INTRODUCTION

Spina bifida is caused by an incomplete closure of the neural tube during the first 3 weeks of pregnancy. The nerves, which are normally embedded in the spinal cord, develop in a hernia sac outside of the spinal cord and are unable to transmit impulses from the brain to extremities and organs below the area of the lesion. Depending on the placement of the hernia, the effects vary in terms of the level of paralysis, numbness and difficulties with bladder and bowel functions. In addition, spina bifida significantly affects the development of the nervous system and the central nervous system.
brain, with deviations in different regions, especially in the corpus callosum, midbrain and cerebellum. More than 80% of individuals with spina bifida also have type II Arnold-Chiari malformation, a deformity of the brainstem and cerebellum that blocks the circulation of spinal fluid, thereby causing hydrocephalus. In hydrocephalus, the ventricles widen with the increased amount of fluid, damaging both white matter and nerve cells in the area surrounding the ventricles. Hydrocephalus results in axonal stretching and pressure on both grey and white matter in the brain, which can cause cognitive difficulties in the areas of perception, memory and attention. Hydrocephalus is often treated immediately after birth, usually by surgically implanting a shunt, which directs the fluid to the abdomen. Untreated hydrocephalus can be fatal, but hydrocephalus in profile infants with spina bifida can also resolve without being shunted. It is an ongoing discussion on how to decide whether to shunt or not.

It is well known that individuals with spina bifida have a characteristic intelligence quotient profile, with superior language function compared with visuoperceptual function. In addition, a commonly observed consequence of spina bifida is delayed social development, dependency in daily life and low levels of participation. Over the last 30 years, neuropsychological research has led to increased knowledge about the cognitive consequences of spina bifida regarding language, perception, attention, memory and executive functions. In order to enhance the independence of persons with spina bifida and by extension well-being—it is important to review and compile relevant research on cognition in order that methods for learning and rehabilitation can be adapted and improved for persons with spina bifida. This scoping review maps original articles and reviews published from 2000 to 2018 that describe cognitive profiles and consequences of spina bifida with and without hydrocephalus in children, adolescents and adults.

The aim of this scoping review was to summarise findings concerning cognitive characteristics in people with spina bifida and explain how cognitive factors influence activities and participation in different areas and stages of life.

2 METHODS

PubMed, Psych INFO, ERIC, Scopus, CINAHL and the Cochrane Library were searched for English language articles published in 2000–2013. This search was commissioned by the association for habilitation in Sweden called Föreningen Habilitering i Sverige as part of describing evidence-based habilitation. This search was compiled and has been published in Swedish on the association’s website. A further search for articles published in 2014–2018 was carried out prior to compiling the scoping review in which results from both searches were presented (Figure 1). For the cognitive characteristics, the following search terms were used, for all databases: spina bifida OR myelomeningocele AND (one of the following for each search): perception, memory, behaviour, executive functions, cognition, ADHD and autism. Search terms for activity and participation were adapted to the International Classification of Functioning and Health (ICF) but also expanded to capture terms used in different professional language. Search terms were thus for each database: spina bifida OR myelomeningocele AND (one of the following for each search) activities of daily life, communication, participation, self-management, self-care, education and depression. In the first stage of searching for papers that described the consequences of activity and participation, the keywords spina bifida or myelomeningocele were combined with the different search terms for everyday life described above and combined with the search term cognition. This did not yield any hits. Instead, articles were searched as described above and all papers were read, to determine whether they had at least one cognitive variable in their design. Cognitive variables were defined as cognitive and executive functions, including processing skills, time perception and time management. The term spina bifida included individuals with myelomeningocele, with and without hydrocephalus of all ages. In 11 articles, lipomyelomeningocele was also included in the demographic description. Intervention studies were excluded.

A total of 211 articles from the first search and 35 articles from the second search were chosen for quality assessment, according to the adapted versions of the McMaster criteria for quantitative and qualitative studies (Table S1). In this adapted version, the two criteria concerning intervention studies were removed. All papers in the first search were assessed by all four authors independently, and the final decision for paper inclusion was made by consensus. In the second search, the first and last authors, BL and MP-D, carried out the search, made the quality assessment and included papers by consensus. In total, 92 papers of which seven reviews fulfilled the criteria of high or moderate quality (Table S1). Many papers discussed several aspects of cognition and consequences in various life domains and child, adolescence and adult perspectives. For this reason, these papers are referred to more than once in the results. To categorise and clarify information in the different papers the International Classification of Functioning and Health (ICF) was used. The ICF is a classification of health components with the purpose of scientifically frame and describe functioning and disability. The ICF classifies different components of health, including Body Functions and Structures, Activity and Participation and Environmental Factors.

Key Notes

- This study summarises findings about cognitive characteristics in spina bifida in relation to daily life in a lifetime perspective.
- Regardless of intellectual level, people with spina bifida have characteristic cognitive profiles including specific executive dysfunctions, which have a major impact on daily life.
- Both assets and difficulties influence school achievement, social abilities and daily life skills and must be considered in order to enhance autonomy and participation.
RESULTS

The results from the 92 included articles were categorised under two components of the ICF, Body Functions and Activity and Participation, over a continuum of the life span.

BODY FUNCTIONS

Mental functions

Of the eight body functions in the ICF, the only one relevant for this search was Mental functions. Mental functions are described as the functions of the brain and are thus related to cognition.7

4.1.1 Intellectual functions

Many papers have described intellectual and cognitive functions in spina bifida. In a study by Lomax-Bream et al.4 91 children with spina bifida and 74 typically developed children were assessed with Bayley Scales of Infant Development II and Preschool Language Scale at ages 12, 18, 26 and 36 months. The children with spina bifida had significantly lower results in all areas. Higher levels of the lesion and shunt-treated hydrocephalus were found to have a negative effect on motor abilities and cognition but not on language functions. In this early childhood, children with and without hydrocephalus had corresponding results.4

In a population-based study of children with spina bifida and hydrocephalus,3 about 30% had average Intelligence Quotient (IQ) (>85), 40% subaverage (IQ 70–84) and 30% had intellectual disabilities (IQ <70). The children had significantly stronger verbal abilities (verbal IQ) compared with visuoperceptual (performance IQ) assessed with intelligence tests such as the Wechsler Preschool and Primary Scale Third Edition and Wechsler Intelligence Scale for Children Third Edition III.

Barf et al.5 assessed IQ, memory, executive functions and reaction time in 168 young adults (16–25 years) by comparing individuals with open and closed spina bifida and with or without hydrocephalus. Of those with open spina bifida and hydrocephalus about one-fifth had an IQ below 70 and scored below average on most cognitive tests. However, those with closed spina bifida and no hydrocephalus performed similarly to the typically developed population. In addition, Wasserman et al.9 found
that socio-economic status, level of lesion and seizure history were associated with neuropsychological functioning in children 8–15 years. In a later review, it was shown that individuals with spina bifida had lower IQ than those without, and that hydrocephalus was a risk factor. It is thus concluded that spina bifida, and especially with added hydrocephalus, has a profound impact on intellectual functions.

### 4.1.2  |  Mental function of language

As early as the first 3 years of life, differences in language development and practice were observed in children with spina bifida compared with typically developed children. In a study by Lomax-Bream et al., the children had significantly delayed development in listening, understanding and communication. Vachha et al. explored vocabulary, syntax and pragmatic abilities in children with spina bifida aged 6–8 years. Compared with typically developed children, children with spina bifida had significant difficulties to understand basic concepts and using language functionally in a social context, so-called pragmatic understanding. The authors suggested that the ability to use grammatically correct language masks the existing difficulties at early ages. Dennis et al. described good abilities in the social use of language, correct grammar both in understanding and use, rich vocabulary and good rhyming, but difficulties in understanding social codes, finding the core content and context and drawing conclusions when reading.

### 4.1.3  |  Perceptual functions

In several experimental studies, Dennis described different aspects of visual perception in children and adolescents with spina bifida. One study described that children aged 8–12 years with spina bifida had significant difficulties in activity-based tasks, such as drawing, tracing labyrinths with a pen and tasks measuring mental rotation ability compared with matched typically developed children. Object-based tasks, such as recognising faces, or objects, or judging size, revealed no difficulties. In four different studies involving children and youths aged 8–19 years, perceptual ability and motor timing were examined in addition to inner automatic attention and inhibition of return. The difficulties correlated with observed reduced cerebellum volume and were attributed to early malformation. The perceptual assets for people with spina bifida were length, surface, size, form, face and figure perception, whereas weaknesses included figure-ground perception, form constancy, geometry and attention orienting.

### 4.1.4  |  Memory functions

Children with spina bifida show early signs of both auditory and visual memory problems. Pike et al. found that children with spina bifida aged 36 months had significant difficulties compared with peers when recalling stories or remembering in which boxes candy was hidden. Boyer reported difficulties in working memory and processing speed in children with spina bifida aged 8–15 years compared with controls. Several studies have reported difficulties in working memory and short-term memory in children and youth, such as shorter memory span and ineffective strategies in verbal learning.

In a Swedish study, visual and auditory memory were assessed in a population of children and adolescents with spina bifida 8–13 years of age. When compared to typically developed children, no differences were found in immediate recall or word recognition; however, both auditory and visual short- and long-term memory tasks were significantly harder for children with spina bifida. Similar results on short-term memory were found in a study including 151 children and youths 7–18 years of age by Hampton et al.

In adults, Barf et al. found significantly lower results on memory tests in 111 young adults with open spina bifida and hydrocephalus compared with 20 young adults with spina bifida without hydrocephalus and 37 with spina bifida occulta. Barnes et al. reported difficulties in the short-term and working memory in 31 young adults with spina bifida, which influenced their reading comprehension. In a study by Jenkinson et al., adults with spina bifida and hydrocephalus were found to have low results for both immediate and delayed memory.

In a thorough analysis of memory functions in adults, Dennis et al. reported an intact working memory with low maintenance and manipulation requirements but impaired on tasks that demanded high information maintenance or manipulation load. Immediate and delayed episodic memory was weak. The prospective memory was affected and significantly more affected in the older individuals. Thus, memory problems observed in childhood populations remained, with the addition of prospective memory difficulties; impaired or poor prospective memory was three times more frequent in the older group over 32 years of age. In addition, Ashley et al. examined aging effects on working memory and inhibitory control in adults 18–56 years old and found that the deficits were stable over age, relative to controls. Treble-Barna et al. found that lower hippocampal volume was associated with reduced prospective memory in adults. Iddon et al. found significantly lower results in tasks measuring both auditory and visual memory in adults with spina bifida and hydrocephalus compared to those who had spina bifida without hydrocephalus or hydrocephalus only. Thus, research supports difficulties with several different aspects of memory in individuals with spina bifida, from early childhood throughout the lifespan.

### 4.1.5  |  Higher-level cognitive function (Executive functions)

In a longitudinal study by Landry et al., 49 children with spina bifida were followed from age three to seven years with focus on understanding social codes, taking others’ perspectives, and the executive
function of adjusting behaviour to feedback. The difficulties the children showed by the age of three years remained until the age of seven. In older children and adolescents’ executive functions were assessed with the parent rating of The Behaviour Rating Inventory of Executive Function (BRIEF) in several studies. All conclude that metacognition in particular posed great difficulties. Lindquist et al. measured executive functions in 16 children 8–13 years. Children with spina bifida had significantly lower results in tasks measuring verbal fluency, flexibility, and organization compared to typically developed children.

Findings by Ashley et al. indicate that disruptions in higher-order cognition, as measured by parent ratings of children and youths (8–18 years) with spina bifida, may be attributed to aberrant development of subcortical grey matter. Both Iddon et al. and Barf et al. described significant difficulties in young adults regarding executive functions such as verbal fluency, working memory, and ability to shift attention, confirming that those difficulties remain from early childhood into adult life.

4.1.6 | Attention functions

Difficulties in attention and activity control, and distractibility have long been observed in individuals with spina bifida and described in many studies. Vaccha et al. had primary caregivers of a cohort of 46 children with spina bifida aged 5–12 years complete the Carey Temperaments Scales questionnaire. They did not find any prominent temperament profiles but instead significantly characteristic behavioural patterns. The children were less adaptable, less persistent when completing a task and more distractible compared with standardised population. In addition, Lindquist et al. observed an increase in inattention/passivity in children 5–12 years old with spina bifida when measured with norms in the Conners Rating Scale (CK).

Dennis et al. found delayed reactions in children and youths with spina bifida on both exogenous and endogenous cues in computerised attention tests compared with matched peers. In tasks examining spatial orientation and possible neglect, individuals with spina bifida made more mistakes and were more attentive to tasks on the left side of the body. They also had delayed inhibition of return, which means a tendency to attach to one stimulus instead of shifting focus to new stimuli. These results suggest that injury in the midbrain creates a delay in endogenous attention, and parietal injury harms the ability to release attention. In a later study, Dennis et al. examined reaction time in individuals aged 18–48 years and found that they were impaired not only in speeded performance but also in the consistency of their performance on tasks that extend over time. Results by Dennis et al. suggested that the corpus callosum and central executive network are important mediators of reaction time, and that grey matter is also important for key neuropsychological function. Further, Swartwout et al. examined attention and persistence in 44 children 7–17 years of age and revealed that the children showed no lack of persistence in a computerised test but one-third had attentional difficulties and omitted many more items. It was concluded that children with spina bifida had spared frontal attentional functions; however, the posterior systems responsible for stimulus orientation had a lower function due to brain injury. The children orientated too slowly to stimuli and were also slower in shifting focus. These findings were confirmed by Vinck et al. who found that children 6–15 years with spina bifida and hydrocephalus had lower results than those with spina bifida without hydrocephalus on attention tasks requiring visual-motor and acquired cognitive abilities, referred to as posterior attention abilities.

In a study of children and youth by Kulesz et al., it was suggested that components of both the orienting and executive control attention networks were impaired in 8–29 years old individuals with spina bifida. Executive control difficulties may emerge from parietal cortical anomalies and reduced frontal and parietal cortical-subcortical white matter pathways, as an effect of congenital hydrocephalus.

In addition, Burmeister et al. revealed that children 7–16 years with spina bifida had attentional deficits related to posterior systems in the brain, as they tended to be inattentive rather than hyperactive. Children with spina bifida were also found to be more distractible and to have problems focusing and organising rather than behaving hyperactively. In a longitudinal study by Holmebeck et al., difficulties in attention showed to be connected to social development and remained from ages 8 to 15 years. The observed attentional problems in individuals with spina bifida have resulted in interest in the presence of attention deficit hyperactivity disorder (ADHD) in this group. De la Torre et al. compared incidence of attention lapses and attentional functioning in 85 children 6–16 years old, 29 with spina bifida, 30 with ADHD and 29 controls. The group with spina bifida differed from the typically developed children, and the group consisting of children with ADHD received lower results than both those groups. The hypothesis from the results was that the performance of the spina bifida group represented a greater involvement in the dorsal attention network vs. predominantly the ventral network in ADHD. Shunt revisions and shunt infections had a negative effect on the performance of children with spina bifida.

Brewer et al. compared children 7–15 years with spina bifida, children with ADHD combined type and typically developed children in tasks measuring focused and sustained attention and shifting of attention. Children with spina bifida had difficulty shifting focus, which is attributed to posterior attentional systems. Children with ADHD had problems with sustained attention, which is regulated by frontal brain systems. Burmeister et al. studied attention and executive functions in 164 children with spina bifida aged 7–16 years old compared with typically developed. Behaviour was assessed with Swanson, Nolan and Pelham Scale, SNAP-IV, executive functions with Behavior Rating Inventory of Executive Function, BRIEF and additional attention tests. In total, 23% of the children with spina bifida fulfilled the criteria for ADHD/inattention, 8% ADHD combined type and 0.5% ADHD hyperactivity. However, the hyperactivity was shown to reflect hyperversatility. Brewer et al. noted, in studying 7–15-year-old children, that in...
comparison to children with spina bifida, children with the impulsive/inattentive form of ADHD combined type could disengage but not sustain attention, whereas children with spina bifida could sustain attention but not disengage. Findings from Rose and Holmbeck’s study on attention and executive functions in adolescents with spina bifida supported these results. Children with spina bifida had difficulties with focused attention rather than sustained, unlike children with ADHD. Swartwout et al. found, in a study with 101 children with spina bifida, that they had good sustained attention over time, which confirms earlier findings that the frontal brain systems are well-preserved. No recent studies are available on possible effects of methylphenidates on the attentional problems in spina bifida. Based on this research, children and young adults with spina bifida had characteristic attentional and activity profiles, not compatible with those observed in children with ADHD.

No studies reporting the presence of autism were found. Stubbierd et al. investigated emotion recognition ability, which is a central aspect of Theory of Mind, in 38 adult participants, where one-third performed poorly compared with normative data. Their results correlated strongly with low verbal IQ.

4.1.7 | Significance of hydrocephalus

As only about 15% of children with spina bifida do not develop hydrocephalus, there were very few separate studies on this group. In most articles, the presence of hydrocephalus or lack thereof was demonstrated in the demographic description of study groups, but the results were described on a whole group basis. In studies comparing children and adults with spina bifida with and without hydrocephalus, the results were concordant: the group with comorbid hydrocephalus had lower results on IQ, auditory and visual memory and executive functions tests. In the Management of Myelomeningocele Study, (MOMS) trial, Houtrow et al. compared outcomes after fetal surgery for three groups of children with spina bifida at 30 months of age: no hydrocephalus, shunted hydrocephalus and nonshunted hydrocephalus. No neurodevelopmental differences were noted among children enrolled in MOMS, but the severity of hydrocephalus was found to be associated with poorer outcomes whether operated prenatally or after birth. It seems that individuals with spina bifida without hydrocephalus constitute an intermediate group with more difficulties than typically developed individuals but less than those with hydrocephalus.

5 | ACTIVITY AND PARTICIPATION

This section contains research on how cognitive functions affect Activity, which describes the execution of tasks and actions, and participation across the lifespan of individuals with spina bifida. The ICF describes nine life domains.

5.1 | Learning and applying knowledge

The domain learning and applying knowledge also includes making decisions and solving problems.

5.1.1 | Reading and writing

Early detected reading and writing difficulties are shown to persist during the lifespan in individuals with spina bifida, as shown in several studies. Pike et al. followed 78 children whose cognitive abilities were initially assessed at 36 months and then again at the age of 9.5 years. They concluded that the young children’s difficulties in language comprehension and reproduction of stories had a direct connection with future problems when comprehending the essentials of a text and answering questions about text content.

English et al. analysed children and adolescents 9–19 years with spina bifida and typically developed controls, whether they adjusted their reading to the purpose and whether a pronounced aim with the reading could facilitate the relationship between working memory and comprehension. Both the spina bifida group and the control adjusted to a faster reading pace when reading for pleasure and slower when searching for facts and answering questions about the content. Verbal working memory was related to how well the text was understood in both groups. However, the group of children with spina bifida had fewer correct answers despite slower reading of factual texts.

Barnes et al. studied the connection between speed of word decoding and reading comprehension in 33 children with hydrocephalus, including 24 with spina bifida, mean age 11.5 and compared with 33 age-matched controls. The group with hydrocephalus decoded as fast as controls but still had lower comprehension of texts. Unlike controls, they did not demonstrate an effect on spelling-sound regularity.

Barnes et al. also investigated reading and writing abilities in 31 young adults with spina bifida and hydrocephalus. They were found to have better decoding abilities than comprehensive, just like children with spina bifida. They also had difficulties in expressing themselves in writing. Writing capacity was less developed in individuals with spina bifida compared with understanding written language. Fluency and writing pace were also lower. Difficulty in writing was present in both children and adults with spina bifida, as they experienced longer reaction times, required more time to learn motor tasks and experienced problems calibrating force, as described by Vinck et al. Problems in reading and writing thus carry over to adulthood.

5.1.2 | Calculating

In a review of early numeracy skills, Raghubar and Barnes stated that difficulties in informal math skills such as verbal counting, knowing the number symbols and recognising and manipulating quantities occurred as early as 36 months in children with spina bifida.
Barnes et al.\textsuperscript{54} have also found that visual-spatial working memory at 36 months and phonological awareness at 60 months partially mediated the group’s effects on mathematical and reading achievement at 8.5 and 9.5 years of age. The children’s limited abilities in fine- and gross motor functions at age three hindered them from effective visually guided exploration of the environment, which negatively influenced mathematical cognition. Barnes et al.\textsuperscript{55} studied mathematical skills in 33 12-year-old children with hydrocephalus compared with matched controls. In division, geometry, estimation, problem-solving and time and money concepts children with hydrocephalus obtained worse scores than controls. In other domains, as knowledge about whole numbers, fractions and decimals and exact measurement, there was no significant difference between the groups. English\textsuperscript{56} and Raghuram et al.\textsuperscript{57} have later confirmed those findings. Barnes et al.\textsuperscript{58} found that individuals with spina bifida with decoding and reading disabilities were those who also had greater difficulties with maths.

Difficulties in calculating and processing speed remained in adults with spina bifida, as described by Dennis\textsuperscript{59}. Both reading and calculating skills are important in everyday life, but the mathematical ability is more important for independence as it is needed, for instance, when handling money, following cooking recipes and making purchases.\textsuperscript{56,59,60}

5.1.3 Making decisions

For decision-making, it is necessary to be able to consider alternatives, make a choice and then evaluate the outcome. Several articles\textsuperscript{561-563} described difficulties in this process in individuals with spina bifida. Friedman\textsuperscript{561} discussed that poor autonomy in decision making in individuals with spina bifida may be related to verbal intelligence. Turminello et al.\textsuperscript{562} stated that executive functions such as planning ability and mental flexibility are highly influential in the development of autonomy. Problems in decision-making also put individuals with spina bifida at risk of being more dependent on parents.

In a study about autonomy, Peny-Dahlstrand et al.\textsuperscript{563} described that children with spina bifida had poor autonomy in everyday life decisions such as choosing clothes for themselves or presents for friends and family, which was related to reduced process skills. In another study, Peny-Dahlstrand et al.\textsuperscript{564} measured participation in school activities among children with spina bifida, which revealed that they seldom participated in decision-making bodies, such as student councils, which related to both low motor skills and low process skills. To summarise, low executive functions, manifested as low process skills, were shown to impede the ability to make decisions.

5.2 General tasks and demands

General tasks and demands, as defined in the ICF, include the carrying out of activities and tasks on a general level and pursuing daily routines, organising a work plan for something, handling stress and adapting one’s behaviour as needed. Six papers\textsuperscript{565-567} showed how cognitive factors impede this life domain in persons with spina bifida.

Peny-Dahlstrand et al. described\textsuperscript{565} that the majority of a geographic cohort of children with spina bifida in Sweden, ages 6–14 had poor skills when carrying out activities such as organising the different steps of a task in time order, initiating new steps in a task, adapting behaviour on their own when problems arose and being able to complete the task within a reasonable time frame, all described as process skills. This was also confirmed in a smaller Norwegian study among adults by Stubberud et al.\textsuperscript{566} Those difficulties were, according to another study by Peny-Dahlstrand,\textsuperscript{563} more disadvantageous for children with spina bifida when achieving autonomy in daily life than their impairments in motor skills. Jacobson et al.\textsuperscript{567} articulated the same observation, as they concluded that difficulty in initiating, which is common among persons with spina bifida, was the most hindering impairment in daily life. They also emphasised that this lack of initiation is often connected to difficulties with prospective memory, which means to remember not only what one is supposed to do, but also the ability to reflect on how to remember what to do, described as metacognitive reasoning.

Friedman et al.\textsuperscript{561} not only described difficulties handling daily routines autonomously but also difficulties handling stress, adapting one’s behaviour to fit into different contexts and struggles in assuming personal responsibility. They found that boys with lower-than-average verbal intelligence were the group that had the most difficulties in this area.

Turminello et al.\textsuperscript{562} pointed out a relationship between low autonomy in daily routines and executive dysfunctions. Difficulties related to planning and impaired mental flexibility often made children with spina bifida dependent on both parents and teachers.

Difficulties in time management, which is important to organise routines, were described by Persson et al.\textsuperscript{568} who found significantly lower time-management skills in children aged 10–17 with spina bifida compared with typically developed children, particularly in orienting to and estimating objective time, understanding time perspective and adapting to time demands.

These results suggest that individuals, both children and adults with spina bifida have difficulties to independently carry out activities from start to finish, to pursue daily routines without being reminded by someone and difficulties planning both complex tasks and time schedules. As such difficulties have a negative impact on all types of activity performance, they affect all other life domains.

5.3 Communication

Communication is a life domain that concerns all types of interactive communication including conversation, sending and receiving messages and using different communication techniques and equipment.\textsuperscript{7} In a systematic review from 2002, Fletcher et al.\textsuperscript{569} presented an overview of the research from the 1990s about language development in children with spina bifida. In the earliest studies, it was argued that children with spina bifida have speech fluency,
even though often with what was earlier described as noncontextual phrases—but more recent studies have shown signs of neuromotor dysarthria and low speed in speech. The review showed that many children with spina bifida can learn vocabulary and grammar, but they exhibit difficulties in terms of flexibility and ability to translate meaning, which poses difficulties when reflecting on previous knowledge with added insights.

Landry et al. stated that early signs of executive dysfunction and social language deficits, such as pragmatic language problems, often influenced the ability of social problem-solving. In addition, Holbein et al. related neuropsychological factors to social behaviours. A group of children and adolescents (8–15 years) with spina bifida had less adaptive social behaviours in peer interaction affected by their difficulties with reciprocal verbal exchanges due to problems in language processing, conversational skills and social cognition. Summarised, the results show that executive dysfunction impedes communication skills in individuals with spina bifida and that this can be observed early in life.

5.4 | Mobility

Mobility is described in the ICF as moving and transferring between places. In its broader sense, mobility is the ability to actively move one’s own body but also to use transportation, both personal, for example, wheelchairs and public buses and trains and to move objects. Few studies have evaluated the relationship between cognition and mobility in spina bifida, as mobility issues have often been related to the level of lesion and gross motor function. The discussion has even been reversed, in the sense that cognitive development is affected by deficits in motor function.

Peny-Dahlstrand stated that the most hindering mobility factor for children with spina bifida was neither the driving of the wheelchair in itself, nor the transportation of objects, but rather the manoeuvring. Difficulties with motor skills that depend on planning abilities and visual perception, such as placing oneself relative to the desired object, impeded the manoeuvring and the ability to calibrate one’s force in the process. This has also been discussed by Vinck et al.

Roebroek et al. explained that cognitive abilities influenced everyday physical activity, where low executive functions were negatively related to how much time was spent on physical activity during the day. Norrlin also found that IQ and executive function were related to mobility. Additionally, Shoenmakers concluded that low cognitive ability was more hindering for mobility than the level of lesion. Vinck et al. did not find any associations between basic motor profile and cognitive functioning when assessed with IQ tests. This confirmed the hypothesis proposed by Peny-Dahlstrand et al., which states that it is not basic motor function that is affected by cognition but rather the motor planning skills and how the person used their motor ability in real-life situations.

When it comes to driving a car independently, this was also related to both communicative and learning abilities in people with spina bifida.

5.5 | Self-care

Self-care as categorised in the ICF includes everything individuals do to take care of themselves, such as taking care of one’s hygiene, feeding, dressing and health self-management (6). Difficulties described in the previously discussed domain of general tasks and demands are carried over into the execution of activities in self-care. Everyday personal care tends to require carrying out certain routines and performing activities that must be done in special sequences at particular times.

Davis et al. showed a delay in the development of autonomy in daily life activities in persons with spina bifida, especially concerning the toilet procedure. Toilet procedure, for most people with spina bifida, means that they need to empty their bladder every 3 h by inserting a catheter through the urethra, so-called clean intermittent catheterisation (CiC). Performing CiC requires both sufficient cognitive, fine motor and gross motor abilities. IQ level was a decisive factor in this process. In order to describe the age of independence in clean intermittent self-catheterisation (CiC) Castillo et al. found that 55% of 200 individuals were able to independently perform CiC at a mean age of 9.45 years. Of those 200, 15% had intellectual disabilities compared with 40% of those unable to perform CiC. Executive dysfunction has been pointed out by O’Hara et al. as the most hindering factor to manage everyday CiC correctly. Jacobsen et al. particularly highlighted difficulties in planning and initiating as the most hindering factor; in other words, the CiC process was often not started or not carried through to the end. Problems with planning also affect time processing abilities, and Donlau et al. found that children with spina bifida who had low time processing abilities might not become independent in carrying out the CiC process. However, according to Gribble et al., time processing ability did not affect the time it took to perform CiC.

Lomax-Bream et al. found that during early childhood, young children with spina bifida developed independence at a slower rate than their peers without known disabilities. Heffelfinger et al. described that executive dysfunction in combination with the severity of spina bifida, by which is meant level of lesion and shunt status, were the two primary determinants of how a child’s functional independence and autonomy were rated with the Functional Independence Measure for Children, WeeFim. They also hypothesised different protective factors but found that so-called good family functioning, as the level of family satisfaction, family resourcefulness and family affection, was not a protective factor concerning dependency. Moreover, Barf et al. found that cognitive level was significantly related to the quality of personal care.

Shoenmakers et al. showed that neurological characteristics, as the level of lesion, and closed or open lesion, were a predictor of the functional independency or dependency in self-care; however,
low IQ was a further deteriorating factor. Peny-Dahlstrand et al. argued that the more developed processing skills the child had, the more autonomous the child was in daily life routines. Still, 50% of the children in their study had very low processing skills, especially skills involving initiation and performance adaptations. Jacobsson et al. confirmed these findings in their research.

Warschausky et al. compared health self-management, transition readiness and adaptive behaviour in persons with cerebral palsy and spina bifida. Individuals with spina bifida had lower scores on the initiation of routines and prospective memory than those with CP. In another article, the authors concluded that group differences in mastery motivation, executive function and adaptive behaviour were not significant, thus highlighting the importance of assessment in developing interventions to promote independence.

Berry et al. have focused on the ability to assume personal health responsibilities and found with both qualitative and quantitative methods that this was difficult for persons with spina bifida, especially for those with a lower IQ. Medical adherence and responsibility in early adolescence were examined by Psihogios et al., who found that impaired gross motor function and low IQ were barriers to assuming medical responsibility, and high family stress and executive dysfunction were barriers to adherence and responsibility. Stern et al. found that deficits in attention and working memory led to increased depressive symptoms in an older cohort of individuals with spina bifida and thus were often associated with less medical adherence.

This, perhaps in particular for people with spina bifida, vital domain of everyday life is, according to these results, highly affected by executive dysfunction, as things often just never get done.

### 5.6 | Domestic life

Domestic everyday activities, such as moving out to live independently, acquiring and taking care of one's own home and helping others in the household are important in managing adult life.

Zuckerman et al. described the importance of good executive functioning to be able to move away from one's parents to live alone. They found that young adults with difficulties planning, solving problems, taking initiative and shifting attention had a lower probability of living independently. Moreover, the authors stated that assessment of executive functions and capabilities in this area predicted the development of independence. Davis and collaborators found that adolescents with spina bifida developed independent life skills, such as doing laundry or cooking, later than typically developed peers, independent of IQ level.

Peny-Dahlstrand et al. showed that children with spina bifida had extensive difficulties when carrying out self-chosen, familiar domestic activities in an efficient and independent way, mainly due to low processing skills. In another study from 2012, Peny-Dahlstrand et al. showed that children with high processing skills were more likely to do home chores. Based on the available research, this domain is also greatly affected by difficulties in initiating and planning.

### 5.7 | Interpersonal interactions and relationships

This domain is about carrying out the actions and tasks required for basic and complex interactions with strangers, friends, relatives, family members and intimate partners in acontextually and socially appropriate manner. Zuckerman et al. argued that several of the milestones in the steps towards independence were difficult for young people with spina bifida, such as finding friends and building romantic and intimate relationships. Executive function was strongly related to these abilities.

Peny-Dahlstrand et al. described that children with spina bifida were rated by their teachers to be less actively involved in social activities at school. There was a strong and significant relation between the child's processing skills, and motor skills and the children's active involvement in play and recess activities. Liptak et al. described a significant relation between the ability to socialise or to go on dates, and cognitive ability in young persons with spina bifida. Additionally, Rose et al. showed that executive dysfunctions reported by their teachers were correlated with social skills in children and young persons with spina bifida.

Holbein et al. concluded that longitudinal analyses supported the hypothesis that neuropsychological skills such as attention and executive functions contributed to the social skill development of youths with spina bifida, beyond family and health factors. Lennon et al. examined the longitudinal relationship between neuropsychological functioning and internalising symptoms such as depression and anxiety as mediated by social competence in youth with spina bifida. Increased neuropsychological functioning was associated with increased social competence, which predicted fewer internalising symptoms 2 years later. Thus, communication difficulties described previously as affected by cognitive, especially executive, dysfunctions, were often obstructing the development of interpersonal interactions and the development of relationships.

### 5.8 | Major life areas

This domain entails abilities to carry out the tasks and actions required to engage in education, work/employment and conducting economic transactions.

In a study by Davis, it was found that even in late adolescence, individuals with spina bifida often did not independently take care of their own money. A lower cognitive level was associated with less independence. This is consistent with a study carried out by Dennis & Barnes, in which they confirmed difficulties in mathematics and demonstrated that for adults with spina bifida, math problems were more hindering in daily life than reading difficulties.

Lindsay et al. (91) emphasised the importance of professional and social support, and school accommodation to enable young people with spina bifida to manage the transition to postsecondary education or work.

Hetherington et al. (59) found a correlation between cognition and self-rated quality of life in spina bifida. Lower cognitive ability was
associated with difficulties related to employment, personal finances, education, recreation and social life, which was also demonstrated by Barf et al. 593 Oakeshott et al. 594 showed in a follow-up study from birth to 40 years that all persons employed had an IQ over 80. One study by Fletcher et al described that communicative shortcomings and executive dysfunctions were hindering factors for persons with spina bifida when succeeding with their education and finding work.

As seen in the other domains, the results show that executive dysfunctions make it difficult for people with spina bifida to be active and take full responsibility for their needs in this domain of life.

5.9 Community, social and civic life

This domain is about the actions and tasks required to engage in organised social life outside the family, in the community and in social and civic areas of life. Few studies have assessed those activities in association with cognitive factors. In a large survey about the transition process into adulthood in the United States, Liptak 576 showed that cognition was one of the variables explaining whether young people with spina bifida participated in community and charity activities or organisations. The other variables were family economic status, gender and technical aids and equipment. The same study demonstrated that the probability of young individuals with spina bifida hanging out with peers in coffee shops or going to the mall was smaller if they had cognitive difficulties. Murray et al. 595 studied normative and risk behaviour in emerging adults with spina bifida and its influence on social adjustment. They reported less lifetime substance use and lower sexual activity than emerging adults in a comparison sample but similar frequencies of smoking cigarettes and marijuana. Higher verbal IQ predicted higher levels of substance use and number of sexual partners. In a review on participation using the ICF-CY as a reference framework, Bakaniené et al. 596 found that only one of 136 studies focused on school participation, whereas the others analysed participation in physical activities. Higher cognitive abilities had a positive impact on physical participation as did lack of a shunt and having no major medical issues. Thus, it seems that the possibilities to participate, for better or worse, in society in general are lower for people with cognitive difficulties.

It seems clear that the cognitive characteristics of spina bifida impact all domains of the ICF component activity and participation.

6 DISCUSSION

The results from this review reveal characteristic cognitive difficulties in individuals with spina bifida in language, perception, memory, executive functions and attention. This has consequences throughout the lifespan in most important areas of life which has a great impact on participation. Some papers showed that the difficulties could be traced early in life, but some of the consequences were not apparent until later in life, for instance during the transition into adulthood.

Further, the results demonstrated that IQ level together with attention, memory and executive functions are significant to manage school, education and work and that executive functions are crucial in how individuals with spina bifida manage their daily lives and health, independent of IQ level. 579,580,586 Motor abilities are important for executing activities, but lacking initiation, planning abilities and time management skills hinder things from getting done. 563,564,568 If activities in health self-management such as performing CiC, are not regularly performed there is a risk for serious medical complications and increased early mortality in SB. 597 Many of these complications are preventable by adhering to self-care routines, such as CiC, or being able to recognise and act on new health-care needs. 598 Problems with working memory, language and reading comprehension and creating texts were frequently shown in studies of both children and adults. 4,11,19,51,52 An often technically good reading ability contrasted with limited reading comprehension.

The life-long challenges experiencing the various problems summarised herein can also leave this group and its families at risk for depression. 599,500 It was shown in the results of this scoping review concerning self-care that deficits in attention and working memory were associated with less medical responsibility via increased depressive symptoms in a cohort with spina bifida followed from childhood through early teens. 588 It seems like when confronted with challenges in the transition process that children with spina bifida are at risk of developing depressive symptoms. 599 Brei et al. 5100 studied depressive symptoms in parents of adolescents and found that those correlated significantly with neuropsychological functions and especially executive functions in their children.

Despite this rather negative picture, a great deal of preventive and supportive efforts can likely support good cognitive, social and psychological development. Based on the results presented in this review, the often-held mindset by professionals and others that parents of individuals with spina bifida are merely being overprotective might need to be reconsidered. However, what really needs to be considered is the abilities and difficulties and how to appropriately guide the individual given challenges that might be present. To be able to do so requires early assessment of cognitive and executive functions and everyday skills. This will allow for the planning of needed support for the optimal development and for the development of age-appropriate independence, to the extent possible. Parents need information on strengths and weaknesses to understand, help and support their children and adolescents adequately and to give them an appropriate and encouraging self-image and a sense of self-efficacy.

The results of this review are also important for professionals providing direct care or planning for the care of individuals with spina bifida. The cognitive component in a condition as complex as spina bifida must not be overlooked when developing care- and rehabilitation programs. Language comprehension and pragmatic ability should also be included in the assessment of the small child, which has not traditionally been recommended given the children’s generally good results on verbal IQ.
Moreover, at school, in-depth information to teachers is necessary, as the child is at risk of being misunderstood and under- or overestimated in terms of ability. Being aware of cognitive difficulties is crucial and has an impact on the psychological well-being in both individuals with spina bifida and their parents. Furthermore, cognitive and executive functions are essential for developing social abilities. According to the results, early play development should be observed in relation to memory and communication. Thus, parents and preschool teachers should be made aware of how to help children play together, take turns and role play.

The risk of developing depressive symptoms could likely be minimised with assistance in understanding cognitive assets and difficulties. This knowledge is quite new, and it is therefore important to offer cognitive and neuropsychological assessments also for the older population with spina bifida. Motor problems, bladder and bowel difficulties, and additional medical conditions are a lot to bear; if cognitive challenges can be reduced, the quality of life may be improved. There is a growing awareness concerning mental health problems in this group. Recently Kritikos et al. published Mental Health Guidelines with accompanying clinical questions to be asked at different ages.

This scoping review did not include intervention studies. However, the results highlight the need for rehabilitation methods and strategies that focus on enhancing or compensating reduced executive abilities which, to our knowledge, only a few studies have examined. One study adapted the environment and activity by using digital reminders, which showed no change in behaviour.

Some studies have tested interventions with goal setting which showed rather promising results. Methods that are person-centred and where the individual develops his or her own goals and cognitive strategies also seemed to be effective. Stubberud et al. have shown positive results on improved executive functioning and better emotional health using the method of Goal management training. Peny-Dahlstrand et al. and Öhrvall et al. have used the Cognitive Orientation to daily Occupational Performance, CO-OP, Approach which contains self-set goals and the use of personal cognitive strategies with promising results. The evidence for different rehabilitation intervention methods needs to be reviewed in a further study. A recent study has published guidelines to enhance self-management of one’s condition for people with spina bifida. We would like to call for similar guidelines regarding intervention methods to reduce the negative effects of cognitive impairment in daily life for people with spina bifida, in which this scoping review can serve as a starting point.

6.1 | Strengths and limitations

This scoping review is comprehensive and provides a description of cognitive characteristics in spina bifida and their manifestations in important areas of life, a combination that has not been published previously but is often requested by individuals with spina bifida, their families and caretakers. In this review, there were few comparisons between different types of spina bifida since most articles described spina bifida as one group. This should be further investigated in future studies.

It is possible that some articles may have been missed in the database searches, especially those concerning consequences in daily life, as they are not always indexed with a cognitive variable as a search term and required full-text reading. Nevertheless, the authors have systematically gone through and summarised a large body of work related to cognition and its consequences as it relates to individuals with spina bifida. Moreover, the authors come from different academic disciplines, psychology and occupational therapy, which hopefully make the proposed recommendations suited for the multidisciplinary teams that support the individual and his or her family.

7 | CONCLUSION

Current comprehensive research is explicit. Regardless of intellectual ability, people with spina bifida have considerable cognitive difficulties that have a profound impact on learning, orientation, social development and ability, daily living skills, work and autonomy, affecting the quality of activity and participation. This may affect medical adherence and responsibility and by extension the prevention of secondary complications.

The motor disability, bladder and bowel problems coupled with cognitive difficulties require people with spina bifida and their families and carers to have support from specialists throughout life. In addition, it is necessary to acknowledge the special needs of this group according to their cognitive profiles in regular health care settings and social support systems.

Most studies included in this work showed that cognitive disabilities greatly affect managing one’s learning, gaining independence in personal care and living and participating in society. It seems that these cognitive disabilities, especially in the area of executive functioning, have an even greater impact on everyday life than the motor disabilities. Consequently, in the future, more focus should be placed on developing interventions to help individuals with spina bifida cope with cognitive disabilities to enhance their independence, self-efficacy, autonomy and participation.

AUTHOR CONTRIBUTION

Dr Lindquist had the primary responsibility for study design and writing the manuscript. Dr Lindquist and associate professor Peny-Dahlstrand were responsible for both data-base searches and analyses of the data. Ms Strinnholm and ms Jacobson participated in the first data-base search and associated analyses. Associate professor Peny-Dahlstrand and mss Strinnholm and Jacobson have participated in respectively contributed to writing the manuscript.

CONFLICT OF INTEREST

The authors have no conflicts of interest to declare. "Supplementary references" is in Appendix S1.
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