The motor impairments of children with cerebral palsy (CP), caused by an inborn or early acquired brain lesion,1 are often accompanied by impaired functioning in other areas, such as cognition. The scope of cognitive impairments varies between and within the spastic, dyskinetic, and ataxic subtypes.2 The identification of cognitive impairments depends upon the quality of the assessments, and the first aim of this study is to review challenges in that regard. In this review, cognitive functioning in children with CP will be discussed from a developmental perspective, on the basis of a review of the literature on cognitive functioning in children with CP.

Assessment of cognition

Two aspects of assessment of cognition need to be addressed: the paucity of studies where a representative sample of all subtypes have been assessed and the challenges involved in reliably assessing cognition in children with impairments that makes test performance difficult. The latter is partly responsible for the former. The Gross Motor Function Classification System (GMFCS),3 a 5-point ordinal scale where level I indicates the least impairment, is typically used to classify motor impairment in the literature discussing cognitive functioning. However, classification of fine motor functioning would have been even more relevant.

There are some epidemiological studies based on data from CP registries4–6 and geographical cohorts7–10 but few where a representative sample is individually assessed.11,12 Without assessment, IQ was estimated on the basis of clinical judgement,8,5 school placement,13 degree of gross motor impairment,10 or interview with parents.8 Assessing cognition in children with motor impairments, including those who are able to perform the tasks of a standardized test of intelligence, is not straightforward. Even small fine motor impairments might influence test scores negatively and lead to an underestimation of IQ.14 Results from timed tests must therefore be interpreted with care, even for children in GMFCS level I. It is even more challenging to assess cognition in children with severe speech and motor impairments.15 The result is that one-third of children in GMFCS levels IV and V are assessed, also in studies aiming to assess a representative sample of children with CP.12,16

The challenges of assessing cognition in the severely affected group leads to cognitive functioning being assumed. Although a correlation between severity of motor and cognitive impairments exists, there is no absolute correspondence;9,11 it is therefore not possible to draw conclusions about cognition from functioning in other areas. Furthermore, it is not necessary as cognition can be assessed using tests with a multiple-choice format and allowing for other means of responding than pointing with a finger. Tests of verbal comprehension, such as the Peabody Picture Vocabulary Test17 and the Test for Reception of Grammar,18 and non-verbal reasoning, such as Raven’s matrices,19 are suitable for adaptation. Alternative response modes include gaze pointing and scanning (Table S1, online supporting information). Comparing standard and alternative response modes, partner-assisted
scanning,20 scanning with switches on a computer,21 the use of frames for gazing pointing,18,22,23 and gaze pointing on a computer20,24–26 have not been found to influence test results. Despite this, children with the most severe motor impairments are described as non-assessible, or test results are provided without information about how tests were adapted.27

**METHOD**

A systematic search of the databases PsycINFO, ERIC, and MEDLINE (Ovid) was performed on 20th to 22nd February 2019. The terms (cognition OR intelligence) were sequentially combined with AND (hemipleg* OR unilateral); AND (dipleg*); AND (quadriplegi*); AND (dyskinetic OR dyskinesia); AND (ataxi*). The search was limited to human children (0–18y) and papers published in English. All abstracts of the 525 hits were browsed. Duplicates, papers not reporting on CP or cognitive functioning, papers published before 1990 (when the International Classification of Diseases, 10th Revision was published), papers where the full text was not available, literature reviews, and case studies were removed, resulting in 111 hits (35 papers on hemiplegia, 31 on diplegia, 16 on quadriplegia, 21 on dyskinesia, and eight on ataxia). Some included information about more than one subtype, leaving 75 unique hits. Two papers, about adults, were removed. Eight papers, which turned up browsing reference lists, were added. From the remaining 81 papers, information about age and subtypes, cognitive areas assessed, and main findings were extracted (Table S2 and Figure S1, online supporting information).

**RESULTS**

Cognitive impairments can be global (expressed as a low IQ score) or specific (pertaining to only one cognitive domain).

**Global cognitive impairment**

There is wide variability in estimates on the proportion of children with CP having an IQ less than 70, but Western countries with national registries report about 30% to 40%.4,5,9 Spastic quadriplegia, epilepsy, severe motor impairment, and brain malformations are associated with more severe cognitive impairments.7,9,11 Disorders of intellectual development can be considered with an IQ less than 70, but the diagnostic criteria2 specifies that there should also be significant impairment in mastery of everyday activities. Difficulties with the latter should not be solely attributable to motor impairment, which complicates the diagnostic process for children in GMFCS levels III to V as there is a lack of appropriate tools for assessing adaptive functioning.28 Further, although a full-scale IQ score might be below 70, it would not be advisable to diagnose intellectual disability if the profile is skewed and functioning is as expected for age in one or more areas. In the only study differentiating between IQ less than 70 and intellectual disability, it was found that although 33% had an IQ less than 70, only 25% qualified for a diagnosis of intellectual disability.11 Reporting only IQ and making assumptions about proportion with intellectual disability on this basis may lead to an overestimation of the prevalence of intellectual disability in the population with CP.

**Giftedness**

Children with CP have an increased risk of cognitive impairments compared with peers, but reporting only this leaves out an important part of the picture. The finding1,29 that children with CP can obtain IQ scores above 120 indicates that some are gifted and need follow-up accordingly.

**Unilateral spastic CP**

Most children with unilateral CP have normal cognition: 81% to 89% are reported to have an IQ greater than 70.49,11 IQ is not significantly different in children with left- and right-sided paresis, or in children born preterm and at term.30–32 Epilepsy is associated with lower IQ.32,33 One-third have specific learning impairments,34 including impairment in visual–spatial cognition,35 acquisition of visual imagery,36 and executive functioning.37 The language functioning in children with right-sided paresis illustrates the plasticity of the developing brain. Contrary to what would happen if adults sustained similar focal brain injuries in the left hemisphere, language is often spared39 and there is no difference in verbal IQ between children with left versus right unilateral brain lesions.30 However, this right hemispheric reorganization of language comes with a cost, as it is associated with lower performance IQ.38

**Bilateral spastic CP: diplegia**

Typically, 67% to 78% of children with diplegia are reported to have an IQ greater than 70.9,11 When the cause of diplegia is periventricular leukomalacia (PVL), IQ is reported to be similar in children born preterm and at term.39 However, in children born at term and with varied aetiology, as few as 39% have an IQ greater than 70.40

Typically, children with diplegia have a skewed profile with normal verbal comprehension and impaired visual–spatial reasoning and non-verbal intelligence.9,41–44 Even though this profile can be observed in 3-year-olds,32 the difference becomes more pronounced as children enter school.45 PVL affects the brain connectivity in the temporal–parietal cortex,46 is particularly frequent in children born preterm,47 and leads to visual–perceptual impairments.38

**What this paper adds**

- Few studies have assessed cognition in a representative sample of children with cerebral palsy.
- Cognition in children with severe motor impairment is often assumed, not assessed.
- Lack of assessment may lead to overestimating the prevalence of intellectual disability.
- Lowered cognitive functioning in older children highlights the need for longitudinal studies.
The relationships between ophthalmological impairments, the extent of white matter injury, visual–perceptual impairment, and non-verbal reasoning are difficult to disentangle, and lack of consensus on definition of core concepts and variability in measurement methods makes comparison across studies challenging. Different concepts, such as cerebral visual impairment (defined as damage to or dysfunction of the retrochiasmatic visual pathways) and visual–perceptual impairment (defined as an impairment in the ability to analyse and process visual information), are sometimes used interchangeably, confounding neurological and functional levels of description.

In children with diplegia, visual–perceptual impairments have been found to be particularly pronounced in those born preterm. For this group, the degree of visual–perceptual impairment is related to white matter thinning. However, PVL has been found not necessarily to lead to visual–perceptual impairment. Performance IQ (which also includes visual–spatial perception, in addition to other abilities such as non-verbal reasoning and processing speed) in those born preterm correlates with white matter integrity. In children born at term, full-scale IQ is related to white matter integrity and severity of PVL, possibly because severe cognitive impairments are more common and thus full-scale IQ correlates with the extent of the brain lesions. The presence of ophthalmological impairments has been reported both to be favored and not to be associated with or fully explain visual–perceptual impairment or constructional dyspraxia.

Visual–spatial impairments and performance IQ are by far the most studied functions in children with diplegia, but specific impairments in attention and executive function have also been reported, especially if there is damage to the anterior corpus callosum in addition to other white matter lesions. Verbal cognition and memory for verbally presented materials are reported to be as expected for age.

Bilateral spastic CP: quadriplegia
In the subgroup with spasticity there is a correlation between degree of motor and cognitive impairment, and up to 90% to 100% with quadriplegia are reported to have an IQ less than 70. However, as formal testing is often reported to be inaccessible to children with quadriplegia, this might represent an overestimate of impairment. Studies in which all children were individually assessed report lower frequencies, around 65%. This implies that there could be more children with quadriplegia who have an IQ in the normal range. Documenting the capabilities of children with quadriplegia requires adapted testing. It has been found that 20% had an IQ greater than 85, illustrating that severe motor impairments can mask cognitive skills.

Dyskinesia
In this second largest subtype, constituting 6% to 15% of the total population with CP, wide variability in standardized scores on tests of non-verbal reasoning (20–129) and verbal comprehension (55–119) is reported. Often 50% to 60% are reported to have an IQ less than 70, but it varies between as few as 25% and as many as 70% to 80%. As in spastic CP, normal cognition is also found in those with the most severe motor impairments. In children with dyskinetic CP this is to be expected, as magnetic resonance imaging (MRI) studies show that lesions in subcortical areas, affecting extrapyramidal pathways, are common.

Studies of cognition including only children with dyskinetic CP are rare, but show that visual perception, language, memory, and executive functions often are as expected for age. Non-vocal children (i.e. children who are not able to use speech as mode of communication) struggle with literacy despite normal cognition. Recently, executive functioning has been the focus in studies of the dyskinetic subtype, but it has not been confirmed that they struggle more in this area than those in the subgroup with spasticity.

Ataxia
Ataxic CP constitutes around 5% to 6% of the CP group, and 42% to 67% are reported to have an IQ less than 70. No studies have focused solely on this group, and knowledge about specific impairments is therefore lacking.

DISCUSSION
Brain lesions and cognitive functioning
Recent MRI studies, particularly those using diffusion tensor imaging which allows more precise descriptions of white matter tracts, have advanced our understanding of the complex relationships between brain lesions and functioning. An acute and severe intrapartum hypoxic–ischaemic insult at term has been linked to dyskinetic CP, while diplegia is often seen in children born preterm with PVL. However, a lesion is visible on MRI in only 85% to 90% of children with CP. All types of lesion (brain malformations and white and grey matter lesions) are found in all subtypes. In children with the same type of lesion, cognition has been found to vary between normal and severe intellectual disability. Studies are mixed in their reporting of correlations between MRI findings and IQ, implying that cognitive functioning cannot be confidently predicted from these scanning technologies. Acute severe perinatal hypoxia–ischaemia can lead to cognitive deficits and no motor impairments. It is well established that in children with unilateral left hemispheric lesions, language might be reorganized in the right hemisphere and might be their strongest skill. However, the developing brain is vulnerable if the lesion is more extensive. In a study of non-vocal children with bilateral CP, the language tracts were visualized using diffusion tensor imaging in the right hemisphere for all five patients regardless of their level of verbal comprehension, but was not visible in the left hemisphere for one child without any
cerebral dysgenesis, more have severe cognitive impairments.\textsuperscript{11,71} This might possibly be linked to epilepsy: more children with a cerebral dysgenesis have epilepsy\textsuperscript{71} and epileptiform activity is negatively correlated with intellectual functioning.\textsuperscript{11,30}

Preterm birth, especially if resulting in infarction, is associated with CP and a risk of cognitive impairment.\textsuperscript{72} However, in children with CP earlier gestational age does not necessarily imply more cognitive challenges. In bilateral spastic CP, the proportion of individuals with severe cognitive impairment increased with increasing gestational age,\textsuperscript{23} and also no relation between cognition and gestational age has been reported.\textsuperscript{11}

Together, this implies that cognitive functioning cannot be inferred from MRI findings alone, nor from information about brain lesion, epilepsy, gestational age at birth, and motor functioning. Instead, the initial brain lesion can be viewed as a constraint on development. Cognitive impairments are the result of reciprocal and continuing interactions between the child and their environment, influenced by the child's opportunities for active exploration and participation.\textsuperscript{74} Knowledge about the risk factors is important because it can lead to awareness about the need for assessment and interventions, and aid in developing follow-up programmes.\textsuperscript{75}

Developmental trends

The panorama of CP has changed over recent decades, with a lower prevalence, proportion of children developing bilateral spastic CP, and incidence of intellectual disability.\textsuperscript{76} However, knowledge about the developmental trajectories of cognition in children with CP is less clear, as there are few longitudinal studies. Of the 81 identified studies included in this review, only nine had a longitudinal design.\textsuperscript{12,11,30,39,42,45,77–79}

These studies show that measures of cognitive functioning at 12 and 18 months of age correlate,\textsuperscript{77} that the skewed cognitive profile of school-aged children with spastic bilateral CP is observable from 3 years of age,\textsuperscript{42} and that after entering school there is an increase in verbal IQ so that it becomes age-average while performance IQ continues to be in the low range.\textsuperscript{45,79} There is a differential development of non-verbal reasoning capacity in children in different GMFCS levels: children in level V not only show initial lower functioning but also increase less with development.\textsuperscript{13} For children with unilateral CP, IQ was stable from 3 to 5 years of age,\textsuperscript{30} while children with the most severe speech and motor impairments did not show the expected increase in non-verbal reasoning from 6 to 12 years of age despite normal cognition.\textsuperscript{78} The development of expressive communication is related to the type of motor impairment, whereas receptive communication is related to IQ.\textsuperscript{13} Following children born preterm and at term, significantly more children with spastic diplegia born preterm were found to develop visual–perceptual impairments.\textsuperscript{19}

Developmental model

The most troubling finding is the lack of age-expected increases in cognitive development in children with severe speech and motor impairments.\textsuperscript{28} This might be explained by the brain lesion, but children with motor impairments also have different experiential backgrounds compared with those who are not restricted in their locomotion. In children restricted in their locomotion, both localization of a brain lesion and restricted upper-limb functioning explain why action-based visual perception is more demanding than object-based perception.\textsuperscript{80} In children expressing themselves using aided communication, their instructions on a construction task included little information about sizes and spatial relations,\textsuperscript{81} further suggesting that allocentric strategies are particularly challenging for severely motor-impaired children. An interaction between the brain lesion and lack of appropriate experiences seems likely.\textsuperscript{74} Applying an embodied cognition framework, the ‘mind must be understood in the context of its relationship to a physical body that interacts with the world.’\textsuperscript{82} Findings supporting this position are that: (1) action planning, which implies that we consider the end point of a movement from the start, is challenging for children with CP and does not improve with age as would be expected;\textsuperscript{83} (2) finger gnosis is important for early numeracy skills, implying that the use of hands, the understanding of numbers, and the perception of space are related;\textsuperscript{84} and (3) constructional dyspraxia in children with CP cannot be related to visual and visual–perceptual impairments.\textsuperscript{43} Thus, the consequences of an initial impairment might be ‘wide reaching with cascading developmental effect on other abilities’,\textsuperscript{85} if measures are not put in place to counteract and minimize the developmental consequences.

Interventions for children with cognitive impairments

It is well established that early interventions are beneficial for alleviating motor impairments in children with CP, but the effect of cognitive training has scarcely been studied.\textsuperscript{86} Also, most studies aim at improving literacy, despite visual–spatial and attentional impairments being more frequent than language impairments.\textsuperscript{27} In a study of executive functioning, no effect of training was found.\textsuperscript{87} This does not imply that one should do nothing. Children with CP attending mainstream school had significantly better progress in mathematics and reading than those attending a special school, despite identical verbal IQ. The finding that the groups differed in the amount of the teaching received, with the children in the mainstream school receiving 1.7 times more, emphasizes the importance of interventions.\textsuperscript{88,89} For the non-vocal group, it is imperative to provide augmentative and alternative communication as early as possible. Otherwise they might be seriously hampered in their development of communication and language and
have severely restricted opportunities for interaction and participation,\(^5^9, ^9^1\) which in turn might have negative cascading effects on their social, academic, emotional, and cognitive development.

**CONCLUSION**

There is a wealth of studies on motor functioning in children with CP. However, even though parents report that learning difficulties are at least as challenging and cognitive functioning plays a greater role for communication, academic functioning, participation, and social functioning, it has been less focused upon.\(^5^9, ^9^1\)

This review illustrates that there are gaps to be filled; few studies have assessed cognitive profiles in a large representative population of children with CP, including also very young children and those with the most severe speech and motor impairments, and there is a need for longitudinal and intervention studies. Some areas, such as visual–spatial abilities and language, are much more focused on, while others, such as memory, are less well researched. This might be because the few studies on memory have not reported specific challenges;\(^4^1, ^4^4\) however, as these studies only include children with milder motor impairments, further research seems warranted.

There seems to be an increasing focus on finding associations between extent and localization of brain lesions and cognitive functions; however, given the evidence of early plasticity as well as the heterogeneity of cognitive functioning in children with similar MRI lesions, it might be questioned whether this is the most useful path forwards. The interplay between brain lesions, sensory deficits, experiential opportunities, and cognitive functioning is complex. It has been investigated whether developmental disregard in children with hemiplegia, a neglect-like disregard of their affected upper limb, is the result of injury to neural networks involved in spatial attention, which are connected to areas involved in motor planning, or the result of lack of use of affected hands during important developmental periods. Monitoring event-related potential during task performance, specific impairments in executive functioning were not found, but general difficulties with performing were. This implied that the executive control processes preceding the motor response were affected, requiring an enhanced cognitive effort in goal-directed behaviour and a developmental delay of executive control mechanisms.\(^9^2\) The same complexity is found when investigating the relationships between visual perception and cognition. Visual–perceptual impairment has been reported to be associated with lower cognitive functioning, found in children with normal cognition, and unrelated to non-verbal cognitive functioning.\(^9^3, ^9^4\)

Future studies of cognitive development and the effect of interventions should therefore take the complex interplay over time between body, brain, and mind into account. Tests need to be adapted, for example using eye-gaze technologies, so that cognitive functioning can be reliably assessed, and not only assumed, in the most severely motor-impaired children.\(^2^0, ^2^4–^2^6\) In the future, brain–computer interfaces might gain importance both for assessment and interventions.\(^9^5\) Furthermore, it might be that not only traditional neuropsychological tests and computerized training tasks, but more naturalistic tests and tasks increasing real-life abilities, such as goal-setting and planning abilities, are needed.

**SUPPORTING INFORMATION**

The following additional material may be found online:

Table S1: Review of studies of adapted assessment of cognition.

Table S2: Eighty-one studies of cognitive functioning in children with cerebral palsy.

Figure S1: PRISMA 2009 flow diagram.

**REFERENCES**


