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Life expectancy for children with cerebral palsy and mental retardation: Implications for life care planning

Richard T. Katz*
Department of Physical Medicine and Rehabilitation, Washington University School of Medicine, St. Louis, MO, USA

Abstract. *Objectives:* Physicians may be asked by attorneys or other patient advocates to help plan for the long-term needs of children with cerebral palsy (CP) and developmental disability (DD). The first step in such planning is to thoroughly examine the literature dealing with life expectancy in these populations. This review paper comprehensively reviews the literature relating to survival in children with cerebral palsy and developmental disability.
Study Selection: A Medline data search was completed using the terms cerebral palsy, life expectancy, survival, as well as other pertinent terms. Further articles were gleaned from bibliographies of pertinent literature.
Data Synthesis: Certain key disabilities can be used to accurately predict life expectancy in children with cerebral palsy and mental retardation. These include: (1) presence and severity of mental retardation, (2) inability to speak intelligible words, (3) inability to recognize voices, (4) inability to interact with peers, (4) severity of physical disability, (5) use of tube feeding, (6) incontinence, and (7) presence and severity of seizures.
Conclusions: Literature review definitively shows that children with CP and DD have a diminished life expectancy, which can be assessed based on simple clinical examination findings.

Keywords: Life care planning, cerebral palsy, life expectancy

1. Introduction

A recent consensus group [33] defined cerebral palsy (CP) as an "umbrella term covering a group of non-progressive, but often changing, motor impairment syndromes secondary to lesions or anomalies of the brain arising in the early stages of its development." Recent studies suggest that CP is a rather common cause of childhood disability, with significant associated disability. According to Newacheck and Taylor [35], greater than 100,000 Americans less than 18 years of age are believed to have some neurologic disability attributed to CP. Approximately 25% of CP children in France and England cannot walk, and 30% are classified as mentally retarded (MR) [16,43]. A simple examination of these statistics suggests that health care practitioners and the social community need to plan for the survival and care of these children [16].

Physicians and others may be asked by attorneys or other patient advocates to help plan for the long-term needs of patients with prenatal or perinatal brain injury. Often for litigation purposes, attorneys or insurance companies want a detailed outline of future expenses with which to negotiate. The initial step in formulating these costs [5,9,13] is to estimate the life expectancy of the child, which has been a source of considerable debate-this topic will be examined extensively in this review. Tables are provided in this paper, from which the physician can glean information relating to mean life expectancy for a variety of groups. This information can be used to form a medical opinion as to life

*Address for correspondence: Richard T. Katz, MD, 4660 Maryland Ave., Suite 250, St. Louis, MO 63108, USA. Tel.: +1 314 361 7384; Fax: +1 314 361 3383; E-mail: pianodoctor@pol.net.

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expectancy that a life care planner can use in planning for future needs.

2. Estimating life expectancy in children with cerebral palsy

There is a considerable amount of epidemiological literature to help life care planners estimate life expectancy-it is surprising that such literature is often left unexplored by parties in negotiation. Children with CP are often grouped into four categories – spastic (approximately 70%), athetoid (approximately 20%), ataxic (approximately 10%), and mixed. Spastic syndromes are most common, and are characterized by muscular hypertonicity and loss of motor control. Spastic syndromes may affect predominantly one side (hemiplegia), both legs (paraplegia), legs greater than arms (diplegia), or all four limbs (quadriplegia or tetraplegia). Athetoid or dyskinetic syndromes are characterized by slow writhing, involuntary movements, and sometimes abrupt, distal, jerky movements. Ataxic syndromes are uncommon ($\sim$10%) and may be marked by weakness, incoordination, wide-based gait, and tremor. Many patients have mixed features.

Through the Mayo Clinic patient data bank, Kudrjavcev et al. [31] identified 60 cases of CP between 1950 and 1976. They stratified CP patients into four groups: (1) mild-functions without marked difficulty and without mechanical aids; (2) moderate-functions with marked difficulty but without mechanical aids; (3) severe-functions only with the assistance of mechanical aids; and (4) very severe–does not function even with the assistance of mechanical aids. Seventy-three percent were spastic, 16% ataxic, and 6% were dyskinetic CP. Survival was calculated for the first 10 years of life. Intelligence was characterized as normal (IQ $>$ 80), mild to moderate MR (IQ 36–80) and severe/profound MR (IQ $<$ 35). For children with severe or profound MR, survival was 68% at 5 years and 54% at 10 years. A life table analysis showed 68% survival at 5 years for normal/mildMR children, and 54% survival at 10 years for severe and profound MR children.

Emond [14] studied the prevalence of children with CP in two cohorts, from the 1958 and 1970 British Births Survey. The prevalence of CP remained constant at 2.5/1000 births. The prevalence at 10 years after birth was higher in the 2nd cohort. All children with CP born in 1970 survived until age 10, while 9 of 40 children born in 1958 were dead before 10 years of age. This study suggests that survival may have improved in children with CP, but more importantly it demonstrates that while obstetrical practices have changed, there was no decrease in the occurrence of CP.

Evans et al. [15,16] have presented two of the more important recent studies dealing with survival in CP. First, they investigated death certificates from 732 children with CP in South East Thames born from 1970–1979. They found that death certificates did not offer accurate data in ascertaining the mortality of children with CP. Evans et al. [16] also prospectively followed cerebral palsy children in South East Thames born 1970–1979, with an average follow-up of 15 years. Immobility (defined as being confined to bed or unable to propel a wheelchair) and severe mental subnormality were the strongest predictors of mortality. Spastic quadriplegia, dyskinesia, and mixed CP were most severely affected. Nearly all ataxic, 90% of dyskinetic, and 72% of quadriplegic children reached 18 years of age. Seizures covaried with mental subnormality and were negatively correlated with survival. Hydrocephalus may also be a negative risk factor. Ninety percent of children in the study survived between 10 and 20 years.

Four studies by Eyman et al. [18–20,52] have added significant insight into the survival of children with CP as well as other developmental disability (DD). In 1990, they reported on 99,543 persons with DD from the California Department of Developmental Services between 1984–1987 [18]. The best predictors of mortality were: (1) deficits in cognitive function, (2) limitations on mobility, (3) incontinence, and (4) inability to eat without assistance. They provided a life table analysis of survival, based on defining three subgroups of children:

Subgroup 1: immobile, not toilet trained, required tube feeding

Subgroup 2: immobile, not toilet trained, but could eat with assistance

Subgroup 3: mobile but not ambulatory and could eat with assistance

A significant error in the data manipulation has been pointed out, resulting in life expectancies which are too low [47]. The *corrected* life table for Subgroups 1, 2, and 3 is listed in Table 1.

According to this data most children in Subgroup 1 would not survive beyond age 10, in Subgroup 2 beyond age 20, and in Subgroup 3 beyond age 50.

Eyman et al. extended these observations in three more recent papers. In the first, they examined the relationship between mortality and the acquisition of basic skills by children and adults with severe disabil-

Table 1
Survival in 3 groups of developmentally disabled children (corrected data)

Age	Subgroup 1: % surviving to age interval	Subgroup 1: life expectancy (years)	Subgroup 2: % surviving to age interval	Subgroup 2: life expectancy (years)	Subgroup 3: % surviving to age interval	Subgroup 3: life expectancy (years)
1	100	11.2	100	25.0	100	42.7
5	66	12.2	85	25.2	95	40.9
10	41	13.1	67	26.2	89	38.5
15	27	13.7	54	26.9	82	36.4
20	18	14.6	45	26.7	77	33.8
25	13	14.8	39	25.7	71	31.6
30	9	14.6	34	23.9	66	28.6
35	6	14.6	30	21.6	62	25.4
40	5	13.3	27	19.1	58	22.1
45	4	12.1	24	16.2	53	18.9
50	3	9.4	21	13.1	46	16.1
55	2	7.5	16	11.5	38	14.2
60	1	6.3	11	10.2	31	11.6
65	<1	4.7	7	9.1	25	9.1
70	<1	3.6	5	7.8	15	8.3
75	<1	4.0	3	6.3	10	6.0
80	<1	2.5	1	6.6	4	7.1
85	<1	2.5	<1	4.0	3	3.3

Adapted from [18].

ities [20]. There were several very important conclusions: (1) subjects who were tube fed and immobile showed very little likelihood of becoming mobile or feeding themselves and had a high probability of death; (2) individuals who had some mobility had a better outcome; (3) self-help skills generally should be achieved by around five years of age if they are likely to occur. After age 6, the most likely outcome for those who were immobile and could not feed themselves was death, or no improvement in self-help skills; (4) there was a very high mortality associated with tube feeding; (5) there was no evidence that these severely involved subjects could be helped by special training, and (6) low IQ was the major deterrent to the acquisition of skills.

In the third study, Eyman et al. [19] once again directly assessed the survival of profoundly disabled people with severe MR. A refinement of the earlier 1990 work, this paper demonstrated that voluntary arm-hand movement and the ability to roll over were associated with improved survival. They again divided their observations into functional categories including mobility (ability to walk, crawl, creep, scoot), rolling (any type), hand use (e.g. grasp), arm use (functional use), toileting skills (trained), eating skills (help with their feeding), and need for tube feeding. Many of the children were also diagnosed with CP. In this study six subgroups were defined:

Subgroup 1: immobile, could not roll, required tube feeding, no arm-hand use

Subgroup 2: immobile, could not roll, required tube feeding, some arm-hand use

Subgroup 3: immobile, could not roll, no arm-hand use, could take food if fed by others

Subgroup 4: immobile, could not roll, could take food if fed by others, some arm-hand use

Subgroup 5: immobile, could roll over, could take food if fed by others, some arm-hand use

Subgroup 6: immobile, some arm-hand use, ability to roll over, tube fed

Several survival charts (which plot additional years children would be expected to live) are presented for children of different ages in each of the different groups.

Table 2 summarizes the median expected additional years of survival for three different age groups (<1, 1–15, and 16–49 years), according to the six subgroups defined above. Note in the table that median survival could not be calculated for all subgroups and all ages. The survival in Subgroup 5, for example, was comparatively good. None of the age groups in Subgroup 5 evidenced a 50% cumulative death rate in the 11-year follow-up period. Another important finding in this study was that "improved medical care has not altered significantly the poor prognosis for those with the most severe impairments."

Anderson [2] criticized Eyman's assessment of the level of mental retardation in subgroups 1–3, and pointed out that many low functioning patients had an outlook as dismal as vegetative patients. Newton [36] objected to Eyman's use of arithmetic means due to outliers within the data set. Strauss [48,57], a future collaborator of Eyman, points out that Eyman had a much

Table 2
Median survival in 6 groups of developmentally disabled children

Subgroup	< 1 year of age	1–15 years of age	16–49 years of age
1	0.9 years	4.8 years	10.4 years
2	1.4 years	5.3 years	Not available
3	1.2 years	5.7 years	10.4 years
4	3.2 years	10.0 years	not available
5	Not available*	Not available*	Not available*
6	8.4	10.9	10.9

*Children in Subgroup 5 had a comparatively good life expectancy. Over 70% of those <1, 70% of those 1–15, and 80% of those greater than 16–49 were alive after 11 years. (Adapted from [19]).

larger data set than other investigators, and was able to assess much lower level of functioning than in previous studies. Thus, low level patients could be better studied than ever before. Also, Eyman eliminated patients that improved from the analysis–these children would have a better prognosis–and thus more accurately rendered the outlook for those children who did not improve.

In 1996 Strauss et al. [52] again attempted to assess predictors of mortality, but also compared risk-adjusted mortality rates for those children living in institutions with those in the community. The population was once again from the California Department of Developmental services, and data comprised over 7000 severely disabled children over a twelve year period. Variables included age, measures of mobility, tube feeding, level of retardation and certain adaptive skills. Confirming their previous studies, reduced mobility and use of tube feeding were the strongest predictors of increased mortality. In addition, newly discovered negative predictors included: (1) lack of hand use, (2) inability to creep/crawl/scoot; (3) inability to speak intelligible words, (4) inability to recognize voices, and (5) inability to interact with peers. The most surprising result was that children cared for within their home residence or within community care facilities had an estimated 25% higher risk-adjusted odds of mortality than those in institutions or health care facilities. This increased relative risk of mortality in community placement has been confirmed in further work by Strauss [53], but has been challenged by other investigators [12,37].

More recently, Strauss et al. [54] have further refined their understanding of the increased mortality associated with tube feeding. The risk was greater in children with less severe rather than those with very severe disabilities, and there was a trend towards reduction in relative risk when the child had a tracheostomy.

Hutton et al. [25] published about the survival of a large cohort of English patients with CP. All children with CP born between 1966 and 1984 to mothers in the Mersey Regional Health Authority were identified, including 1258 subjects with idiopathic CP. The 25 year survival was 89.3% for females and 86.9% for males overall.

They studied three types of disability in the population: (1) ambulation (assistance needed for propulsion), (2) manual dexterity (unable to feed and dress), and (3) mental ability (IQ $<$ 50). For subjects with no severe functional disabilities, 20 year survival was 99%. Children who were severely disabled in all three functional groups had a 20 year survival of 50%. Functional disability was the strongest predictor of survival, while gestational age and birthweight predicted little. Girls had slightly greater survival. The percentage surviving to 27 years of age was 90%, 80%, and 44% for children with one, two, and three severe disabilities, respectively. This data is summarized in Table 3.

Hutton et al. [24] again examined survival in a cohort of CP children in a follow-up study. Data on sex, birth weight, gestation age, and disability were recorded. Cognitive disability was defined as poor if IQ $<$ 50. Manual disability was severe if children were unable to feed or dress independently. Those who did not walk independently were regarded as having severe ambulatory disability. The authors claim that "more than half of even the most severely disabled individuals live to age 35." However the authors them selves acknowledge that their survival data differ markedly from others due to the "composition of study populations and the choice of definitions of disability." Indeed, in the "Commentary" which follows the article, Lewis Rosenbloom noted that the authors "could have provided more detail about the patterns and degrees of disability that correlate with very limited survival." This criticism is mirrored by Eyman [17], who, in a letter to the editor, notes that the reason for Hutton et al's more optimistic life expectancies (in the first study) was due to the failure to differentiate the severity of certain key disabilities. To differentiate patients into many levels of functioning requires very large sample groups, which Hutton's studies did not provide. Furthermore,

Table 3
Survival in children with cerebral palsy according to 3 key disabilities

Age	No disability (% survival)	One disability (% survival)	Two disabilities (% survival)	Three disabilities (% survival)
5	99.9	100	95.2	85.1
10	99.7	96.8	92.1	70.0
15	99.0	95.5	90.4	54.5
20	98.8	93.5	84.7	50.3
25	98.8	90.6	80.5	44.2
27	98.8	90.6	80.5	44.2

Adapted from [25].

Hutton's data can be misconstrued. As Strauss [55] has noted, the most severely impaired children had to survive until age 5 before they were included in the analysis, resulting in an overly optimistic picture for this group. Most recently, Hutton and Pharoah have demonstrated in the same cohort that severe visual impairment is an additional risk factor for early demise, although they did not clarify the type of visual impairment (opthalmic versus cerebral) [26].

Median term survival rates have been calculated for 3189 children born with CP in British Columbia between 1952 and 1989 by Crichton et al. [8]. They noted four categories of CP (quadriplegia/diplegia, hemiplegia/monoplegia, athetosis, other), seizure type (generalized, partial with/without secondary generalization, infantile spasms, unclassified), and presence of MR (non-existent/mild, moderate, severe/profound). The data clearly support the adverse effects of mental retardation, epilepsy, and type of cerebral palsy on survival. The spastic quadriplegia group had the worst prognosis. Overall survival rate at thirty years for all children in the study was at least 87%.

Strauss et al. [58] studied 12,709 infants with cerebral palsy and assessed survival based on two hundred risk factors.

The most powerful prognostic factors for survival were mobility and feeding skills. While 90% of those with fair motor and eating skills reached adulthood, the prognosis for those without those skills was poorer. Among children unable to lift their heads, median survival was seven additional years for those tube fed, and 14 years for those requiring feeding entirely by caregivers. For those with at least some mobility, results were slightly better and are summarized in Table 4.

Strauss and Shavelle [56] also sought to determine predictors of mortality in adults with cerebral palsy. They studied all persons from the California Department of Developmental Services older than 15.

The most important risk factors were deficits in mobility and feeding. Table 5 demonstrates life expectancy by age and cohort for eight stratified groups, and is markedly diminished in those with key functional disabilities. In contrast, life expectancies for the highest functioning groups were on average only five years less than the general population.

A study by Plioplys et al. [41] studied 371 children with cerebral palsy in three RN staffed nursing facilities in the Chicago area. The study was interesting in that it divided its study population into groups based on functional abilities in the manner of Eyman [18,20] in order to compare statistical results. Survival rates for groups 1 through 4 were notably longer than in the Eyman studies, reaching both statistical as well as clinical significance. However, the study is marred by profound weaknesses. The study population is small; only 251 children had significant mental impairment and cerebral palsy. The number of children in groups 4, 5 and 6 were only 11, 9, and 2, respectively. The study population was all inpatient, a notable distinction to the thousands of patients in the California cohort. Details of the data analysis are not provided, leaving the reader totally uninformed of the statistical methods used. The subject count seems confusing: at one point 371 children had cerebral palsy, yet later in the paper only 367. The paper does not consider the vast number of studies included in this review, concentrating only on the studies of Eyman. The study does not note any difference in outcome of children with tracheostomy, in contrast to conflicting studies in the literature [46,59]. However, the article does support the role of severity of mental retardation, presence and severity of epilepsy, and the importance of arm movement/mobility/rolling as important factors in predicting survival. The choice of journal for publication is questionable as outside the literary arena of most persons interested in this topic area.

Blair et al. [4] have recently reported on life expectancy children from an Australian Registry of 2014 CP children. The data confirms that profoundly mentally retarded children with CP do not live into adulthood, and that intellectual disability was the single strongest predictor of survival. The severity of physical

Table 4
Survival in children with cerebral palsy according to 3 key disabilities

	5 year survival	10 year survival	15 year survival
Lifts head and chest, partial rolling			
Tube fed	73%	62%	62%
Fed by others	93%	87%	79%
Some self-feeding	98%	95%	91%
Full rolling, does not walk unaided			
Tube fed	89%	85%	80%
Fed by others	97%	94%	90%
Some self-feeding	98%	97%	95%
Walks ten feet unaided			
Tube fed	89%	Inadequate data	Inadequate data
Fed by others	98%	98%	91%
Some self-feeding	100%	99%	99%

Adapted from [58].

Table 5
Life expectancy in adults with cerebral palsy according to key disabilities

Sex/Age	Cannot lift head		Lifts head			Rolls/sits			General
	TF	FBO	TF	FBO	SF	TF	FBO	SF	population
Female									
15 y	15.4	21.3	21.1	27.7	43.4	25.0	39.2	52.7	65.0
30 y	13.4	23.8	16.2	28.1	33.0	21.5	34.1	40.1	50.4
45 y	*	20.4	16.1	20.6	23.6	23.3	24.1	29.4	36.2
Male									
15 y	11.7	16.9	16.7	22.8	38.8	20.3	34.3	48.7	58.3
30 y	12.0	22.0	14.7	26.2	31.0	19.8	32.1	38.0	44.5
45 y	*	17.5	13.6	17.7	20.6	20.3	21.0	26.3	31.1

*Group too small for reliable computations.
TF = tube fed, FBO = fed by others, SF = self-feed.
Adapted from [56].

impairment was the second most important factor that profoundly impacted life expectancy. Indeed, when Shavelle et al. [44] compared the data from their California data base, the Australia data base, and the English data base (see Hutton studies above) while controlling for IQ and severity of CP, they found strikingly similar results among the three data sets. Finally, Blair [4] compared different cohorts of children with CP and found no evidence of increased duration of survival since the 1950's despite advances in medical care.

Several investigators have investigated what are the causes of excess mortality in persons with cerebral palsy. Strauss et al. [50] reported on a very large data set 4028 deaths and found causes related to the respiratory and circulatory systems, as well as certain cancers. Williams [60], Blair [4], and Reddihough [42] confirm the respiratory system as a leading cause of mortality, as well as neurological complications (seizures and hydrocephalus). Results of Plioplys [41] were similar.

Estimating life expectancy in children with mental retardation (MR). Several studies have also examined the life expectancy of children with MR, distinct from those with developmental disability with mental retardation. While technically not part of the "cerebral palsy" population, these studies also have bearing on the prognostication of life expectancy in a related group. Using present nomenclature, MR is categorized according to intellectual impairment into mild (IQ level 50–55 to 70), moderate (IQ level 35–40 to 50-55), severe (IQ level 20–25 to 35–40), and profound MR (IQ level below 20 or 25) [1,10]. Many recent publications prefer the term "intellectual disability".

Heaton-Ward [23] studied the life expectancy of inpatients with MR, including 108 females and 105 males. The age at death was strongly correlated to IQ. The average age of death for "non-Mongoloid" persons (i.e. mental retardation not associated with the genetic defect Trisomy 21 or Down Syndrome) was 29.0/32.0 years for males/females with IQ < 25, and 39.5/45.2 for males/females with IQ 26–50.

McCurley et al. [32] examined the average age at death in persons with MR and compared his data to earlier studies (summarized in Table 6). All individuals with MR had a significantly decreased life expectancy, which decreased dramatically with the severity of the

Table 6
Average age at death in three older studies of adults with mental retardation

Survey	Mild MR		Moderate MR		Severe MR	
	male*	female*	male	female	male	female
Primrose (1966)	45.1	47.5	45.1	46.2	25.2	25.0
Heaton Ward (1968)			40.0	49.8	28.8	29.3
McCurley (1972)	58.1	49.8	37.1	40.8	17.3	14.0

*As used by these authors, feeble-minded designates the "moron" (outdated term) or "mildly retarded" group of mentally handicapped individuals.
Adapted from [32].

mental deficit. They included patients cared for in a hospitalized as well as a community setting. In this study, patients in a community care program had a better prognosis. Specific data for patients with CP showed the mean age of death was 8.3 for males and 10.5 for females.

The life expectancy of mentally retarded persons in Canadian institutions from 1966–1968 was published by Balakrishnan and Wolf [3]. This study primarily involved more severely retarded individuals. For the profoundly retarded, the average age of death was approximately 23.5/23.7 for males and females. For the severely and moderately retarded, average age of death was 37.9 and 42.5 years for males and females, respectively. These results were more recently replicated in a facility within the southwestern United States [38].

Kaveggia [29] performed a survival analysis of 1915 severely and profoundly mentally retarded individuals who were institutionalized at the Central Wisconsin Center. Survivors of CP comprised 442 of the total studied, which included a wide gamut of etiologies for mental retardation. On admission, the median remaining lifetime was 8.81 ± 4.16 years for residents with inborn errors of metabolism, 18.56 ± 4.26 years for residents with primary CNS malformations, and 24.66 ± 4.05 years for residents with unknown syndromes.

Chaney et al. [7] reviewed 1146 patients who had died and were autopsied from 1944–1983 from Lanterman State Hospital, a South Carolina hospital for persons with MR. They evaluated survival by reported central nervous system "insult". Of those with perinatal insult, 34.6% survived beyond age 21. Similarly, 29.6% of prenatal, 16.7% of postnatal, and 47.4% of those in whom time of insult was uncertain survived beyond age 21. When considering all groups, 34.1% survived beyond age 21. This study is limited by the wide variety of etiologies causing MR. Furthermore, even in those cases with the diagnosis of "perinatal asphyxia," recent literature suggests that the ability of an investigator or caregiver to accurately determine the time of central nervous system insult is highly limited, suggesting the information in this study must be cautiously interpreted [14,34]. As stated by Kuban and Leviton [30], "epidemiologic studies . . . have not provided any reasons to change the impression that our ability to identify modifiable, presumed causes of cerebral palsy is limited".

Table 7
Key disabilities diminishing life expectancy in children with cerebral palsy

Presence and severity of mental retardation
Inability to speak intelligible words
Inability to recognize voices
Inability to interact with peers
Physical disability
Limitations on mobility
Inability to propel wheelchair
Inability to roll over
Inability to creep/crawl/scoot
Lack of upper extremity function
Inability to eat without assistance
Tube feeding
Incontinence
Presence and severity of seizures

Simila et al. [45] prospectively followed 12,058 children from Finland, Oulu, and Lapland, born in 1966 (96% of all children born) until they had reached the age of 17. One hundred and sixty-five children had MR–97 severe (IQ < 50), 68 mild (IQ 50–70). The death rate was 158/1000 for those with MR as compared with 22/1000 for those with normal intelligence.

In one of the most methodologically sound and clinically important studies to date on the survival of those with MR, Dupont et al. [11] studied the mortality, life expectancy, and causes of death of individuals with mild MR in Denmark. A total of 7314 persons were included. Even in the mild MR population, the 50% survival for men and women was approximately 65 and 75 years of age, respectively, significantly less than a "normal" population. When examining those who had already survived to 10 years of age, only 50% survived to be 68–70 years. When comparing those who were and were not institutionalized, the type of residence had no effect on life expectancy. Indeed, while Plioplys et al. [41] argue that institutionalized patients have

longer life expectancy due to the availability of acute care medical services, Chaney and Eyman [6] argue that mortality of children with MR who are institutionalized has increased as less severely impaired children have been mainstreamed into the community. Hayden [22] reviewed the literature on this subject and summarizes the articles as "conflicting".

Wolf and Wright [61] examined changes in life expectancy of persons with MR in Canadian institutions using life table analysis. Less than 50% of the persons with profound MR population lived beyond 41 years, and only 50% of persons with moderate/severe MR lived beyond 58 years. Even in the group with borderline/mild CP, only 50% lived beyond age 67. Wolf calculated that the life expectancies for institutionalized Canadian children with MR (who had reached five years of age from 1976–1978) was 39.6 for profoundly retarded, 48.2 for moderately and severely retarded, and 58.0 years for borderline and mildly retarded. There had been a mild increase in life expectancy between an earlier cohort (1966–1968) and the most recent cohort (1976–1978) studied.

Kastner et al. [28] studied mortality among 1300 people with mental retardation who lived in the community. They noted only 14 deaths in a four-year period, several of which were considered "avoidable" by standardized criteria. Surprisingly, their mortality rate of 4/1000 over the four years was lower than in the general population.

Forsgren [21] studied a cohort of all children with MR living in a non-institutionalized setting within a single Swedish province for a span seven years. The standardized mortality ratio was 1.6 the general population in those with MR only, 5.0 greater in those with MR and epilepsy, and 5.8 greater in those with MR, epilepsy, and cerebral palsy. The mortality increase was seen in both those with partial seizures which generalized, and those which did not. The highest mortality was seen in those who always generalized from the onset–8.1. Pneumonia was the most common cause of death.

Strauss and Eyman [51] presented a life table analysis of persons with mental retardation, and compared groups with and without Down Syndrome from a data base from the California Department of Developmental Services. Survivors were stratified as mild, moderate, severe, and profound mental retardation. Non-Down Syndrome children with mild or moderate mental retardation had life expectancies only had life expectancies slightly shorter than those in the general population. Less than half of severely mentally retarded children (non-Down) survived beyond age 60, and less than half of profoundly mentally retarded children survived beyond age 25. While respiratory disease was the most common cause of death, gastrointestinal disease (perforation, ruptured appendix, diverticulitis, fecal impaction, ischemic bowel), was also a notable cause [49].

Patja et al. [39] studied 2366 persons from the Finnish Population Register Centre who were available for follow-up from a study performed in 1962 with a mean follow-up of 26.9 years. For persons with mild MR, life expectancy did not differ from the general population. In the moderate MR group, the decrement was slight, but in the severe and profound groups there was a 19–35% diminishment in life expectancy. The results of Janicki et al. [27] have been similar. Patja [40] also found the three most common causes of death to be cardiovascular disease, respiratory disease, and neoplasms.

3. Conclusions

A systematic review of the literature dealing with life expectancy in children with CP and MR reveals:

1. Life span of the child is curtailed by the presence of certain key disabilities, summarized in Table 7.
2. Decreased cognitive abilities are associated with diminished lifespan, even in the absence of physical impairment.
3. Life expectancy for physically and mentally disabled persons has increased slightly with time.
4. Institutionalized children do not have shorter life spans than those cared for in a non-institutionalized setting.
5. Improvements in medical care have not clearly altered the poor prognosis for the most severely disabled children.

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