Change in ambulatory ability of adolescents and young adults with cerebral palsy

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This study aimed to determine the probability that a child with cerebral palsy (CP) will lose or gain ambulatory ability through adolescence and young adulthood. We analyzed retrospectively data from 1987 to 2002 on Californians with CP initially aged 10 years (SD 0.9y; *n*=7550 [4304 males, 3246 females]) and 25 years (SD 0.8y; *n*=5721 [3261 males, 2460 females]) who had varying levels of ambulatory ability (initial Gross Motor Function Classification System Levels I–IV). We used the Aalen–Johansen estimator to estimate probabilities of transition to other levels of ambulatory ability in the future. Those who walked and climbed stairs without difficulty at age 10 had only a 23% chance of decline (to requiring a handrail to manage stairs, or worse) 15 years later. Those who ambulated with some difficulty but did not use a wheelchair had a significant chance (33%) of improvement (to being able to walk unsteadily alone at least 3m or better) and only a small chance (11%) of becoming non-ambulatory. Those who used a wheelchair were more likely to lose ambulatory ability (34%) or die (6%). Those who walked and climbed stairs well at age 25 were likely to maintain that ability 15 years later (76%), while those needing support to climb stairs were more likely to lose ability. Improvement in ambulation after age 25 was unlikely. Children and young adults with CP are likely to maintain their ambulatory ability during their next 15 years. Some who ambulate with difficulty at age 10 may improve through adolescence, but those who use a wheelchair are more likely to decline. By age 25 improvement in ambulation is unlikely and decline more likely. Most, however, will not change over the next 15 years.

See end of paper for list of abbreviations.

Though cerebral palsy (CP) is generally considered to be a static neurological condition, gross motor skills of persons with CP do evolve over time. Age, general health, and therapeutic or surgical interventions may all contribute to changes in functional abilities throughout childhood.[1–6] Functional abilities are related to health and self-reliance, and their pattern of development is important for anticipating needs and planning future care.

Numerous studies have documented changes in ambulation and other gross motor functions for children and adults with CP.[7–13] For example, a recent study illustrated that prospects for future ambulation of 2- to 3-year-old non-ambulatory children with CP vary according to their level of gross motor functioning (e.g. rolling and sitting ability).[9] The study also suggested that children who remained non-ambulatory by 8 years of age had little chance of walking after that. Other studies have also suggested that improvement in motor function may plateau by age 8.[10] At least one study found children continuing to gain ambulatory ability as late as age 9.[12] An investigation into the decline in function of older adults with CP found that most who walked well alone at age 60 lost some ambulatory ability over the next 15 years.[11]

Various physical therapies or other interventions intended to improve ambulatory ability in children with CP have been considered. These have included surgery,[14] ankle–foot orthoses,[15] strength training,[16,17] and electrical stimulation.[18] Few of these studies examined the question of change in ambulatory status in adolescence or adulthood, though one study did find that strength training can improve ambulatory ability even in adolescents,[16] while another found no improvement after surgical intervention for children over 12 years of age.[14]

In this study we investigated whether the ambulatory ability of persons with CP continues to evolve throughout adolescence and into adulthood, and whether the chance of future improvement or decline depends on current levels of ambulatory ability.

Method

We selected two retrospective cohorts of persons with CP: children aged 9 to 12 years (mean age 9y 10mo [SD 11mo], henceforth, age 10y), and adults aged 24 to 27 years (mean age 24y 6mo [SD 10mo], henceforth, age 25y). Participants were drawn from persons with CP who received services from the Department of Developmental Disabilities (DDS) in the State of California between January 1987 and December 2002. Services provided by the DDS include medical treatment, occupational or physical therapy, case management, and social services for all state residents with a substantive disability resulting from CP. CP is defined as either 'a non-progressive lesion or disorder in the brain occurring during intrauterine life or the perinatal period and characterized by paralysis, spasticity, or abnormal control of movement or posture which is manifest prior to 2 to 3 years of age', or 'other significant motor dysfunction appearing prior to age 18'.[19]

Individuals receiving services from the DDS are evaluated approximately every 12 months with a structured interview known as the Client Development Evaluation Report (CDER).[20] This instrument contains more than 200 psychological, medical, functional, behavioral, and cognitive items. From these assessments we extracted information regarding each person's ambulatory status, stair climbing ability, and wheelchair use. The reliability of the functional items in the CDER

has been assessed previously and judged satisfactory.[21,22]

We identified CDERs completed during the study period for persons with CP at 10 or 25 years of age. If a person received two evaluations within the age range (9–12y or 24–27y), we began follow-up with the earlier evaluation. We were interested in congenitally acquired CP, and, therefore, excluded persons with diagnoses suggesting CP of postnatal origin (e.g. traumatic brain injury, near drowning, motor vehicle accident, brain tumor, other acquired injuries) as well as those with significant concomitant diagnoses (e.g. autism, Down syndrome, degenerative disorders). The final study groups at ages 10 and 25 years comprised the remaining participants. We obtained mortality information from electronic files of the California Department of Health Services.[23]

In classifying current and future ambulatory ability in persons with CP we considered three functional items on the CDER: 'ambulation', 'climbing stairs', and 'wheelchair mobility'. Based on clinical considerations and initial explorations of outcomes, we ultimately considered the following 4-point scale of ambulatory ability: (1) walks well alone at least 6 meters, balances well, and moves up and down stairs without the need for a handrail; (2) walks well alone at least 6 meters and balances well, but moves up and down stairs only with the use of a handrail; (3) walks unsteadily alone at least 3 meters, or walks only with assistive devices; and (4) does not walk.

For individuals at a given level on this scale we sought to determine the likelihood of their being at the same or any other level, or deceased, in the future. Persons initially at level 4 (does not walk) included many individuals with severe impairment, some of whom could not roll, sit, or crawl. Even among those who could crawl or sit, the chance of gaining ambulatory ability after age 10 was extremely small. Therefore, we did not consider level 4 further as a starting state, choosing instead to focus on those who had at least some ambulatory ability by age 10 or 25. As children and adults initially at level 3 included a significant number who were also using a wheelchair on a regular basis, we further stratified this starting level according to wheelchair use (yes/no). Thus, we finally considered four starting states corresponding to levels 1, 2, and 3 above, with level 3

Table I: Demographic characteristics of children and adults at start of follow-up

Characteristic	*Children (n=7550) (mean age 10y)*	*Adults (n=5721) (mean age 25y)*
Sex		
Female	43	46
Male	57	54
Race/ethnicity		
Asian	3	3
Black	10	10
Hispanic	35	24
White	39	5
Other	4	2
Unspecified	9	8
Type of motor dysfunction		
Spasticity	54	50
Ataxia	6	10
Dyskinesis (dystonia or athetosis)	3	5
Hypotonia	15	7
Other (including mixed)	17	1
Unknown	6	9
Location of motor dysfunction		
Quadriplegia	36	32
Diplegia	19	14
Hemiplegia	19	23
Monoplegia	2	3
Triplegia	1	1
Paraplegia	4	6
Other (including mixed)	12	11
Unknown	5	9
Hand use		
Has no functional use of hand	1	1
Uses raking motion or grasps with hand	9	10
Uses thumb and fingers of hand in opposition	19	18
Uses fingers independently of each other	71	71
Stair climbing		
Does not move up and down stairs	15	7
Moves up and down stairs with help	25	20
Moves up and down stairs with handrail independently	38	40
Moves up and down stairs without need for handrail	20	32
No opportunity to use stairs	2	1
Wheelchair use		
Sits in wheelchair but does not move by self	9	6
Assists in moving wheelchair	4	3
Moves self with some difficulty in steering	6	6
Moves or guides chair independently and smoothly	5	9
Does not use wheelchair	76	77

All figures are column percentages.

Table II: Initial distribution of stair climbing ability and wheelchair use by level of ambulatory ability in children aged 10

	Ambulatory ability		
	Walks with support (n=2002)	*Walks unsteadily alone at least 3m (n=1418)*	*Walks well alone at least 6m, balances well (n=4130)*
Stair climbing			
Does not move up or down stairs	49	7	1
Moves up and down stairs with help	39	44	12
Moves up and down stairs with handrail	6	44	52
Moves up and down stairs without handrail	1	3	35
No opportunity to use stairs	5	1	1
Wheelchair use			
Sits in wheelchair, does not move by self	28	7	1
Assists in moving wheelchair	12	3	1
Moves self with some difficulty in steering	16	5	1
Moves chair independently and smoothly	14	3	1
Does not use wheelchair	30	82	96

All figures are column percentages.

stratified according to need for wheelchair (yes/no). In addition to wheelchair use, we initially stratified each starting group by a number of other variables (e.g. type or location of CP, presence or absence of epilepsy) to determine possible impact of the variables on long-term probabilities of change in ambulatory status after age 10 or 25 years.

STATISTICAL ANALYSIS

For each starting group (at level 1, 2, and 3 with or without wheelchair use) the probabilities of being in a given state (1–4, or dead) any number of years in the future were estimated using the Aalen–Johansen estimator.[24] The estimator was developed as an extension of the usual Kaplan-Meier estimator to account for multiple live states (or multiple dead states as in a competing risks model).[25] The Kaplan-Meier estimator is a non-parametric estimator of the probability of survival to a given time. It is the product of estimated conditional probabilities of survival calculated as $1 - d_j/r_j$, where r_j is the number of individuals at risk (i.e. alive and not censored) just prior to time t_j, and d_j is the number dying at time t_j. The probability of dying by time t_j is equal to 1 minus the estimated probability of survival to time t_j, and the two figures can be calculated simultaneously as the product of 2×2 matrices of estimated conditional probabilities of transitions from live to live, live to dead, dead to live (probability 0), and dead to dead (probability 1). In an analogous manner, the Aalen–Johansen estimator provides estimates of the probabilities of transition to any one of a number of different live states (in the present case, four levels of ambulatory ability), or death, by any future time t_j for persons in a given state at time t_0. The estimator is a product of $n \times n$ matrices, where n is the total number of states, alive or dead (in the present study, $n=5$). Diagrams based on the Aalen–Johansen estimator have been used previously to describe the likelihood of change in ambulatory status of persons with CP.[9,11] As applied here, the estimator is consistent, regardless of any Markov assumption about the transition probabilities, and more efficient than other proposed estimators.[26] Variances of the estimated transition probabilities are approximated by a formula (involving matrices) analogous to that for the Kaplan-Meier estimator, and we have used these to determine confidence intervals (CI). Statistical analyses were carried out in SAS (version 6.12) with S-PLUS (version 4.0) used for graphics. In particular, matrix manipulations were performed using the Interactive Matrix Language of SAS.

Ethical approval

This study received approval from the Institutional Review Board of the Office of Research Affairs, University of California at Riverside.

Results

After exclusions, the final cohorts included 7550 participants at age 10, and 5721 participants at age 25. Table I displays demographic characteristics of the participants. The most common type of CP was spasticity, and the most common anatomical distribution was quadriplegia. More than 70% of participants had good use of their hands (used fingers independently of each other) and nearly 80% did not use a wheelchair at the beginning of follow-up.

By selection, all participants in both age groups were initially able to ambulate at some level (with supportive devices or better). Within each level of ambulation, however, participants in both age groups had varying levels of stair climbing ability

Table III: Initial distribution of stair climbing ability and wheelchair use by level of ambulatory ability in adults age 25

	Ambulatory ability		
	Walks with support (n=1090)	*Walks unsteadily alone at least 3m (n=1095)*	*Walks well alone at least 6m, balances well (n=3536)*
Stair climbing			
Does not move up or down stairs	31	4	<1
Moves up and down stairs with help	47	37	6
Moves up and down stairs with handrail	13	55	44
Moves up and down stairs without handrail	4	3	49
No opportunity to use stairs	5	1	<1
Wheelchair use			
Sits in wheelchair, does not move by self	20	7	1
Assists in moving wheelchair	12	3	<1
Moves self with some difficulty in steering	21	7	<1
Moves chair independently and smoothly	30	7	2
Does not use wheelchair	16	75	96

All figures are column percentages.

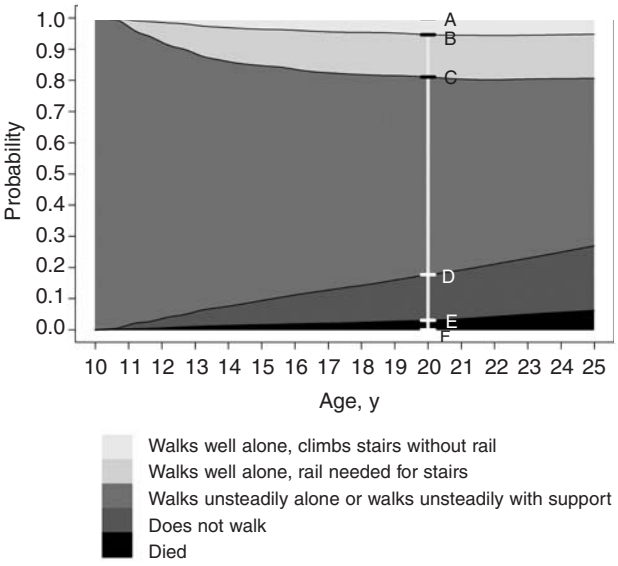

Figure 1: *Subsequent walking ability for 3420 persons with cerebral palsy aged 10 years who initially walked unsteadily alone at least 3 meters, or who walked with support. At age 20, probabilities (95% confidence interval) are 5% (4–7%) for level 1 (segment A–B), 14% (12–15%) for level 2 (segment B–C), 63% for remaining in level 3 (segment C–D), 14% for level 4 (segment D–E), and 4% for dying (segment E–F).*

or wheelchair use. Table II gives the distribution at start of follow-up of stair climbing ability and wheelchair use, stratified by ambulatory ability.

Figure 1 shows the prognostic chart for study participants initially able to walk with support, or unsteadily alone at least 10 meters (i.e. level 3 of the scale described above) at age 10 years. Different levels of walking ability are indicated by shades of gray, the lightest shade being the highest level of ability and the darkest shade being the lowest. Death is indicated in black. The probability of being in a given state at a given age in the future can be determined. A vertical line at age 20 is shown in Figure 1 for illustration. Thus, for a 10-year-old initially at level 3, the probability (with 95% CI) of being in the various states (or dead) at age 20, are: 5% (4–7%) at level 1; 14% (12–15%) at level 2; 63% (61–66%) at level 3 (i.e. no change); 14% (12–16%) at level 4; and 4% (1–6%) died (see Fig. 1).

Figure 2 shows the prognostic charts for participants aged 10 years who are initially at levels 1, 2, and 3, with level 3 further stratified by wheelchair use (some use, or none). Figure 3 shows comparable charts for study participants initially aged 25. Reading from Figure 2d, 10-year-olds who initially walked and climbed stairs without difficulty were very likely (77%) to maintain that level of ambulation at age 25. Those who walked well alone at least 20 meters and balanced well, but required some assistance with stair climbing (Fig. 2c), were about as likely to improve as decline in ambulatory ability, but were most likely to stay the same (54%). For participants who had more difficulty walking at age 10 (unsteadily alone at least 10 meters, or only with support), there were two very different outlooks depending on whether they initially used a wheelchair. Those who did need a wheelchair (Fig. 2a) had a substantial chance (34%) of losing their ambulatory ability by age 25, while those who had not begun to use a wheelchair (Fig. 2b) had a very small chance of regressing, and a better chance (32% by age 25) of improving in ambulatory ability.

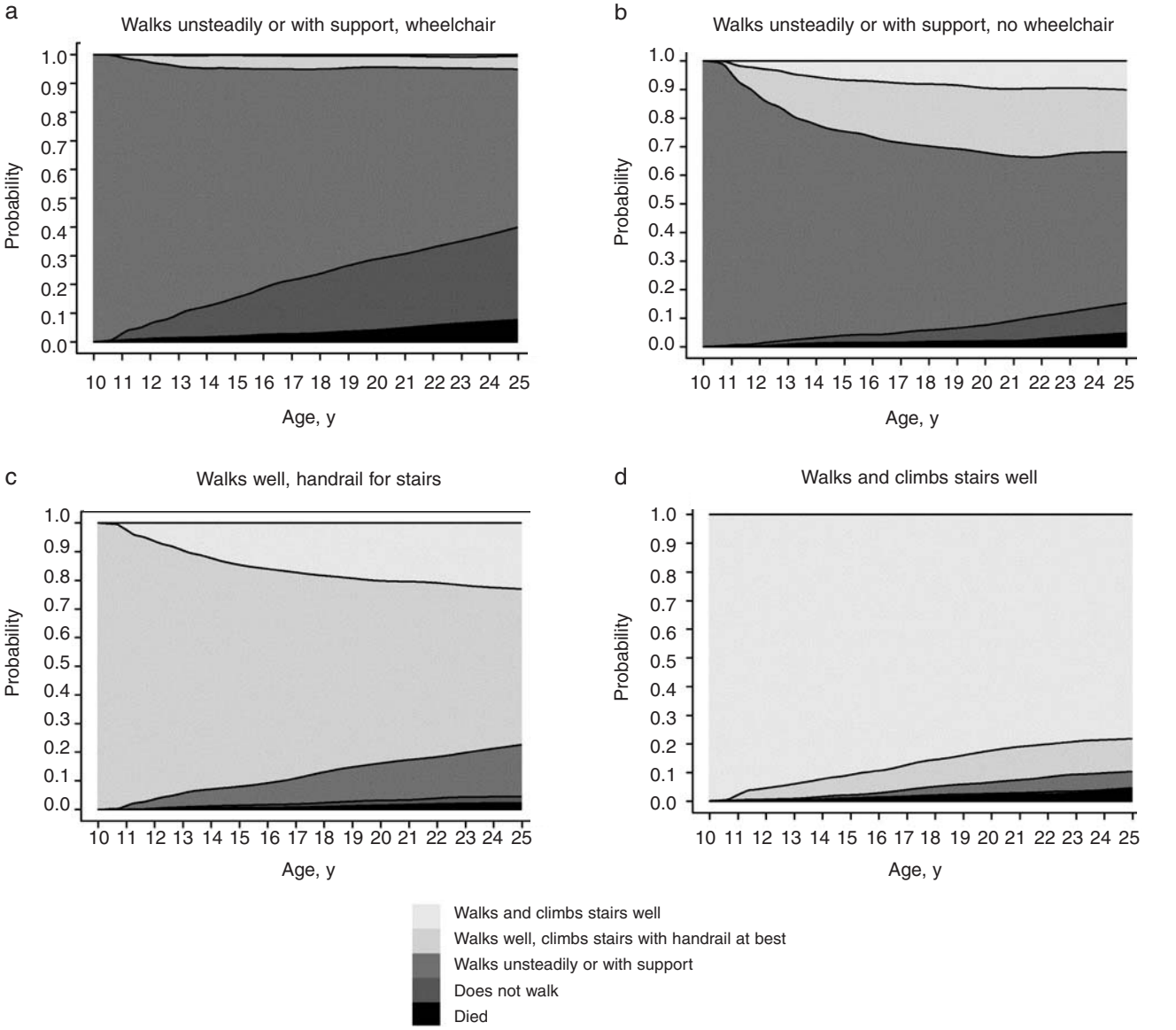

Figure 2: *Subsequent walking ability for persons with cerebral palsy aged 10 years with various initial levels of ambulatory ability. Those who walked unsteadily alone, or with supportive devices, were stratified according to wheelchair use.*

For participants beginning the study at age 25, similar results were observed (Fig. 3). Generally, however, participants beginning the study at age 25 had an even greater chance of remaining at their initial level of ability, and a smaller chance of improvement than for the younger groups. For those who walked without support (Figs. 3c and 3d) there was a slightly greater chance of decline in function than for the 10-year-olds. Those who walked unsteadily or with supportive devices (Figs. 3a and 3b) again showed a substantial difference in outcome depending on initial wheelchair use with those who used a wheelchair initially having virtually no chance of improvement and a far greater chance of decline than those who at age 25 had not begun using a wheelchair. For both 10-year-olds and 25-year-olds, further stratification of initial groups by other variables (e.g. type of CP, distribution of limb involvement, or presence of seizure activity) did not result in significant differences in estimated probabilities.

Discussion

We find that persons with CP at both age 10 and age 25 years, who initially walk without difficulty, have a high likelihood of maintaining that ability over the next 15 years. This is not surprising as those with the mildest form of CP would be expected to have natural histories not unlike those of the general population: ambulatory ability does not decline for most people throughout early adulthood.

Children with CP who have some difficulty with ambulation at age 10 (i.e. who walk well but require some support or assistance in climbing stairs) are equally likely to improve in their stair climbing, or decline in their walking, over the next 15 years. By age 25, however, the chance of further improvement is very small, while the chance of losing ability

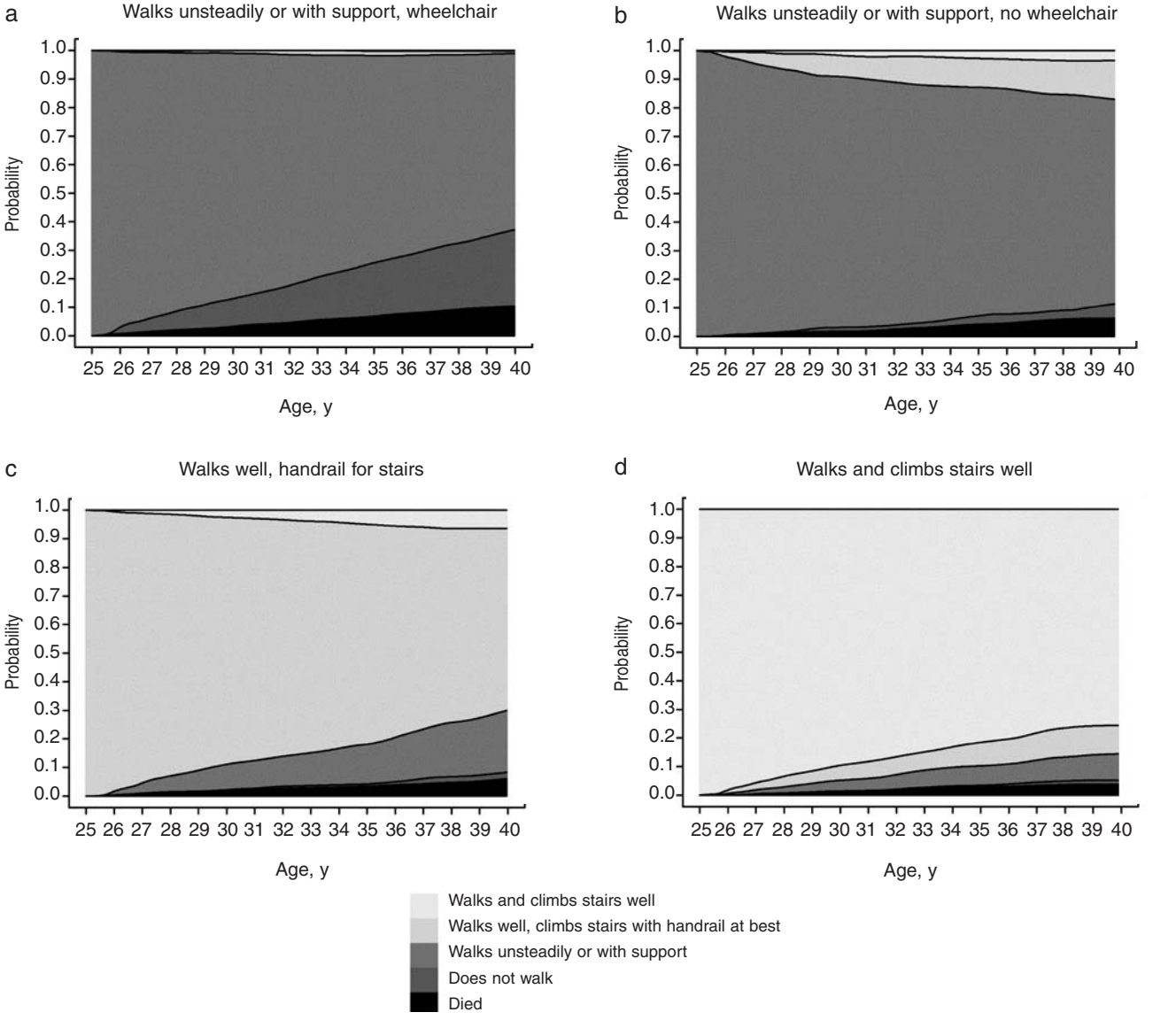

Figure 3: *Subsequent walking ability for persons with cerebral palsy aged 25 years with various initial levels of ambulatory ability. Those who walked unsteadily alone, or with supportive devices, were stratified according to wheelchair use.*

is significant. Few 10-year-olds or 25-year-olds at GMFCS Level II (walks well, uses support or assistance in stair climbing) initially lost their ambulatory ability completely.

Children with CP aged 10 who walk with some difficulty (i.e. walk 3 meters unsteadily alone, or walk only with support) have roughly equal chances of gaining or losing ambulatory ability, though again, most likely, no change will occur. That they show a significant chance of improvement, however, is a bit surprising, as it has been suggested that gross motor functioning in children with CP changes little after age 8.[10] The chance of further improvement in ambulatory ability is even greater for those not using a wheelchair, and the chance of decline in this group is small.

Mortality was relatively low over the 15-year study period for all groups considered. Not surprisingly, mortality was highest for those with the lowest initial ambulatory ability (GMFCS Level III: walks unsteadily alone, or only with support).

Our study is subject to a number of potential limitations. First, it is possible that children or adults with milder motor dysfunction may have been preferentially lost to follow-up, perhaps declining further DDS services due to perceived lack of need. This would result in a study population that includes a greater proportion of more severely affected adolescents or young adults, and the chance of declining in ambulatory ability may be overestimated as a result.

Also, we lack specific information on the type or frequency of wheelchair use. Such information could have led to further useful stratification of the groups initially ambulating with some difficulty. Details on ambulation and stair climbing are also limited to those provided in the CDER and, again, it is possible that further and more detailed information (e.g. specifics regarding gait or posture) may have had some influence on future improvement or decline. We also lacked information on a number of other factors that may have an impact on future ambulatory ability in persons with CP. Examples include surgical or therapeutic interventions, psychological factors, and specific attributes of gait or ankle motion. It would be of interest to know how these might affect long-term ambulatory status in CP.

While we did not detect significant differences in outcome according to a number of variables other than wheelchair use (e.g. type or location of CP, presence of seizure activity), in many cases numbers within a specific group were small. It does seem plausible, for example, that different types of CP may have somewhat different progressions of ambulatory ability. Future researchers should not be discouraged by our failure to detect such differences here.

We have used a definition of CP available in the California DDS data. This definition, as well as our exclusion of persons with diagnoses suggesting CP of postnatal origin and persons with significant concomitant diagnoses (see Method) should be considered when comparing our results with other available or future research.

The above limitations are offset by strengths, including the large numbers, and the ability to provide prognostic information on several levels of ambulation over time.

These findings may be important for parents, doctors, and life care planners involved in the long-term care of persons with CP. Clearly there is potential in this population for maintaining or even improving ambulatory ability throughout adolescence and young adulthood.

Accepted for publication 14th February 2007.

Acknowledgments

Provision of data from the California Department of Developmental Disabilities and Department of Health Services is gratefully acknowledged.

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List of abbreviations

CDER	Client Development Evaluation Report
DDS	Department of Developmental Disabilities