Stability of motor function and associated impairments between childhood and adolescence in young people with cerebral palsy in Europe

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ABBREVIATIONS

BFMF	Bimanual Fine Motor Function
MACS	Manual Ability Classification system
SPARCLE	Study of Participation of Children with Cerebral Palsy Living in Europe

AIM The aim of the study was to investigate whether impairments associated with cerebral palsy were stable between childhood and adolescence.
METHOD The Study of Participation of Children with Cerebral Palsy Living in Europe (SPARCLE) longitudinal study was conducted in nine European regions. In total, 818 children aged 8 to 12 years were randomly selected from population-based registers; 594 (73%) were followed up at the age of 13 to 17 years (344 males, 250 females; median age 10y 4mo) Research associates visited them in their homes and recorded their motor function and additional impairments. Stability of impairment was assessed using the weighted kappa coefficient.
RESULTS The proportion of participants whose level of impairment remained unchanged varied from 63% for fine motor function to 98% for hearing. For gross motor function, communication, and cognitive level, the kappa and the lower bound of its 95% confidence interval (CI) were above 0.75, indicating stability between childhood and adolescence; for fine motor function and feeding, the kappa was above 0.75 but the lower bound of the 95% CI was below 0.75, indicating probable stability; for seizures and vision, the kappa was below 0.75, although the upper bound of the 95% CI was above 0.75, indicating possible change; for hearing the kappa and its entire CI were below 0.75, indicating change. Overall, 81% of participants had no seizures in childhood, of whom 93% were seizure-free in adolescence.
INTERPRETATION Motor function and additional impairments were generally stable between childhood and adolescence.

Cerebral palsy (CP) is the most common serious motor impairment in children and adolescents.[1] Although it is the result of a brain lesion which is non-progressive, function may change over time. A recent update of the definition of CP included activity limitations and associated impairments such as cognitive impairment, epilepsy, and problems with behaviour, hearing, vision, feeding, and communication.[2]

Longitudinal data on gross motor function (using the Gross Motor Function Classification System [GMFCS]) for children and adolescents with CP have been reported.[3–5] Limited information on hand function and associated impairments has been reported from smaller studies.[6,7]

In this paper, we investigated stability over time not only of motor function, but also of associated impairments between childhood and adolescence from a large sample of population-based data.

METHOD

The study is part of the wider Study of Participation of Children with Cerebral Palsy Living in Europe (SPARCLE) project, which aims to identify factors that influence participation and quality of life in children and adolescents with CP in Europe. Nine regions in seven countries participated. Information on the children was available from population-based CP registers from eight regions across Europe,[1] and one further region recruited children from multiple sources. The methods and background data from SPARCLE1 and SPARCLE2 have been described.[8–11]

The participants were born between 31 July 1991 and 1 April 1997. Children and their parents were visited at home on two occasions by research associates from each region, first at the age of 8 to 12 years (SPARCLE1) and then when they were aged 13 to 17 years (SPARCLE2). The research associates received joint training before the study. During the visit, the research associate completed

the impairment form after observing the young person and from information provided by a parent. The same form was used on each occasion to record information on gross motor function (GMFCS), hand function (Bimanual Fine Motor Function [BFMF]), seizures, communication, feeding, cognitive level, vision, and hearing. Cognitive level was estimated by combining the information from the CP register at age 4 years, neuropsychological assessment if available, current school performance, parent information, and questions on learning and understanding if in doubt. Seizure frequency in the last year and the use of anticonvulsants were recorded.

Ethics approval was obtained from ethics committees in each country.

Sample characteristics

Of the 818 children who participated in SPARCLE1, 594 (73%) agreed to participate in SPARCLE2. These children constituted the study sample. The number of participants, percentage of males and females, and median age in SPARCLE1 and SPARCLE2 for each region are shown in Table I.

The categorization of impairment and the proportion of children and adolescents in each category are shown in Table II. At the group level, the distribution of impairment was similar in childhood and adolescence.

Statistical analysis

We report the numbers and percentages of young people at each level of impairment in childhood and adolescence. We plotted the number at each combination of childhood and adolescent impairment levels.

We used the weighted kappa statistic with its 95% confidence interval (CI) to summarize the agreement, corrected for chance, between the childhood and adolescent levels of impairment.

Table I: Number and median age of participants by region

Region	Total	Male, %	Female, %	SPARCLE1 median age	SPARCLE2 median age
North England, UK	80	64	36	10.5	15.0
West Sweden	68	57	43	10.5	15.6
Northern Ireland, UK	85	59	41	10.3	15.1
South-east France	50	58	42	10.6	14.8
South-west Ireland	74	53	47	10.2	14.8
East Denmark	77	53	47	10.5	15.5
Central Italy	41	54	46	10.4	15.4
South-west France	55	69	31	10.3	14.7
North-west Germany	64	56	44	10.1	14.4
Total	594	58	42	10.3	15.0

SPARCLE, Study of Participation of Children with Cerebral Palsy Living in Europe.

What this paper adds

- Motor function is generally stable in CP between childhood and adolescence.
- Additional impairments in CP are generally stable.
- Any changes are usually small.
- Most children who are seizure-free in childhood remain seizure-free in adolescence.

Any children for whom data on any type of impairment were missing were excluded from the analysis of that impairment.

Statistical analyses were performed using STATA software, Release 12 (StataCorp, College Station, TX, USA).

RESULTS

Figure 1 shows, for each impairment, the numbers of young people (represented by the size of the circles) at each combination of childhood level of impairment and adolescent level of impairment. If all children had remained at the same level of impairment in adolescence, all circles would be on the diagonal of each graph. In most children, level did not change, but some children showed either more or less impairment. In total, 70% of the children were assessed as the same GMFCS level on each occasion. The change in GMFCS level occurred mainly in children categorized as levels II to IV in childhood, with 46% of such children showing a change at adolescence. Of those originally classified as GMFCS levels I and V, only 15% and 4% respectively, showed a change in level. Of the 8- to 12-year-olds, 81% of the children were seizure-free, and 93% of these remained seizure-free in adolescence. Overall, 76% of the young people were seizure-free at both ages.

Table III shows, for each impairment, the percentage of children who remained stable between childhood and adolescence. For all impairments, most young people remained at the same level of impairment in childhood and adolescence. This stability of impairment was confirmed by the weighted kappa statistics. For gross motor function, communication, and cognitive level, both the estimated kappa and the lower bound of its 95% CI were above 0.75, indicating excellent agreement between impairment in childhood and adolescence. For fine motor function and feeding, the kappa was above 0.75 but the lower bound of the 95% CI was below this level, indicating some uncertainty about the strength of agreement. For seizures and vision, the kappa was below 0.75, although the upper bound of the 95% CI was above this level, indicating possible change in these impairments. For hearing, the kappa and its entire CI were below 0.75, suggesting change in this impairment.

The proportion that changed for the better was not significantly different from the proportion that changed for the worse for all impairments, except hearing (Table III). The significant (p=0.04) difference for hearing was based on only one child who needed hearing aids in childhood but not in adolescence and eight children who needed

Table II: Impairments and activity limitations in SPARCLE1 and SPARCLE2

	SPARCLE1, *n* (%)	SPARCLE2, *n* (%)
Gross motor function		
I. Walks without restrictions, limitations in more advanced gross motor skills	176 (30)	204 (34)
II. Walks without restrictions, limitations walking outdoors and in the community	132 (22)	105 (18)
III. Walks with assistive mobility devices, limitations walking outdoors and in the community	102 (17)	76 (13)
IV. Self-mobility with limitations, children are transported or use power mobility outdoors and in the community	85 (14)	78 (13)
V. Self-mobility is severely limited, even with the use of assistive technology	99 (17)	131 (22)
Bimanual fine motor function		
I. One hand manipulates without limitation; the other hand normally or with limitation in fine skills such as buttons, writing, knife, and fork	201 (34)	206 (35)
II. EITHER one hand manipulates without limitation; the other hand can grasp OR both hands are limited in fine skills such as buttons, writing, knife, and fork	162 (27)	136 (23)
III. Child needs help with tasks. EITHER one hand manipulates without limitation; the other hand can only hold or do even less OR one hand is limited in fine skills; the other hand can only grasp	95 (16)	109 (18)
IV. Child needs help and adapted equipment. EITHER both hands can only grasp OR one hand can grasp; other hand can only hold or do even less	71 (12)	73 (12)
V. Child always needs total human assistance, even with adaptations. Both hands can only hold or do even less	65 (11)	67 (11)
Missing	0 (0)	3 (1)
Seizures		
1. No seizures, no medication	427 (72)	418 (71)
2. No seizures, medication	55 (9)	63 (11)
3. Seizures <1 a month	48 (8)	47 (8)
4. Seizures >1 a month and <1 a week	32 (5)	21 (4)
5. Seizures >1 a week	32 (5)	39 (7)
Missing	0 (0)	6 (1)
Communication		
1. Normal	341 (57)	349 (59)
2. Difficulties, but uses speech	102 (17)	91 (15)
3. Uses non-speech for formal communication	73 (12)	77 (13)
4. No formal communication	78 (13)	73 (12)
Missing	0 (0)	4 (1)
Feeding		
1. No problems	429 (72)	448 (76)
2. Feeds orally with difficulty	131 (22)	99 (17)
3. Partial or complete non-oral feeding	34 (6)	44 (7)
Missing	0 (0)	3 (1)
Cognitive level		
1. IQ >70 Normal	289 (49)	274 (46)
2. IQ 50–70	138 (23)	154 (26)
3. IQ <50	162 (28)	165 (28)
Missing	5 (1)	1 (0)
Vision		
1. Useful vision	553 (93)	548 (92)
2. Blind or no useful vision	41 (7)	46 (8)
Hearing		
1. Does not need hearing aids	583 (98)	575 (97)
2. Needs hearing aids. Profound or severe loss, >70 decibels	10 (2)	18 (3)
Missing	1 (0)	1 (0)

SPARCLE, Study of Participation of Children with Cerebral Palsy Living in Europe.

hearing aids in adolescence but not in childhood. Changes in all impairments were in most cases of only one level. The proportion that remained stable was generally higher if the impairment was categorized on fewer levels. For all impairments, changes in level were more likely to occur in children who were in one of the intermediate levels in childhood than in those in the top or bottom level.

DISCUSSION

Although, by definition, the underlying lesion in CP is non-progressive, the consequences of CP may change over time. We found that the levels of impairment of most children did not change between childhood and adolescence (Table III) despite the physical growth, increased risk of contractures, and muscle stiffness, pain and fatigue of adolescence.[5] Among children whose impairments changed, most changed by only one level, and similar proportions of deterioration and improvement were recorded. A change in level was more likely in children with intermediate levels of impairment than in those in the top or bottom levels: an unremarkable finding because children with an intermediate level could change in either direction, whereas those in the top and bottom levels could change in only one direction. The proportion of children who changed level tended to be higher for those impairments with more levels: again unremarkable because a classification with more levels is likely to be more finely tuned.

Strengths and weaknesses

The participants from eight regions were sampled from population-based registers, and in one region the participants came from multiple sources. The SPARCLE sample is large. Although all research associates received joint training, home visits were not undertaken by the same person on each occasion. The evaluation of gross and fine motor function was made by the research associate in conjunction with a parent. Parents can reliably classify their child to a GMFCS level.[12] Information on the additional impairments was reported by parents.

The GMFCS is a valid and reliable classification of gross motor function for children and adolescents.[13] Validation of the BFMF classification of hand function was based on a review of the literature and the experience of experts, but its

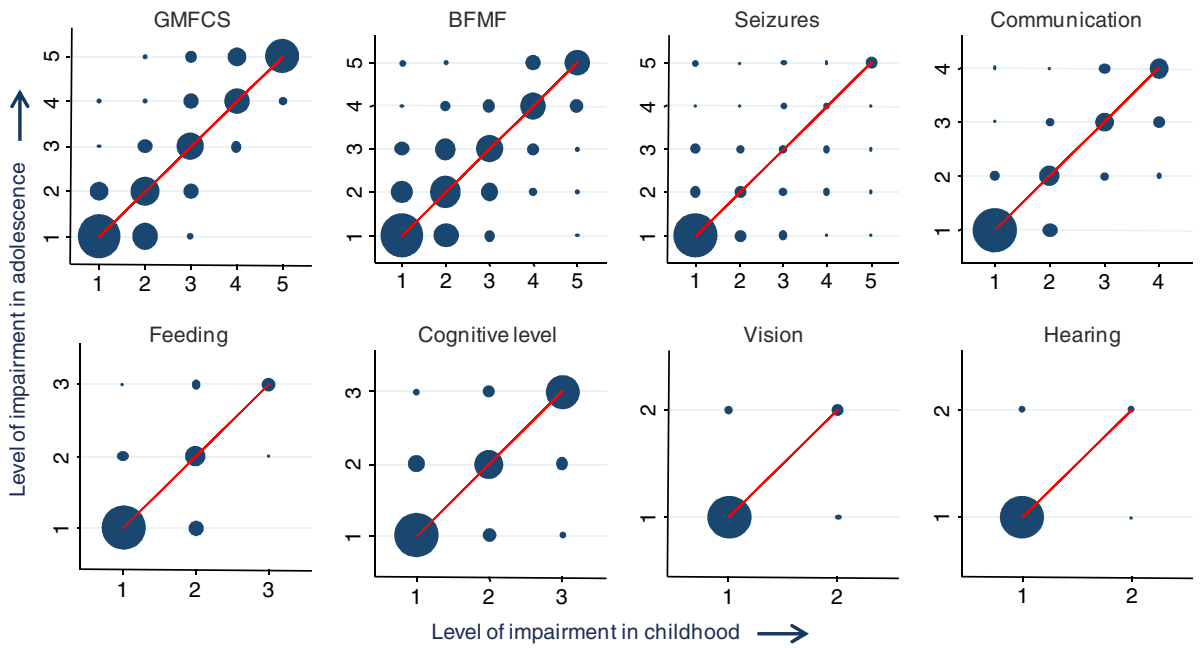

Figure 1: Number of children by level of impairment in childhood (SPARCLE1) and adolescence (SPARCLE2). SPARCLE, Study of Participation of Children with Cerebral Palsy Living in Europe; GMFCS, Gross Motor Function Classification System; BFMF, Bimanual Fine Motor Function.

Table III: Stability of impairment between childhood and adolescence

				Kappa		% who changed for:			% who changed:		% who moved from:		
Impairment	Nr of levels	*n*[a]	Overall % who remained stable	Mean	95% CI	Better	Worse	*p*[b]	One level	Two levels or more	Top level	Middle levels	Bottom level
Gross motor function	5	594	70	0.91	0.83–0.99	14	16	0.26	28	3	15	46	4
Bimanual fine motor function	5	591	63	0.82	0.74–0.90	17	19	0.50	30	7	26	46	22
Seizures	5	588	76	0.73	0.65–0.81	13	11	0.59	14	10	10	69	25
Communication	4	590	82	0.90	0.82–0.98	10	7	0.08	16	1	6	36	29
Feeding	3	591	86	0.79	0.71–0.87	8	6	0.26	14	0	6	44	9
Cognitive level	3	588	83	0.85	0.77–0.93	7	10	0.07	16	1	15	28	12
Vision	2	594	96	0.71	0.63–0.80	2	2	0.40	4	–	3	–	22
Hearing	2	592	98	0.66	0.58–0.74	0	1	0.04	2	–	1	–	10

[a]Number of adolescents for whom this impairment was recorded in both SPARCLE1 and SPARCLE2. [b]Significance of difference between percentages that changed for better and for worse.

reliability has not been reported.[14] If the reliability of a measure is lower, the estimated kappa is likely to be lower and its 95% CI is likely to be wider. The lower bound of the estimated kappa for BFMF was 0.74, indicating some uncertainty about the agreement between childhood and adolescent classification, which could reflect either unsatisfactory reliability of the measure or true change. The BFMF classification has been used in research and in registers, such as the Surveillance of Cerebral Palsy in Europe (SCPE),[1] but, in clinical practice, the Manual Ability Classification system (MACS)[15] is often used. When the SPARCLE study began, the MACS was not available.

There is now a standardized classification of communication in children with CP, but it, also, was not available when SPARCLE began.[16]

Although kappa has been used to analyse agreement while allowing for chance,[17,18] it has the disadvantage that its value depends on the proportion of participants in each category.[19] Hence, it is misleading to compare kappas from different studies, where the prevalence of the categories differs.

Although only 594 out of the 818 (73%) families who participated in SPARCLE1 also participated in SPARCLE2, overall, non-response did not vary significantly with any type of impairment.[11] Therefore, it is unlikely that our estimates of the kappa would have been different if all the original families had been followed up.

Comparisons with other studies

The GMFCS[13] has five levels and emphasizes function in sitting and walking. In population-based studies from wes-

tern Sweden, approximately 65% of children with CP walked without restrictions (GMFCS levels I–II),[14,20] and at these levels motor function did not generally change over time. However, for GMFCS levels III to V there was a peak of motor function around 6 to 8 years of age before decline occurred.[4,5] Gains in gross motor function in childhood and declines in adolescence and young adulthood, especially in non-walking children, have been described in large samples.[3,5,17] In our study, 70% of the children remained on the same GMFCS level over the 4-year period between visits. Palisano et al.[17] studied stability of the GMFCS in a sample of 610 children, aged 16 months to 13 years at baseline, who were followed up for between 6 months and 52 months (average 33mo) and assessed two to seven times at 6- to 12-month intervals. They found that GMFCS level was the same on all occasions in 73%, and had changed by one level in 16%, by two levels in 8.4%, by three levels in 2.5%, and by four levels in 0.2% at any of the assessments. These results are similar to ours, although the distribution of GMFCS levels in their sample was significantly ($p<0.05$) different from that in our sample.

Stability of hand function in children with CP has been studied much less frequently. Holmefur et al.[21] assessed 1- to 5-year-old children with unilateral CP several times and concluded that the development of manual skills in CP does not follow the same course as gross motor function. For the majority of the adolescents in our study, BFMF remained the same as in childhood.

Although the studies have been few, it seems that hand function, speech, vision, hearing, and cognition are stable over time.[6,7]

In a 15-year follow-up of children with epilepsy by Geerts et al.,[22] non-idiopathic epilepsy, which is usually the type in children with CP, and young age at onset were associated with a poorer seizure outcome. In our study, it is encouraging that most children without seizures (with or without medication) in SPARCLE1 were still seizure-free as adolescents. On the other hand, three-quarters of the children with weekly seizures still had the same seizure frequency as adolescents. There seems to be a large group of children with CP that may never develop seizures, and a small group whose seizures are difficult to control.

CONCLUSION

We found that motor function and associated impairments were, in general, stable between childhood and adolescence in young people with CP.

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