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ORIGINAL ARTICLE

Prevalence, birth, and clinical characteristics of dyskinetic cerebral palsy compared with spastic cerebral palsy subtypes: A Norwegian register-based study

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Abstract

Aim: To study the prevalence, birth, and clinical characteristics of children with dyskinetic cerebral palsy (CP) in Norway compared with spastic quadriplegic CP and other spastic CP subtypes.

Method: Data on children born from 1996 to 2015 were collected from the Norwegian Quality and Surveillance Registry for Cerebral Palsy and the Medical Birth Registry of Norway.

Results: One hundred and seventy (6.8%) children had dyskinetic CP. The birth prevalence decreased during 1996 to 2015 from 0.21 to 0.07 per 1000 livebirths ($p < 0.001$). Dyskinetic CP was more often associated with term/post-term birth, and motor and associated impairments were more severe compared with spastic bilateral and unilateral CP, but less severe than spastic quadriplegic CP. On neuroimaging, grey matter injuries were most prevalent in dyskinetic CP (mainly basal ganglia/thalamus) and spastic quadriplegic CP (mainly cortico-subcortical), white matter injuries in spastic bilateral, and white and grey matter injuries were equally common in spastic unilateral CP. Normal neuroimaging and brain maldevelopment were present in 25% of children with dyskinetic CP.

Interpretation: The decrease in birth prevalence of dyskinetic CP was probably due to improved antenatal and perinatal care. Potential sentinel events at term were more common in dyskinetic CP than other spastic CP subtypes. However, probable antenatal aetiologies were most prevalent. Motor and associated impairments were less severe in children with dyskinetic CP compared with spastic quadriplegic CP.

Dyskinetic cerebral palsy (CP) is a rare CP subtype, accounting for 6% to 17% of children with CP.[1,2] Motor impairments are typically more severe and associated impairments more common in children with dyskinetic CP compared with spastic CP.[3,4] Sentinel events during delivery are well-known risk factors for dyskinetic CP,[5] and on neuroimaging basal ganglia/thalamus lesions are characteristic.[4,5]

Although the overall birth prevalence of CP has decreased during the past two decades in high-income countries,[6] the prevalence of dyskinetic CP in Europe increased from 0.08 per 1000 livebirths in the 1970s to 0.14 in the 1990s.[7] In western Sweden, dyskinetic CP increased from 0.14 per 1000 livebirths in 1976 to 0.25 in 2005, followed by a trend towards decrease in 2010.[8] A similar trend was observed in a Norwegian study.[9] However, whether the decrease in the prevalence of dyskinetic CP during 2005 to 2010 was simply a fluctuation or whether it has continued to decrease is unknown.

The Surveillance of Cerebral Palsy in Europe (SCPE) classification of CP subtypes includes two subtypes characterized

Abbreviations: MACS, Manual Ability Classification System; NorCP, Norwegian Quality and Surveillance Registry for Cerebral Palsy; SCPE, Surveillance of Cerebral Palsy in Europe..

by spasticity as the dominating symptom: spastic unilateral and spastic bilateral. In contrast, the International Statistical Classification of Diseases and Related Health Problems, 10th Revision (ICD-10) includes three subtypes of spastic CP: hemiplegic, diplegic, and quadriplegic, whereby the last two are included in the SCPE spastic bilateral subtype. However, while spastic diplegic CP, the most prevalent spastic bilateral subtype, is associated with preterm birth and less severe fine motor impairments than dyskinetic CP, spastic quadriplegic CP shares many features with dyskinetic CP. The prevalence of spastic quadriplegic CP has been stable around 0.15 per 1000 livebirths,[1,8,10,11] and motor and associated impairments are severe. Sentinel events during delivery are risk factors for both dyskinetic CP and spastic quadriplegic CP.[5]

The increasing birth prevalence of dyskinetic CP and stable prevalence of spastic quadriplegic CP towards the end of the 20th century,[7] despite considerable improvements in obstetric and perinatal care, have led some authors to conclude that these CP subtypes are mainly antenatal in origin and can rarely be prevented.[13] In 1998, an international consensus statement defined dyskinetic CP or spastic quadriplegic CP as essential criteria in defining an acute intrapartum event.[13] However, perinatal health has improved and the overall prevalence of CP has continued to decrease during the first decade of the 21st century, suggesting improved antenatal, perinatal, and neonatal care.[9] Therefore, updated information on the birth prevalence, birth characteristics, and clinical characteristics of dyskinetic CP and spastic CP, in particular spastic quadriplegic CP, was of interest.

Considering this background, our aims were to update the birth prevalence of dyskinetic CP in Norway, including temporal trends, and to describe birth and clinical characteristics. An additional aim was to compare the findings with children with spastic CP, primarily spastic quadriplegic CP.

What this paper adds

- Birth prevalence of those with dyskinetic and spastic bilateral cerebral palsy (CP) in Norway decreased between 1996 and 2015.
- Potential sentinel events at term were more common in dyskinetic CP.
- Nonetheless, probable antenatal aetiologies were most prevalent in dyskinetic CP.
- Basal ganglia/thalamus lesions were more common in dyskinetic than spastic quadriplegic CP.
- Motor and associated impairments were milder in dyskinetic than spastic quadriplegic CP.

METHOD

Study design and participants

Children with CP born from 1996 to 2015 and registered in the Norwegian Quality and Surveillance Registry for Cerebral Palsy (NorCP) as of 31st December 2020 were included. The NorCP is a national consent-based medical quality registry collecting information on children with CP born from 1996 onwards. Paediatricians at all 21 habilitation centres in Norway record data at the time of diagnosis, at age 5 years, and at age 15 to 17 years.[14] NorCP includes clinical data on approximately 90% of all children with CP born during the study period, and aggregated data per CP subtype and birth year for the remaining percentage from four validation studies.[14,15] Data from the NorCP Five Year Registration Form were linked to the Medical Birth Registry of Norway using unique 11-digit identification numbers. The Medical Birth Registry of Norway is a mandatory national health registry collecting pre-pregnancy, antenatal, perinatal, and neonatal information on all deliveries in Norway since 1967.[16]

Variables

CP was defined according to Rosenbaum et al.,[17] and CP subtypes according to SCPE guidelines (confirmed at age 5 years).[18] However, to address our additional aims, spastic bilateral CP was further categorized into spastic diplegic (ICD-10 code G80.1) and quadriplegic (ICD-10 code G80.0) subtypes. The use of ICD-10 codes is universally mandatory in Norway, and, in general, Norwegian neuropaediatricians follow the definitions as indicated in textbooks (i.e. spastic diplegic CP is characterized by spasticity predominantly affecting the lower extremities more than upper extremities, whereby spastic quadriplegic CP is characterized by spasticity of all four limbs).[19] Neuroimaging findings were classified by the reporting paediatrician according to pre-defined categories on the registration form in use since birth year 1999, and later recoded according to the main categories of the Magnetic Resonance Imaging Classification System.[12] Gross and fine motor function were classified using the Gross Motor Function Classification System (GMFCS)[20] and Manual Ability Classification System (MACS).[21] Although the registration form in use for children born from 1996 to 1998 did not include the GMFCS, their gross motor function was retrospectively classified into levels I to II, III, IV, and V.[18] Information on MACS was only available from birth year 1999 onwards. Associated impairments were classified as epilepsy by the current use of antiseizure medication, severe vision impairment as blind less than 6/60 (<0.1) after correction on the best eye, and severe hearing impairment as loss greater than 70 dB before correction in the best ear. The need for gastrostomy tube feeding was used as a proxy for severe eating difficulties and speech impairment was classified according to the Viking Speech Scale.[22] Cognitive function was classified as normal, learning disorder (ICD-10 F80–81, F83), intellectual disability (ICD-10 F70–73, F79) or clinical assessment, and non-testable.

Gestational age was categorized into less than 28 weeks, 28 to 31 weeks, 32 to 36 weeks, 37 to 41 weeks, and more than 41 weeks. Preeclampsia was defined as new-onset maternal hypertension (blood pressure ≥ 140/90 mmHg) after the

20th week of pregnancy and new-onset proteinuria and/or other new-onset organ damage. Pregnancies with more than one fetus were categorized as multiple births. Birth position was classified as cephalic or non-cephalic (i.e. breech, transverse, abnormal, and other), and birthweight was categorized as less than, equal to, or more than 2500 g. Apgar scores at 5 minutes were categorized into 0 to 3, 4 to 6, and 7 to 10. Complications during labour and delivery including abruptio placentae, uterine rupture, umbilical cord complications, and brachial plexus birth palsy/clavicle fracture were defined as yes/no. 'Potential sentinel events' were defined as one (or more) complications during labour and delivery (all gestational ages and > 36 weeks). However, as umbilical cord complications do not necessarily imply a sentinel event, it was combined with an Apgar score at 5 minutes of less than 7.

Statistical analyses

Differences in proportions between CP subtypes were analysed using Pearson's χ^2 test and linear-by-linear association for continuous variables, or with Fischer's exact test where appropriate. Birth prevalence was calculated by dividing the total number of children with CP in each CP subtype by the total number of livebirths, per birth year (Table S1). Logistic regression with birth year as a covariate was used to estimate time trends in the birth prevalence of dyskinetic CP and spastic CP per 1000 livebirths for children born from 1996 to 2015. Nonlinear trends were accounted for by using fractional polynomials with birth year as a covariate.[23] The number of livebirths was retrieved from the Medical Birth Registry of Norway statistics bank.[16] Children born abroad and/or with postneonatally acquired CP, occurring after 28 days of life and up to 2 years of age, were excluded from analyses of prevalence and birth characteristics. To explore the validity of spastic bilateral CP (diplegic vs quadriplegic), we studied the distribution of manual performance (MACS levels I–III vs MACS levels IV–V). Any p-values less than 0.05 were considered statistically significant, and 95% confidence intervals (CIs) reported where relevant. Missing data are reported in or under each table. Analyses were performed using SPSS version 26 (IBM Corp, Armonk, NY, USA), with the exception of logistic regression using Stata version 16 (StatCorp, College Station, TX, USA).

Ethics

NorCP is governed by the Regulations of Medical Quality Registers in Norway and is consent-based. The Regional Committee for Medical Research Ethics Central Norway approved this study (2011/754).

RESULTS

The total number of livebirths in Norway from 1996 to 2015 was 1 187 688. Among these, 2540 children were registered in the NorCP. A registration form had not yet been received for 45 children. Thus 2495 children (57% males) were included in the study, and of these 1039 (41.6%) had spastic unilateral CP, 1131 (45.3%) spastic bilateral CP, 170 (6.8%) dyskinetic CP, and 104 (4.2%) ataxic CP, while in 51 (2.0%) children the subtype was unspecified. According to the ICD-10 classification, 778 (31.2%) had spastic diplegic CP and 353 (14.1%) had spastic quadriplegic CP. The validity analyses of the last two subtypes showed that 96% of children with spastic diplegic CP were classified in MACS levels I to III, while 89% with spastic quadriplegic CP were in MACS levels IV or V ($\kappa = 0.85$) (Table S2). Among the 2340 children with spastic and dyskinetic CP, 323 were born abroad and/or with postneonatal acquired CP. Figure S1 shows the total number of children with CP included/excluded in each analysis.

Prevalence

The overall birth prevalence of spastic and dyskinetic CP in Norway from 1996 to 2015 was 2.04 per 1000 livebirths (95% CI 1.96–2.12). The prevalence of dyskinetic CP was 0.16 per 1000 livebirths (95% CI 0.14–0.18), decreasing from an average 0.21 in 1996 to 2006 to 0.10 in 2007 to 2015 (Figure 1). Assuming a linear model, the annual reduction was 4.5% ($p < 0.001$). In comparison, the prevalence of spastic quadriplegic CP was 0.30 per 1000 livebirths (95% CI 0.27–0.33), decreasing from an average 0.40 in 1996 to 2006 to 0.18 in 2007 to 2015 (5.4% annual reduction; $p < 0.001$) (Figure 1). The prevalence of spastic diplegic CP was 0.68 per 1000 livebirths (95% CI 0.64–0.73), decreasing from an average 0.83 in 1996 to 2006 to 0.50 in 2007 to 2015 (4.5% annual reduction; $p < 0.001$), while spastic unilateral CP held stable at around 0.90 per 1000 livebirths (95% CI 0.85–0.95) (Figure 1).

Birth characteristics

A higher proportion of children with dyskinetic CP were born at term or later, with a birthweight of at least 2500 g, and after uterine rupture compared with spastic quadriplegic CP (Table 1). Compared with spastic diplegic CP, term birth, low Apgar scores at 5 minutes, uterine rupture, brachial plexus birth palsy/clavicle fracture, umbilical cord complications, and potential sentinel events (including gestational age > 36 weeks) were more common among children with dyskinetic CP (Table 1).

Clinical characteristics

Neuroimaging was available in 1501 (72.6%) of the 2068 children with dyskinetic CP and spastic CP born from 1999 to 2015 (Table 2). Of these, the proportion of children with brain maldevelopment did not differ between the CP subtypes, while white matter injuries were less common among children with dyskinetic CP compared with

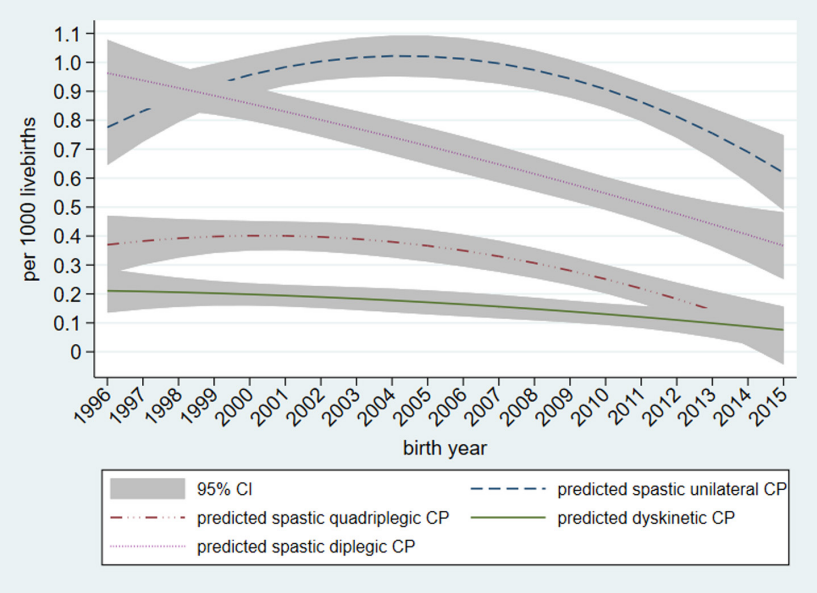

FIGURE 1 Trends in the birth prevalence of dyskinetic, spastic bilateral cerebral palsy (CP) (diplegic and quadriplegic), and unilateral CP in Norway. Each line represents predicted CP prevalence using logistic regression with fractional polynomials, and the shaded area indicates 95% confidence intervals (CI).

spastic quadriplegic CP and the other spastic CP subtypes. Grey matter injuries were present in about 50% of children with dyskinetic CP and between 43% and 45% in children with spastic quadriplegic CP or spastic unilateral CP, but only in 15% with spastic diplegic CP. Within the subcategories of grey matter injuries, basal ganglia/thalamus lesions were most prevalent in children with dyskinetic CP, cortico-subcortical lesions in children with spastic quadriplegic CP, and arterial infarctions in children with spastic unilateral CP. Nineteen children with dyskinetic CP (16%) had normal brain MRI findings, similar to children with spastic diplegic CP, which was significantly higher than in children with spastic quadriplegic CP and spastic unilateral CP (Table 2). Eleven children (9.3%) with dyskinetic CP had multiple injuries according to the main Magnetic Resonance Imaging Classification System categories, compared with 19 (8.1%) with spastic quadriplegic CP ($p = 0.840$), 15 (3.2%) with spastic diplegic CP ($p = 0.007$), and 31 (4.6%) with spastic unilateral CP ($p = 0.058$) (Table S3).

Children with dyskinetic CP were more often able to walk without assistive devices and handle objects with their hands independently (GMFCS levels I and II, MACS levels I and II) (Table 3), and had less associated impairments compared with children with spastic quadriplegic CP (Table 4). This included normal cognitive abilities, although 19% of children with dyskinetic CP and 36% with spastic quadriplegic CP were considered non-testable and 31% and 24% respectively had missing data. Compared with children with spastic unilateral and diplegic CP, children with dyskinetic CP had more severe motor and associated impairments (Tables 3 and 4).

DISCUSSION

The birth prevalence of children with dyskinetic CP in Norway born from 1996 to 2015 was low and decreased significantly. Most were born as singletons, near term with birthweight greater than 2500 g. Among those born near term or later, one in four had evidence of a potential sentinel event at delivery. On MRI, grey matter injuries were present in half of the children with dyskinetic CP, and among these basal ganglia/thalamus lesions were the dominating lesion, present in four of five children, consistent with an insult near term.[12] However, neuroimaging results (i.e. maldevelopment, white matter, and normal findings) also suggested that the insult was of antenatal origin in about half of the children with dyskinetic CP.[12] Motor impairments were in general severe and associated impairments were common, although about 20% of children with dyskinetic CP were able to walk independently (GMFCS levels I–II), and 29% used their hands independently (with adaptation in some situations) (MACS levels I–III).

Compared with children with spastic quadriplegic CP, children with dyskinetic CP were more often born around term, when potential sentinel events occurred more often. On neuroimaging, white matter injuries were less common, while basal ganglia/thalamus lesions, but also normal findings, were more common in children with dyskinetic CP. Finally, motor impairments were less severe and associated impairments were less common when compared with spastic quadriplegic CP.

Among children with spastic unilateral and dyskinetic CP, white matter injuries and evidence of stroke were more common, while potential sentinel events were less common

TABLE 1 Birth characteristics among children with dyskinetic CP compared with spastic bilateral (quadriplegic and diplegic) and unilateral CP born in Norway between 1996 and 2015.

SCPE CP subtypes			Spastic bilateral								
ICD-10 CP subtypes	Dyskinetic		Quadriplegic			Diplegic			Spastic unilateral		
	n	%	*n*	%	*p*[a]	*n*	%	*p*[b]	*n*	%	*p*[c]
Sex											
Male	80	54.8	174	61.3		414	60.4		513	56.9	
Female	66	45.2	110	38.7		271	39.6		389	43.1	
Total	146	100	284	100	0.196	685	100	0.207	902	100	0.638
Gestational age											
<28 weeks	9	6.3	23	8.3		100	14.9		62	7.1	
28–31 weeks	7	4.9	40	14.4		167	24.9		83	9.5	
32–36 weeks	21	14.7	41	14.7		155	23.1		128	14.6	
37–41 weeks	93	65.0	160	57.6		227	33.8		543	61.8	
≥42 weeks	13	9.1	14	5.0		22	3.3		62	7.1	
Total	143	100	278	100	0.006	671	100	<0.001	878	100	0.137
Missing	3	2.1	6	2.1		14	2.0		24	3.0	
Multiple birth	12	8.2	40	14.1		115	16,8		99	11.0	
Singleton	134	91.8	244	85.9		570	83.2		803	89.0	
Total	146	100	284	100	0.077	685	100	0.009	902	100	0.315
Preeclampsia											
Yes	7	4.8	17	6.0		62	9.1		66	7.3	
No	139	95.2	267	94.0		623	90.9		836	92.7	
Total	146	100	284	100	0.610	685	100	0.091	902	100	0.267
Cephalic presentation											
Yes	113	77.4	235	82.7		529	77.2		739	81.9	
No	33	22.6	49	17.3		156	22.8		163	18.1	
Total	146	100	284	100	0.181	685	100	0.964	902	100	0.193
Birthweight											
<2500 g	31	21.5	106	38.0		407	60.3		264	29.5	
≥2500 g	113	78.5	173	62.0		268	39.7		631	70.5	
Total	144	100	279	100	0.001	675	100	<0.001	895	100	0.049
Missing	2	1.4	5	1.8		10	1.5		7	0.8	
Apgar score at 5 minutes											
0–3	33	23.4	56	20.1		38	5.7		20	2.3	
4–6	30	21.3	53	19.1		105	15.6		67	7.6	
7–10	78	55.3	169	60.8		528	78.7		797	90.2	
Total	141	100	278	100	0.556	671	100	<0.001	884	100	<0.001
Missing	5	3.4	6	2.1		14	2.0		18	2.0	
Abruptio placentae											
Yes	6	4.1	20	7.0		38	5.5		29	3.2	
No	140	95.9	264	93.0		647	94.5		873	96.8	
Total	146	100	284	100	0.227	685	100	0.481	902	100	0.577
Uterine rupture											
Yes	5	3.4	2	0.7		1	0.1		0	0	
No	141	96.6	282	99.3		684	99.9		902	100	
Total	146	100	284	100	0.048	685	100	<0.001	902	100	<0.001

(Continues)

TABLE 1 (Continued)

SCPE CP subtypes			Spastic bilateral								
ICD-10 CP subtypes	Dyskinetic		Quadriplegic			Diplegic			Spastic unilateral		
	n	%	*n*	%	*p*[a]	*n*	%	*p*[b]	*n*	%	*p*[c]
BPBP/clavicle fracture											
Yes	4	2.7	2	0.7		3	0.4		7	0.8	
No	142	97.3	282	99.3		682	99.6		895	99.2	
Total	146	100	284	100	0.186	685	100	0.006	902	100	0.031
Umbilical cord complications											
Yes	31	21.2	49	17.3		86	12.6		147	16.3	
No	115	78.8	235	82.7		599	87.4		755	83.7	
Total	146	100	284	100	0.315	685	100	0.006	902	100	0.141
Potential sentinel events,[d] all gestational ages											
Yes	27	18.5	41	14.4		54	7.9		47	5.2	
No	119	81.5	243	85.6		631	92.1		855	94.8	
Total	146	100	284	100	0.275	685	100	<0.001	902	100	<0.001
Potential sentinel events,[d] gestational age > 36 weeks											
Yes	25	17.1	26	9.2		15	2.2		20	2.2	
No	121	82.9	258	90.8		670	97.8		882	97.8	
Total	146	100	284	100	0.016	685	100	<0.001	902	100	<0.001

Abbreviations: CP, cerebral palsy; BPBP, brachial plexus birth palsy; ICD-10, International Statistical Classification of Diseases and Related Health Problems, 10th Revision; SCPE, Surveillance of Cerebral Palsy in Europe.

[a]Comparison of dyskinetic and spastic quadriplegic CP.

[b]Comparison of dyskinetic and spastic diplegic CP.

[c]Comparison of dyskinetic and spastic unilateral CP.

[d]Potential sentinel events is the presence of one (or more) of the following complications: abruptio placentae, uterine rupture, BPBP/clavicle fracture, or an umbilical cord complication in a newborn with Apgar scores at 5 minutes <7.

and motor and associated impairments were less severe than among children with dyskinetic CP.

The strengths of the study are the prospectively recorded information, the high number of children within each CP subtype, and the documented high quality of data in the NorCP.[14,15] The classification of the main CP subtypes and motor and associated impairments were based on SCPE guidelines and validated scales. However, the ICD-10 CP subtypes are less well defined, and therefore some misclassification of spastic diplegic CP as spastic quadriplegic CP and vice versa is likely. Also, in the instance when dyskinesia (involuntary movements) and spasticity (increased muscle tone) coexisted, the neuropaediatricians needed to determine the child's CP subtype based on the dominating feature. On the other hand, explorative analyses suggested it to be unlikely that such misclassification could explain the main findings of the secondary aim of this study. The results for cognitive ability should be interpreted with caution because a large proportion of children with dyskinetic CP or spastic quadriplegic CP were either not appropriately assessed (non-testable) or had missing data. Caution is also required when interpreting the results about complications during delivery and potential sentinel events. While abruptio placentae and uterine rupture are clear evidence of potential sentinel events, umbilical cord complications may not necessarily lead to a sentinel event. Therefore, umbilical cord complications were included as a potential sentinel event only when the child was born with a low Apgar score at 5 minutes. Additionally, because shoulder dystocia was not collected in the Medical Birth Registry of Norway, we included brachial plexus birth palsy and clavicle fracture as proxies. Although potential sentinel events should be interpreted with caution, our results are unlikely to be due to systematic misclassification between the CP subtypes. A further limitation is that information on the use of therapeutic hypothermia was not available.

The birth prevalence of dyskinetic CP in this study was 0.16 per 1000 livebirths, identical to a study of Danish children born from 1999 to 2007,[3] and similar to an estimated 0.17 in Icelandic children born from 1999 to 2003.[24] However, the prevalence of dyskinetic CP was significantly lower than in western Sweden, around 0.25 to 0.30 per 1000 in children born from 1990 to 2010.[8] The higher birth prevalence of dyskinetic CP in western Sweden compared with the three other Nordic countries is difficult to explain, considering the similar healthcare systems and that all are SCPE partners. However, it may be noted that in our study fewer children with dyskinetic CP had neuroimaging findings consistent with hypoxic–ischaemic events at term than in two studies in western Sweden.[8,25] The prevalence of dyskinetic CP was lower, around 0.11 per 1000, in a study of Canadian children born from 1999 to 2002.[1]

TABLE 2 MRICS results in children with dyskinetic CP compared with spastic bilateral (quadriplegic and diplegic) and unilateral CP in Norway, born between 1999 and 2015.

SCPE CP subtypes			Spastic bilateral								
ICD-10 CP subtypes	Dyskinetic		Quadriplegic			Diplegic			Spastic unilateral		
	n	%	*n*	%	*p*[a]	*n*	%	*p*[b]	*n*	%	*p*[c]
Children included in study	151	6.9	307	14.0		669	30.5		941	43.0	
Children with MRI	118	78.1	234	76.2		475	71.0		674	71.6	
MRICS categories[d]											
A Maldevelopment											
Yes	11	9.3	26	11.1		37	7.8		56	8.3	
No	107	90.7	208	88.9	0.605	438	92.2	0.585	618	91.7	0.715
B White matter injury											
Yes	32	27.1	88	37.6		277	58.3		305	45.3	
No	86	72.9	146	62.4	0.050	198	41.7	<0.001	369	54.7	<0.001
C Grey matter injury											
Yes	63	53.4	106	45.3		73	15.4		291	43.2	
No	55	46.6	128	54.7	0.151	402	84.6	<0.001	383	56.8	0.040
D Miscellaneous											
Yes	5	4.2	13	5.6		15	3.2		14	2.1	
No	113	95.8	221	94.9	0.596	460	96.8	0.561	660	97.9	0.157
E Normal											
Yes	19	16.1	20	8.5		89	18.7		39	5.8	
No	99	83.9	214	91.5	0.033	386	81.3	0.507	635	94.2	<0.001
MRICS subcategories of grey matter injuries[d]											
C1 Basal ganglia/thalamus lesions											
Yes	48	78.7	35	36.5		25	33.8		59	20.1	
No	13	21.3	61	63.5	<0.001	49	66.2	<0.001	234	79.9	<0.001
C2 Cortico-subcortical lesions											
Yes	18	29.5	70	72.9		29	39.2		41	14.0	
No	43	70.5	26	27.1	<0.001	45	60.8	0.240	252	86.0	0.003
C3 Arterial infarctions											
Yes	6	9.8	17	17.7		26	35.1		215	73.4	
No	55	90.2	79	82.3	0.174	48	64.9	<0.001	78	26.6	<0.001

Abbreviations: CP, cerebral palsy; ICD-10, International Statistical Classification of Diseases and Related Health Problems, 10th Revision; MRICS, Magnetic Resonance Imaging Classification System; SCPE, Surveillance of Cerebral Palsy in Europe.

[a]Comparison of dyskinetic and spastic quadriplegic CP.

[b]Comparison of dyskinetic and spastic diplegic CP.

[c]Comparison of dyskinetic and spastic unilateral CP.

[d]Seventy-six of 1501 (5.1%) children had multiple injuries of the main categories, and 65 of 524 (12.4%) with grey matter injuries had multiple grey matter lesions. For details, see Table S2.

TABLE 3 GMFCS and MACS levels among children with dyskinetic CP compared with spastic bilateral (quadriplegic and diplegic) and unilateral CP in Norway, born between 1996 and 2015.

SCPE CP subtypes			Spastic bilateral								
ICD-10 CP subtypes	Dyskinetic		Quadriplegic			Diplegic			Spastic unilateral		
	n	%	*n*	%	*p*[a]	*n*	%	*p*[b]	*n*	%	*p*[c]
GMFCS level[d]											
I–II	20	12.1	8	2.3		501	66.4		990	97.2	
III	16	9.7	12	3.5		147	19.5		13	1.3	
IV	54	32.7	78	22.9		99	13.1		4	0.4	
V	75	45.5	243	71.3		7	0.9		11	1.1	
Total	165	100	341	100	<0.001	754	100	<0.001	1018	100	<0.001
Missing	5	2.9	12	3.4		24	3.1		21	2.0	
MACS level[e]											
I	2	1.4	3	1.1		322	53.6		435	49.5	
II	15	10.8	9	3.4		162	27.0		340	38.7	
III	23	16.5	17	6.4		92	15.3		95	10.8	
IV	34	24.5	58	21.7		21	3.5		9	1.0	
V	65	46.8	180	67.4		4	0.7		0	0.0	
Total	139	100	267	100	<0.001	601	100	<0.001	879	100	<0.001
Missing	12	7.9	40	13.0		68	10.2		62	6.6	

Abbreviations: CP, cerebral palsy; GMFCS, Gross Motor Function Classification System; ICD-10, International Statistical Classification of Diseases and Related Health Problems, 10th Revision; MACS, Manual Ability Classification System; SCPE, Surveillance of Cerebral Palsy in Europe.

[a]Comparison of dyskinetic and spastic quadriplegic CP.

[b]Comparison of dyskinetic and spastic diplegic CP.

[c]Comparison of dyskinetic and spastic unilateral CP.

[d]Born 1996–2015.

[e]Born 1999–2015.

The birth prevalence of dyskinetic CP in Norway decreased during the study period. In a previous study of children with CP born in Norway from 1996 to 2010, the birth prevalence of dyskinetic CP held relatively stable around 0.2 per 1000 livebirths until 2006/2007, followed by a decrease towards 2010. A similar decreasing trend towards 2010 was also reported in Sweden.[8] Our results confirm a continued declining trend in Norway.

Regarding birth characteristics, our results are largely in line with other European studies.[3,7,26] On neuroimaging, the proportions of children with dyskinetic CP with grey matter (53.1%) or white matter (27.1%) injuries in our study were somewhat higher than in a European study including children born from 1999 to 2009 (39% and 21% respectively). The proportions with maldevelopments (13%), miscellaneous (11%), and normal findings (17%) were slightly higher in the European study than in ours.[27] However, the authors in the European study did not describe grey matter lesions in detail.[27]

The distribution of gross motor and associated impairments was also largely consistent with results in another European study of children with dyskinetic CP born from 1990 to 2006.[26] Typical cognitive abilities were reported in 30% of children with dyskinetic CP in the European study, slightly lower than the 36% in our study. However, our study had 31% missing cognition results, compared with 16.5% in the European study.

The decrease in birth prevalence of dyskinetic CP is most likely due to general improvements in antenatal, obstetric, and neonatal care. However, it is noteworthy that, around 2007, Scandinavian guidelines recommended that improved ST segment analysis techniques could provide better surveillance of the infant during delivery,[28] and Norwegian guidelines recommended the use of therapeutic hypothermia as the treatment of choice for newborn infants with mild to severe hypoxic–ischaemic encephalopathy.[29] An alternative explanation could be a systematic change over time in the classification of CP subtypes. However, this explanation is less likely because we found that the prevalence of both spastic bilateral quadriplegic and diplegic CP decreased significantly during the study period.

Grey matter injuries, in particular basal ganglia/thalamus and cortico-subcortical lesions, were common in children with dyskinetic CP. The basal ganglia/thalamus are important areas for learning, automating, and modulating movements through interaction with other subcortical areas, different parts of the cerebral cortex, and cerebellum.[30] Thus, the lesions in these regions are reasonable explanations for the movement disorder seen in dyskinetic CP. Moreover, owing to high metabolic requirements, these regions are particularly vulnerable to hypoxaemia at term.[31] Our finding that, among children with dyskinetic CP born around term, 40% had basal ganglia/thalamus lesions,

TABLE 4 Associated impairments among children with dyskinetic CP compared with spastic bilateral (quadriplegic and diplegic) and unilateral CP in Norway, born between 1996 and 2015.

SCPE CP subtypes			Spastic bilateral								
ICD-10 CP subtypes	Dyskinetic		Quadriplegic			Diplegic			Spastic unilateral		
	n	%	*n*	%	*p*[a]	*n*	%	*p*[b]	*n*	%	*p*[c]
Total children	170	6.8	353	14.1		778			1039	41.6	
Epilepsy											
Yes	55	37.2	210	67.7		100	14.3		145	15.1	
No	93	62.8	100	32.3		597	85.7		813	84.9	
Total	148	100	310	100	<0.001	697	100	<0.001	958	100	<0.001
Missing	22	12.9	43	12.2		81	10.4		81	7.8	
Severe vision impairment											
Yes	5	3.6	82	30.9		15	2.2		14	1.5	
No	135	96.4	183	69.1		655	97.8		932	98.5	
Total	140	100	265	100	<0.001	670	100	0.355	946	100	0.078
Missing	30	17.6	88	24.9		108	13.9		93	9.0	
Severe hearing impairment											
Yes	8	6.3	22	8.9		12	1.9		7	0.8	
No	120	93.8	226	91.1		629	98.1		868	99.2	
Total	128	100	248	100	0.374	641	100	0.004	875	100	<0.001
Missing	42	24.7	105	29.7		137	17.6		164	15.8	
Gastrostomy tube											
Yes	59	42.4	162	55.9		34	5.1		8	0.9	
No	80	57.6	128	44.1		628	94.9		885	99.1	
Total	139	100	290	100	0.009	662	100	<0.001	893	100	<0.001
Missing	31	18.2	63	17.8		116	14.9		146	14.1	
Viking Speech Scale level											
I	4	3.0	11	3.7		387	59.9		669	76.3	
II	15	11.1	17	5.8		125	19.3		139	15.8	
III	36	26.7	46	15.6		94	14.6		54	6.2	
IV	80	59.3	221	74.9		40	6.2		15	1.7	
Total	135	100	295	100	0.016	646	100	<0.001	877	100	<0.001
Missing	35	20.6	58	16.4		132	17.0		162	15.6	
Cognitive functioning											
Normal	42	35.6	20	7.4		370	63.5		599	73.0	
Learning disorder	10	8.5	13	4.8		74	12.7		120	14.6	
Intellectual disability	44	37.3	138	51.3		115	19.7		94	11.5	
Non-testable	22	18.6	98	36.4		24	4.1		7	0.9	
Total	118	100	269	100	<0.001	583	100	<0.001	820	100	<0.001
Missing	52	30.6	84	23.8		195	25.1		219	21.1	

Abbreviations: CP, cerebral palsy; ICD-10, International Statistical Classification of Diseases and Related Health Problems, 10th Revision; SCPE, Surveillance of Cerebral Palsy in Europe.

[a]Comparison of dyskinetic and spastic quadriplegic CP.

[b]Comparison of dyskinetic and spastic diplegic CP.

[c]Comparison of dyskinetic and spastic unilateral CP.

suggests that the lesions were most likely caused by acute or chronic hypoxaemia late in pregnancy or at delivery.[12,27,31,32] Taking potential sentinel events into consideration, an overall interpretation of our results could be that an acute hypoxaemic event during delivery was in the causal pathway leading to dyskinetic CP in about 17% of the children.

In some of these cases, a sentinel event may have added to already existing brain lesions, such as maldevelopments or white matter lesions. The MRI findings suggest that in nine children such a 'cascade' may have been in the causal pathway leading to dyskinetic CP (Table S3).

The proportion of children with dyskinetic CP caused by acute or chronic hypoxaemia late in pregnancy cannot be estimated from our data. However, the finding that only 17% of children with dyskinetic CP born at term may have had a sentinel event, and the proportion of children with a normal MRI (16%), brain maldevelopment (9%), and white matter lesions (27%), suggests that in the vast majority, dyskinetic CP was not caused by events occurring during term delivery.

Compared with children with dyskinetic CP, those with spastic quadriplegic CP were more often born preterm and MRI findings more often showed white matter lesions. The proportion of children with potential sentinel events was lower among children with spastic quadriplegic CP when the analyses were restricted to children born near term. Thus, taken together, our results indicate that antenatal insults may be even more common in children with spastic quadriplegic CP than in those with dyskinetic CP.

Cortico-subcortical lesions were more common and basal ganglia/thalamic lesions less common while motor and associated impairments were more severe in spastic quadriplegic CP compared with dyskinetic CP. Thus, we speculate that cortico-subcortical lesions may be stronger predictors of the severity of impairments than basal ganglia/thalamus lesions.

A matter of concern is the large proportion of children with dyskinetic CP as well as with spastic quadriplegic CP where cognitive abilities were considered non-testable or missing. Studies have reported that it is a challenge to assess cognition in children with severe motor and associated impairments, and therefore appropriate habilitation of children with dyskinetic CP and spastic quadriplegic CP may be hampered.[33,34] However, it is also possible that some children with CP were tested for cognitive abilities, but the results were not reported to the NorCP.

CONCLUSION

The birth prevalence of dyskinetic CP in Norway was low and decreased significantly from 1996 to 2015. The decrease is encouraging because it suggests that general improvements in antenatal, obstetric, and neonatal care may indeed prevent some cases of dyskinetic CP. In fewer than 20% of children with dyskinetic CP an acute event at term delivery was considered a possible cause, although basal ganglia/thalamus lesions were present in 40% of the children. Motor impairments were in general severe and associated impairments were common. There were significant differences in both birth and clinical characteristics between children with dyskinetic CP and those with spastic quadriplegic CP. Children with spastic quadriplegic CP were more often born preterm, potential sentinel events at term were less common, they more often had white matter injuries or cortico-subcortical lesions, and motor and associated impairments were more severe. A concern is that around half of the children with dyskinetic CP and 60% with spastic quadriplegic CP either were considered non-testable or not registered with information on cognition.

DATA AVAILABILITY STATEMENT

The data that support the findings of this study are available on request from the corresponding author. The data are not publicly available due to privacy or ethical restrictions.

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SUPPORTING INFORMATION

The following additional material may be found online:

Figure S1: Total number of children registered in the in the NorCP born from 1996 to 2015.

Table S1: Total number of liveborn children in Norway and with pre/perinatal CP per birth year and per subtype.

Table S2: Distribution of MACS levels I to III versus levels IV to V in children with spastic bilateral CP (diplegic and quadriplegic).

Table S3: Distribution of Magnetic Resonance Imaging Classification System results in children with cerebral palsy born 1999 to 2015.

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