small numbers).

Expected survival proportions for the UK population, matched to the Bristol CP adult subcohort by sex, decade of birth, and conditional on being alive at age 20 years, allowed comparisons to the general population. Expected survival estimates were computed using the relative survival package, *surv2* (http://www.cancerregistry.fi/surv2/), and using English life tables (source: Government Actuary's Department, English life tables). Relative risks of death to the UK population allow evaluations of changes in risk of death due to CP with increasing age. Point estimates (available at yearly intervals) were smoothed in Splus using a spline smoother.

Underlying causes of death are classified according to the 17 main chapters of the International Classification of Diseases (ICD), 9th revision. The 10 deaths classified by the ICD 10th revision are also classified according to these broad groupings. Frequencies of cases where CP (ICD 9th revision code

343-) was mentioned anywhere on the death certificate are also computed. Proportions expected in each of the categories for the UK general population (year of death 2001) are also given for comparison (Office for National Statistics 2001).

We present results for both the Bristol and Mersey cohort in the following section, but defer discussion of the comparative Mersey data to the final section.

## Results

The CP cohort consists of 506 individuals with early impairment CP born between 1932 and 1969, of whom 471 (93%) were successfully flagged with NHSCR. An initial summary of the 471 cases of CP flagged with NHSCR is presented in Table I.

Age at first assessment was missing for a small proportion of the patients (21 out of 471). All patients from the 1930s cohort were seen during their teenage years, whereas the majority of the 1950s cohort were seen for the first time before their fifth birthday. By assuming completeness up to 1959 and excluding births before 1940, the dataset is reduced to 388 individuals (110 deaths).

The adult subcohort of 341 individuals (66 deaths), excludes all individuals who died before their 20th birthday (10 from the 1940s and 37 from the 1950s). For the incident cohort, we include those born between 1954 and 1959, excluding deaths before age 2 years. The sample size for the Bristol incident cohort is 106 (27 deaths).

The Mersey cohort consists of 245 adults (39 deaths), of whom two individuals could not be traced. When considering only people who attained adulthood, the sample size for the Mersey cohort reduced to 214 (12 deaths). The Mersey incident cohort consisted of 243 individuals (39 deaths).

### SUMMARY STATISTICS

Basic summary statistics for the subcohort of adult survivors, and the incident cohorts, are presented in Table II. Male:female ratios vary slightly between the two decades, from males consisting of 62% of the sample in the 1940s to an equal distribution in the 1950s. The proportions in the very-low-birthweight category (<1500g) were similar between the decades, at about 12%. The 1940s had a higher proportion of infants falling into the normal birthweight category (73%). More infants were also born at term in the 1940s (71% compared with 64% in the 1950s). IQ was missing for almost 40% of participants in the 1950s and 7% in the 1940s. For those participants where IQ was known, about 20% of them had severe cognitive disability during the 1940s, whereas just 7% did in the 1950s. Proportions of participants with a recorded severe manual, ambulatory, or visual disability were very low (less than 5% for the 1950s), despite proportions of missing data for these variables being much less than that for IQ.

### SURVIVAL ESTIMATES

Survival estimates conditional on being alive at age 20 years are presented in Table III. Expected survival rates for the general population are consistently higher than those observed in the CP cohorts. Survival outcome was similar between the two birth cohorts. Male survival was worse than female, for example 81 versus 89% at 50 years ($p=0.05$). Interestingly, it is only in the 1940s cohort that this difference between male and female survival existed: in the 1950s survival of the males and females was almost identical. Infants born at term have a poorer survival outlook than those born preterm, for example 83 versus 89% at age 50 years ($p=0.23$). This relation is not as consistent between infants with normal and low birthweight, but is likely to be affected by small numbers of deaths

**Table I: Cases of early impairment CP ($n$=471) by age at first assessment and decade of birth**

| *Decade of birth* | *Age first assessed (y)* | | | | |
|---|---|---|---|---|---|
| | *<6* | *6–10* | *11–15* | *16–20* | *Unknown* |
| 1930s ($n$=31) | – | – | 14 | 14 | 3 |
| 1940s ($n$=174) | 59 | 78 | 30 | 1 | 6 |
| 1950s ($n$=214) | 179 | 25 | 2 | 1 | 7 |
| 1960s ($n$=52) | 45 | 2 | – | – | 5 |

Includes all years of birth, but only those participants who were successfully flagged with the National Health Service Central Register (NHSCR).

**Table II: Basic summary statistics for adult subcohorts and incident cohorts, $n$ (%)**

| | *Adult subcohorts* | | | *Incident cohorts* | |
|---|---|---|---|---|---|
| | *1940s* | *1950s* | *MC* | *1954–1959* | *MC* |
| Numbers | 164 | 177 | 214 | 106 | 243 |
| Deaths | 41 | 25 | 12 | 27 | 39 |
| Sex | | | | | |
| Male | 98 (62) | 93 (50) | 115 (54) | 48 (45) | 131 (54) |
| Female | 66 (38) | 84 (50) | 99 (46) | 58 (55) | 112 (46) |
| Birthweight, g | | | | | |
| Mean | 2810 | 2600 | 2780 | 2600 | 2873 |
| Missing | 9 | 3 | 0 | 3 | 2 |
| <1500 | 17 (11) | 23 (13) | 18 (8) | 12 (12) | 18 (8) |
| 1500–2499 | 25 (16) | 49 (28) | 49 (23) | 30 (29) | 21 (21) |
| ≥2500 | 113 (73) | 102 (59) | 147 (69) | 61 (59) | 172 (71) |
| Gestational age, wks | | | | | |
| Mean | 36 | 37 | 38 | 37 | 38 |
| Missing | 14 | 8 | 3 | 7 | 4 |
| <32 | 14 (9) | 23 (14) | 12 (6) | 13 (13) | 12 (5) |
| 32–36 | 30 (20) | 37 (22) | 40 (19) | 21 (21) | 40 (17) |
| ≥37 | 106 (71) | 109 (64) | 159 (75) | 65 (66) | 187 (78) |
| IQ level | | | | | |
| Missing | 12 | 66 | 0 | 58 | 5 |
| <50 | 34 (22) | 9 (7) | 60 (28) | 5 (10) | 81 (34) |
| ≥50 | 118 (78) | 102 (93) | 154 (72) | 43 (90) | 157 (66) |
| Manual dexterity | | | | | |
| Missing | 15 | 39 | 2 | 34 | 4 |
| Severe | 5 (3) | 2 (1) | 29 (14) | 1 (1) | 52 (22) |
| Not severe | 144(97) | 136(99) | 183(86) | 71(99) | 187(78) |
| Ambulation | | | | | |
| Missing | 13 | 37 | 0 | 33 | 2 |
| Severe | 10 (7) | 4 (3) | 48 (22) | 3 (4) | 74 (31) |
| Not severe | 141 (93) | 136 (97) | 166 (78) | 70 (96) | 167 (69) |
| Vision | | | | | |
| Missing | 10 | 19 | 33 | 19 | 56 |
| Severe | 5 (3) | 6 (4) | 8 (4) | 5 (6) | 9 (5) |
| Not severe | 149 (97) | 152 (96) | 173 (96) | 82 (94) | 178 (95) |

MC, Mersey cohort. Adult subcohorts are conditional on survival to age 20 years; incident cohorts conditional on survival to age 2 years. Percentages, with reference only to cases for which level of covariate is known (i.e. excluding missing cases), are given in parentheses.

and individuals. The largest difference between the covariate groups exists between those with severe cognitive disability compared with those who do not have a severe cognitive disability, for example 60 versus 91% respectively at age 50 years ($p<0.01$).

For the birth period 1954–1959, where it is possible to make survival predictions from childhood, 78% were observed to survive until their 30th birthday (Table IV).

**Table III: Estimated survival percentages, conditional on being alive at age 20 years, for the Bristol cohort**

| | *30y* | *40y* | *50y* | *60y* |
|---|---|---|---|---|
| Observed and expected | | | | |
| Bristol | 94 (91–97) | 91 (88–94) | 84 (80–88) | 70 (61–79) |
| Mersey | 98 (96–100) | 93 (89–97) | – | – |
| English life table | 99 | 98 | 96 | 90 |
| Decade | | | | |
| 1940s | 95 (91–98) | 91 (86–96) | 84 (78–90) | 70 (60–80) |
| 1950s | 94 (90–98) | 91 (87–95) | 85 (79–91) | – |
| Sex | | | | |
| Male | 93 (89–97) | 89 (85–94) | 81 (75–87) | 70 (60–80) |
| Female | 96 (93–99) | 93 (89–97) | 89 (84–94) | 69 (49–88) |
| Birthweight, g | | | | |
| <1500 | 98 (93–100) | 98 (93–100) | 85 (73–97) | – |
| 1500–2499 | 92 (86–98) | 90 (83–97) | 84 (75–93) | 69 (50–88) |
| ≥2500 | 94 (91–97) | 89 (85–93) | 84 (79–89) | 68 (57–79) |
| Gestational age, wks | | | | |
| <32 | 92 (83–100) | 92 (83–100) | 78 (63–93) | – |
| 32–36 | 97 (93–100) | 96 (91–100) | 89 (81–97) | 78 (64–92) |
| ≥37 | 94 (91–97) | 89 (85–93) | 83 (78–88) | 67 (56–78) |
| IQ level | | | | |
| Missing | 90 (83–97) | 87 (80–94) | 76 (65–87) | 42 (6–78) |
| <50 | 79 (67–91) | 67 (53–81) | 60 (45–75) | 56 (40–72) |
| ≥50 | 90 (97–100) | 98 (96–100) | 91 (87–95) | 78 (67–89) |

English life table refers to comparable expected survival in general population. Participants who died before age 20 years are excluded from calculations. 95% confidence intervals are given in parentheses.

### RELATIVE RISKS OF DEATH COMPARED WITH THE GENERAL POPULATION

At each given age in years (from 20 to 60 years, where a death occurred) we can estimate the increase in risk of death at that particular age for the cohort of adults with CP compared with the general population (matched by year of birth and sex) using relative risks of death. The relative risk of death for the CP cohort compared with the general population (Fig. 1) was higher for women than men. At age 25 years the Bristol CP adult subcohort had an increased risk of death over 10 times that of the general population. For both men and women this increased relative risk decreased with age although, at all ages, they faced an increased risk relative to the general population. For the female cohort there was some indication that the relative risk increased again after age 50 years.

For those patients known not to have a severe ambulatory, manual, cognitive, or visual disability (196 individuals, 24 deaths), relative risks of death compared with the general population (no stratification by sex because of the small number of deaths) were also above one.

### CAUSE OF DEATH

Of the 66 participants flagged with NHSCR who had died, death certificates were available for all but one individual (where the date of death was known, but the death draft could not be traced). Mention of the condition 'infantile cerebral palsy' (ICD 9th revision code 343-) occurred for nine individuals (14%). Of those individuals dying during their twenties and thirties the most common underlying causes of death were diseases of the respiratory system (Table V). For those dying in their forties and fifties, few deaths were attributable to diseases of the respiratory system; rather, the most common underlying causes of death were diseases of the circulatory system, neoplasms, and diseases of the digestive and nervous systems. Six deaths were attributable to accidental causes.

## Discussion

Patients with the severest forms of CP face the worst prognosis, and many will die before they attain adulthood. However, conditional on surviving to age 20 years, the survival outlook

**Table IV: Estimated survival proportions, from age 2 years, for period of births 1954–1959**

| | *5y* | *10y* | *20y* | *30y* | *40y* | *50y* |
|---|---|---|---|---|---|---|
| All participants | 97 (94–100) | 91 (86–96) | 83 (76–90) | 78 (70–86) | 76 (68–84) | 74 (66–82) |
| Males | 95 (89–100) | 85 (76–94) | 78 (67–89) | 74 (63–85) | 72 (60–83) | – – |
| Females | 100 (100–100) | 98 (93–100) | 90 (81–99) | 83 (72–93) | 79 (67–91) | 76 (64–88) |
| Term delivery | 95 (84–98) | 91 (74–92) | 83 (69–90) | 79 (65–85) | 75 (62–84) | 73 – |
| Normal birthweight | 95 (90–100) | 89 (81–97) | 79 (69–89) | 74 (63–85) | 72 (61–83) | 70 (59–81) |
| Mersey cohort | 98 (96–100) | 95 (92–98) | 90 (86–94) | 88 (84–92) | 84 (79–89) | – – |

Participants who died before age 2 years are excluded from these calculations. 95% confidence intervals are given in parentheses.

reported here is good. Survival rates for a cohort of children born today with CP may not necessarily be extrapolated from this study, but it is likely that, conditional on levels of impairment, they will have a better outlook than children born 50 or more years ago. The survival outlook is associated with the severity of intellectual disability. Even those with no severe disabilities have a higher risk of death than the general population. Other factors affecting outlook are birthweight, gestational age, and sex.

Deaths due to diseases of the respiratory system are much more prevalent than in the general population, especially for those who die before age 40 years, whereas deaths due to injuries and accidents occur much less often than would be expected for this age group in the general population. This may explain why men had a smaller relative risk of death compared with the general population than their female counterparts. Higher proportions of deaths in adults over 30 years of age result from cancers and diseases of the circulatory system. Deaths due to diseases of the digestive system are more prevalent than would be expected in the general population for those over 40 years, as are deaths due to diseases of the nervous system. Deaths due to CP (both as the underlying cause and CP mentioned anywhere on the death certificate) occur less often than reported elsewhere (Evans et al. 1990, Maudsley et al. 1999, Hutton et al. 2000), although this would be consistent with our observed finding of an increase in the number of deaths due to circulatory problems and cancer with increasing age.

Only one other cohort that we are aware of (Strauss and Shavelle 1998a) allowed such long-term follow-up of people with CP. General findings here are consistent with the Californian cohort, where it was also found that excess risks of death over the general population decreased with age (Strauss and Shavelle 1998b). The Californian study did not, however, report any increase in relative risk of death after age 50 years for the female cohort, as we found here. It is while people with disabilities are in their adulthood that their carers face general increases in morbidity and mortality. This may pose problems for the care of the adult disabled, and these changes in social circumstances may even cause increases in risk of death. No such increase was observed for the males, and in addition males were observed to have a lower relative risk than their female counterparts. This would be consistent with the hypothesis that elderly males with medical problems do better than elderly females with similar medical problems, due to female partners who act as carers.

Interestingly, the Californian study has reported a decline in ambulatory function for persons over the age of 60 years along with a poorer life expectancy for those individuals who had lost mobility (Strauss et al. 2004). Our finding of fewer deaths due to injuries and accidents than would be expected in the general population is in contrast to that of the Californian study (Strauss et al. 1999).

No difference between male and female survival outcome has been reported from studies of childhood CP survival. Both the Californian adult survival cohort (Strauss and Shavelle 1998b) and results here have found significant differences between male and female adult survival outcomes. The underlying mortality excess risk for males over females in the general population, peaks over the age range 20–40 years (much of which is attributable to accidental causes of death). This difference between males and females in the general population over this age range may help to explain observed differences between male and female CP survival rates. Furthermore, differences in life expectancies between males and females have decreased over recent decades, and so are likely to be more noticeable in a 1950s cohort than a 1990s cohort.

### LIMITATIONS OF THE STUDY

The particular nature of this study, with regard to its retrospective ascertainment and basis on case referral, may have implications for bias in various directions. Results must be interpreted in light of possible ascertainment biases. Clearly the limitations of this study show the value of investment in well-defined cohort studies, the full implications of which will not be realized for decades to come.

The retrospective nature of the data collection is perhaps our most serious concern. All participants in the 1930s cohort were assessed for the first time before their 20th birthdays. Those born later were assessed for the first time before their 16th birthdays, with two exceptions. However, this does not ensure complete ascertainment by 20 years of age: the upper age limit for paediatric referrals might be 16 years. The clear

**Table V: Percentages of cases by underlying cause of death and age at death**

| *Classification group* | *Age at death, y* | | | |
|---|---|---|---|---|
| | *20–29* | *30–39* | *40–49* | *50–59* |
| Neoplasms, II | 11 (10) | 0 (16) | 24 (27) | 21 (40) |
| Nervous system, VI | 0 (7) | 0 (5) | 14 (4) | 21 (3) |
| Circulatory system, VII | 6 (5) | 17 (9) | 19 (19) | 21 (27) |
| Respiratory system, VIII | 50 (3) | 42 (3) | 10 (4) | 0 (5) |
| Digestive system, IX | 11 (2) | 8 (5) | 19 (10) | 21 (10) |
| Accidental causes, VII E800–E985 | 11 (56) | 8 (44) | 5 (23) | 14 (9) |
| Others | 11 (14) | 25 (16) | 9 (12) | 2 (5) |
| Number of deaths | 18 | 12 | 21 | 14 |

The expected percentages for each group, based on UK 2001 deaths, are given in parentheses.

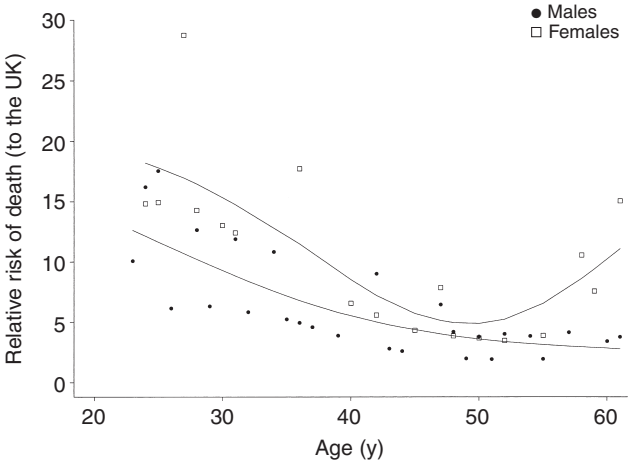


**Figure 1:** *Relative risk of death for CP cohort compared with general population.*

under-ascertainment for the 1930s cohort may well indicate ascertainment problems for other birth cohort years, such as the earlier part of the 1940s.

Limited information made it impossible to extract a geographically defined cohort, and it is expected that the children with severe disabilities out-of-region were more likely to have been referred than those with minimal disabilities. Indeed, on a subset of the cohort it has been reported that about 40% of the children referred were born outside of the city of Bristol, and that of these two in five were quadriplegic, as opposed to one in five of those born within the city of Bristol (Woods 1957).

The actual proportions of missing information on severity are not high. However, severe impairment is recorded for a lower proportion of individuals than in other cohorts. The less severely impaired may not feel it necessary to seek a referral during their later childhood years. During the earlier part of the 20th century, it was also common for children with severe disabilities to be placed in residential institutions and, as such, may not have come under the care of the Bristol centre. Furthermore, much of the data on the punchcards pertain to the initial consultation only. Those individuals born during the 1950s onwards were often seen as very young infants and, as such, it may not have been possible to make an informed estimate as to the level of severity. Those born in the earlier decades have an assessment date at an older age, where degrees of disability would have been easier to assess. Level of impairment cannot, therefore, be assumed to be missing at random. Less broad spectrums for the severely impaired are of interest, for example distinguishing those who cannot roll over or lift their head from those who cannot walk, but were not available from the case notes.

### COMPARISONS WITH OTHER COHORTS

Most of the survival rates reported here are conditional on having been alive at age 20 years, and are, therefore, not comparable to other published UK results. For this reason, the Mersey cohort was also included here as a comparison. For both the adult subcohorts, and the survival from childhood, survival up to age 40 years is better for the Mersey cohort than the Bristol cohorts. This is consistent with the potential over-ascertainment of participants with severe disabilities in the Bristol cohorts, due to delays in ascertainment and partial identification of the out-of-region cases.

Alternatively this might reflect poorer social and clinical management, or be the result of secular changes in proportions of children with severe disabilities. A smaller proportion of infants from the Bristol cohort fell into the normal birthweight and at term groups than the Mersey cohort, and fewer infants had a recorded severe cognitive disability. The relatively small sample sizes may explain these observed differences, or they may again be due to secular changes. Those children born preterm or with a low birthweight were also found to have a better survival than those born at term or with a normal birthweight, although these differences were not significant. Although seemingly counter-intuitive, this is consistent with previous findings for the Mersey cohort (Hutton and Pharoah 2002).

### PATTERNS OF CARE IN BRISTOL

Quality of care might affect outcomes and survival. Generalizability of these results is, therefore, dependent on the nature and generalizability of care in the Bristol region during the period of this study. In 1941 Dr Grace Woods set up a clinic in Bristol for infants with some abnormality. Although before the advent of the National Health Service in 1948, all mothers were free to bring their children to this clinic. From 1951 onwards a special clinic for children with CP was established at the Bristol Children's Hospital. In 1952 the Claremont School for children with CP was established in Bristol. At the same time, the Spastics Society (SCOPE) opened residential schools for children with CP or other special needs. In 1959 the Bristol work centre opened when the first young adults were leaving the Claremont School. Infants with CP and their families living in the Bristol area may have received care above that of other areas during these periods of time.

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