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Review

Hydrocephalus: A neuropsychological and theoretical primer

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ABSTRACT

Hydrocephalus is a common neurological condition, the hallmark feature of which is an excess in production, or accumulation, of cerebrospinal fluid in the ventricles. Although it is associated with diffuse damage to paraventricular brain areas, patients are broadly typified by a particular pattern of cognitive impairments that include deficits in working memory, attention, and spatial abilities. There have, however, been relatively few neuropsychological accounts of the condition. Moreover, theories of the relationship between aetiology and impairment appear to have emerged in isolation of each other, and proffer fundamentally different accounts. In this primer, we aim to provide a comprehensive and contemporary overview of hydrocephalus for the neuropsychologist, covering cognitive sequelae and theoretical interpretations of their origins. We review clinical and neuropsychological assays of cognitive profiles, along with the few studies that have addressed more integrative behaviours. In particular, we explore the distinction between congenital or early-onset hydrocephalus with a normal-pressure variant that can be acquired later in life. The relationship between these two populations is a singularly interesting one in neuropsychology since it can allow for the examination of typical and atypical developmental trajectories, and their interaction with chronic and acute impairment, within the same broad neurological condition. We reflect on the ramifications of this for our subject and suggest avenues for future research.

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1. Introduction

Hydrocephalus is a neurological condition that is generally characterised by an increase in the volume of cerebrospinal fluid (CSF). This causes ventricular swelling that exerts pressure on the brain and skull, causing widespread damage to neural structures (Fletcher, Dennis, & Nothrup, 2000; Loveday & Edginton, 2011). The aim of the present review is to provide a contemporary overview of the functional and aetiological characteristics of this condition. In doing so, we hope that its profile might be raised to a position of greater prominence within academic neuropsychology. Hydrocephalus is a well-researched and well-understood condition within the clinical sciences, but it has not achieved quite the same level of awareness within the behavioural sciences, despite possessing characteristics that make it of particular psychological interest. Some of these characteristics are rooted in concepts of functional impairment, sparing, and dissociation, whilst others touch upon broader notions of typical and atypical developmental trajectories, and acute (and treatable) versus chronic manifestations. Hydrocephalus, therefore, offers a rare opportunity to bridge a theoretical gap between traditional patient-based neuropsychology and the study of developmental disorders of cognition (which has previously, and not uncontroversially, been considered in terms of 'developmental neuropsychology'; Johnson & de Haan, 2015).

One reason why hydrocephalus may have, thus far, avoided the mainstream neuropsychological limelight is that it can be considered a relatively new condition – i.e. it is only since the introduction of the shunt procedure in the 1950s that the life expectancy of patients has increased sufficiently for them to represent an extant population (with extant cognitive impairments). In turn, this historical context has constrained development of the scientific literature on hydrocephalus. Research in the early 1900s focused on how to best diagnose and treat the condition (Aschoff, Kremer, Hashemi, & Kunze, 1999), and research on cognitive function largely focused on children. Later studies, on the other hand, have been more likely to include people of varying ages (including adults) due to the advances in treatment. This profile has created some variability in cognitive outcomes between studies, even though some functions appear to be consistently impaired (e.g., executive function, fine motor function).

Hydrocephalus carries the potential to occupy a particularly interesting role in neuropsychology. Whilst our understanding of conditions such as hemispatial neglect or Balint's syndrome are gleaned from individuals that we presume to have had typical pre-morbid function, our understanding of cognitive impairment associated with, for example, Fragile X or Turner's syndromes comes from individuals who have, by definition, developed atypically from the outset. Hydrocephalus offers an intriguing opportunity to study the same condition from the perspective of both typical and atypical developmental trajectories. The latter is perhaps the most usual, with hydrocephalus being evident at birth (or before), most commonly as a result of neural tube defects or ventricular aetiology. However, a normal pressure variant of the condition can appear in adulthood, typically around middle age, leading to cognitive impairment in the face of a typical developmental trajectory. It is not easy to generate other neuropsychological conditions that share such a unique profile (although developmental impairments of reading or face processing may be considered similar to their clinical equivalents), and we shall discuss the potential implications.

Table 1 – Glossary of terms.

Condition	Acronym
Hydrocephalus	HC
Spina bifida	SB
Spina bifida with hydrocephalus	SB + HC
Spina bifida aperta with hydrocephalus	SBA + HC
Spina bifida meningomyelocele with hydrocephalus	SBM + HC
Spina bifida, many of whom have hydrocephalus	SB ± HC
Spina bifida with no hydrocephalus	SB-HC
Hydrocephalus with no spina bifida	HC-SB
Normal pressure hydrocephalus	NPH
Idiopathic normal pressure hydrocephalus	iNPH

This detailed overview of hydrocephalus will focus on knowledge that will likely be of most interest to the neuropsychologist. As such, we will spend more time on the unique cognitive and behavioural sequelae of the condition than we shall its clinical intricacies, although we direct the reader to useful sources for the latter. Our review begins with a broad description of the aetiology of hydrocephalus, which is followed by a discussion of its relationship with spina bifida. We then move on to characterising its cognitive profile, as informed by careful neuropsychological study of basic cognitive processes. This will be complemented by coverage of the relatively sparse research that has addressed more integrative behaviours in patients with hydrocephalus, such as spatial navigation. Finally, we will return to the more theoretical and philosophical implications associated with studying this population (or populations), and suggest some avenues for future endeavour. Throughout the text we will use acronyms for the variety of conditions reviewed and, owing to their relative complexity, these are detailed in Table 1.

2. Congenital and early-onset hydrocephalus

Early-onset hydrocephalus (HC) develops within the first two years of life, although most of these cases are congenital in nature. Congenital HC is often caused by a type of neural tube defect (i.e. a prenatal spinal cord malformation, leading to a lesion in the spinal cord; see Frey & Hauser, 2003) known as *spina bifida* (SB). SB has different subtypes and its most common (and mildest) form, spina bifida occulta, only results in mild clinical symptoms (Boone, Parsons, Lachmann, & Sherwood, 1985). HC is often the consequence of spina bifida meningomyelocele (SBM), a type of SB that commonly presents with a Chiari II malformation (Dennis, Landry, Barnes, & Fletcher, 2006; Fletcher et al., 2000). SBM will also frequently result in sensory and motor impairments of the lower limbs, neurogenic bladder and bowel leading to incontinence, and early-onset puberty (Cholley et al., 2001; Hochhaus, Butenandt, Schwarz, & Ring-Mrozik, 1997) all of which can result in long-term difficulties for some patients.

There are a variety of additional subtypes of early-onset HC, which are defined according to the aetiology and progression of the condition and can make classification more complex (Oi, 2010; Tully & Dobyns, 2014). For example, when HC results from an obstruction or blockage within the ventricular system, it is known as 'obstructive hydrocephalus' or 'non-communicating hydrocephalus'. The blockage may be located in the ventricles, cerebral aqueduct (i.e. aqueductal stenosis), subarachnoid space, or arachnoid villi (Del Bigio, 1993). Obstructive forms of HC can then be further classified depending on where the obstruction is located (Oi, 2010), although several points of obstruction in one patient are possible. In some cases, the CSF levels become balanced following removal of the obstruction and this is referred to as 'arrested hydrocephalus'. These patients do not usually require ongoing treatment but retain the residual damage (if any) due to initial expansion of the ventricles (Fletcher et al., 2000). Another subtype is 'communicating hydrocephalus', which can develop without a blockage as might happen if there are problems with the production or reabsorption of CSF (Dandy & Blackfan, 1913; Erickson, Baron, & Fantie, 2001; Sakka, Coll, & Chazal, 2011). Further causes of HC include infection, Dandy-Walker Syndrome, adhesions, and intraventricular haemorrhage associated with premature birth (Fletcher et al., 2000). See Tully and Dobyns (2014) for a more detailed review of aetiologies in congenital and childhood-onset variants of the condition. HC also appears to have a genetic causal component, which is starting to be more well-understood in recent work (Berker, Goldstein, Lorber, Priestley, & Smith, 1992; Jin et al., 2020; Kahle, Kulkarni, Limbrick, & Warf, 2016; Munch et al., 2012; Zhang, Williams, & Rigamonti, 2006).

2.1. *Prevalence and physiology*

While congenital and early-onset HC can be prevented by consumption of folic acid during pregnancy (MRC Vitamin Study Research Group, 1991; O'Dell et al., 1948; Overholser, Whitley, O'Dell, & Hogan, 1954; Wald, 2004; Wald & Bower, 1995), it remains a common neurological condition. It has a prevalence of .47–.85 per 1000 births (Garne et al., 2010; Isaacs et al., 2018; Persson, Hagberg, & Uvebrant, 2005), although estimates vary across time and geography (Loveday & Edginton, 2011). Perhaps more strikingly, HC accounts for approximately 29.4% of paediatric neurosurgical admissions (Green, Pereira, Kelly, Richards, & Pike, 2007), although recent estimates are more conservative for example, 8.9/100 was found in Perenc, Guzik, Podgórska-Bednarz, & Drużbicki, 2022.

The overt physiological characteristics of the brain in HC somewhat mirror the aetiological mechanisms detailed above. Accordingly, a hallmark feature of HC is enlarged ventricles (Toma, 2015), which are associated with widespread neurological damage, particularly to periventricular areas (Fig. 1). These include the corpus callosum (Fletcher et al., 1996, 1997; Jinkins, 1991), cerebellum (Dennis et al., 2004), corticospinal tract, fornix, hippocampal structures, alveus, fimbria, parts of the basal ganglia, and dorsal cortex (Del Bigio et al., 2003). There is also evidence of altered white matter connectivity, including hyperconnectivity between the ventral attention and default mode network and lower fractional anisotropy in posterior white matter (Adam, Ghahari, Morton, Eagleson, & de Ribaupierre, 2022). The extent of damage depends on the rate of ventricular dilation (Del Bigio, 1993), and its distribution can also relate to aetiological factors. So, for example, SBM is most likely to lead to HC because of CSF blockage at the third ventricle, and it is associated with thinner posterior cerebral cortex (compared to anterior regions), particularly on the right hemisphere (Fletcher et al., 2005; Loveday & Edginton, 2011).

2.2. *Treatment*

The most common type of treatment for HC is the surgical implantation of a ventriculoperitoneal shunt. During this procedure, a mechanical tube is inserted into the brain, diverting the excess CSF to a different body cavity (usually abdominal) where it is absorbed by the lining (Toma, 2015). Shunting is often an effective treatment, although it has a high incidence of associated problems, including infection and shunt malfunction, which require revisions. This is despite recent advances and improvements in shunt design and techniques with antibiotic impregnated shunt materials and adjustable valves to resolve dynamic pressure changes (Kahle et al., 2016; Reddy, Bollam, Shi, Guthikonda, & Nanda, 2011). Furthermore, repeated shunt revisions have been reported to be associated with poorer cognitive outcomes (Arrington et al., 2016; Barf et al., 2003; Brewer, Fletcher, Hiscock, & Davidson, 2001; Dennis & Barnes, 2002; Dennis et al., 2007). As a result, the option of treating HC without a shunt is taken if this is available, as it avoids the long-term risks associated with shunting.

In the cases of obstructive HC, another widely-used form of surgical treatment is known as ETV (endoscopic third ventriculostomy; Recinos, Jallo, & Recinos, 2012). ETV aims to restore the opening for CSF to exit the ventricles into the subarachnoid space. However, ETV can also result in complications (Schroeder, Niendorf, & Gaab, 2002), particularly when ETV is performed early in life (Navarro et al., 2006), and higher mortality rates (Hader et al., 2002), although this appears to vary with aetiology (Erşahin & Arslan, 2008) as well as time of surgery. For recommendations on the cases where ETV is appropriate, please see Yadav, Parihar, Pande, Namdev, and Agarwal (2012). For a review of the remaining questions in the treatment of HC, see Kahle et al. (2016).

2.3. *Cognitive function*

Although there are a variety of aetiological origins for congenital and early-onset variants of HC, assays of associated functional impairments present a relatively consistent picture across individuals. However, since insights have been gleaned from a mixture of developmental and adult samples, there are not always accounts of the same cognitive functions in both cohorts of patient. In this section, we begin by presenting a broad cognitive profile of the condition, primarily informed by characterising performance across batteries of standardised neuropsychological tests. This is aimed at providing a general account of those domains within which patients exhibit impairment, and those that might be considered unimpaired or 'spared'. We then explore some of

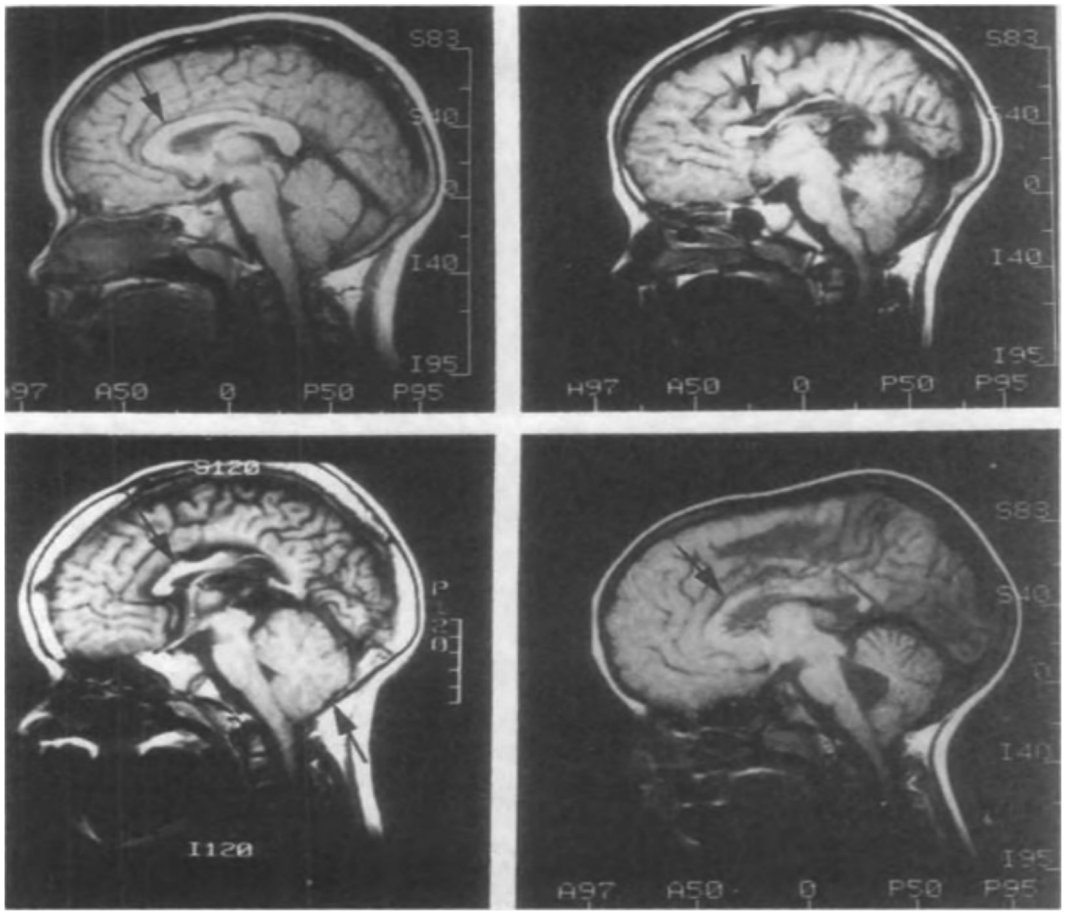

Fig. 1 – Early-onset hydrocephalus. Top left panel: typically-developing child. Top right panel: Child with aqueductal stenosis and shunted hydrocephalus. Bottom left panel: Child with spina bifida meningomyelocele, Chiari II malformation, and shunted hydrocephalus. Bottom right panel: Child with prematurity/intraventricular haemorrhage and shunted hydrocephalus. Reprinted from *Syndrome of nonverbal learning disabilities: Neurodevelopmental manifestations* by Fletcher, Brookshire, Bohan, Brandt, and Davidson (1995). Reprinted with permission of Guilford Press.

the more in-depth examinations of specific cognitive impairments that serve to refine our understanding of particular domains.

2.3.1. *Cognitive profile*

Previous research in children has found that HC is associated with lower scores on fluid intelligence measures (Ayr, Yeates, & Enrile, 2005; Erickson et al., 2001; Vinck, Maassen, Mullaart, & Rotteveel, 2006), and with a general sparing of verbal abilities, relative to non-verbal reasoning abilities (Dennis et al., 1981; Fletcher et al., 1995, 1997; Ito et al., 1997; Lindquist, Uvebrant, Rehn, & Carlsson, 2009; Riva et al., 1994; Wills, Holmbeck, Dillon, & McLone, 1990). Non-verbal difficulties have been attributed to differences in motor function associated with HC (Erickson et al., 2001). Adults with HC have also been reported to score lower on measures of fluid intelligence than patients with SB but without HC (SB-HC; e.g., on Raven's progressive matrices: Barf et al., 2003; 111 patients with HC compared with 57 patients without). However, there have been some inconsistencies in reports of IQ profiles in adult samples, with Hommet et al. (1999) reporting no differences between young adults with HC and young adults with SB-HC in either verbal intelligence (VIQ) or performance intelligence (PIQ). Moreover, Hommet et al. (2002) found no difference between VIQ and PIQ in a sample of young adults with HC + SB, and reported global IQ within the typical range, although this may be attributable to the small number individuals tested ($N = 10$).

While research into the exact patterns of spared and impaired functions associated with HC is ongoing, there has been some promising theoretical work regarding the cognitive profile of HC (Dennis & Barnes, 2010; Dennis et al., 2006; Fletcher & Dennis, 2009). Before discussing this, it is worth noting that these observations are based on HC with a specific aetiology – i.e. SBM. First and foremost, these reviews have acknowledged that SBM + HC results in a varied cognitive profile, consisting of specific functions that are either intact or impaired (see also, e.g., Loveday & Edginton, 2011; Iddon, Morgan, Loveday, Sahakian, & Pickard, 2004). Dennis and colleagues argue that domain-specific difficulties arise from domain-general difficulties. Accordingly, they propose that general impairments in movement, timing, and exogenous attentional orienting (also known as posterior attention; e.g., Posner & Petersen, 1990; Posner, 2016) lead to specific difficulties in aspects of visuospatial ability, language, and mathematics (discussed in more detail in the next section).

These three domain-general difficulties are thought to be dissociable, and reliant on the status of different brain areas. For example, movement difficulties in the upper/lower limbs and the eyes relate to damage to the spinal cord, cerebellum, and midbrain areas controlling eye movements. Difficulties in perception and production of timing are also related to cerebellar damage, while attentional difficulties depend on the status of midbrain, posterior cortex, and corpus callosum. These impairments have been proposed to map on to processing bias, with strengths in 'associative processing' and difficulties with 'assembled processing' (Dennis & Barnes, 2010; Dennis et al., 2006; Fletcher & Dennis, 2009). So-called 'associative processing' is reliant on formation of associations, changes in response to stimulus repetition, and information categorisation (e.g., recognising faces, comprehending familiar words). In turn, 'assembled processing' refers to the ability to assemble input across domains based upon on-line responding over time (e.g., mental rotation, perceiving perceptual relations). Assembled processing, therefore, operates on the products of associative processing, and is thought to mediate the relationship between domain-general and domain-specific deficits. Relative strength in each domain is proposed to rely on associative processing, and difficulties in assembled processing, though neither associative nor assembled processing are directly observable. These authors argue that HC moderates the cognitive difficulties caused by SBM and specifically impairs assembled processing.

There have been two comprehensive neuropsychological examinations of SB + HC and SB-HC, which have further clarified the cognitive profile of patients. The first published example was from Barf et al. (2003), who examined three groups of young adults aged 16–25 years with either SB occulta, SB aperta without HC (SBA-HC), or SB aperta with HC (SBA + HC). They found that SB occulta and SBA-HC groups performed similarly, and within the typical range, on all tasks included in their study: fluid intelligence (Raven's Standard Progressive Matrices), memory (Wechsler Memory Scale), immediate and delayed verbal memory (Dutch version of the California Verbal Learning Test), executive function (letter fluency, Wisconsin-Card-Sorting and Trail-Making Tests), and reaction time (decision reaction time and motor reaction time). The SBA + HC group scored lower than the other two groups on the majority of fluid intelligence and memory measures including Raven's Progressive Matrices, Wechsler's Memory Scale, and verbal learning. They also scored lower on executive functioning tests, including Wisconsin Card Sorting and Trail-Making Part B, as well as a reaction time task (decision reaction time). The SBA + HC group also scored lower than SB occulta group (but not SBA-HC group) on measures of motor speed (motor reaction time) and letter fluency.

A similar study was reported by Iddon et al. (2004), who administered a comprehensive battery of neuropsychological tests to typical adults and three groups of patients: those with HC only, those with SB-HC (precise variants were not reported), and those with SB + HC. They found that all groups performed similarly on the Eyes Test of Emotional Judgement, and all patient groups performed similarly (but lower than controls) on verbal and visual recognition memory. For the rest of the measures, the two groups of patients with HC (HC only, SB + HC) scored lower than the other groups. These tasks included verbal learning ability, delayed verbal recall, spatial working memory, spatial recognition memory, measures of psychomotor speed, and the interdimensional extradimensional shift task, measuring attentional switching. In contrast, the group with SB-HC performed within the typical range, and similarly to controls, on all measures except for category fluency.

Based on these patterns of results, both Barf et al. (2003) and Iddon et al. (2004) concluded that HC alone is responsible for the cognitive profile of this population, while SB does not appear to have strong negative effects on cognition. The contrast between this position and that proffered by Dennis and colleagues (Dennis & Barnes, 2010; Dennis et al., 2006; Fletcher & Dennis, 2009) does not seem to have been identified in the current literature, and we will return to this debate later.

2.3.2. *Domain-specific cognitive impairments*

Whilst neuropsychological batteries allow for a broad overview of cognitive abilities, they are somewhat limited by the tests contained therein, and cannot always provide a finer grain of measurement for specific abilities. In this section, we aim to provide a more in-depth summary of impairments that have been reported within specific cognitive domains (see also: Dennis et al., 2006; Dennis & Barnes, 2010; Erickson, Baron, & Fantie, 2001; Loveday & Edginton, 2011).

2.3.2.1. Processing speed. Processing speed usually refers to the rate at which we acquire, process, and respond to new information (Weiss, Saklofske, Coalson, & Raiford, 2010). It is one of the factors of the intelligence quotient (Wechsler, 1991), and can affect general cognitive skills such as the ability to make computations, carry out conversations, or set goals. Children with HC have been found to score lower on measures of processing speed (Boyer et al., 2006; Calhoun & Mayes, 2005), and difficulties appear to persist into adulthood (Lindquist, Persson, Fernell, & Uvebrant, 2011). For example, Ayr et al. (2005) found that children with SBM + HC had a lower processing speed index compared to children with traumatic brain injuries or orthopaedic injury. This supplemented the findings of Jacobs, Northam, and Anderson (2001), who reported that children with SBM + HC had lower estimates of processing speed than typically-developing children. Importantly, this was also true for a measure that did not require motor skill – i.e. the Controlled Oral Word Association Test (COWAT: Gaddes & Crockett, 1975). Vinck et al. (2006) also observed lower processing speed indices in children with SB + HC and Chiari II malformation compared with children with SB-HC without the Chiari II malformation. However, those participants had lower performance on all tests included in the study, and when the authors excluded participants with global cognitive impairment (VIQ <75), they did not observe significant differences in processing speed between these groups. Although this implies an alternative basis for impairments of processing speed, Iddon et al. (2004) also found that adults with HC-SB (and with VIQ >75) scored lower than participants with SB-HC and typical adults on simple measures of psychomotor speed, and also on measures of psychomotor speed within more complex psychological tasks involving sequencing. For a summary of research discussed in this subsection, please see Table 2.

Table 2 – Summary of presented literature on processing speed.

Processing speed	
Author	Findings
Ayr et al. (2005)	Lower processing speed index (WISC–III processing speed) in children with SBM + shunted HC compared with children with traumatic brain injury or orthopaedic injury.
Boyer, Yeates, and Enrile (2006)	Lower processing speed indices in children with SBM + shunted HC compared with healthy siblings. (N = 31 SBM + HC, N = 27 healthy siblings). All children with SBM + shunted HC scored >70 on the Wechsler Intelligence Scale for Children-Third Edition (WISC-III; Wechsler, 1991).
Calhoun and Mayes (2005)	Lower processing speed index and perceptual organisation index (PSI & POI) in N = 19 children with SB + HC compared with mean population value of 100.
Iddon et al. (2004)	Adults with HC-SB (and with VIQ >75) scored lower than participants with SB-HC and typical adults on simple measures of psychomotor speed, and also on measures of psychomotor speed within more complex tasks involving sequencing.
Jacobs et al. (2001)	Children with SBM + HC had lower estimates of processing speed than typically-developing children.
Lindquist et al. (2011)	Majority of adults with HC had results below average on the PSI. Out of the 24 adults with HC, six had a low result, and eight in the lower normal zone while 10 were normal or over.
Vinck et al. (2006)	Children with SB and Chiari II malformation had lower processing speed compared with children with SB without Chiari II malformation. However, when participants with global cognitive impairment (VIQ <75) were excluded from the study, no significant differences in processing speed were observed.

PSI: processing speed index from the Wechsler Intelligence Scale for Children-Third Edition (WISC-III; Wechsler, 1991).
VIQ: Verbal Intelligence Quotient.

2.3.2.2. Attention. HC has been associated with difficulties in attention, evidenced in the finding that children with congenital HC have a greater incidence of Attention Deficit Hyperactivity Disorder (ADHD) compared to the typical population (Burmeister et al., 2005). Fletcher et al. (1997) also found that approximately 50% of children with arrested HC and 47% with shunted HC had attentional problems. However, the type of attentional problems experienced by children with HC appears to be different from ADHD. For example, Brewer et al. (2001) found that on a sustained attention task, the reaction times of children with ADHD slowed over the number of blocks compared with typically-developing children, while children with HC were slower than typically-developing children at the beginning, but at similar level to typically-developing children towards the end of the task. Children with ADHD also had a greater number of perseverative errors on the Wisconsin Card Sorting task than children with HC or typical children. Children with HC on the other hand, had more non-perseverative errors and were more likely to fail to maintain a set once a category was achieved. Attentional impairments have been explicitly linked to the aetiology of HC – in typical foetal development, the occipital horns of the lateral ventricles enlarge at a faster rate than other ventricular areas, and so increased CSF pressure and ventricular volume in HC produces damage that spreads in a posterior to anterior direction (Brewer et al., 2001; Fletcher et al., 1996; see also Van Roost, Solymosi, & Funke, 1995). This has led to the suggestion that HC leads to impairments in posterior attention (Brewer et al., 2001; Dennis et al., 2005a, 2005b, 2006; Posner, Petersen, Fox, & Raichle, 1988; Rose & Holmbeck, 2007).

Posner's model of attention (Posner, 2016; Posner & Petersen, 1990) describes two separate but interconnected systems, distinguished by function and cerebral locus. The anterior system is responsible for goal-directed and volitional attentional allocation, maintaining vigilance and sustaining attention, and is mainly dependent on frontal and parietal areas. A posterior system subserves largely bottom-up functions driven by environmental salience, including attentional orienting and focusing, selecting information from sensory input, attentional disengagement and shifting, and is dependent mainly on the midbrain and posterior parietal cortex. Children with SBM + HC have been proposed to have difficulty with attentional processes subserved by the posterior attentional system, namely stimulus-driven orienting and shifting attention (Brewer et al., 2001; Dennis et al., 2005a, 2005b, 2006; Posner et al., 1988; Rose & Holmbeck, 2007). However, they have also been found to struggle with anterior aspects of attention as well, including maintaining sustained attention (see review by Erickson et al., 2001). Swartwout et al. (2008) further found that children with SBM + HC had a greater number of omission errors on a continuous-performance vigilance task, compared to typically-developing children and children with HC due to aqueductal stenosis, and a greater number of commission errors than typically-developing children. Overall, this evidence indicates that HC is associated with widespread effects on attention.

There is evidence for attentional difficulties in adults with HC as well, particularly relating to switching – for example, they have been shown to score lower on the Wisconsin Card Sorting compared with adults with SB aperta and occulta (Barf

et al., 2003; attention switching is discussed further below in relation to executive function). Iddon et al. (2004) found that adults with HC scored lower than adults with SB and typically-developed control participants on attentional switching (i.e. on an intra dimensional/extra dimensional set shift task). There is, however, a lack of research investigating posterior attentional difficulties in adults with HC and further studies are required to characterise comprehensively the attentional difficulties experienced in HC across the lifespan. See Table 3 for a summary of the studies discussed in this section.

2.3.2.3. Learning and memory. HC appears to be associated with difficulties in several important aspects of both verbal and non-verbal learning and memory. Barf et al. (2003) found that adults with SBA + HC scored lower than those with SBA-HC or SB occulta on the Wechsler Memory Scale and a verbal learning test (total recall of list A across five trials, learning rate of additional items across the five trials, and delayed verbal memory), which, together, assess different types of memory, both immediate and delayed. Iddon et al. (2004) also found that adults with SB + HC had lower performance than those with only SB on immediate and delayed verbal recall and on both spatial and verbal learning (Hopkins Verbal Learning Test and the Cambridge Neuropsychological Test Automated Battery).

These sparings in verbal recognition memory in both adults (Dennis et al., 2007) and children (Lindquist, Persson, Uvebrant, & Carlsson, 2008; Yeates et al., 1995) have been previously reported. Implicit memory also appears to be relatively spared in children with SBM + HC (shunted) (Yeates & Enrile, 2005, see also Del Bigio et al., 2003 for findings on hydrocephalic rats), although this remains to be investigated in adults. See Table 4 for a summary of the literature presented here.

2.3.2.4. Language and verbal reasoning. Language is generally viewed as an area of relative strength for people with HC, although there is a varied profile within this domain. Language abilities have been studied extensively in children, where HC has been associated with proficiency in word decoding and pronunciation, difficulty in writing, memory (Barnes, Faulkner, & Dennis, 2001), adaptive use of context (Huber-Okrainec, Blaser, & Dennis, 2005; Tew, 1979), and certain aspects of reading comprehension (Barnes et al., 2001; Barnes, Faulkner, Wilkinson, & Dennis 2004). Hampton et al. (2013) found that children with HC had only small differences compared with typical control participants on reading and vocabulary measures, while Ayr et al. (2005) found that children with SBM + HC (shunted) did not differ from Traumatic Brain Injury and Orthopaedic Injury groups on a reading subtest. Vinck et al. (2006) found that children with SB and Chiari II malformation scored similarly on verbal skills than children with SB without the Chiari II malformation. On the other hand, Fletcher et al. (1997) found that children with shunted HC performed worse on language measures, with lower reading and spelling scores, and Dennis, Jacennik, and Barnes (1994) found that children with HC had difficulties with storytelling. Children with SB ± HC (38/49 were SB + HC and 11/49 were SB-HC) have also been shown to struggle with context-relevant speech compared with typical children (Tew, 1979), and children with SBM ± HC have been found to have difficulties with understanding idioms requiring context change, but not those which do not require contextual change, compared with same-age peers (Huber-Okrainec et al., 2005).

Barnes, Dennis, and Hetherington (2004) investigated whether difficulties in writing and reading comprehension that they had previously found in children with SB (e.g., Barnes et al., 2001; Barnes, Faulkner, Wilkinson, & Dennis, 2004) persist into adulthood. They found that, similarly to children, adults with SB + HC scored lower than the population mean on reading comprehension measures and on constructing written sentences, but with a relatively high reading accuracy. It is worth noting, however, that their writing measure may have been confounded by issues with writing speed, and was shown to be predicted by fine motor function. This may point to a role for SB in impairments of writing and, to our knowledge, there have been no published investigations of writing and reading abilities in adults with HC but without SB. See Table 5 for a summary of the literature presented here.

2.3.2.5. Spatial and motor abilities. Children with HC have been consistently observed to have lower visuospatial performance than typically-developing children (e.g., Dennis, Fletcher, Rogers, Hetherington, & Francis, 2002, 2005a, 2005b; Fletcher et al., 1997). One complication, however, is that it can be difficult to interpret the cause of spatial difficulties in HC, because many tasks rely upon a motor response (Dennis et al., 2002; Loveday & Edginton, 2011; Simms, 1987a). Children with HC have been shown to perform lower than controls on motor tasks (Hampton et al., 2013), fine motor coordination, visuospatial motor tasks (Fletcher et al., 1996, 1997), and action-based visual perception (Dennis et al., 2002). It has been argued that this is likely dependent on the presence of SB (Hetherington & Dennis, 1999), and the difficulties have also been found to persist into adulthood (Barf et al., 2003). It does, however, appear that spatial performance is impaired even on tests that do not rely on motor ability. For example, it has been found that children with HC had lower visuospatial performance on the judgement of line orientation test, which does not involve a motor component (Fletcher et al., 1995). Of note here is the suggestion that lower visuospatial performance in children with SB may be due to difficulties in visual matching rather than the spatial aspects of the task (Mammarella, Cornoldi, & Donadello, 2003).

Irrespective of the precise cause, HC has been associated with deficits in spatial memory, spatial learning, and spatial working memory. For example, Iddon et al. (2004) found that adults with HC had lower performance on measures of spatial working memory and spatial recognition memory than adults with SB-HC, and typical controls. Buckley and Smith (2013) found that adults with HC performed worse on spatial learning and memory tasks than typical controls, and young adults with SB + HC (shunted) have also been found to score lower on spatial memory measures, compared with typical controls and population estimates (Dennis et al., 2007). Not all aspects of spatial processing are impaired with HC however – for example, evidence suggests that some aspects of spatial processing, such as categorical coding of spatial relations

Table 3 – Summary of presented literature on attention.

Attention	
Author	Findings
Barf et al. (2003)	Adults with SB aperta and HC scored lower on the Wisconsin Card Sorting Task compared with adults with SB Aperta and SB Occulta without HC.
Brewer et al. (2001)	Children with HC made more non-perseverative errors and were more likely to fail to maintain a set once a category was achieved on a Wisconsin Card Sorting task than children with ADHD or typically-developing children. Children with ADHD had a greater number of perseverative errors on the Wisconsin Card Sorting task than children with HC or typical children. On a sustained attention task, children with HC were slower than typically-developing children at the beginning, but at similar level to typically-developing children towards the end of the task. Children with ADHD slowed over the number of blocks compared with typically-developing children. On a visual orienting and detection task, children with HC had slower RTs for invalid and null cues to targets presented in the left visual field as opposed to the right visual field (for covert attentional shifts; 100 ms).
Burmeister et al. (2005)	Children with congenital HC had a greater incidence of ADHD compared to the typical population (31% vs upper limit of 17% in the general population). This was mostly the inattentive type (23%, distractability, lack of focus, disorganisation), while other types (hyperactive, impulsive and combined) were at the level of the general population.
Fletcher et al. (1997)	Examined how many children with HC met psychometric criteria for and attention problems (CBLC Attention Problems Scale; Achenbach, 1991a). 50% of children with arrested HC and 47% with shunted HC had attentional problems compared with 30% in children born preterm with no hydrocephalus and 13% in full-term children.
Dennis et al. (2005a)	On a cued orienting task, responses were required to a stimulus which was validly or invalidly cued by either an endogenous (luminance change) or exogenous (arrow/word indicating direction) cue. Children with SBM + shunted HC were found to have slower RTs compared with age-matched typically-developing control participants, especially for invalid cues at the vertical (as opposed to horizontal) plane. Children with SBM + HC also had a higher cost of attentional disengagement (defined as the difference in median RT on valid and invalid trials) to exogenous cues in short interval from cue to target (200 ms).
Dennis et al. (2005b)	On a cued orienting task, responses were required to a stimulus which was validly or invalidly cued by an endogenous (luminance change) cue. For targets presented in the vertical plane, children with SBM ± HC showed greater attentional disengagement cost (misdirecting attention on invalidly-cued trials at 200 ms interval from cue to target) and smaller disinhibition of return (longer RTs to invalidly-cued trials on 1000 ms interval from cue to target) compared with age-matched, typically-developing controls.
Dennis et al. (2006)	Review paper discussing impairments of orienting attention in SBM.
Iddon et al. (2004)	Adults with HC scored lower than adults with SB and typical control participants on an intra dimensional/extra dimensional set shift task requiring attentional switching.
Rose and Holmbeck (2007)	Used the Cognitive Assessment System (CAS; Naglieri & Das, 1997) and the Behavior Rating Inventory of Executive Function (BRIEF; Gioia, Isquith, Guy, & Kenworthy, 2000) and found that children with SB showed more impairment on focused visual attention (CAS Number Detection & CAS Receptive Attention), but no differences on a measure of sustained attention (BRIEF Sustain).
Swartwout et al. (2008)	Children with SBM + HC had a greater number of omission errors on a continuous-performance vigilance task, compared to typically-developing children and children with HC due to aqueductal stenosis, and a greater number of commission errors than typically-developing children.

RT: reaction time.

Table 4 – Summary of presented literature on learning and memory.

Learning and memory	
Author	Findings
Barf et al. (2003)	Adults with SBA + HC scored lower than those with SBA-HC or SB occulta on the Wechsler Memory Scale and a verbal learning test (total recall of list A across five trials, learning rate of additional items across the five trials, and delayed verbal memory).
Dennis et al. (2007)	Young adults with SBM + HC did not differ on episodic immediate verbal memory, which involved story recognition.
Iddon et al. (2004)	Adults with SB + HC had lower performance than those with SB only on immediate and delayed verbal recall and on spatial and verbal learning (Hopkins Verbal Learning Test and the Cambridge Neuropsychological Test Automated Battery).
Lindquist et al. (2008)	No differences in performance on an auditory-verbal recognition between children with HC only, children with SBM + HC, and age- and gender-matched typically-developing children, all of whom had average results according to test norms.
Yeates and Enrile, 2005	Children with SBM + HC (shunted) showed a perceptual and semantic priming effect of similar magnitude to that of children with severe traumatic brain injuries or orthopaedic injuries.
Yeates et al. (1995)	Children with SBM + HC (shunted) were found to have similar verbal recognition as typical children (matched on age, gender, ethnic background, and whenever possible on the standard score of Vocabulary subtest of Wechsler Intelligence Scale for Children). However, children with SBM + HC had worse verbal recall than typical children (California Verbal Learning Test).

Table 5 – Summary of presented literature on language and verbal reasoning.

Language and verbal reasoning	
Author	Findings
Ayr et al. (2005)	Children with SBM + HC (shunted) did not differ from Traumatic Brain Injury and Orthopaedic Injury groups on a reading subtest.
Barnes, Dennis, and Hetherington (2004)	Adults with SB + HC scored lower than the population mean on reading comprehension measures and on constructing written sentences, but higher on word identification. Reading and listening comprehension did not significantly differ from the population mean.
Barnes et al. (2001)	Children with HC were faster at word identification than passage comprehension. However, they had a smaller effect of spelling-sound regularity compared with typically-developing children.
Barnes, Faulkner, Wilkinson, and Dennis (2004)	Children with HC were poorer than typically-developing children at suppressing contextually-irrelevant meanings. While they were able to integrate previously-read information to understand a new sentence, they were more disadvantaged with greater amount of text between the meaningful and ambiguous sentences.
Dennis et al. (1994)	Children with HC produced oral texts which were less cohesive and less coherent compared with typically-developing children.
Fletcher et al. (1997)	Children with shunted HC performed worse than those with arrested HC on language measures, including reading, writing, and spelling.
Hampton et al. (2013)	Children with HC had only small differences compared with typical control participants on reading and vocabulary measures (compared with larger differences on spatial and motor tasks, and verbal memory).
Huber-Okrainec et al. (2005)	Children with SBM ± HC showed difficulties with the adaptive use of context: they were found to have difficulties with understanding idioms requiring context change, but not those that do not require contextual change, compared with same-age peers.
Tew (1979)	Children with SB ± HC (38/49 were SB + HC and 11/49 were SB-HC) had difficulties in the adaptive use of context: they were found to struggle with context-relevant speech compared with typical children.
Vinck et al. (2006)	Children with SB and Chiari II malformation had similar scores on verbal skills as children with SB without the Chiari II malformation.

Table 6 – Summary of presented literature on spatial and motor abilities.

Spatial and motor abilities	
Author	Findings
Barf et al. (2003)	Adults with SBA + HC had longer motor responses (time taken to lift a finger from the rest button to touching the reaction button) than adults with SB occulta.
Buckley and Smith (2013)	Adults with HC performed worse on spatial learning and memory tasks than typical controls, including on tasks of probabilistic cueing, route learning, and path integration.
Dennis et al. (2002)	Children with SB + HC had lower visuospatial performance than typically-developing children, particularly for action-based visual perception.
Dennis et al. (2005a)	Children with SBM + HC (shunted) had a higher cost of attentional disengagement and slower reaction times on a cued orienting task compared with age-matched typically-developing control participants (especially for invalid cues at the vertical plane).
Dennis et al. (2005b)	Children with SBM ± HC showed greater attentional disengagement cost and a smaller disinhibition of return on a cued orienting task compared with age-matched, typically-developing controls.
Dennis et al. (2007)	Young adults with SB + HC (shunted) scored lower on spatial memory (Tic Tac, from the MicroCog™ battery of computerized tests of cognition), compared with population estimates.
Dennis and Barnes (2010)	Review paper on the cognitive profile of SBM ± HC, summarising the findings of studies that have investigated aspects of spatial processing. This includes the finding that categorical coding of spatial relations between objects (i.e. "to the right", "behind", or "next to") remain intact in children with SBM ± HC, and that they are relatively proficient at spatial orientating when using landmarks.
Fletcher et al. (1995)	Children with HC had lower visuospatial performance on the judgement of line orientation test, Berry test of visuo-motor integration, and fine motor coordination (Grooved Pegboard).
Fletcher et al. (1996)	Children with shunted HC performed lower than controls on a task of fine motor skills (Grooved Pegboard).
Fletcher et al. (1997)	Children with HC had lower visuospatial performance (Beery Test of Visual-Motor Integration) and fine motor skills (Grooved Pegboard and Purdue Pegboard) than typically-developing children.
Hampton et al. (2013)	Children with HC performed lower than controls on motor (Purdue Pegboard) and spatial (Judgement of Line Orientation) tasks.
Iddon et al. (2004)	Adults with HC had lower performance on measures of spatial working memory and spatial recognition memory than adults with SB-HC, and typical controls.
Mammarella et al. (2003)	Children with SB had lower performance on a visuospatial working memory task involving visual matching (the House Visual Span Task), but not spatial working memory (Forwards and Backwards Corsi Blocks). The authors suggest that lower visuospatial performance in children with SB may be due to difficulties in visual rather than the spatial aspects of the task.

between objects (i.e. "to the right", "behind", or "next to") remain intact in children with SBM ± HC, and they have also been reported to be relatively proficient at spatial orientating when using landmarks (Dennis & Barnes, 2010). See Table 6 for a summary of the literature presented in this section.

2.3.2.6. Mathematical ability. Children with HC have been found to exhibit only small differences when compared with typical children on mathematics problems (Hampton et al., 2013), although they still have been shown to struggle with particular aspects of mathematics and numeracy (Fletcher et al., 1997). For example, Barnes et al. (2002) found that while children with HC had similar levels of fact-retrieval and visuospatial mathematical errors as typical children, they displayed lower performance on geometry, mental computation, and applied mathematics measures. They were also found to make more procedural errors on a mathematics task compared with typical children. These findings have been echoed by Ayr et al. (2005), who found that children with SBM + HC (shunted) had similar numbers of knowledge-based (i.e. mathematical facts) and visuospatial errors compared with children with orthopaedic injuries, but had lower arithmetic performance than children with traumatic brain injuries and orthopaedic injuries. After accounting for age and group, arithmetic performance was predicted by working memory, processing speed, and declarative memory, in this study. Performance on a subtraction task was predicted by declarative memory and planning skills. Processing speed and declarative memory were stronger predictors for younger children while visuospatial skills were a stronger predictor for older children.

There is some indication that these difficulties persist into adulthood. For example, Dennis and Barnes (2002) found that adults with SB + HC showed difficulties with mathematical

Table 7 – Summary of presented literature on mathematical ability.

Mathematical ability	
Author	Findings
Ayr et al. (2005)	Children with SBM + HC (shunted) had similar numbers of knowledge-based (i.e. mathematical facts) and visuospatial maths errors, but lower arithmetic performance compared with children with orthopaedic injuries.
Barnes et al. (2002)	Children with HC had similar levels of fact-retrieval and visuospatial mathematical errors as typical children, however, they displayed lower performance on geometry, mental computation, and applied math measures. They also made more procedural errors on a mathematics task compared with typical children.
Dennis and Barnes (2002)	Young adults with SB + HC scored significantly lower than population mean adjusted for age and/or education on measures of mathematical reasoning, including computational accuracy and speed, mathematical problem-solving accuracy and speed, and functional numeracy.
Fletcher et al. (1997)	Preterm children with HC had lower arithmetic scores than preterm children with arrested HC.
Hampton et al. (2013)	Children with HC had only small differences in performance on mathematics problems compared with typical control participants (with larger differences on spatial and motor tasks, and verbal memory).

reasoning, including computational accuracy and speed, mathematical problem-solving accuracy and speed, and functional numeracy, however more research is needed to confirm this. See Table 7 for a summary of the literature presented in this section.

2.3.2.7. Working memory and executive function. HC results in difficulties with executive function. Fletcher et al. (1996) found that children with SB + HC showed some difficulties on executive tasks including the Stroop Task, Tower of London Task, and Wisconsin Card Sorting Task. Adolescents with HC (Mahone, Zabel, Levey, Verda, & Kinsman, 2002) and SB (Tuminello, Holmbeck, & Olson, 2012), have been found to have lower executive functioning than typically-developing adolescents, even when differences in intellectual functioning are controlled for (Rose & Holmbeck, 2007). Lindquist et al. (2008) investigated cognitive function in 36 children with HC, 16 of whom had SBM. This study found that IQ was related to short-term memory and executive function measures. The authors also found that patients scored significantly lower than controls on executive function tasks, which included verbal fluency, Tower of London Task, Rey–Osterrieth Complex Figure task, and the Trail-Making Test. The HC only and SBM + HC groups did not differ on these measures, although it is important to acknowledge that similar scores do not necessarily mean that similar processing strategies were involved (e.g., featural vs holistic). In fact, Lindquist et al. (2009) found that children with SBM-HC performed higher than children with SBM + HC on the same tasks of executive function. They also found that when the two children in the SBM-HC group with IQ of less than 70 were excluded, the SBM-HC group performed at similar levels to controls on verbal fluency, the Tower of London task, and the Trail-Making Test (as well as immediate and delayed auditory-verbal learning and immediate spatial learning). This led the authors to conclude that SB alone is not a major determinant of neuropsychological impairments, and they commented on the large variability in cognitive outcomes for children with SBM-HC.

Studies have also used parent and teacher reports to investigate executive functioning abilities in children with HC. Rose and Holmbeck (2007) found that adolescents with SB (71% of whom were shunted for HC) scored lower on the multidimensional neurocognitive measure assessing focused visual attention and planning ability – the Cognitive Assessment System (CAS; Naglieri & Das, 1997) and the Behavior Rating Inventory of Executive Function (BRIEF; Gioia et al., 2000), a measure of executive functioning assessed by parent and teacher ratings. Adolescents with SB scored lower on the Initiate, Sustain, and Working Memory subscales of the BRIEF, with no differences on the Organize and Plan subscales. Differences on the Initiate and Working Memory subscales remained significant even when accounting for differences in general intellectual ability (Peabody Picture Vocabulary Test – Revised; Dunn & Dunn, 1981). Shunt status (acting as a proxy for the presence of HC) was related to the performance on the BRIEF (parent report). This study also found that executive functioning was related to measures of social competence: regardless of group, participants' average BRIEF scores predicted social skills based on parent and teacher reports (SSRS–Social Skills Rating System; Gresham & Elliott, 1990) and teacher reports of social competence (Self-Perception Profile for Children measure, or SPPC; Harter, 1985), while CAS also predicted one of the social competence measures (teacher-reported SPPC). Evidence also suggested that executive function mediated the relationship between group (SB ± HC versus typical) and parent-reported social competence and skill.

Tarazi, Zabel, and Mahone (2008) also found that children and adolescents with SB + HC (shunted) had lower executive functioning as rated by parents on the BRIEF, compared with typical individuals. In addition, there was an age by group interaction on the subscales comprising the Behavioural Regulation Index, which suggested that scores of typical individuals, but not the SB + HC group, increased with age. Similarly, Brown et al. (2008) also found that adolescents with SBM ± HC scored lower on the BRIEF metacognition subscales

Table 8 – Summary of presented literature on working memory and executive function.

Working memory and executive function	
Author	Findings
Barf et al. (2003)	Adults with HC scored lower on executive function Trail-Making Test Part B, and Wisconsin Card Sorting, compared with adults with SB aperta and occulta.
Brown et al. (2008)	Adolescents with SBM ± HC scored lower on the BRIEF metacognition subscales (Initiation, Working Memory, Plan/Organise, Organisation of Materials, but not Monitor subscale), controlling for Full Scale IQ and age. Metacognition index scores were significantly predicted by age, number of shunt revisions, and history of seizures.
Buckley and Smith (2013)	Adults with HC scored lower on verbal and visuospatial working memory indices than typical adults.
Boyer et al. (2006)	Children with SB + HC had lower working memory performance than typically-developing siblings.
Dennis and Barnes (2002)	Young adults with SB + HC had lower indices of working memory than population mean.
Dennis et al. (2007)	Young adults with SB + HC (shunted) had lower indices of working memory compared with the population mean.
Fletcher et al. (1996)	Children with SB + HC (shunted) showed some difficulties on executive tasks including the Stroop Task, Tower of London Task, and Wisconsin Card Sorting Task compared with children with arrested HC, non-HC patients, and typically-developing participants (however these differences were smaller relative to differences on fine motor skills).
Iddon et al. (2004)	Lower indices of spatial working memory in adults with HC (with or without SB) compared with adults with SB only, and control participants.
Lindquist et al. (2008)	Children with HC (16/36 also had SBM) scored significantly lower than controls on executive function tasks (verbal fluency, Tower of London Task, Rey–Osterrieth Complex Figure task, and the Trail-Making Test). However, there were no significant differences between the groups on visuospatial (Corsi block test) and auditory-verbal (Digit span) working memory.
Lindquist et al. (2009)	Children with SBM without HC performed higher than children with SBM + HC on the tasks of executive function which included verbal fluency, Tower of London Task, Rey–Osterrieth Complex Figure task, and the Trail-Making Test. When two children in the SBM without HC group with IQ < 70 were excluded, this group performed at similar levels to controls on verbal fluency, the Tower of London task, and the Trail-Making Test (as well as immediate and delayed auditory-verbal learning and immediate spatial learning).
Mahone et al. (2002)	Adolescents with HC had lower estimates of executive functioning (self-reported and parent-reported) than typically-developing participants.
Rose and Holmbeck (2007)	Adolescents with HC scored lower on executive functioning measures than typically-developing adolescents, even when differences in intellectual functioning were controlled for.
Tarazi et al. (2008)	Children and adolescents with SB + HC (shunted) had lower executive functioning as rated by parents on the BRIEF, compared with typical individuals. There was also an age by group interaction on the subscales comprising the Behavioural Regulation Index, which suggested that scores of typical individuals, but not the SB + HC group, increased with age.
Tuminello et al. (2012)	Adolescents with SB (71% of whom also had shunted HC) were found to have lower executive functioning than typically-developing adolescents.

(Initiation, Working Memory, Plan/Organise, Organisation of Materials, but not Monitor subscale), controlling for Full Scale IQ and age. Metacognition index scores were also significantly predicted by age, number of shunt revisions, and history of seizures. Tuminello et al. (2012) also examined executive function using the BRIEF and the CAS measures in adolescents with SB, 71% of whom also had shunted HC. They found that the patient group scored lower on these measures. Furthermore, lower sequencing ability on the CAS and executive functioning parent reports were related to higher levels of observed child dependency and lower levels of intrinsic motivation as reported by teachers. Performance measures of executive functioning on the other hand, predicted levels of child dependency and maternal intrusiveness.

Adults with HC have also been shown to have lower executive functioning than typical adults, although evidence of this is scant compared to that from child cohorts. In one apparent observation, Barf et al. (2003) found that adults with HC scored lower on executive function Trail-Making Test Part B, and Wisconsin Card Sorting, compared with adults with SB aperta and occulta.

The findings regarding working memory are also mixed. While most studies have found lower indices of working memory in HC and SB (Boyer et al., 2006; Buckley & Smith,

Table 9 – Summary of presented literature on mental imagery.

Mental imagery	
Author	Findings
Dennis et al. (2002)	Children with SB + HC scored lower than typically-developing children on a mental rotation task (non-significant trend).
Jansen-Osmann et al. (2008)	Children with SB ± HC (19/20 had shunted HC) had poorer performance on a mental rotation task than typically developing children.
Lehmann and Jansen (2013)	Children with HC had faster performance on mental rotation tasks than SB + HC children – the two groups were matched in age, cognitive processing speed, and gender. The reaction time of the group with SB-HC did not differ from that of control participants.
Lehmann and Jansen (2012)	Tested 19 children with SB ± HC (18/19 had shunted HC), half of whom received juggling training for 1 h per week, for a duration of 8 weeks. This resulted in decreased reaction time in the mental rotation task compared with children who did not receive the training.
Wiedenbauer and Jansen-Osmann (2007)	Tested 19 children with SB ± HC (18/19 had shunted HC) and typically-developing controls. Found that the group with SB ± HC had lower performance on a mental rotation task, however they improved after manual rotation training to a greater extent than controls.

2013; Dennis & Barnes, 2002; Dennis et al., 2007; Iddon et al., 2004), one study found evidence for comparable verbal and visuospatial memory span between children with HC, children with SB + HC, and typically-developing children (Lindquist et al., 2008). See Table 8 for a summary of the literature presented in this section.

2.3.2.8. Mental imagery. Mental imagery has primarily been examined in the context of mental object rotation, and is another example of an ability that has, hitherto, only been investigated in children with HC, and not in adults. Children with SB + HC (Dennis et al., 2002; Jansen-Osmann, Wiedenbauer, & Heil, 2008) have demonstrated poorer accuracy and speed of mental rotation than typically developing children. Interestingly, Lehmann and Jansen (2013) found that children with HC performed faster on mental rotation tasks than SB + HC children – the two groups were matched in age, cognitive processing speed, and gender. Furthermore, the reaction time of the group with SB-HC did not differ from that of control participants.

Importantly, mental rotation abilities appear to be malleable to training. Wiedenbauer and Jansen-Osmann (2007) found that mental rotation abilities could be improved by manual rotation training in children with SB ± HC, resulting in greater improvements than in typically developing children. Similar increases in mental rotation performance could also be seen after juggling training: Lehmann and Jansen (2012) tested 19 children with SB ± HC, half of whom received juggling training for 1 h per week, for a duration of 8 weeks. This resulted in decreased reaction time in the mental rotation task compared with children who did not receive the training. These studies provide encouraging indications that mental rotation speed can be improved in children with SB, and sit alongside other demonstrations (e.g., motor skills in SB: Edelstein et al., 2004) of the effective application of training as an intervention. See Table 9 for a summary of the literature presented in this section.

2.3.2.9. Emotion and behaviour. HC has been shown to be associated with elevated anxiety and depression levels (Lindquist, Carlsson, Persson, & Uvebrant, 2006; Loveday & Edginton, 2011; Oliveira, Rotta & Pinto, 2014; Urban & Rabe-Jabłońska, 2013). Zimmerman et al. (2020) found that 2.5% of children with HC scored in the severe depression range, 5% in the moderate range, and 12.5% in the mild range. On the other hand, 2.5% of the children scored in the severe anxiety range, 17.5% in the moderate range, and 20% in the mild range. They also found that headache burden was significantly associated with anxiety and fatigue. Dicianno et al. (2008) recommend that adults with SB should receive regular screening for depression though recent work from the authors of this paper suggests that anxiety in HC is more prevalent and problematic for the HC population (Edginton, Iddon, Loveday, Pickard, & Morgan, 2009). Wall, Kestle, Fulton, and Gale (2021) found that children with hydrocephalus had more difficulties with social-emotional functioning, compared with normative data. Despite these affective correlates, people with HC appear to demonstrate typical levels of emotional recognition. For example, whilst agenesis of the corpus callosum has been associated with poorer recognition of emotions from upright faces (Bridgman et al., 2014), Iddon et al. (2004) found that adults with HC were able to recognise displayed emotion, performing similarly to typically-developed adults and those with SB on the Eyes Test of Emotional Judgement. Similarly, children with SB + HC also have been shown to have typical face-recognition ability (Dennis et al., 2002).

HC has, however, been associated with difficulties in behavioural regulation (Dennis et al., 2006), disinhibition (Mazzini et al., 2003), and poorer self-monitoring (Lacy, Baldassarre, Nader, & Frim, 2012). Fletcher et al. (1997) found that children with shunted HC scored lower than children with arrested HC on adaptive behaviour scales, social competence, and cognitive development measures. Whilst clinical reports of children with HC describe individuals that are sociable, curious, and lively (Koval, 2004; Loveday & Edginton, 2011, see also Dennis et al., 2006), this is has not yet been explored as a potential strength in the literature (although see Dennis & Barnes, 2010). Further assessment of social cognition in people with HC is clearly of importance, and it has been suggested that measures of social cognition

Table 10 – Summary of presented literature on emotion and behaviour.

Emotion and behaviour	
Author	Findings
Dennis et al. (2006)	Review paper discussing impaired behavioural regulation in people with SBM ± HC.
Dennis et al. (2002)	Children with SB + HC performed as well as controls on a face-recognition task.
Dicianno et al. (2008)	Recommend that adults with SB should receive regular screening for depression.
Edginton et al. (2009)	Anxiety (rather than depression) in HC was more prevalent and problematic for the HC population.
Fletcher et al. (1997)	Children with shunted HC scored lower than children with arrested HC on adaptive behaviour scales, social competence, and cognitive development measures.
Iddon et al. (2004)	Adults with HC were able to recognise displayed emotion, performing similarly to typically-developed adults and those with SB-HC on the Eyes Test of Emotional Judgement.
Koval (2004)	State that children with SB are friendly and sociable.
Lacy et al. (2012)	Children with HC (shunted) had lower executive functioning and more self-monitoring difficulties than healthy peers.
Lindquist et al. (2006)	Found that a proportion of children with HC had elevated anxiety problems.
Loveday and Edginton (2011)	A review paper highlighting that HC is associated with elevated anxiety and depression levels and that children with HC are lively and talkative.
Mazzini et al. (2003)	The severity of HC was associated with behavioural disinhibition in people who developed hydrocephalus after severe traumatic brain injury.
Oliveira, Rotta, and Pinto (2014)	71% of 35 NPH patients had psychiatric disorders, including anxiety, depression, and psychotic syndromes.
Urban & Rabe-Jabłońska, 2013	Discussed a case study of a patient with depression complicated by HC.
Wall et al. (2021)	Found that children with hydrocephalus had more difficulties with social-emotional functioning using the Behavior Assessment System for Children, Third Edition (BASC-3), compared with normative populations.
Zimmerman et al. (2020)	2.5% children with HC scored in the severe depression range, 5% in the moderate range, and 12.5% in the mild range. Furthermore, 2.5% of the children scored in the severe anxiety range, 17.5% in the moderate range, and 20% in the mild range.

should be more widely included in studies on neuropsychological functioning in atypical development (Dennis et al., 2014). See Table 10 for a summary of the literature presented in this section.

2.4. *Higher-level/integrative behaviours*

Although more focused assays of cognitive abilities have extended our understanding of the impairments associated with HC beyond clinical batteries, they are still relatively constrained by their use of standardised tasks. The utility of such an approach is clear, since it allows for an accurate assessment of a given function, and it is underpinned by knowledge of how other populations tend to perform on the same tasks. However, they provide less insight into higher-level cognitive operations, or behaviours that rely on the integration of cognitive abilities. This is necessary to generate a more nuanced account of the cognitive impairments associated with a neurological condition, and they also provide a pathway to understanding how those impairments affect everyday function in the real world, rather than performance on isolated and rather abstract measures (see, for example: Humphreys & Riddoch, 2013). Few published studies seem to have moved beyond the clinical characterisation of spared and impaired functions, but here we describe those that have attempted to understand more complex behaviours. Interestingly, they seem to have converged on understanding exploratory behaviour within a spatial context, thus providing insights into some aspects of everyday living for people with HC.

We have already outlined that HC is associated with difficulties in spatial memory and learning (Buckley & Smith, 2013; Dennis et al., 2007; Iddon et al., 2004). Its impact on more complex behaviour is anecdotally confirmed by patients, carers, and clinicians, who report that people with HC often experience difficulties in everyday navigation, including severe disorientation, problems with following directions, lack of awareness of spatial locations (even when driving), getting lost when routes or landmarks are changed, and difficulty retracing routes. This, in turn, leads to significant stress, anxiety, and worry. Despite this, investigations into real-world spatial behaviours have been scarce, with the following exceptions. Simms (1987a) tested in-car route memory of young adults with SB + HC, who were asked to direct the driver to follow a previously-observed route. They found that participants with SB + HC made a greater number of errors on this task, and had more difficulty when marking the route taken on a map than typical participants. Similarly, Wiedenbauer and Jansen-Osmann (2006) found that, controlling for performance IQ, children with SB ± HC (all but one

participant were SB + HC, shunted) had lower route-learning performance than PIQ-matched typically-developing children, taking more trials to reach criterion and making more incorrect turns at test. Landmark memory, on the other hand, seemed unimpaired as the clinical sample recalled a similar number of landmarks as typical children. The authors also found that age at which children learned to walk was correlated with the number of learning trials required to reach criterion, but not with the number of errors made on the maze route-learning task during the test trial, or the number of landmarks recalled. This suggests that motor ability impacts some aspects of spatial navigation and particularly route-learning abilities. These findings were replicated by Jansen-Osmann et al. (2008), who found that the age at which children learned to walk was correlated with performance in the same maze route-learning task in children with SB ± HC (all but one participant were SB + HC, shunted). In addition, the age at which children learned to walk was also correlated with small-scale visuospatial abilities, including spatial memory and attention to detail (Children's Embedded Figures Test). Together, these observations suggest an important role for motor ability development on further spatial cognition, echoing the arguments made by Dennis and colleagues (Dennis & Barnes, 2010; Dennis et al., 2006; Fletcher & Dennis, 2009).

Buckley and Smith (2013) reported a study of spatial navigational behaviour, and some of its underlying abilities, in a sample of adults with HC-SB. Their battery included: a route learning task, where participants watched a video of a route, were asked to draw it on an unmarked map, and were then tested on recognition memory for landmark objects; a path integration task, in which they were blindfolded, led along two arms of a right-angled triangle, and then asked to return to the starting location along the hypotenuse; and, a monitor-based search task in which the location of a hidden target was defined by a probabilistic cue (i.e. 80% of trials in one hemifield). The authors found that people with HC scored lower than typical adults on all of these spatial tasks. They also scored lower on verbal working memory, visuospatial working memory, and fluid intelligence (i.e. Raven's Progressive Matrices) measures. Fluid intelligence significantly predicted performance on the spatial learning task and route learning, while spatial working memory predicted performance on the path integration task, providing some insight into the cognitive abilities that underpin differences in navigational behaviour.

Naturally, the more heterogeneous the measure, the more likely one is to find some discrepancies between findings. It is, therefore, interesting to note that whilst Wiedenbauer and Jansen-Osmann (2006) reported intact landmark memory, this was an area of relative weakness for participants in the Buckley and Smith (2013) study. That said, there are some core differences between studies that may account for this. For example, the former investigated children with SB + HC, while the latter tested a sample of adults with HC-SB. Wiedenbauer and Jansen-Osmann (2006) required participants to actively explore a desktop virtual environment and then probed recall for landmarks whilst participants were retracing their steps. In contrast, Buckley and Smith (2013) presented participants with a video of a route through a virtual environment and then probed recognition memory for landmarks (i.e. targets vs. foils). This may point to useful distinctions between recognition and recall, and it is also possible that participants in the Wiedenbauer and Jansen-Osmann (2006) paradigm also benefited from the influence of context to aid recall, whereas items in the Buckley and Smith (2013) study were probed in isolation.

Another real-world spatial behaviour that has been examined in people with HC is driving. Simms (1991) found that recently-qualified drivers with SB + HC had taken longer to learn to drive than typically-developing control participants, and also took longer to become independent drivers after passing the test. They also had more difficulty with planning, remembering, and following routes compared with typical participants and individuals with cerebral palsy. In addition, participants with SB + HC reported a greater number of difficulties during their tuition than the other groups, which fell into three main categories: (1) road procedures, e.g., reversing, navigating junctions; (2) car controls, e.g., steering, indicating; and, (3) visual/cognitive factors, e.g., dazzle from oncoming cars, concentration. In addition, 17% of SB + HC drivers reported that they often travelled along unfamiliar routes, compared to 29% of cerebral palsy participants and 47% of typically-developing participants. Participants with SB + HC also had difficulties with route planning and route following, including drawing a route map, remembering a route, finding alternative routes, planning a route, following directions, and finding places on a map. Both cerebral palsy and SB + HC participants had more difficulties learning to operate car controls than typical adults.

Simms (1987b) also explored driving outcomes in young adults with SB and HC using a longitudinal approach. The participants in this study were first assessed for driving potential, and then a three-year follow-up investigated their driving status. It was found that nearly half of this group passed their driving tests. However, intriguingly, driving status did not relate to neuropsychological performance. This suggests that there may be a gap in what cognitive tests can tell us about the real-world difficulties of people with HC and their underlying abilities. Equally, it may be worth considering whether the self-report nature of Simms's (1991) study could have affected the results. If so, these findings would indicate that cognitive abilities as measured by neuropsychological testing are poor predictors of self-reported driving difficulties – perhaps partly due to low insight into cognitive abilities, as well as differences in the level of measurement (i.e. behaviourally-assessed cognitive skill versus real-world behavioural outcomes). These findings feed into the debates within cognitive neuropsychology about the usefulness of neuropsychological measures for predicting real-life performance, and the need to design instruments that can bridge the gap between diagnosis and behavioural functioning (Dennis et al., 2014; Rabin, Burton, & Barr, 2007; see Olson, Jacobson, & Van Oot, 2013 for a discussion specifically on paediatric neuropsychological measures).

Nevertheless, evidence from Buckley and Smith (2013), Wiedenbauer and Jansen-Osmann (2006), and Jansen-Osmann et al. (2008) suggests that neuropsychological assessment can be useful for elucidating functional relationships between different cognitive domains and complex

Table 11 – Summary of presented literature on higher-order behaviours.

Higher-order behaviours	
Author	Findings
Barnes, Dennis, and Hetherington (2004)	In young adults with SB + HC, writing fluency predicted social language communication, personal living independence, and community living indices on the Scales of Independent Behaviour-Revised. Writing performance was itself predicted by VIQ, verbal memory span (forward), working memory (backward digit span), and fine motor function.
Buckley and Smith (2013)	Adults with HC-SB had lower performance on a route learning task, path integration task, and a monitor-based search task in which the location of a hidden target was defined by a probabilistic cue (i.e. 80% of trials in one hemifield), compared with typically-developed adults. They also scored lower on verbal working memory, visuospatial working memory, and fluid intelligence (i.e. Raven's Progressive Matrices) measures. Fluid intelligence significantly predicted performance on the spatial learning task and route learning, while spatial working memory predicted path integration performance.
Jansen-Osmann et al. (2008)	In children with SB ± HC (all but one participant were SB + HC, shunted), the age at which children learned to walk was correlated with performance in a maze route-learning task. The age at which children learned to walk was also correlated with small-scale visuospatial abilities, including spatial memory and attention to detail (Children's Embedded Figures Test).
Loomis et al. (1994)	In adults with SB, verbal memory, attention and concentration, and adaptive behaviour were related to paid employment status.
Simms (1987a)	Young adults with SB + HC made a greater number of errors than typically-developing participants on a task in which they were asked to direct the driver to follow a previously-observed route. They also had more difficulty when marking the route taken on a map.
Simms (1987b)	Nearly half of young adults with SB + HC passed their driving test over the 3-year follow-up period, but driving status did not relate to performance on neuropsychological tests.
Simms (1991)	Recently-qualified drivers with SB + HC were found to have taken longer to learn to drive than typically-developing control participants, and also took longer to become independent drivers after passing the test. They also had more difficulty with planning, remembering, and following routes compared with typical and cerebral palsy participants. In addition, participants with SB + HC reported a greater number of difficulties during their tuition, and after passing their test travelled along unfamiliar routes less often as well as having a greater number of difficulties with route planning and route following.
Wiedenbauer and Jansen-Osmann (2006)	Controlling for performance IQ, children with SB ± HC (all but one participant were SB + HC, shunted) were found to have lower route-learning performance than PIQ-matched typically-developing children, with unimpaired landmark memory. Age at which children learned to walk was correlated with the number of learning trials required to reach criterion, but not with the number of errors made on the maze route-learning task during the test trial, or the number of landmarks recalled.

behaviours, both in HC and in typical cognition. Loomis, Lindsey, Javornisky, and Monahan (1994), for example, found that skills in verbal memory, attention and concentration, and adaptive behaviour (i.e. sets of practical, conceptual, and social abilities that enable people to cope with environmental demands, such as awareness and understanding of one's surroundings, ability to engage in regular economic and social life, ability to maintain one's basic health and safety) were related to paid employment status in adults with SB. A further study of adults with SB + HC by Barnes, Dennis, and Hetherington (2004) found that writing fluency predicted social language communication, personal living independence, and community living indices on the Scales of Independent Behaviour-Revised (Bruininks, Woodcock, Weatherman, & Hill, 1996). Writing performance was itself predicted by VIQ, verbal memory span (forward), working memory (backward digit span), and fine motor function, which further strengthens links between assays of behavioural substrates. See Table 11 for a summary of the literature presented in this section.

2.5. *Theoretical questions*

As may already be apparent, research into HC has predominantly (and incorrectly) treated participants with HC and SB as a single population (Loveday & Edginton, 2011), making interpretation of the underlying cause of any difficulties unclear. This applies to much of the research discussed in the previous sections, including factors such as processing speed, implicit memory, reading, writing, mathematical skills, and

mental rotation. Furthermore, there is a theoretical disagreement on whether HC or SB is primarily responsible for the cognitive difficulties seen in this group. The work by Maureen Dennis and colleagues, discussing the varied cognitive profile of people of HC (Dennis & Barnes, 2010; Dennis et al., 2006; Fletcher & Dennis, 2009), proposes that, in individuals with SBM + HC, SB is primarily responsible for the cognitive difficulties experienced, with HC being a moderator for the severity of these difficulties. Similarly, Vinck et al. (2006) argue that because HC is not independent of SB, HC alone cannot explain the deficits in the cognitive profile of this group. This is supported by Lehmann and Jansen (2013), who found that children with HC performed faster on mental rotation tasks than children with SB + HC, suggesting effects of SB on performance. Furthermore, Dennis and colleagues (Dennis & Barnes, 2010; Dennis et al., 2006; Fletcher & Dennis, 2009) propose that physical difficulties associated with SB (i.e., movement of the limbs or eyes) affect the development of the neurocognitive system. These difficulties are suggested to prevent children from exploring, learning, and integrating information and, therefore, affect the development of abilities such as memory, visual search, coordination of movement, and attention.

However, an alternative position in the literature is that HC, rather than SB, is directly responsible for the cognitive difficulties experienced in these patients. In the large-scale investigations of the HC cognitive profile in adulthood, it has been found that people with HC, with and without SB, perform worse than people with SB only, or typically developing controls (Barf et al., 2003; Iddon et al., 2004; see also Loveday & Edginton, 2011). This is supported by findings in developmental cohorts. For example, Lindquist et al. (2008) found that children with SB + HC, and children with HC only, had lower performance on verbal and visuospatial tasks than typically-developing children. Lindquist et al. (2009) found that when the two children in the SBM-HC group with IQ of less than 70 were excluded, the SBM-HC group performed at similar levels to controls on measures of short and long-term memory and executive functions. In addition, Hetherington and Dennis (1999) found that the extent of the motor difficulties varies with the cause of the HC: motor difficulties appear to be greater when HC is of congenital onset – i.e. caused by SB or aqueductal stenosis, compared with infantile-onset HC caused by infection, adhesions, or intraventricular haemorrhage. This evidence suggests that HC is primarily responsible for the cognitive difficulties experienced, while SB is primarily responsible for any motor and sensory-related difficulties. This proposal is also consistent with the differential physical effects of SB and HC, although further research is required to establish which is the case.

Perhaps more fundamentally, it is unclear whether each of these theoretical positions outlined above has been stated with an awareness of the existence of the other, since they have both been proffered in relative isolation and without reference to the alternative. As a result, this review is, perhaps, the first time that their opponent positions have been identified.

There is one other theoretical perspective that bears outlining here. Helen Williams (2008) proposed a theory that unifies the causes and development of HC and SB, based on the mechanical and biological aspects of pre-natal development. The key to this theory is the close coupling of CSF and venous pressure. If one is increased (e.g., due to movement), the other will decrease to maintain homeostatic levels of pressure in the central nervous system (CNS). An obstruction (e.g., due to Chiari malformation) will create greater pressure fluctuations as the pressure in the head and spine will fluctuate independently of each other. Localised pressure increases will have an adverse effect on venous drainage and rapid pressure increases can lead to obstructions, which in turn can lead to greater CSF accumulation and further pressure increase. As a result of this circular nature between increase in venous pressure and CSF accumulation, HC will progress. Raised spinal pressure will result in syringomyelia and raised cranial pressure will result in HC. Raised CNS pressure also opposes neural tube closure. Where raised pressure interacts with mesodermal growth or progenitor-cell migration restriction, a failure of neural tube closure will occur. Abnormal mesodermal growth at different stages of development is proposed to result in different morphological features of NTD closure.

By explicitly identifying these competing accounts, we hope to inspire a more integrative approach to the study of HC, and one that overtly brings theoretical debate to the table. A related problem in the characterisation of the cognitive effects of HC is that much of the relevant research has been conducted on children, rather than on adults. This includes extant reviews on the cognitive profile of HC in clinical presentation (Dennis & Barnes, 2010; Dennis et al., 2006; Fletcher & Dennis, 2009). It is, therefore, essential that accounts, and the theoretical claims outlined above, are verified in adult samples. While there are indications that some difficulties (e.g., processing speed, attentional switching, memory, language, spatial, and motor abilities) persist into adulthood, the research into this is relatively sparse. Difficulties experienced by adults may be different from those identified in children as some skills may improve with maturation, and adults may be able to employ additional compensatory strategies to mitigate against deficits. It is also clear that a longitudinal approach would be particularly beneficial in this context.

An additional aspect of research into HC may also be of relevance here, and this is that most studies that have selected participants based on IQ frequently exclude participants with a verbal IQ of 70 points or lower. While we acknowledge that a minimum level of IQ is required for people to complete neuropsychological measures at all, including only a subset of patients may lead to an underestimation of the severity of the condition and its effects (a similar point is made by Barf et al., 2003). People with higher IQ, for example, may be able to employ compensatory strategies for their deficits more easily. The complex relationship between IQ and ability in HC and SB can be illustrated by the study by Vinck et al. (2006). They found that whilst children with SB + HC with Chiari II malformation had lower processing speed than children with SB + HC but without Chiari II malformation, those differences disappeared when they excluded participants with VIQ <75. Similarly, Lindquist et al. (2009) found that when participants with full-scale IQ of less than 70 were excluded, differences in short-term memory and executive function disappeared between children with SBM without HC

and typically-developing groups. Accordingly, it may be recommendable for published studies to clearly report how many people were excluded according to IQ, so that the relative proportion within cohorts can be accurately gauged. However, in the long term, it would be most useful to devise measurements that are appropriate for a broader range of abilities, so that the severity of cognitive impairments might be more comprehensively characterised.

2.6. *Relationships between cognitive and neural function*

Although the neurological footprint of HC is far from localised, the condition does offer some opportunity to associate neural damage with cognitive function. Ventricular enlargement is often used as a marker for severity of HC and yet, despite some evidence to the contrary (Venkataramana & Mukundan, 2011), ventricle size does not appear to be correlated with symptom severity or cognitive function (Buckley et al., 2012; Warf et al., 2009). Instead, cognitive ability appears to be better predicted by brain volume, which does not itself correlate with ventricle size (Mandell, Kulkarni, Warf, & Schiff, 2015). Corpus callosum damage, on the other hand, does appear to be linked with cognitive function, and callosal abnormalities are a key hallmark of HC (Jinkins, 1991). Early studies investigating the relationship between the size of the corpus callosum and cognitive function found relationships with intelligence measures, visuospatial abilities, and motor function (Fletcher et al., 1992, 1996). However, these relationships are not necessarily straightforward. Dennis et al. (2005c) found a correlation between corpus callosum volume and a leftward bias in line bisection in typically-developing children, but not in children with SBM, suggesting that the relationships between brain areas and function can vary between patient and typically-developing populations. The complexity of these relationships is further illustrated by the findings of Mataró et al. (2007), who reported a correlation between corpus callosum size and cognitive functioning in adult patients with the normal pressure variant of HC (which we shall turn to next), who were due to undergo shunting. They found that the genu of the corpus callosum was related to psychomotor speed, clinical and daily life activity functioning, while the rostral body and the splenium correlated with frontal functions. They also found that the size of the corpus callosum increased after shunting. Importantly, greater increases in corpus callosum size were related to lesser improvement in cognitive function. Investigating HC can, therefore, help to uncover and elucidate these complex relationships between neural and cognitive function. This would also benefit from more strategic comparisons, such as comparisons between patients with discrete callosal lesions and patients with HC.

3. Normal Pressure Hydrocephalus

At the beginning of this review we alluded to the fact that HC is a singularly interesting neurological condition since it can be present from birth (or soon thereafter), but can also suddenly appear in adulthood, in the form of Normal Pressure Hydrocephalus (NPH). NPH was first described by Hakim and Adams (1965), and its occurrence is usually secondary to infection, tumour, or trauma (Daou, Klinge, Tjoumakaris, Rosenwasser, & Jabbour, 2016), although a more common and less well-understood variant is idiopathic in origin (Hellström et al., 2007; Philippon, 2005; Relkin, Marmarou, Klinge, Bergsneider, & Black, 2005). NPH is associated with ventricular enlargement (Fig. 2), normal-pressure CSF, and three main behavioural symptoms: gait/balance disturbance, cognitive disturbance/dementia, and urinary incontinence (Hellström, Klinge, Tans, & Wikkelsø, 2012; Passler et al., 2017), which are used as a basis for diagnosis, together with radiological evidence from CT or MRI scanning (Nakajima et al., 2021; Relkin et al., 2005). It has been considered a separate disorder from congenital or childhood-onset HC although, as in congenital/early-onset HC, there appears to be a possible genetic contribution to NPH (Cusimano et al., 2011; McGirr & Cusimano, 2016).

3.1. *Prevalence and physiology*

The prevalence estimates of NPH tend to vary (Zaccaria et al., 2020), and this is likely to be related to the comorbidities associated with NPH when it is secondary to more apparent or acute circumstances. Most international studies of prevalence have, therefore, taken particular focus on the idiopathic variant (iNPH: Andersson et al., 2019; Brean & Eide, 2008; Iseki et al., 2009; Klassen & Ahlskog, 2011; Pyykkö et al., 2018; Tanaka, Yamaguchi, Ishikawa, Ishii, & Meguro, 2009). A 10-year follow-up of a prospective population-based study estimated the incidence of iNPH for people at 70 or over to be at 1.2/1000 people per year (Iseki et al., 2014). The higher rates of iNPH found by Iseki et al. (2014) compared with other estimates, combined with the observation that most of the prevalence studies of iNPH are based on examining incident cases in hospitals, led Martín-Láez, Caballero-Arzapalo, López-Menéndez, Arango-Lasprilla, and Vázquez-Barquero (2015) to suggest that iNPH is severely underdiagnosed.

Most of the research on the physiology of NPH has focused on the features of the CSF using lumbar punctures, and the ventricular and white matter changes using imaging, in order to aid diagnosis (e.g., Capone, Bertelson, & Ajtai, 2019; Kimura, Tanaka, & Yoshinaga, 1992; Mataró et al., 2007; Pyykkö et al., 2014; Raftopoulos et al., 1992; Silverberg, Mayo, Saul, Rubenstein, & McGuire, 2003; Tanaka, Kimura, Nakayama, Yoshinaga, & Tomonaga, 1997; Wang, Zhang, Hu, Ding, & Wang, 2020). Lumbar drainage involves removal of 30–60 mL of CSF, which can itself lead to symptom improvement (Gavrilov et al., 2019). As with early-onset HC, a hallmark feature of NPH is enlarged ventricles (not attributable to general aging, cerebral atrophy, or congenital enlargement) (Relkin et al., 2005). The brain morphology indicative of NPH includes: smaller collosal angle, narrowing of the posterior half of the cingulate sulcus, wider Sylvian fissures, focally widened sulci, third ventricular walls that are parallel or bow outward (Capone et al., 2019), and Evan's index of >0.3 or comparable (Relkin et al., 2005). People with NPH have larger ventricles than individuals with other types of dementias (Kitagaki et al., 1998). As in early-onset/congenital HC the ventricle expansion affects periventricular areas, particularly periventricular white matter and periventricular grey matter

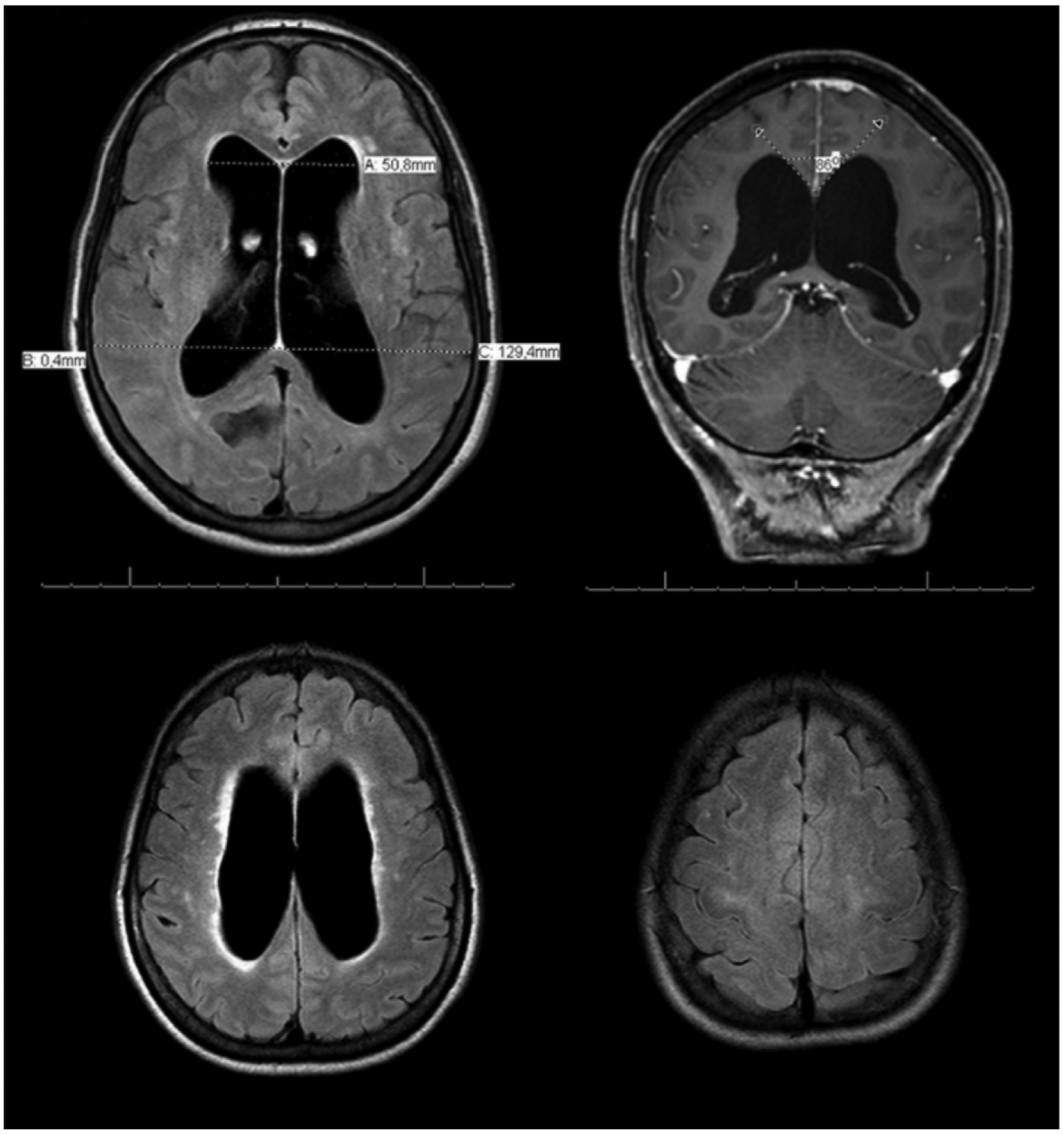

Fig. 2 – MRI scan of a patient with Normal pressure hydrocephalus. Reprinted from Oliveira, Nitrini, and Román (2019).

such as frontal cortical and subcortical areas, hippocampus, thalamus, and basal ganglia (Kazui, 2008; Peterson et al., 2019). For further information on clinical manifestations of NPH, see a review by Gavrilov et al. (2019).

3.2. *Treatment and diagnosis*

NPH can be treated with a shunt. As with early-onset HC, shunting can lead to improvement and even reversal of symptoms, but complications can also occur (Bugalho, Alves, & Ribeiro, 2013; Caixeta, 2007; Chaudhry et al., 2007; Daou et al., 2016; Hebb & Cusimano, 2001; Mataró et al., 2003, 2007; Oliveira et al., 2019). Shunting results in a high rate of success, particularly if patients receive shunts soon after diagnosis (Andrén, Wikkelsø, Tisell, & Hellström, 2014) and are selected based on likelihood of responsiveness (Marmarou, Young, et al., 2005; Poca et al., 2005; Stein, 2001; Williams & Malm, 2016). Endoscopic third ventriculostomy (ETV) has also been used in select cases with varied effectiveness, but further research is needed to determine its success relative to shunting (Oliveira et al., 2019).

It is recommended that a combination of clinical assessment, medical history, and neuroimaging should be used for diagnosis (Oliveira et al., 2019; Relkin et al., 2005). However, given that NPH commonly occurs in an elderly population, symptoms that resemble those of NPH (e.g., urinary problems; hip and knee pain affecting gait) are highly common and it is important to distinguish symptoms caused by NPH as opposed to typical aging (Malm et al., 2013). In addition, NPH can also co-occur with, and resemble, some neurodegenerative disorders common in the older population, including several forms of dementia (Cabral et al., 2011; Jingami et al., 2015; Krauss et al., 1997; Malm et al., 2013; Pomeraniec, Bond, Lopes, & Jane, 2016; Savolainen, Paljärvi, & Vapalahti, 1999; Tullberg et al., 2002). As a result, NPH can be difficult to diagnose (Relkin et al., 2005) and, as previously stated, this is further complicated by multiple comorbidities that can additionally occur with NPH. Hypertension and Type II

diabetes mellitus are the most common (Pyykkö et al., 2018), but they also include schizophrenia (Vanhala et al., 2019), cerebral palsy (Albright, Ferson, & Carlos, 2005), cerebrovascular disease (Bech-Azeddine, Høgh, Juhler, Gjerris, & Waldemar, 2007; Boon, et al., 1999), and cervical myelopathy (Naylor et al., 2020), among others (see Malm et al., 2013 for a full list of comorbidities). The presence of comorbidities can affect treatment outcomes (Bugalho et al., 2013). Given that NPH is treatable and early diagnosis may facilitate the best treatment outcomes (Andrén et al., 2014; Hejl, Høgh, & Waldemar, 2002), it is very important to distinguish NPH from other similar conditions.

3.3. *Cognitive function*

NPH usually presents with gradually worsening cognitive symptoms relating to memory recall, attention, executive functioning, and processing speed (e.g., Caltagirone, Gainotti, Masullo, & Villa, 1982; De Mol, 1978, 1986; Hurley, Bradley, Latifi, & Taber, 1999; Kanno et al., 2012; Krauss & Strupp, 2003), as well as spatial and motor symptoms (Adams, 1975; De Mol, 1977, 1978). However, very few studies have directly investigated cognitive functioning in NPH relative to typical adults. An exception to this is a report by Hellström et al. (2007), who found that people with iNPH had consistently lower performance on a range of neuropsychological measures, compared with typical adults. Their battery included measures of: psychomotor speed and inhibition (Stroop task); wakefulness (target reaction time); manual dexterity and motor speed (Grooved Pegboard task); immediate and delayed verbal recall (Rey Auditory Verbal Learning Test); and working memory and working memory capacity (forwards and backwards digit span). They also found that performance on these measures correlated with the severity of physical symptoms, particularly gait, balance, and increase of daily sleep. The presence of additional health conditions (diabetes, hypertension, and cardiovascular disease) was associated with poorer performance on approximately half of these neuropsychological measures.

Perhaps not surprisingly, given the concerns about NPH diagnosis and treatment, most of the research on the cognitive function in NPH has focused on two main directions: (1) investigating the differences between NPH and other clinical conditions, for the purpose of more accurate diagnosis, and (2) how to best predict treatment outcomes.

3.4. *Comparison between NPH and Alzheimer's disease*

NPH has most frequently been compared to Alzheimer's disease (AD), typically revealing that people with NPH appear to be particularly impaired on measures of executive functioning, but have superior immediate and delayed verbal, visual, and auditory memory performance (Kanno et al., 2012; Kazui, 2008; Miyoshi et al., 2005; Ogino et al., 2006). Miyoshi et al. (2005) compared iNPH patients with age and gender-matched AD patients on cognitive function and gait-related measures. They found that people with iNPH had poorer performance on tests of executive function (Frontal Assessment Battery; Dubois, Slachevsky, Litvan, & Pillon, 2000) and verbal fluency, and performance also correlated significantly with the degree of gait impairment in patients with iNPH, measured by the number of steps taken to walk a distance of 20 m. A review paper by Kazui (2008) also noted that frontal lobe functions such as attention, psychomotor speed, verbal fluency, working memory, and executive function, appear to be disproportionately severely affected in patients with iNPH, while the impairments in memory are disproportionally mild, compared with AD. They also observed that iNPH is associated with relatively preserved recognition memory as opposed to recall. Similarly, Kanno et al. (2012) compared AD and iNPH patients and found that iNPH was associated with lower scores on tests of executive functioning, including the following measures: counting forwards and backwards; the Frontal Assessment Battery; verbal fluency; Stroop; and, digit span. Ogino et al. (2006) compared patients with iNPH and AD on standardised tests of neuropsychological functioning, which included: the cognitive part of the Alzheimer's Disease Assessment Scale, consisting of orientation and visuoconstruction subtests (Rosen, Mohs, & Davis, 1984); the Wechsler Memory Scale—Revised (general memory, delayed recall, attention/concentration); and, the Wechsler Adult Intelligence Scale-Revised (information, digit span, vocabulary, arithmetic, comprehension, similarities, picture completion, picture arrangement, block design, object assembly, digit symbol substitution, verbal IQ, performance IQ). They found that the iNPH group scored lower on the attention/concentration index of the Wechsler Memory Scale, as well as on the digit span, arithmetic, block design and digit symbol substitution subtests of the Wechsler Adult Intelligence Scale. On the other hand, patients with AD scored lower on the orientation subtest of the Alzheimer's Disease Assessment Scale and on measures of general memory and delayed recall of Wechsler Memory Scale. The findings of these studies are summarised in Table 12.

3.5. *Symptom improvement after shunting*

It is generally found that a large proportion of patients with NPH respond well to shunting, with rapid improvement of symptoms. Polled data from Hebb and Cusimano (2001) indicated that 59% shunted iNPH patients obtain a measurable benefit, with a 6% complication rate. A later investigation (Toma, Papadopoulos, Stapleton, Kitchen, & Watkins, 2013) indicated positive improvement in 71% of patients with an average of 1% mortality, with up to 82% success rate when considering data from 2005 to 2010 (articles published in the last five years of the dates of their literature search). Marmarou, Young, Aygok, Tsuji, Yamamoto, and Dunbar (2005) found that symptom improvement following lumbar drainage of CSF predicted shunt outcome with the accuracy of 88%. Motor symptoms are generally associated with the highest rates of improvement, whereas the improvement rate of cognitive symptoms is less certain and more variable (Petersen, Mokri, & Laws, 1985; Raftopoulos et al., 1994). For example, while Poca et al. (2005) found that 33% of shunted patients with NPH improved in their cognitive function, Thomsen, Børgesen, Bruhn, and Gjerris (1986) found this figure to be at 40%, and Mataró et al. (2007) at 65%. Similarly, Raftopoulos et al. (1994) found cognitive improvement after shunting in 66.6% of their patients after 1 year, whilst

Table 12 – Summary of presented literature on comparisons between NPH and Alzheimer's Disease.

Comparison between NPH and Alzheimer's Disease	
Author	Findings
Kanno et al. (2012)	iNPH was associated with lower scores on executive functioning measures (counting forwards and backwards; the Frontal Assessment Battery; verbal fluency; Stroop; and, digit span) compared with AD patients.
Kazui (2008)	Review paper noting that iNPH is associated with more severe impairments with frontal lobe functions such as attention, psychomotor speed, verbal fluency, working memory, and executive function, but disproportionally mild impairments in memory compared with AD. They also noted that iNPH is associated with relatively preserved recognition memory (as opposed to recall).
Miyoshi et al. (2005)	Participants with iNPH had poorer performance on tests of executive function (Frontal Assessment Battery; Dubois et al., 2000) and verbal fluency than age and gender-matched people with AD. Performance correlated significantly with the degree of gait impairment in patients with iNPH.
Ogino et al. (2006)	iNPH participants scored lower on the attention/concentration index of the Wechsler Memory Scale—Revised, as well as on the digit span, arithmetic, block design and digit symbol substitution subtests of the Wechsler Adult Intelligence Scale. Patients with AD scored lower on the orientation subtest of the Alzheimer's Disease Assessment Scale and on measures of general memory and delayed recall of Wechsler Memory Scale—Revised.

iNPH: Idiopathic Normal Pressire Hydrocephalus.
AD: Alzheimer's Disease.

Hellström et al. (2008) found cognitive improvement in 80.8% of their shunted patients. Whilst variability in these findings could be attributable to differences between patients or measures, there is also evidence to suggest that mild symptoms are more susceptible to improvement than severe symptoms (Hamilton et al., 2010; Marmarou, Young, Aygok, Tsuji, Yamamoto, & Dunbar, 2005). This is, presumably, because more severe symptomatology is associated with a greater degree of neural damage, which is less likely to be remediated by shunting (e.g., Iddon et al., 2004).

Some studies have investigated which cognitive functions are most likely to improve post-shunting in iNPH, and there is evidence for improvement in immediate and delayed verbal recall, psychomotor speed, and visuoconstructive abilities. Smaller improvements have also been observed for attention, recognition memory, inhibition (Stroop task), semantic fluency, and visuospatial ability, with inconsistent findings regarding working memory. For example, Thomas et al. (2005) found that 52.3% of iNPH patients showed overall neurocognitive improvement post-shunt, and significant improvement in psychomotor speed and verbal memory. Lower improvement rates were seen for those who scored more than one standard deviation lower than the mean on immediate verbal recall pre-shunt, and even more so if they additionally had low performance on executive functioning or visuoconstructive performance. Duinkerke, Williams, Rigamonti, and Hillis (2004) found significant group improvements at follow-up after 1 year of shunting on tests of verbal memory as well as in one test of psychomotor speed. Chaudhry et al. (2007) administered a battery of tests measuring verbal memory, complex figures, visuospatial performance, verbal fluency, psychomotor and motor speed, as well as fine motor coordination. They found that shunting resulted in improved performance on all cognitive tests for which performance was impaired at baseline, including immediate and delayed verbal learning and Trail-Making Test Part A, with the exception of recognition memory. The cognitive improvements post-shunt significantly predicted improvements at 3–6 months after surgery. No improvements were found in executive function, which they reasoned was due to the fact that half of the patients scored within normal range. Mataró et al. (2003) found shunting to be associated with significant improvement in verbal memory (RAVLT learning), visuoconstructive functioning (Block Design), psychomotor speed (Trail-Making Test Part A and pegboard right hand), and daily life activities scale (Informant's Test). However, many attentional and memory tests (working memory, visual reproduction, Trail-Making Part B, Stroop, semantic fluency, line orientation, and NPH behavioural scales) did not yield significant improvement. Hellström et al. (2008) included measures of psychomotor speed, wakefulness, manual dexterity and motor speed, immediate and delayed verbal recall, inhibition, working memory and working memory capacity, and a simple reaction time task, in which participants had to respond to changes in the colour of the screen, to investigate differences in iNPH patients before and after shunting. They found that shunting resulted in higher performance on all of the tasks with the exception of simple reaction time and forwards digit span. Peterson et al. (2019) found that shunting was associated with improvements in verbal learning (Hopkins Verbal Learning Test) and semantic fluency post-shunt in NPH patients. Phonemic fluency and delayed verbal learning measures, as well as Mini-Mental State Examination, depression, and apathy scores did not differ pre- and post-shunt. Finally, Golomb et al. (2000) found that for patients with NPH both with and without AD, shunting resulted in improvements in gait and urinary

Table 13 – Summary of presented literature on symptom improvement after shunting.

Symptom improvement after shunting	
Author	Findings
Chaudhry et al. (2007)	Shunting resulted in improved performance on all cognitive tests included in the study for which performance was impaired at baseline (immediate and delayed verbal learning and Trail-Making Test Part A, with the exception of recognition memory) in people with NHP. The cognitive improvements post-shunt significantly predicted improvements at 3–6 months after surgery. No improvements found in executive function.
da Rocha et al. (2021)	Tap Test in participants with iNPH, resulted in improvements in orientation, RAVLT, Rey Complex Figure Test, Stroop test, Mini Mental State Examination. Some of the tests improved to the level that was not statistically different from controls (A1, B1, A6 of RAVLT, immediate and delayed reproduction of Rey Complex Figures, Stroop Test).
Duinkerke et al. (2004)	Significant improvements in iNPH patients following shunting at follow-up after 1 year of shunting on tests of verbal memory as well as on one test of psychomotor speed.
Golomb et al. (2000)	Shunting resulted in improvements in gait and urinary control, as well as cognitive functioning indicated by a composite cognitive score (combined performance on cognitive deterioration scale and the Mini Mental State Examination) for patients with NPH both with and without AD.
Hamilton et al. (2010)	Better improvements after shunting for people with mild symptoms as opposed to moderate or severe symptoms.
Hebb and Cusimano (2001)	Literature review reporting that 59% shunted iNPH patients improve, with a 6% complication rate.
Hellström et al. (2008)	Cognitive improvement in 80.8% of shunted iNPH patients on measures of psychomotor speed, wakefulness, manual dexterity and motor speed, immediate and delayed verbal recall, inhibition, working memory (backwards digit span), but not on working memory (forwards digit span) and a simple reaction time task.
Marmarou, Young, Aygok, Tsuji, Yamamoto, and Dunbar (2005)	Outcome following lumbar drainage of CSF predicted shunt outcome in patients with NPH with the accuracy of 88%, and sensitivity of 95. Greater improvement was found for those whose symptoms were mild as opposed to severe.
Mataró et al. (2003)	Shunting of NPH patients was associated with significant improvement in verbal memory (RAVLT learning), visuoconstructive functioning (Block Design), psychomotor speed (Trail-Making Part A and pegboard right hand), and daily life activities scale (Informant's Test). However, working memory, visual reproduction, Trail-Making Part B, Stroop, semantic fluency, line orientation, and NPH behavioural scales) did not yield significant improvement.
Mataró et al. (2007)	65% of shunted patients with iNPH improved in their cognitive function, including measures of verbal memory, visuospatial tasks (line orientation and block design), frontal lobe functions (verbal fluency and digit span backward), and all included measures of psychomotor speed.
Poca et al. (2005)	33% of shunted patients with NPH improved in their cognitive function.
Petersen et al. (1985)	Motor symptoms are generally associated with the highest rates of improvement, whereas the improvement rate of cognitive symptoms is less certain and more variable.
Peterson et al. (2019)	Shunting was associated with improvements in verbal learning (immediate/learning parts of the Hopkins Verbal Learning Test) and semantic fluency post-shunt in NPH patients. Phonemic fluency and delayed verbal learning measures, as well as Mini Mental State Examination, depression, and apathy scores did not differ pre- and post-shunt.
Raftopoulos et al. (1994)	Gait improvement was seen in 95% of people with iNPH in a 1-year follow-up after shunting, while 66.6% of people improved in their cognitive function 1 year after shunting.
Savolainen et al. (2002)	Only one neuropsychological test – word recognition – distinguished the patients who improved after a shunt from those who did not, although shunting consistently improved gait and urinary symptoms.
Thomas et al. (2005)	52.3% of iNPH patients showed overall neurocognitive improvement post-shunt, and significant improvement in psychomotor speed and verbal memory. Lower improvement rates were seen for those who scored more than one standard deviation lower than the mean on immediate verbal recall pre-shunt, and even more so if they additionally had low performance on executive functioning or visuoconstructive performance.

Table 13 – (*continued*)

Symptom improvement after shunting	
Author	Findings
Thomsen et al. (1986)	16/40 of shunted patients with NPH improved in their cognitive function.
Toma et al. (2013)	Review paper documenting positive improvement in 71% of NPH patients after shunting, with an average of 1% mortality, with up to 82% success rate when considering data from 2005 to 2010 (articles published in the last five years of the dates of their literature search).

control, as well as cognitive functioning indicated by a composite cognitive score, which combined performance on cognitive deterioration scale and the Mini Mental State Examination. These findings suggest that shunting can be useful in reducing symptoms for people with NPH even in the presence of AD.

It is unclear whether neuropsychological testing can predict improvements after shunting. Savolainen, Hurskainen, Paljärvi, Alafuzoff, and Vapalahti (2002) found that only one neuropsychological test – word recognition – distinguished the patients who improved after a shunt from those who did not, although shunting consistently improved gait and urinary symptoms. A recent study by da Rocha et al. (2021) investigated the effects of a Spinal Tap Test (Lumbar Drainage Test) on cognition. They were the first to administer the Tap Test twice, with systematic neuropsychological testing administered after each Tap Test. They found that in participants with iNPH, several measures improved from baseline to measures administered after first and second Tap Test. These were Orientation, RAVLT, Rey Complex Figure Test, Stroop test, and Mini Mental State Examination. As such, while progress has been made, further work is needed to determine the optimal method for selecting patients for shunting and to determine how effective shunting is based on severity of symptoms, including the use of double-blind randomised control trials. We also require further evidence to document the neuroanatomical basis of cognitive impairments in iNPH (Kazui, 2008, but see Tullberg, Hellström, Piechnik, Starmark, & Wikkelsö, 2004), which would benefit from participants being tested alongside a control group who do not undergo shunting (Chaudhry et al., 2007). The findings of the studies discussed in this section are summarised in Table 13.

3.6. *Theoretical questions*

As outlined previously, iNPH results in difficulties with gait and executive functions, including verbal fluency, attention and concentration, and working memory. This pattern of difficulty closely resembles that observed in early-onset HC. Consideration of the commonalities between early-onset and normal pressure HC could help us understand the effects that HC, in its own right, has on the brain as well as the cognitive and motor systems. Despite this, little work has compared the two forms of HC directly. The exception to this is a study by Donnet, Schmitt, Dufour, Giorgi, and Grisoli (2004), who compared people with NPH and a group with aqueductal stenosis (a congenital cause of HC) on performance on a variety of cognitive tasks. They found that NPH patients had greater impairments on executive functioning tasks but had higher memory indices (particularly delayed recall) than the aqueductal stenosis group. Performance on the other measured constructs was comparable in both groups (forwards and backwards digit span, short-term memory, other delayed memory indices). However, the groups differed in age, and sample sizes were relatively small (i.e. $N = 10$ per group). It is, of course, important to acknowledge the difficulties associated with recruiting participants from these vulnerable groups, but research with larger samples and analyses accounting for age are needed.

The relative lack of research comparing NPH and congenital/early-onset HC likely reflects the differing needs of these two groups. Those for whom HC is diagnosed early in life will require support with schooling, home life, and development, while those for whom HC is diagnosed later in life will require support for more focused needs, such as the work environment, as well as disentangling the diagnosis between iNPH and other similar disorders. Nevertheless, comparing these two types of HC has enormous theoretical relevance to neuropsychology and our understanding of developmental disorders of cognition. In particular, it offers an important opportunity to study the same condition from the perspective of both typical and atypical developmental trajectories. The latter is perhaps the most usual, with HC being evident at birth (or even before) for aetiological reasons detailed earlier. These individuals, when successfully treated, will have an atypical developmental trajectory, and some cognitive processes may never operate at a typical level. However, NPH appears in adulthood, and successfully treated individuals might experience cognitive impairment in the face of a typical developmental trajectory – i.e. they will suddenly experience difficulties in cognitive processes that were, up until that point, likely to be typical (see Dennis et al., 2014). It is not easy to generate other conditions that share such a unique profile, and it suggests that we should more formally account for age of acquisition when considering the relationship between a clinical condition and its effects on cognitive function.

Pursuing this research could also help examine how poorer motor or spatial abilities (potentially due to SB) in congenital/early-onset HC affect further development of the cognitive system. Dennis and colleagues (Dennis & Barnes, 2010; Dennis et al., 2006; Fletcher & Dennis, 2009) propose that, over and above the effects of HC on the brain and cognitive function, reduced spatial and motor abilities will have additional effects on functional development by restricting exploration and further learning. Supporting this, Wiedenbauer and Jansen-Osmann (2006) found that the age at which children learned to walk was correlated with the number of learning trials required to reach performance criterion. Donnet et al. (2004)

also reported cognitive differences between an NPH sample and participants with aqueductal stenosis. A comparison between congenital/early-onset HC and NPH with larger samples would help to inform this important debate, as it would highlight the effects of HC with and without the associated effects on cognitive development.

Administering neuropsychological measures to both typically-developed individuals and people with HC might also inform our understanding of typically-developing cognitive processes and functions. For example, if particular features are correlated in both groups (e.g., if fluid intelligence predicts spatial learning), this would mean that the neural damage associated with HC does not interfere with nature or extent of the relationship between these two features. On the other hand, if two features were correlated in the typical group, but not in the HC group, this may suggest differences in the substrates of performance in the HC group. For example, certain tasks may be solved in a different manner, using different abilities or strategies. If the two features were correlated in the HC group only, this would suggest that a particular function has been recruited for performance (e.g., verbal memory span is utilised for writing) in the HC group only, possibly because the typical basis for solving this task has been attenuated by brain damage.

3.7. *Co-morbidity*

As discussed previously, NPH is often comorbid with a variety of different conditions (Malm et al., 2013), including dementia, hypertension, Type II diabetes (Pykkö et al., 2018), schizophrenia (Vanhala et al., 2019), cerebral palsy (Bech-Azeddine et al., 2007), cerebrovascular disease (Boon, et al., 1999), and cervical myelopathy (Naylor et al., 2020), as well as general aging. Given that these complex and varying comorbidities are known to affect cognitive function, they will inevitably influence the cognitive profiles in NPH. This is further complicated by methodological limitations such as different selection criteria and inconsistent follow-up procedures and intervals (Klinge, Marmarou, Bergsneider, Relkin, & Black, 2005). On the other hand, SB is the most common cause and comorbidity in early-onset HC, and as discussed earlier, it has not been clearly established whether it affects cognitive functioning in people with HC. HC can also be caused by Chiari-II malformation, which is another comorbid condition. Evidence indicates that participants with HC and Chiari-II malformation have lower performance on tasks of visuospatial function, visual analysis and synthesis, and verbal memory and verbal fluency, compared with participants with HC without the Chiari-II malformation, but similar processing speed, non-verbal memory, and verbal skill (Vinck et al., 2006). While the presence of these comorbidities could be responsible for the conflicting data regarding cognitive functioning between early-onset HC and NPH, there is a lack of studies investigating the effects of comorbidities, particularly in early-onset HC.

4. Summary and recommendations

In this paper, we have attempted to present a representative and contemporary insight into a common neurological condition. In doing so, our aim was not only to provide an overview of its neuropsychological ramifications, but also to highlight some of the challenges and debates associated with understanding HC. Although the condition is relatively well understood from a clinical perspective, there is much that remains to be elucidated, both in terms of its precise cognitive correlates (in the various forms that HC can take) but also the neurological and aetiological factors that are responsible for them. The literature currently contains the seeds of an important theoretical debate regarding the precise origins of cognitive impairment. Moreover, the full impact of studying a condition from the perspective of a typical (NPH) or atypical (congenital/early-onset HC) developmental trajectory could make a ground-breaking contribution to neuropsychological thinking. However, these seeds can only germinate if they are fed and watered by continued careful patient testing.

The need for further research is, however, not simply an academic one. Research into the cognitive and behavioural outcomes of adults with neurodevelopmental disorders is thought to be lacking in general (Barnes Dennis, & Hetherington, 2004), and it has also been argued that adults with HC, in particular, do not tend to receive the same level of support as other clinical groups (e.g., Sawyer et al., 1998), or compared to children with SB + HC. For example, a co-ordinated interdisciplinary team-based clinical approach for managing the condition has been shown to be fruitful for children with SB, but is reported to be lacking in adults (Dicianno et al., 2008; Morgan, Blackburn, & Bax, 1995). One further barrier that people with HC face is that, due to their varied cognitive profile, they have a general tendency to present as highly articulate, which can mask other cognitive difficulties. Coupled with low awareness of HC and its effects in patients, their families and significant others, as well as general health professionals, this can prevent patients from getting access to required support. Targeted research, liaising with clinicians, and public engagement undertakings are all necessary steps to extend understanding and awareness and lead to an improvement in the quality of life for this population.

HC research could further benefit from adopting a dimensional approach to understanding clinical difference. The traditional categorical approach to clinical research compares typical and atypical groups of people and has been criticised for excluding the dimensionality of clinical conditions, which emphasise degree of deficit (Graham & Madigan, 2016). This is highly relevant to HC, which has varied aetiology, varied rates of ventricle expansion, and variability in the extent of neural damage. Karmiloff-Smith (1998) underlines the importance of researching clinical conditions over and above just mapping the patterns of sparing and impairments. According to this viewpoint, even in cases where no differences in performance exist between typical and atypical groups, the underlying cognitive processes that result in those outcomes may still differ.

Our understanding of HC is particularly complicated by the distribution of insights across developmental and adult samples. In the study of attentional difficulties, for example, both anterior and posterior processes have been implicated in children (Brewer et al., 2001; Erickson et al., 2001; Fletcher et al., 1996; Swartwout et al., 2008), whereas executive

processes such as switching and sequencing have been highlighted in adults (Barf et al., 2003; Iddon et al., 2004). Further research is also needed to classify the memory profile of HC, with particular reference to working memory and immediate memory, as well as the sparing of implicit memory in adults. The spatial and motor profile of HC requires fuller examination, and a clearer dissociation from the impact of SB; this may benefit from using more translational approaches that explicitly divide functions into their sub-domains (e.g., in the context of navigation, see: Smith, 2015). Such approaches would shed more light on the precise types of spatial difficulties experienced in HC and, more importantly, highlight a clearer, more nuanced pathway to cognitive intervention.

In tandem with these calls for more research, it is clear from this review that a finer grain of understanding of HC, both in terms of its unique cognitive profile and its functional sequelae, is emerging in the literature. Similarly, approaches to rehabilitating these aspects of the condition are also developing. The multidisciplinary approach to treatment attempts to provide an individualised, holistic strategy that addresses the complex physical, emotional and behavioural needs of individuals across the lifespan. However, despite the presence of specialised expertise, and the resources devoted to development of targeted support for individuals with HC (and their carers), there is a lack of evidence for HC-specific cognitive rehabilitation interventions reported in the literature (Castellani et al., 2021). Of course, the potential for cognitive training has been supported by studies we have mentioned earlier – i.e. specific spatial-ability training for individuals with HC can improve spatial processing and generalise to mental rotation and mental imagery (Lehmann & Jansen, 2012; Wiedenbauer & Jansen-Osmann, 2007) and a range of process-specific navigation strategies can generalise to support individuals with HC navigate in the real world (Buckley & Smith, 2013).

Current clinical practice has, rather, been established to align with the documented evidence for cognitive rehabilitation in TBI and stroke, based on process-specific interventions designed to improve impairments in processing speed, attentional processing, learning and memory, spatial abilities, numeracy and executive function, which may generalise and transfer to higher-order cognitive function (Cicerone et al., 2005, 2008, 2019). Recommendations from the systematic review of randomised control trials that met the criteria to support processing speed and attention include the use of Attentional Processing Training, Metacognitive Training, and the development of compensatory strategies with a trained therapist to support real world function (Cicerone et al., 2019). The review highlighted the efficacy of errorless learning, memory strategies, external memory aids (e.g., paging systems; assistive technologies), in conjunction with awareness, self-monitoring and problem-solving strategies. These techniques have been found to improve learning, memory and executive function in TBI (Wilson, Emslie, Quirk, Evans, & Watson, 2005) and in stroke (Fish, Manly, Emslie, Evans, & Wilson, 2008). A randomised control trial conducted by Cicerone et al. (2008) evaluated a multidisciplinary rehabilitation programme and a comprehensive-holistic neuropsychological rehabilitation programme. Notably, both interventions improved neuropsychological function in individuals with TBI, although there were more functional gains observed in the holistic rehabilitation group that included psychological support. More recent studies (Edginton et al., 2009; Loveday & Edginton, 2011) have documented the impact of the high levels of anxiety in HC, coupled with the misunderstanding of the unique cognitive profile, and the need to offer psychoeducation, psychological support, and specific cognitive strategies on an individual and/or group basis. In response Edginton et al. (2014) designed and delivered a 6-week programme that was conducted within a community and a multidisciplinary hospital setting leading to the inclusion of cognitive behavioural therapy and mindfulness within the clinic protocol for individuals with HC.

We hope that this detailed review will stimulate further research to inform the development and systematic evaluation of tailored cognitive rehabilitation interventions to support real world function for individuals with HC across the lifespan in a range of education, workplace and community settings.

Author Contributions

Tara Zaksaite: Writing – original draft preparation, **Catherine Loveday:** Writing – review & editing, **Trudi Edginton:** Writing – review & editing, **Hugo J. Spiers:** Writing – review & editing, **Alastair D. Smith:** Writing – original draft preparation, Supervision.

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Declaration of competing interest

None.

Data availability

No data was used for the research described in the article.

REFERENCES

Achenbach, T. M. (1991). *Manual for the child behavior checklist.* Burlington: University of Vermont, Department of Psychiatry.

Adam, R., Ghahari, D., Morton, J. B., Eagleson, R., & de Ribaupierre, S. (2022). Brain network connectivity and executive function in children with infantile hydrocephalus. *Brain Connectivity*. https://doi.org/10.1089/brain.2021.0149 [published online ahead of print, 2022 Apr. 21].

Adams, R. D. (1975). Recent observations on normal pressure hydrocephalus. *Schweizer Archiv für Neurologie, Neurochirurgie und Psychiatrie, 116*(1), 7–15.

Albright, A. L., Ferson, S., & Carlos, S. (2005). Occult hydrocephalus in children with cerebral palsy. *Neurosurgery,*

56(1), 93–97. https://doi.org/10.1227/01.NEU.0000144779.32401.A2

Andersson, J., Rosell, M., Kockum, K., Lilja-Lund, O., Söderström, L., & Laurell, K. (2019). Prevalence of idiopathic normal pressure hydrocephalus: A prospective, population-based study. *Plos One, 14*(5), Article e0217705. https://doi.org/10.1371/journal.pone.0217705

Andrén, K., Wikkelsø, C., Tisell, M., & Hellström, P. (2014). Natural course of idiopathic normal pressure hydrocephalus. *Journal of Neurology, Neurosurgery, and Psychiatry, 85*(7), 806–810. https://doi.org/10.1136/jnnp-2013-306117

Arrington, C. N., Ware, A. L., Ahmed, Y., Kulesz, P. A., Dennis, M., & Fletcher, J. M. (2016). Are shunt revisions associated with IQ in congenital hydrocephalus? A meta-analysis. *Neuropsychology Review, 26*(4), 329–339. https://doi.org/10.1007/s11065-016-9335-z

Aschoff, A., Kremer, P., Hashemi, B., & Kunze, S. (1999). The scientific history of hydrocephalus and its treatment. *Neurosurgical Review, 22*(2), 67–93. https://doi.org/10.1007/s101430050035

Ayr, L. K., Yeates, K. O., & Enrile, B. G. (2005). Arithmetic skills and their cognitive correlates in children with acquired and congenital brain disorder. *Journal of the International Neuropsychological Society: JINS, 11*(3), 249–262. https://doi.org/10.1017/S1355617705050307

Barf, H. A., Verhoef, M., Jennekens-Schinkel, A., Post, M. W. M., Gooskens, R. H. J. M., & Prevo, A. J. H. (2003). Cognitive status of young adults with spina bifida. *Developmental Medicine and Child Neurology*, 45(12), 813–820. https://doi.org/10.1017/S0012162203001518

Barnes, M., Dennis, M., & Hetherington, R. (2004a). Reading and writing skills in young adults with spina bifida and hydrocephalus. *Journal of the International Neuropsychological Society: JINS, 10*(5), 655–663. https://doi.org/10.1017/S1355617704105055

Barnes, M. A., Faulkner, H. J., & Dennis, M. (2001). Poor reading comprehension despite fast word decoding in children with hydrocephalus. *Brain and Language, 76*(1), 35–44. https://doi.org/10.1006/brln.2000.2389

Barnes, M. A., Faulkner, H., Wilkinson, M., & Dennis, M. (2004b). Meaning construction and integration in children with hydrocephalus. *Brain and language, 89*(1), 47–56. https://doi.org/10.1016/S0093-934X(03)00295-5

Barnes, M. A., Pengelly, S., Dennis, M., Wilkinson, M., Rogers, T., & Faulkner, H. (2002). Mathematics skills in good readers with hydrocephalus. *Journal of the International Neuropsychological Society: JINS, 8*(1), 72–82. https://doi.org/10.1017/S1355617702811079

Bech-Azeddine, R., Høgh, P., Juhler, M., Gjerris, F., & Waldemar, G. (2007). Idiopathic normal-pressure hydrocephalus: Clinical comorbidity correlated with cerebral biopsy findings and outcome of cerebrospinal fluid shunting. *Journal of Neurology, Neurosurgery, and Psychiatry, 78*(2), 157–161. https://doi.org/10.1136/jnnp.2006.095117

Berker, D. E., Goldstein, D. G., Lorber, J., Priestley, D. B., & Smith, D. A. (1992). Reciprocal neurological developments of twins discordant for hydrocephalus. *Developmental Medicine and Child Neurology, 34*(7), 623–632. https://doi.org/10.1111/j.1469-8749.1992.tb11493.x

Boone, D., Parsons, D., Lachmann, S. M., & Sherwood, T. (1985). Spina bifida occulta: Lesion or anomaly? *Clinical Radiology, 36*(2), 159–161. https://doi.org/10.1016/S0009-9260(85)80100-8

Boon, A. J., Tans, J. T., Delwel, E. J., Egeler-Peerdeman, S. M., Hanlo, P. W., Wurzer, H. A., et al. (1999). Dutch normal-pressure hydrocephalus study: The role of cerebrovascular disease. *Journal of Neurosurgery, 90*(2), 221–226. https://doi.org/10.3171/jns.1999.90.2.0221

Boyer, K. M., Yeates, K. O., & Enrile, B. G. (2006). Working memory and information processing speed in children with myelomeningocele and shunted hydrocephalus: Analysis of the children's paced auditory serial addition test. *Journal of the International Neuropsychological Society: JINS, 12*(3), 305. https://doi.org/10.1017/S1355617706060425

Brean, A., & Eide, P. K. (2008). Prevalence of probable idiopathic normal pressure hydrocephalus in a Norwegian population. *Acta Neurologica Scandinavica, 118*(1), 48–53. https://doi.org/10.1111/j.1600-0404.2007.00982.x

Brewer, V. R., Fletcher, J. M., Hiscock, M., & Davidson, K. C. (2001). Attention processes in children with shunted hydrocephalus versus attention deficit-hyperactivity disorder. *Neuropsychology, 15*(2), 185–198. https://doi.org/10.1037/0894-4105.15.2.185

Bridgman, M. W., Brown, W. S., Spezio, M. L., Leonard, M. K., Adolphs, R., & Paul, L. K. (2014). Facial emotion recognition in agenesis of the corpus callosum. *Journal of Neurodevelopmental Disorders, 6*(1), 1–14. https://doi.org/10.1186/1866-1955-6-32

Brown, T. M., Ris, M. D., Beebe, D., Ammerman, R. T., Oppenheimer, S. G., Yeates, K. O., et al. (2008). Factors of biological risk and reserve associated with executive behaviors in children and adolescents with spina bifida myelomeningocele. *Child Neuropsychology, 14*(2), 118–134. https://doi.org/10.1080/092970490911324

Bruininks, R. H., Woodcock, R. W., Weatherman, R. F., & Hill, B. K. (1996). *Scales of independent behavior–revised (SIB–R)*. Chicago: Riverside Publishing.

Buckley, M. G., & Smith, A. D. (2013). Evidence for spatial navigational impairments in hydrocephalus patients without spina bifida. *Brain and Cognition, 83*(1), 132–141. https://doi.org/10.1016/j.bandc.2013.07.010

Buckley, R. T., Yuan, W., Mangano, F. T., Phillips, J. M., Powell, S., McKinstry, R. C., … Limbrick, D. D. (2012). Longitudinal comparison of diffusion tensor imaging parameters and neuropsychological measures following endoscopic third ventriculostomy for hydrocephalus: Case report. *Journal of Neurosurgery: Pediatrics, 9*(6), 630–635. https://doi.org/10.3171/2012.2.PEDS11331

Bugalho, P., Alves, L., & Ribeiro, O. (2013). Normal pressure hydrocephalus: A qualitative study on outcome. *Arquivos de Neuro-Psiquiatria, 71*, 890–895. https://doi.org/10.1590/0004-282X20130173

Burmeister, R., Hannay, H. J., Copeland1, K., Fletcher, J. M., Boudousquie, A., & Dennis, M. (2005). Attention problems and executive functions in children with spina bifida and hydrocephalus. *Child Neuropsychology, 11*(3), 265–283. https://doi.org/10.1080/092970490911324

Cabral, D., Beach, T. G., Vedders, L., Sue, L. I., Jacobson, S., Myers, K., et al. (2011). Frequency of Alzheimer's disease pathology at autopsy in patients with clinical normal pressure hydrocephalus. *Alzheimer's & Dementia, 7*(5), 509–513. https://doi.org/10.1016/j.jalz.2010.12.008

Caixeta, L. (2007). What matters in white matter dementia? *Dementia & Neuropsychologia, 1*(2), 131–139. https://doi.org/10.1590/S1980-57642008DN10200004

Calhoun, S. L., & Mayes, S. D. (2005). Processing speed in children with clinical disorders. *Psychology in the Schools, 42*(4), 333–343. https://doi.org/10.1002/pits.20067

Caltagirone, C., Gainotti, G., Masullo, C., & Villa, G. (1982). Neurophysiological study of normal pressure hydrocephalus. *Acta Psychiatrica Scandinavica, 65*(2), 93–100. https://doi.org/10.1111/j.1600-0447.1982.tb00827.x

Capone, P. M., Bertelson, J. A., & Ajtai, B. (2019). Neuroimaging of normal pressure hydrocephalus and hydrocephalus. In L. L. Mechtler, & R. W. Evans (Eds.), *Neuroimaging, an issue of*

neurologic clinics E-book (pp. 171–183). Philadelphia, Pensylvania: Elsevier.

Castellani, G. B., Miccoli, G., Cava, F. C., Salucci, P., Colombo, V., Maietti, E., et al. (2021). From shunt to recovery: A multidisciplinary approach to hydrocephalus treatment in severe acquired brain injury rehabilitation. *Brain Sciences, 12*(1), 3. https://doi.org/10.3390/brainsci12010003

Chaudhry, P., Kharkar, S., Heidler-Gary, J., Hillis, A. E., Newhart, M., Kleinman, J. T., … Williams, M. A. (2007). Characteristics and reversibility of dementia in normal pressure hydrocephalus. *Behavioural Neurology, 18*(3), 149–158. https://doi.org/10.1155/2007/456281

Cholley, F., Trivin, C., Sainte-Rose, C., Souberbielle, J. C., Cinalli, G., & Brauner, R. (2001). Disorders of growth and puberty in children with non-tumoral hydrocephalus. *Journal of Pediatric Endocrinology and Metabolism, 14*(3), 319–328. https://doi.org/10.1515/jpem.2001.14.3.319.

Cicerone, K. D., Dahlberg, C., Malec, J. F., Langenbahn, D. M., Felicetti, T., Kneipp, S., … Catanese, J. (2005). Evidence-based cognitive rehabilitation: Updated review of the literature from 1998 through 2002. *Archives of Physical Medicine and Rehabilitation, 86*(8), 1681–1692.

Cicerone, K. D., Goldin, Y., Ganci, K., Rosenbaum, A., Wethe, J. V., Langenbahn, D. M., … Harley, J. P. (2019). Evidence-based cognitive rehabilitation: systematic review of the literature from 2009 through 2014. *Archives of physical medicine and rehabilitation, 100*(8), 1515–1533. https://doi.org/10.1016/j.apmr.2019.02.011.

Cicerone, K. D., Mott, T., Azulay, J., Sharlow-Galella, M. A., Ellmo, W. J., Paradise, S., et al. (2008). A randomized controlled trial of holistic neuropsychologic rehabilitation after traumatic brain injury. *Archives of Physical Medicine and Rehabilitation, 89*(12), 2239–2249. https://doi.org/10.1016/j.apmr.2008.06.017

Cusimano, M. D., Rewilak, D., Stuss, D. T., Barrera-Martinez, J. C., Salehi, F., & Freedman, M. (2011). Normal-pressure hydrocephalus: Is there a genetic predisposition? *The Canadian Journal of Neurological Sciences. Le Journal Canadien Des Sciences Neurologiques, 38*(2), 274–281. https://doi.org/10.1017/S031716710001146X

Dandy, W. E., & Blackfan, K. D. (1913). An experimental and clinical study of internal hydrocephalus. *Journal of the American Medical Association, 61*(25), 2216–2217.

Daou, B., Klinge, P., Tjoumakaris, S., Rosenwasser, R. H., & Jabbour, P. (2016). Revisiting secondary normal pressure hydrocephalus: Does it exist? A review. *Neurosurgical Focus, 41*(3), 1–12. https://doi.org/10.3171/2016.6.FOCUS16189. E6.

De Mol, J. (1977). Neuropsychological study of mental troubles in normal pressure hydrocephaly and their short term evolution after spinal fluid derivation. *Acta Psychiatrica Belgica, 77*(2), 228–253.

De Mol, J. (1978). Psychic disturbance in normal pressure hydrocephalus. *Acta Neurologica Belgica, 78*(6), 321–340.

De Mol, J. (1986). Neuropsychological symptomatology in normal pressure hydrocephalus. *Schweizer Archiv fur Neurologie und Psychiatrie (Zurich, Switzerland: 1985), 137*(4), 33–45.

Del Bigio, M. R. (1993). Neuropathological changes caused by hydrocephalus. *Acta Neuropathologica, 85*(6), 573–585. https://doi.org/10.1002/ddrr.94

Del Bigio, M. R., Wilson, M. J., & Enno, T. (2003). Chronic hydrocephalus in rats and humans: White matter loss and behavior changes. *Annals of Neurology: Official Journal of the American Neurological Association and the Child Neurology Society, 53*(3), 337–346. https://doi.org/10.1002/ana.10453

Dennis, M., & Barnes, M. (2002). Math and numeracy in young adults with spina bifida and hydrocephalus. *Developmental Neuropsychology, 21*(2), 141–155. https://doi.org/10.1207/S15326942DN2102_2

Dennis, M., & Barnes, M. A. (2010). The cognitive phenotype of spina bifida meningomyelocele. *Developmental Disabilities Research Reviews, 16*(1), 31–39. https://doi.org/10.1002/ddrr.89

Dennis, M., Edelstein, K., Copeland, K., Frederick, J., Francis, D. J., Hetherington, R., et al. (2005a). Covert orienting to exogenous and endogenous cues in children with spina bifida. *Neuropsychologia, 43*, 976–987. https://doi.org/10.1016/j.neuropsychologia.2004.08.012

Dennis, M., Edelstein, K., Copeland, K., Frederick, J., Francis, D. J., Hetherington, R., et al. (2005b). Space-based inhibition of return in children with spina bifida. *Neuropsychology, 19*, 456–465. https://doi.org/10.1037/0894-4105.19.4.456

Dennis, M., Edelstein, K., Frederick, J., Copeland, K., Francis, D., Blaser, S. E., … Fletcher, J. M. (2005c). Peripersonal spatial attention in children with spina bifida: Associations between horizontal and vertical line bisection and congenital malformations of the corpus callosum, midbrain, and posterior cortex. *Neuropsychologia, 43*(14), 2000–2010. https://doi.org/10.1016/j.neuropsychologia.2004.10.014

Dennis, M., Edelstein, K., Hetherington, R., Copeland, K., Frederick, J., Blaser, S. E., … Fletcher, J. M. (2004). Neurobiology of perceptual and motor timing in children with spina bifida in relation to cerebellar volume. *Brain: a Journal of Neurology, 127*(6), 1292–1301. https://doi.org/10.1093/brain/awh154

Dennis, M., Fitz, C. R., Netley, C. T., Sugar, J., Harwood-Nash, D. C., Hendrick, E. B., … Humphreys, R. P. (1981). The intelligence of hydrocephalic children. *Archives of Neurology, 38*(10), 607–615. https://doi.org/10.1001/archneur.1981.00510100035004

Dennis, M., Fletcher, J. M., Rogers, T., Hetherington, R., & Francis, D. J. (2002). Object-based and action-based visual perception in children with spina bifida and hydrocephalus. *Journal of the International Neuropsychological Society: JINS, 8*(1), 95. https://doi.org/10.1017/S135561771020094

Dennis, M., Jacennik, B., & Barnes, M. A. (1994). The content of narrative discourse in children and adolescents after early-onset hydrocephalus and in normally developing age peers. *Brain and Language, 46*(1), 129–165. https://doi.org/10.1006/brln.1994.1008

Dennis, M., Jewell, D., Drake, J., Misakyan, T., Spiegler, B., Hetherington, R., … Barnes, M. (2007). Prospective, declarative, and nondeclarative memory in young adults with spina bifida. *Journal of the International Neuropsychological Society: JINS, 13*(2), 312–323. https://doi.org/10.1017/S1355617707070336

Dennis, M., Landry, S. H., Barnes, M., & Fletcher, J. M. (2006). A model of neurocognitive function in spina bifida over the life span. *Journal of the International Neuropsychological Society: JINS, 12*(2), 285–296. https://doi.org/10.1017/S1355617706060371

Dennis, M., Spiegler, B. J., Simic, N., Sinopoli, K. J., Wilkinson, A., Yeates, K. O., … Fletcher, J. M. (2014). Functional plasticity in childhood brain disorders: When, what, how, and whom to assess. *Neuropsychology Review, 24*(4), 389–408. https://doi.org/10.1007/s11065-014-9261-x

Dicianno, B. E., Kurowski, B. G., Yang, J. M. J., Chancellor, M. B., Bejjani, G. K., Fairman, A. D., … Sotirake, J. (2008). Rehabilitation and medical management of the adult with spina bifida. *American Journal of Physical Medicine & Rehabilitation, 87*(12), 1027–1050. https://doi.org/10.1097/PHM.0b013e31818de070

Donnet, A., Schmitt, A., Dufour, H., Giorgi, R., & Grisoli, F. (2004). Differential patterns of cognitive impairment in patients with aqueductal stenosis and normal pressure hydrocephalus. *Acta Neurochirurgica, 146*(12), 1301–1308. https://doi.org/10.1007/s00701-004-0384-3

Dubois, B., Slachevsky, A., Litvan, I., & Pillon, B. F. A. B. (2000). The FAB: A frontal assessment battery at bedside. *Neurology, 55*(11), 1621–1626. https://doi.org/10.1212/WNL.55.11.1621

Duinkerke, A., Williams, M. A., Rigamonti, D., & Hillis, A. E. (2004). Cognitive recovery in idiopathic normal pressure

hydrocephalus after shunt. *Cognitive and Behavioral Neurology: Official Journal of the Society for Behavioral and Cognitive Neurology, 17*(3), 179–184. https://doi.org/10.1097/01.wnn.0000124916.16017.6a

Dunn, L. M., & Dunn, L. M. (1981). *Peabody picture vocabulary test-revised (PPVT-R)*. Circle Pines, MN: American Guidance Service.

Edelstein, K. I. M., Dennis, M., Copeland, K. I. M., Frederick, J. O. N., Francis, D., Hetherington, R., … Fletcher, J. M. (2004). Motor learning in children with spina bifida: Dissociation between performance level and acquisition rate. *Journal of the International Neuropsychological Society: JINS, 10*(6), 877–887. https://doi.org/10.1017/S1355617704106085

Edginton, T., Iddon, J., Loveday, C., Pickard, J., & Morgan, R. (2009). Cognitive and psychological sequelae of hydrocephalus and spina bifida: Correlating subjective data and objective neuropsychological data to establish insight and inform clinical intervention and guidelines. *Cerebrospinal Fluid Research, 6*(S2). https://doi.org/10.1186/1743-8454-6-s2-s7

Edginton, T., Loveday, C., Cohen, G., Yaz, G., Wing, A., & Morgan, R. (2014). Addressing self-awareness of neuropsychological function and anxiety in individuals with Hydrocephalus. In *Oral presentation at The Society for Research into Hydrocephalus and Spina Bifida, Uppsala, Sweden*.

Erickson, K., Baron, I. S., & Fantie, B. D. (2001). Neuropsychological functioning in early hydrocephalus: Review from a developmental perspective. *Child Neuropsychology, 7*(4), 199–229. https://doi.org/10.1076/chin.7.4.199.8737

Erşahin, Y., & Arslan, D. (2008). Complications of endoscopic third ventriculostomy. *Child's Nervous System, 24*(8), 943–948. https://doi.org/10.1007/s00381-008-0589-5

Fish, J., Manly, T., Emslie, H., Evans, J. J., & Wilson, B. A. (2008). Compensatory strategies for acquired disorders of memory and planning: Differential effects of a paging system for patients with brain injury of traumatic versus cerebrovascular aetiology. *Journal of Neurology, Neurosurgery, and Psychiatry, 79*(8), 930–935. https://doi.org/10.1136/jnnp.2007.125203

Fletcher, J. M., Bohan, T. P., Brandt, M. E., Brookshire, B. L., Beaver, S. R., Francis, D. J., … Miner, M. E. (1992). Cerebral white matter and cognition in hydrocephalic children. *Archives of Neurology, 49*(8), 818–824. https://doi.org/10.1001/archneur.1992.00530320042010

Fletcher, J. M., Bohan, T. P., Brandt, M. E., Kramer, L. A., Brookshire, B. L., Thorstad, K., … Baumgartner, J. E. (1996). Morphometric evaluation of the hydrocephalic brain: Relationships with cognitive development. *Child's Nervous System, 12*(4), 192–199. https://doi.org/10.1007/BF00301250

Fletcher, J. M., Brookshire, B. L., Bohan, T. P., Brandt, M. E., & Davidson, K. C. (1995). Early hydrocephalus. In B. P. Rourke (Ed.), *Syndrome of nonverbal learning disabilities: Neurodevelopmental manifestations* (pp. 206–238). The Guilford Press.

Fletcher, J. M., Copeland, K., Frederick, J. A., Blaser, S. E., Kramer, L. A., Northrup, H., … Dennis, M. (2005). Spinal lesion level in spina bifida: A source of neural and cognitive heterogeneity. *Journal of Neurosurgery: Pediatrics, 102*(3), 268–279. https://doi.org/10.3171/ped.2005.102.3.0268

Fletcher, J. M., & Dennis, M. (2009). Spina bifida and hydrocephalus. In K. O. Yeates, M. D. Ris, H. G. Taylor, & B. F. Pennington (Eds.), *Pediatric neuropsychology: Research, theory, and practice*. Guilford press.

Fletcher, J. M., Dennis, M., & Nothrup, H. (2000). Hydrocephalus. In K. O. Yeates, M. D. Ris, & H. G. Taylor (Eds.), *Pediatric neuropsychology: Research, theory, and practice* (pp. 25–46). New York, US: The Guilford Press.

Fletcher, J. M., Landry, S. H., Bohan, T. P., Davidson, K. C., Brookshire, B. L., Lachar, D., … Francis, D. J. (1997). Effects of intraventricular hemorrhage and hydrocephalus on the long-term neurobehavioral development of preterm very-low-birthweight infants. *Developmental Medicine and Child Neurology, 39*(9), 596–606. https://doi.org/10.1111/j.1469-8749.1997.tb07495.x

Frey, L., & Hauser, W. A. (2003). Epidemiology of neural tube defects. *Epilepsia, 44*, 4–13. https://doi.org/10.1046/j.1528-1157.44.s3.2.x

Gaddes, W. H., & Crockett, D. J. (1975). The Spreen-Benton Aphasia Tests, normative data as a measure of normal language development. *Brain and Language, 2*(3), 257–280. https://doi.org/10.1016/S0093-934X(75)80070-8

Garne, E., Loane, M., Addor, M. C., Boyd, P. A., Barisic, I., & Dolk, H. (2010). Congenital hydrocephalus–prevalence, prenatal diagnosis and outcome of pregnancy in four European regions. *European Journal of Paediatric Neurology: Ejpn: Official Journal of the European Paediatric Neurology Society, 14*(2), 150–155. https://doi.org/10.1016/j.ejpn.2009.03.005

Gavrilov, G. V., Gaydar, B. V., Svistov, D. E., Korovin, A. N., Samarcev, I. P., Churilov, L., et al. (2019). Idiopathic normal pressure hydrocephalus (Hakim-Adams syndrome): Clinical symptoms, diagnosis and treatment. *Psychiatria Danubina, 31*(suppl 5), 737–744.

Gioia, G. A., Isquith, P. K., Guy, S. C., & Kenworthy, L. (2000). *Behavior rating inventory of executive function: Professional manual*. Odessa, FL: Psychological Assessment Resources, Inc.

Golomb, J., Wisoff, J., Miller, D. C., Boksay, I., Kluger, A., Weiner, H., … Graves, W. (2000). Alzheimer's disease comorbidity in normal pressure hydrocephalus: Prevalence and shunt response. *Journal of Neurology, Neurosurgery, and Psychiatry, 68*(6), 778–781. https://doi.org/10.1136/jnnp.68.6.778

Graham, S. A., & Madigan, S. (2016). Bridging the gaps in the study of typical and atypical cognitive development: A commentary. *Journal of Cognition and Development, 17*(4), 671–681. https://doi.org/10.1080/15248372.2016.1200898

Green, A. L., Pereira, E. A., Kelly, D., Richards, P. G., & Pike, M. G. (2007). The changing face of paediatric hydrocephalus: A decade's experience. *Journal of Clinical Neuroscience, 14*(11), 1049–1054. https://doi.org/10.1016/j.jocn.2006.11.004

Gresham, F. M., & Elliott, S. N. (1990). *Social skills rating system: Manual*. Circle Pines, MN: American Guidance Services Inc.

Hader, W. J., Drake, J., Cochrane, D., Sparrow, O., Johnson, E. S., & Kestle, J. (2002). Death after late failure of third ventriculostomy in children. *Journal of Neurosurgery, 97*(1), 211–215. https://doi.org/10.3171/jns.2002.97.1.0211

Hakim, S., & Adams, R. D. (1965). The special clinical problem of symptomatic hydrocephalus with normal cerebrospinal fluid pressure: Observations on cerebrospinal fluid dynamics. *Journal of the Neurological Sciences, 2*, 307–327. https://doi.org/10.1016/0022-510X(65)90016-X

Hamilton, R., Patel, S., Lee, E. B., Jackson, E. M., Lopinto, J., Arnold, S. E., … Trojanowski, J. Q. (2010). Lack of shunt response in suspected idiopathic normal pressure hydrocephalus with Alzheimer disease pathology. *Annals of neurology, 68*(4), 535–540. https://doi.org/10.1002/ana.22015

Hampton, L. E., Fletcher, J. M., Cirino, P., Blaser, S., Kramer, L. A., & Dennis, M. (2013). Neuropsychological profiles of children with aqueductal stenosis and spina bifida myelomeningocele. *Journal of the International Neuropsychological Society: JINS, 19*(2), 127–136. https://doi.org/10.1017/S1355617712001117

Harter, S. (1985). *Manual for the self-perception profile for children: Revision of the perceived competence scale for children*. Denver, CO: University of Denver.

Hebb, A. O., & Cusimano, M. D. (2001). Idiopathic normal pressure hydrocephalus: A systematic review of diagnosis and outcome. *Neurosurgery, 49*(5), 1166–1186. https://doi.org/10.1097/00006123-200111000-00028

Hejl, A., Høgh, P., & Waldemar, G. (2002). Potentially reversible conditions in 1000 consecutive memory clinic patients. *Journal*

of Neurology, Neurosurgery, and Psychiatry, 73(4), 390–394. https://doi.org/10.1136/jnnp.73.4.390

Hellström, P., Edsbagge, M., Archer, T., Tisell, M., Tullberg, M., & Wikkelsø, C. (2007). The neuropsychology of patients with clinically diagnosed idiopathic normal pressure hydrocephalus. *Neurosurgery, 61*(6), 1219–1228. https://doi.org/10.1227/01.neu.0000306100.83882.81

Hellström, P., Edsbagge, M., Blomsterwall, E., Archer, T., Tisell, M., Tullberg, M., et al. (2008). Neuropsychological effects of shunt treatment in idiopathic normal pressure hydrocephalus. *Neurosurgery, 63*(3), 527–536. https://doi.org/10.1227/01.NEU.0000325258.16934.BB

Hellström, P., Klinge, P., Tans, J., & Wikkelsø, C. (2012). A new scale for assessment of severity and outcome in i NPH. *Acta Neurologica Scandinavica, 126*(4), 229–237. https://doi.org/10.1111/j.1600-0404.2012.01677.x

Hetherington, R., & Dennis, M. (1999). Motor function profile in children with early onset hydrocephalus. *Developmental Neuropsychology, 15*(1), 25–51. https://doi.org/10.1080/87565649909540738

Hochhaus, F., Butenandt, O., Schwarz, H. P., & Ring-Mrozik, E. (1997). Auxological and endocrinological evaluation of children with hydrocephalus and/or meningomyelocele. *European Journal of Pediatrics, 156*(8), 597–601. https://doi.org/10.1007/s004310050672.

Hommet, C., Billard, C., Gillet, P., Barthez, M. A., Lourmiere, J. M., Santini, J. J., … Autret, A. (1999). Neuropsychologic and adaptive functioning in adolescents and young adults shunted for congenital hydrocephalus. *Journal of Child Neurology, 14*(3), 144–150. https://doi.org/10.1177/088307389901400302

Hommet, C., Cottier, J. P., Billard, C., Perrier, D., Gillet, P., De Toffol, B., … Autret, A. (2002). MRI morphometric study and correlation with cognitive functions in young adults shunted for congenital hydrocephalus related to spina bifida. *European Neurology, 47*(3), 169–174. https://doi.org/10.1159/000047977

Huber-Okrainec, J., Blaser, S. E., & Dennis, M. (2005). Idiom comprehension deficits in relation to corpus callosum agenesis and hypoplasia in children with spina bifida meningomyelocele. *Brain and Language, 93*(3), 349–368. https://doi.org/10.1016/j.bandl.2004.11.002

Humphreys, G. W., & Riddoch, M. J. (2013). *To see but not to see: A case study of visual agnosia*. Psychology Press.

Hurley, R. A., Bradley, W. G., Jr., Latifi, H. T., & Taber, K. H. (1999). Normal pressure hydrocephalus: Significance of MRI in a potentially treatable dementia. *The Journal of Neuropsychiatry and Clinical Neurosciences, 11*(3), 297–300. https://doi.org/10.1176/jnp.11.3.297

Iddon, J. L., Morgan, D. J. R., Loveday, C., Sahakian, B. J., & Pickard, J. D. (2004). Neuropsychological profile of young adults with spina bifida with or without hydrocephalus. *Journal of Neurology, Neurosurgery, and Psychiatry, 75*(8), 1112–1118. https://doi.org/10.1136/jnnp.2003.029058

Isaacs, A. M., Riva-Cambrin, J., Yavin, D., Hockley, A., Pringsheim, T. M., Jette, N., … Hamilton, M. G. (2018). Age-specific global epidemiology of hydrocephalus: Systematic review, metanalysis and global birth surveillance. *PLos One, 13*(10), Article e0204926. https://doi.org/10.1371/journal.pone.0210851

Iseki, C., Kawanami, T., Nagasawa, H., Wada, M., Koyama, S., Kikuchi, K., … Kato, T. (2009). Asymptomatic ventriculomegaly with features of idiopathic normal pressure hydrocephalus on MRI (AVIM) in the elderly: A prospective study in a Japanese population. *Journal of the Neurological Sciences, 277*(1–2), 54–57. https://doi.org/10.1016/j.jns.2008.10.004

Iseki, C., Takahashi, Y., Wada, M., Kawanami, T., Adachi, M., & Kato, T. (2014). Incidence of idiopathic normal pressure hydrocephalus (iNPH): A 10-year follow-up study of a rural community in Japan. *Journal of the Neurological Sciences, 339*(1–2), 108–112. https://doi.org/10.1016/j.jns.2014.01.033

Ito, J. I., Saijo, H., Araki, A., Tanaka, H., Tasaki, T., Cho, K., et al. (1997). Neuroradiological assessment of visuoperceptual disturbance in children with spina bifida and hydrocephalus. *Developmental Medicine and Child Neurology, 39*(6), 358–392. https://doi.org/10.1111/j.1469-8749.1997.tb07451.x

Jacobs, R., Northam, E., & Anderson, V. (2001). Cognitive outcome in children with myelomeningocele and perinatal hydrocephalus: A longitudinal perspective. *Journal of Developmental and Physical Disabilities, 13*(4), 389–405. https://doi.org/10.1007/s00381-010-1311-y

Jansen-Osmann, P., Wiedenbauer, G., & Heil, M. (2008). Spatial cognition and motor development: A study of children with spina bifida. *Perceptual and Motor Skills, 106*(2), 436–446. https://doi.org/10.2466/pms.106.2.436-446

Jin, S. C., Dong, W., Kundishora, A. J., Panchagnula, S., Moreno-De-Luca, A., Furey, C. G., … Kahle, K. T. (2020). Exome sequencing implicates genetic disruption of prenatal neurogliogenesis in sporadic congenital hydrocephalus. *Nature Medicine, 26*(11), 1754–1765. https://doi.org/10.1038/s41591-020-1090-2

Jingami, N., Asada-Utsugi, M., Uemura, K., Noto, R., Takahashi, M., Ozaki, A., … Kinoshita, A. (2015). Idiopathic normal pressure hydrocephalus has a different cerebrospinal fluid biomarker profile from Alzheimer's disease. *Journal of Alzheimer's Disease, 45*(1), 109–115. https://doi.org/10.3233/JAD-142622

Jinkins, J. R. (1991). Clinical manifestations of hydrocephalus caused by impingement of the corpus callosum on the falx: An MR study in 40 patients. *AJNR. American Journal of Neuroradiology, 12*(2), 331–340.

Johnson, M. H., & de Haan, M. (2015). *Developmental cognitive neuroscience: An introduction*. UK: Blackwell Publishing Ltd.

Kahle, K. T., Kulkarni, A. V., Limbrick, D. D., Jr., & Warf, B. C. (2016). Hydrocephalus in children. *Lancet, 387*(10020), 788–799. https://doi.org/10.1016/S0140-6736(15)60694-8

Kanno, S., Saito, M., Hayashi, A., Uchiyama, M., Hiraoka, K., Nishio, Y., … Mori, E. (2012). Counting-backward test for executive function in idiopathic normal pressure hydrocephalus. *Acta Neurologica Scandinavica, 126*(4), 279–286. https://doi.org/10.1111/j.1600-0404.2012.01644.x

Karmiloff-Smith, A. (1998). Development itself is the key to understanding developmental disorders. *Trends in Cognitive Sciences, 2*, 389–398. https://doi.org/10.1016/S1364-6613(98)01230-3

Kazui, H. (2008). Cognitive impairment in patients with idiopathic normal pressure hydrocephalus. *Brain and Nerve = Shinkei Kenkyu no Shinpo, 60*(3), 225–231.

Kimura, M., Tanaka, A., & Yoshinaga, S. (1992). Significance of periventricular hemodynamics in normal pressure hydrocephalus. *Neurosurgery, 30*(5), 701–705. https://doi.org/10.1097/00006123-199205000-00009

Kitagaki, H., Mori, E., Ishii, K., Yamaji, S., Hirono, N., & Imamura, T. (1998). CSF spaces in idiopathic normal pressure hydrocephalus: Morphology and volumetry. *AJNR. American Journal of Neuroradiology, 19*(7), 1277–1284.

Klassen, B. T., & Ahlskog, J. E. (2011). Normal pressure hydrocephalus: How often does the diagnosis hold water? *Neurology, 77*(12), 1119–1125. https://doi.org/10.1212/WNL.0b013e31822f02f5

Klinge, P., Marmarou, A., Bergsneider, M., Relkin, N., & Black, P. M. (2005). Outcome of shunting in idiopathic normal-pressure hydrocephalus and the value of outcome assessment in shunted patients. *Neurosurgery, 57*(suppl_3), S2–S40.

Koval, V. H. (2004). *Comprehension of facial emotion and facial display rules in children with spina bifida*. Unpublished MA dissertation: University of Toronto.

Krauss, J. K., Regel, J. P., Vach, W., Orszagh, M., Jüngling, F. D., Bohus, M., et al. (1997). White matter lesions in patients with idiopathic normal pressure hydrocephalus and in an age-matched control group: A comparative study. *Neurosurgery, 40*(3), 491–496. https://doi.org/10.1097/0006123-199703000-00011

Krauss, J. K., & Strupp, M. (2003). Normal pressure hydrocephalus. In T. Brandt, L. R. Caplan, J. Dichgans, H. C. Diener, & C. Kennard (Eds.), *Neurological disorders* (pp. 1113–1122). Academic Press. https://doi.org/10.1016/B978-012125831-3/50272-0.

Lacy, M., Baldassarre, M., Nader, T., & Frim, D. (2012). Parent ratings of executive functioning in children with shunted hydrocephalus. *Pediatric neurosurgery, 48*(2), 73–79. https://doi.org/10.1159/000339313

Lehmann, J., & Jansen, P. (2012). The influence of juggling on mental rotation performance in children with spina bifida. *Brain and Cognition, 80*(2), 223–229. https://doi.org/10.1016/j.bandc.2012.07.004

Lehmann, J., & Jansen, P. (2013). Mental rotation performance in children with hydrocephalus both with and without spina bifida. *Developmental Neuropsychology, 38*(7), 433–444. https://doi.org/10.1080/87565641.2013.820304

Lindquist, B., Carlsson, G., Persson, E. K., & Uvebrant, P. (2006). Behavioural problems and autism in children with hydrocephalus. *European Child & Adolescent Psychiatry, 15*(4), 214–219. https://doi.org/10.1007/s00787-006-0525-8

Lindquist, B., Persson, E. K., Fernell, E., & Uvebrant, P. (2011). Very long-term follow-up of cognitive function in adults treated in infancy for hydrocephalus. *Child's Nervous System, 27*(4), 597–601. https://doi.org/10.1007/s00381-010-1311-y

Lindquist, B., Persson, E. K., Uvebrant, P., & Carlsson, G. (2008). Learning, memory and executive functions in children with hydrocephalus. *Acta Paediatrica, 97*(5), 596–601. https://doi.org/10.1111/j.1651-2227.2008.00747.x

Lindquist, B., Uvebrant, P., Rehn, E., & Carlsson, G. (2009). Cognitive functions in children with myelomeningocele without hydrocephalus. *Child's Nervous System, 25*(8), 969–975. https://doi.org/10.1007/s00381-009-0843-5

Loomis, J. W., Lindsey, A., Javornisky, J. G., & Monahan, J. J. (1994). Measures of cognition and adaptive behavior as predictors of adjustment outcomes in young adults with spina bifida. *European Journal of Pediatric Surgery: Official Journal of Austrian Association of Pediatric Surgery= Zeitschrift fur Kinderchirurgie, 4*, 35–36.

Loveday, C., & Edginton, T. (2011). Spina bifida and hydrocephalus. In A. S. Davis (Ed.), *Handbook of pediatric neuropsychology* (pp. 769–782). Springer Publishing Company.

Mahone, E. M., Zabel, T. A., Levey, E., Verda, M., & Kinsman, S. (2002). Parent and self-report ratings of executive function in adolescents with myelomeningocele and hydrocephalus. *Child Neuropsychology, 8*(4), 258–270. https://doi.org/10.1076/chin.8.4.258.13510

Malm, J., Graff-Radford, N. R., Ishikawa, M., Kristensen, B., Leinonen, V., Mori, E., … Relkin, N. R. (2013). Influence of comorbidities in idiopathic normal pressure hydrocephalus—research and clinical care. A report of the ISHCSF task force on comorbidities in INPH. *Fluids and Barriers of the CNS, 10*(1), 1–14. https://doi.org/10.1186/2045-8118-10-22

Mammarella, N., Cornoldi, C., & Donadello, E. (2003). Visual but not spatial working memory deficit in children with spina bifida. *Brain and Cognition, 53*(2), 311–314. https://doi.org/10.1016/S0278-2626(03)00132-5

Mandell, J. G., Kulkarni, A. V., Warf, B. C., & Schiff, S. J. (2015). Volumetric brain analysis in neurosurgery: Part 2. Brain and CSF volumes discriminate neurocognitive outcomes in hydrocephalus. *Journal of Neurosurgery: Pediatrics, 15*(2), 125–132. https://doi.org/10.3171/2014.9.PEDS12427

Marmarou, A., Bergsneider, M., Klinge, P., Relkin, N., & Black, P. M. (2005). The value of supplemental prognostic tests for the preoperative assessment of idiopathic normal-pressure hydrocephalus. *Neurosurgery, 57*(suppl_3), S2–S28. https://doi.org/10.1227/01.NEU.0000168184.01002.60

Marmarou, A., Young, H. F., Aygok, G. A., Sawauchi, S., Tsuji, O., Yamamoto, T., et al. (2005). Diagnosis and management of idiopathic normal-pressure hydrocephalus: A prospective study in 151 patients. *Journal of Neurosurgery, 102*(6), 987–997. https://doi.org/10.3171/jns.2005.102.6.0987

Martín-Láez, R., Caballero-Arzapalo, H., López-Menéndez, L.Á., Arango-Lasprilla, J. C., & Vázquez-Barquero, A. (2015). Epidemiology of idiopathic normal pressure hydrocephalus: A systematic review of the literature. *World Neurosurgery, 84*(6), 2002–2009. https://doi.org/10.1016/j.wneu.2015.07.005

Mataró, M., Matarin, M., Poca, M. A., Pueyo, R., Sahuquillo, J., Barrios, M., et al. (2007). Functional and magnetic resonance imaging correlates of corpus callosum in normal pressure hydrocephalus before and after shunting. *Journal of Neurology, Neurosurgery, and Psychiatry, 78*(4), 395–398. https://doi.org/10.1136/jnnp.2006.096164

Mataró, M., Poca, M. A., Matarín, M. D. M., Catalan, R., Sahuquillo, J., & Galard, R. (2003). CSF galanin and cognition after shunt surgery in normal pressure hydrocephalus. *Journal of Neurology, Neurosurgery, and Psychiatry, 74*(9), 1272–1277. https://doi.org/10.1136/jnnp.74.9.1272

Mazzini, L., Campini, R., Angelino, E., Rognone, F., Pastore, I., & Oliveri, G. (2003). Posttraumatic hydrocephalus: A clinical, neuroradiologic, and neuropsychologic assessment of long-term outcome. *Archives of Physical Medicine and Rehabilitation, 84*(11), 1637–1641. https://doi.org/10.1053/S0003-9993(03)00314-9

McGirr, A., & Cusimano, M. D. (2016). Does idiopathic normal pressure hydrocephalus (iNPH) run in families? *Journal of the Neurological Sciences, 368*, 128–129. https://doi.org/10.1016/j.jns.2016.06.054

Miyoshi, N., Kazui, H., Ogino, A., Ishikawa, M., Miyake, H., Tokunaga, H., … Takeda, M. (2005). Association between cognitive impairment and gait disturbance in patients with idiopathic normal pressure hydrocephalus. *Dementia and Geriatric Cognitive Disorders, 20*(2–3), 71–76. https://doi.org/10.1159/000085858

Morgan, D. J., Blackburn, M., & Bax, M. (1995). Adults with spina bifida and/or hydrocephalus. *Postgraduate Medical Journal, 71*(831), 17–21. https://doi.org/10.1136/pgmj.71.831.17

MRC Vitamin Study Research Group. (1991). Prevention of neural tube defects: Results of the MRC vitamin study. *Lancet, 338*, 131–137.

Munch, T. N., Rostgaard, K., Rasmussen, M. L. H., Wohlfahrt, J., Juhler, M., & Melbye, M. (2012). Familial aggregation of congenital hydrocephalus in a nationwide cohort. *Brain: a Journal of Neurology, 135*(8), 2409–2415. https://doi.org/10.1093/brain/aws158

Naglieri, J. A., & Das, J. P. (1997). *Cognitive assessment system: Interpretive handbook*. Itasca, IL: Riverside Publishing.

Nakajima, M., Yamada, S., Miyajima, M., Ishii, K., Kuriyama, N., Kazui, H., … Arai, H. (2021). Guidelines for management of idiopathic normal pressure hydrocephalus: Endorsed by the Japanese society of normal pressure hydrocephalus. *Neurologia Medico-Chirurgica, 61*(2), 63–97. https://doi.org/10.2176/nmc.st.2020-0292

Navarro, R., Gil-Parra, R., Reitman, A. J., Olavarria, G., Grant, J. A., & Tomita, T. (2006). Endoscopic third ventriculostomy in children: Early and late complications and their avoidance. *Child's Nervous System, 22*(5), 506. https://doi.org/10.1007/s00381-005-0031-1

Naylor, R. M., Lenartowicz, K. A., Graff-Radford, J., Jones, D. T., Cutsforth-Gregory, J. K., Graff-Radford, N. R., et al. (2020). High

prevalence of cervical myelopathy in patients with idiopathic normal pressure hydrocephalus. *Clinical Neurology and Neurosurgery, 197*, Article 106099. https://doi.org/10.1016/j.clineuro.2020.106099

O'dell, B. L., Whitley, J. R., & Hogan, A. G. (1948). Relation of folic acid and vitamin A to incidence of hydrocephalus in infant rats. *Proceedings of the Society for Experimental Biology and Medicine, 69*(2), 272–275. https://doi.org/10.3181/00379727-69-16689

Ogino, A., Kazui, H., Miyoshi, N., Hashimoto, M., Ohkawa, S., Tokunaga, H., … Takeda, M. (2006). Cognitive impairment in patients with idiopathic normal pressure hydrocephalus. *Dementia and Geriatric Cognitive Disorders, 21*(2), 113–119. https://doi.org/10.1159/000090510

Oi, S. (2010). Hydrocephalus research update—controversies in definition and classification of hydrocephalus. *Neurologia Medico-chirurgica, 50*(9), 859–869. https://doi.org/10.2176/nmc.50.859

Oliveira, L. M., Nitrini, R., & Román, G. C. (2019). Normal-pressure hydrocephalus: A critical review. *Dementia & Neuropsychologia, 13*, 133–143. https://doi.org/10.1590/1980-57642018dn13-020001

Oliveira, M. F., Oliveira, J. R., Rotta, J. M., & Pinto, F. C. (2014). Psychiatric symptoms are present in most of the patients with idiopathic normal pressure hydrocephalus. *Arquivos de Neuro-psiquiatria, 72*, 435–438. https://doi.org/10.1590/0004-282X20140047

Olson, K., Jacobson, K. K., & Van Oot, P. (2013). Ecological validity of pediatric neuropsychological measures: Current state and future directions. *Applied Neuropsychology: Child, 2*(1), 17–23. https://doi.org/10.1080/21622965.2012.686330

Overholser, M. D., Whitley, J. R., O'Dell, B. L., & Hogan, A. G. (1954). The ventricular system in hydrocephalic rat brains produced by a deficiency of vitamin B12 or of folic acid in the maternal diet. *The Anatomical Record, 120*(4), 917–933. https://doi.org/10.1002/ar.1091200407

Passler, J. S., Doty, R. L., Dolske, M. C., Louis, P. G. S., Basignani, C., Pepe, J. W., et al. (2017). Olfactory ability in normal pressure hydrocephalus as compared to Alzheimer's disease and healthy controls. *Journal of the Neurological Sciences, 372*, 217–219. https://doi.org/10.1016/j.jns.2016.11.049

Perenc, L., Guzik, A., Podgórska-Bednarz, J., & Drużbicki, M. (2022). Somatic development disorders in children and adolescents affected by syndromes and diseases associated with neurodysfunction and hydrocephalus treated/untreated surgically. *International Journal of Environmental Research and Public Health, 19*(9), 5712. https://doi.org/10.3390/ijerph19095712

Persson, E. K., Hagberg, G., & Uvebrant, P. (2005). Hydrocephalus prevalence and outcome in a population-based cohort of children born in 1989–1998. *Acta Paediatrica, 94*(6), 726–732. https://doi.org/10.1111/j.1651-2227.2005.tb01972.x

Petersen, R. C., Mokri, B., & Laws, E. R., Jr. (1985). Surgical treatment of idiopathic hydrocephalus in elderly patients. *Neurology, 35*, 307–311. https://doi.org/10.1212/WNL.35.3.307

Peterson, K. A., Mole, T. B., Keong, N. C., DeVito, E. E., Savulich, G., Pickard, J. D., et al. (2019). Structural correlates of cognitive impairment in normal pressure hydrocephalus. *Acta Neurologica Scandinavica, 139*(3), 305–312. https://doi.org/10.1111/ane.13052

Philippon, J. (2005). Normal pressure" hydrocephalus. *Psychologie & Neuropsychiatrie du Vieillissement, 3*(1), 53–61. PMID: 15814326.

Poca, M. A., Mataró, M., Matarín, M., Arikan, F., Junqué, C., & Sahuquillo, J. (2005). Good outcome in patients with normal-pressure hydrocephalus and factors indicating poor prognosis. *Journal of Neurosurgery, 103*(3), 455–463. https://doi.org/10.3171/jns.2005.103.3.0455

Pomeraniec, I. J., Bond, A. E., Lopes, M. B., & Jane, J. A. (2016). Concurrent Alzheimer's pathology in patients with clinical normal pressure hydrocephalus: Correlation of high-volume lumbar puncture results, cortical brain biopsies, and outcomes. *Journal of Neurosurgery, 124*(2), 382–388. https://doi.org/10.3171/2015.2.JNS142318

Posner, M. I. (2016). Orienting of attention: Then and now. *The Quarterly Journal of Experimental Psychology, 69*(10), 1864–1875. https://doi.org/10.1080/17470218.2014.937446

Posner, M. I., & Petersen, S. E. (1990). The attention system of the human brain. *Annual review of neuroscience, 13*(1), 25–42. https://doi.org/10.1146/annurev.ne.13.030190.000325

Posner, M. I., Petersen, S. E., Fox, P. T., & Raichle, M. E. (1988). Localization of cognitive operations in the human brain. *Science, 240*(4859), 1627–1631. https://doi.org/10.1126/science.3289116

Pyykkö, O. T., Lumela, M., Rummukainen, J., Nerg, O., Seppälä, T. T., Herukka, S. K., … Leinonen, V. (2014). Cerebrospinal fluid biomarker and brain biopsy findings in idiopathic normal pressure hydrocephalus. *Plos One, 9*(3), Article e91974. https://doi.org/10.1371/journal.pone.0091974

Pyykkö, O. T., Nerg, O., Niskasaari, H. M., Niskasaari, T., Koivisto, A. M., Hiltunen, M., … Leinonen, V. (2018). Incidence, comorbidities, and mortality in idiopathic normal pressure hydrocephalus. *World Neurosurgery, 112*, e624–e631. https://doi.org/10.1016/j.wneu.2018.01.107

Rabin, L. A., Burton, L. A., & Barr, W. B. (2007). Utilization rates of ecologically oriented instruments among clinical neuropsychologists. *The Clinical Neuropsychologist, 21*(5), 727–743. https://doi.org/10.1080/13854040600888776

Raftopoulos, C., Chaskis, C., Delecluse, F., Cantrainet, F., Bidauti, L., & Brotchi, J. (1992). Morphological quantitative analysis of intracranial pressure waves in normal pressure hydrocephalus. *Neurological Research, 14*(5), 389–396. https://doi.org/10.1080/01616412.1992.11740091

Raftopoulos, C., Deleval, J., Chaskis, C., Leonard, A., Cantraine, F., Desmyttere, F., et al. (1994). Cognitive recovery in idiopathic normal pressure hydrocephalus: A prospective study. *Neurosurgery, 35*, 397–404. https://doi.org/10.1227/00006123-199409000-00006

Recinos, P. F., Jallo, G. I., & Recinos, V. R. (2012). Endoscopic third ventriculostomy. In A. Quiñones-Hinojosa (Ed.), *Schmidek and sweet operative neurosurgical techniques: Indications, methods, and results* (6th ed., pp. 1143–1150). Elsevier Saunders. https://doi.org/10.1016/C2011-1-05132-9.

Reddy, G. K., Bollam, P., Shi, R., Guthikonda, B., & Nanda, A. (2011). Management of adult hydrocephalus with ventriculoperitoneal shunts: Long-term single-institution experience. *Neurosurgery, 69*(4), 774–781. https://doi.org/10.1227/NEU.0b013e31821ffa9e

Relkin, N., Marmarou, A., Klinge, P., Bergsneider, M., & Black, P. M. (2005). Diagnosing idiopathic normal-pressure hydrocephalus. *Neurosurgery, 57*(suppl_3), S2–S4. https://doi.org/10.1227/01.NEU.0000168185.29659.C5

Riva, D., Milani, N., Giorgi, C., Pantaleoni, C., Zorzi, C., & Devoti, M. (1994). Intelligence outcome in children with shunted hydrocephalus of different etiology. *Child's Nervous System, 10*(1), 70–73. https://doi.org/10.1007/BF00313588

da Rocha, S. F. B., Kowacs, P. A., de Souza, R. K. M., Pedro, M. K. F., Ramina, R., & Teive, H. A. G. (2021). Serial Tap test of patients with idiopathic normal pressure hydrocephalus: Impact on cognitive function and its meaning. *Fluids and Barriers of the CNS, 18*(1), 1–11. https://doi.org/10.1186/s12987-021-00254-3

Rose, B. M., & Holmbeck, G. N. (2007). Attention and executive functions in adolescents with spina bifida. *Journal of Pediatric Psychology, 32*(8), 983–994. https://doi.org/10.1093/jpepsy/jsm042

Rosen, W. G., Mohs, R. C., & Davis, K. L. (1984). A new rating scale for Alzheimer's disease. *The American Journal of Psychiatry, 141*(11), 1356–1364. https://doi.org/10.1176/ajp.141.11.1356

Sakka, L., Coll, G., & Chazal, J. (2011). Anatomy and physiology of cerebrospinal fluid. *[European Annals of Otorhinolaryngology, Head and Neck Diseases Electronic Resource], 128*(6), 309–316. https://doi.org/10.1016/j.anorl.2011.03.002

Savolainen, S., Hurskainen, H., Paljärvi, L., Alafuzoff, I., & Vapalahti, M. (2002). Five-year outcome of normal pressure hydrocephalus with or without a shunt: Predictive value of the clinical signs, neuropsychological evaluation and infusion test. *Acta Neurochirurgica, 144*(6), 515–523. https://doi.org/10.1007/s00701-002-0936-3

Savolainen, S., Paljärvi, L., & Vapalahti, M. (1999). Prevalence of alzheimer's disease in patients investigated for presumed normal pressure hydrocephalus: A clinical and neuropathological study. *Acta Neurochirurgica, 141*(8), 849–853. https://doi.org/10.1007/s007010050386

Sawyer, S. M., Collins, N., Bryan, D., Brown, D., Hope, M. A., & Bowes, G. (1998). Young people with spina bifida: Transfer from paediatric to adult health care. *Journal of Paediatrics and Child Health, 34*(5), 414–417. https://doi.org/10.1046/j.1440-1754.1998.00265.x

Schroeder, H. W., Niendorf, W. R., & Gaab, M. R. (2002). Complications of endoscopic third ventriculostomy. *Journal of Neurosurgery, 96*(6), 1032–1040. https://doi.org/10.3171/jns.2002.96.6.1032

Silverberg, G. D., Mayo, M., Saul, T., Rubenstein, E., & McGuire, D. (2003). Alzheimer's disease, normal-pressure hydrocephalus, and senescent changes in CSF circulatory physiology: A hypothesis. *Lancet Neurology, 2*(8), 506–511. https://doi.org/10.1016/S1474-4422(03)00487-3

Simms, B. (1987a). The route learning ability of young people with spina bifida and hydrocephalus and their able-bodied peers. *Surgery in Infancy and Childhood, 42*, 53–56.

Simms, B. (1987b). A 3-year follow-up of the driving status of 32 young adults with spina bifida. *International disability studies, 9*(4), 177–180.

Simms, B. (1991). The car use of young drivers with spina bifida and hydrocephalus. *European Journal of Pharmaceutical Sciences: Official Journal of the European Federation for Pharmaceutical Sciences, 1*(S1), 31–34. https://doi.org/10.1055/s-2008-1042536

Smith, A. D. (2015). Spatial navigation in autism spectrum disorders: A critical review. *Frontiers in Psychology, 6*(31). https://doi.org/10.3389/fpsyg.2015.00031

Stein, S. C. (2001). Normal-pressure hydrocephalus: An update. *Neurosurgery Quarterly, 11*(1), 26–35.

Swartwout, M. D., Cirino, P. T., Hampson, A. W., Fletcher, J. M., Brandt, M. E., & Dennis, M. (2008). Sustained attention in children with two etiologies of early hydrocephalus. *Neuropsychology, 22*(6), 765. https://doi.org/10.1037/a0013373

Tanaka, A., Kimura, M., Nakayama, Y., Yoshinaga, S., & Tomonaga, M. (1997). Cerebral blood flow and autoregulation in normal pressure hydrocephalus. *Neurosurgery, 40*(6), 1161–1167. https://doi.org/10.1097/0006123-199706000-00009

Tanaka, N., Yamaguchi, S., Ishikawa, H., Ishii, H., & Meguro, K. (2009). Prevalence of possible idiopathic normal-pressure hydrocephalus in Japan: The osaki-Tajiri project. *Neuroepidemiology, 32*(3), 171–175. https://doi.org/10.1159/000186501

Tarazi, R. A., Zabel, T. A., & Mahone, E. M. (2008). Age-related differences in executive function among children with spina bifida/hydrocephalus based on parent behavior ratings. *The Clinical Neuropsychologist, 22*(4), 585–602. https://doi.org/10.1080/13854040701425940

Tew, B. (1979). The "cocktail party syndrome" in children with hydrocephalus and spina bifida. *British Journal of Disorders of Communication, 14*(2), 89–101. https://doi.org/10.3109/13682827909011349

Thomas, G., McGirt, M. J., Woodworth, G., Heidler, J., Rigamonti, D., Hillis, A. E., et al. (2005). Baseline neuropsychological profile and cognitive response to cerebrospinal fluid shunting for idiopathic normal pressure hydrocephalus. *Dementia and Geriatric Cognitive Disorders, 20*(2–3), 163–168. https://doi.org/10.1159/000087092

Thomsen, A. M., Børgesen, S. E., Bruhn, P., & Gjerris, F. (1986). Prognosis of dementia in normal-pressure hydrocephalus after a shunt operation. *Annals of Neurology: Official Journal of the American Neurological Association and the Child Neurology Society, 20*(3), 304–310. https://doi.org/10.1002/ana.410200306

Toma, A. K. (2015). Hydrocephalus. *Neurosurgery, 33*(8), 384–389. https://doi.org/10.1016/j.mpsur.2015.05.006

Toma, A. K., Papadopoulos, M. C., Stapleton, S., Kitchen, N. D., & Watkins, L. D. (2013). Systematic review of the outcome of shunt surgery in idiopathic normal-pressure hydrocephalus. *Acta Neurochirurgica, 155*(10), 1977–1980. https://doi.org/10.1007/s00701-013-1835-5

Tullberg, M., Hellström, P., Piechnik, S. K., Starmark, J. E., & Wikkelsö, C. (2004). Impaired wakefulness is associated with reduced anterior cingulate CBF in patients with normal pressure hydrocephalus. *Acta Neurologica Scandinavica, 110*(5), 322–330. https://doi.org/10.1111/j.1600-0404.2004.00325.x

Tullberg, M., Hultin, L., Ekholm, S., Månsson, J. E., Fredman, P., & Wikkelsø, C. (2002). White matter changes in normal pressure hydrocephalus and binswanger disease: Specificity, predictive value and correlations to axonal degeneration and demyelination. *Acta Neurologica Scandinavica, 105*(6), 417–426. https://doi.org/10.1034/j.1600-0404.2002.01189.x

Tully, H. M., & Dobyns, W. B. (2014). Infantile hydrocephalus: A review of epidemiology, classification and causes. *European Journal of Medical Genetics, 57*(8), 359–368. https://doi.org/10.1016/j.ejmg.2014.06.002

Tuminello, E. R., Holmbeck, G. N., & Olson, R. (2012). Executive functions in adolescents with spina bifida: Relations with autonomy development and parental intrusiveness. *Child Neuropsychology, 18*(2), 105–124. https://doi.org/10.1080/09297049.2011.590470

Urban, M., & Rabe-Jabłońska, J. (2013). Undiagnosed normal pressure hydrocephalus as a cause of diagnostic difficulties in a patient with depression. *Journal of Psychiatric Practice, 19*(6), 501–504. https://doi.org/10.1097/01.pra.0000438191.04786.5f

Van Roost, D., Solymosi, L., & Funke, K. (1995). A characteristic ventricular shape in myelomeningocele-associated hydrocephalus? A CT stereology study. *Neuroradiology, 37*(5), 412–417. https://doi.org/10.1007/BF00588025

Vanhala, V., Junkkari, A., Korhonen, V. E., Kurki, M. I., Hiltunen, M., Rauramaa, T., … Leinonen, V. (2019). Prevalence of schizophrenia in idiopathic normal pressure hydrocephalus. *Neurosurgery, 84*(4), 883–889. https://doi.org/10.1093/neuros/nyy147

Venkataramana, N. K., & Mukundan, C. R. (2011). Evaluation of functional outcomes in congenital hydrocephalus. *Journal of Pediatric Neurosciences, 6*(1), 4–12. https://doi.org/10.4103/1817-1745.84399

Vinck, A., Maassen, B., Mullaart, R., & Rotteveel, J. (2006). Arnold-Chiari-II malformation and cognitive functioning in spina bifida. *Journal of Neurology, Neurosurgery, and Psychiatry, 77*(9), 1083–1086. https://doi.org/10.1136/jnnp.2005.075887

Wald, N. J. (2004). Folic acid and the prevention of neural-tube defects. *New England Journal of Medicine, 350*(2), 101–102. https://doi.org/10.1056/NEJMp038186

Wald, N. J., & Bower, C. (1995). Folic acid and the prevention of neural tube defects. *BMJ (Clinical Research ed.), 310*(6986), 1019–1020.

Wall, V. L., Kestle, J. R., Fulton, J. B., & Gale, S. D. (2021). Social-emotional functioning in pediatric hydrocephalus: Comparison

of the hydrocephalus outcome questionnaire to the behavior assessment system for children. *Journal of Neurosurgery: Pediatrics, 28*(5), 572–578. https://doi.org/10.3171/2021.5.PEDS2178

Wang, Z., Zhang, Y., Hu, F., Ding, J., & Wang, X. (2020). Pathogenesis and pathophysiology of idiopathic normal pressure hydrocephalus. *CNS Neuroscience & Therapeutics, 26*(12), 1230–1240. https://doi.org/10.1111/cns.13526

Warf, B., Ondoma, S., Kulkarni, A., Donnelly, R., Ampeire, M., Akona, J., … Nsubuga, B. K. (2009). Neurocognitive outcome and ventricular volume in children with myelomeningocele treated for hydrocephalus in Uganda. *Journal of Neurosurgery: Pediatrics, 4*(6), 564–570. https://doi.org/10.3171/2009.7.PEDS09136

Wechsler, D. (1991). Wechsler intelligence scale for children. In *WISC-III): Manual* (3rd ed.). San Antonio, TX: The Psychological Corporation. https://doi.org/10.1007/978-1-4615-1185-4_11.

Weiss, L. G., Saklofske, D. H., Coalson, D. L., & Raiford, S. E. (2010). Theoretical, empirical and clinical foundations of the WAIS-IV index scores. In *WAIS-IV clinical use and interpretation* (pp. 61–94). Academic Press. https://doi.org/10.1016/B978-0-12-375035-8.10003-5.

Wiedenbauer, G., & Jansen-Osmann, P. (2006). Spatial knowledge of children with spina bifida in a virtual large-scale space. *Brain and Cognition, 62*(2), 120–127. https://doi.org/10.1016/j.bandc.2006.04.003

Wiedenbauer, G., & Jansen-Osmann, P. (2007). Mental rotation ability of children with spina bifida: What influence does manual rotation training have? *Developmental Neuropsychology, 32*(3), 809–824. https://doi.org/10.1080/87565640701539626

Williams, H. (2008). A unifying hypothesis for hydrocephalus, Chiari malformation, syringomyelia, anencephaly and spina bifida. *Cerebrospinal Fluid Research, 5*(7). https://doi.org/10.1186/1743-8454-5-7

Williams, M. A., & Malm, J. (2016). Diagnosis and treatment of idiopathic normal pressure hydrocephalus. Continuum. *Continuum: Lifelong Learning in Neurology, 22*(2 Dementia), 579–599. https://doi.org/10.1212/CON.0000000000000305

Wills, K. E., Holmbeck, G. N., Dillon, K., & McLone, D. G. (1990). Intelligence and achievement in children with myelomeningocele. *Journal of Pediatric Psychology, 15*(2), 161–176. https://doi.org/10.1093/jpepsy/15.2.161

Wilson, B. A., Emslie, H., Quirk, K., Evans, J., & Watson, P. (2005). A randomized control trial to evaluate a paging system for people with traumatic brain injury. *Brain Injury, 19*(11), 891–894. https://doi.org/10.1080/02699050400002363

Yadav, Y. R., Parihar, V., Pande, S., Namdev, H., & Agarwal, M. (2012). Endoscopic third ventriculostomy. *Journal of Neurosciences in Rural Practice, 3*(2), 163. https://doi.org/10.4103/0976-3147.98222

Yeates, K. O., & Enrile, B. G. (2005). Implicit and explicit memory in children with congenital and acquired brain disorder. *Neuropsychology, 19*(5), 618–628. https://doi.org/10.1037/0894-4105.19.5.618

Yeates, K. O., Enrile, B. G., Loss, N., Blumenstein, E., & Delis, D. C. (1995). Verbal learning and memory in children with myelomeningocele. *Journal of Pediatric Psychology, 20*(6), 801–815. https://doi.org/10.1093/jpepsy/20.6.801

Zaccaria, V., Bacigalupo, I., Gervasi, G., Canevelli, M., Corbo, M., Vanacore, N., et al. (2020). A systematic review on the epidemiology of normal pressure hydrocephalus. *Acta Neurologica Scandinavica, 141*(2), 101–114. https://doi.org/10.1111/ane.13182

Zhang, J., Williams, M. A., & Rigamonti, D. (2006). Genetics of human hydrocephalus. *Journal of Neurology, 253*(10), 1255–1266. https://doi.org/10.1007/s00415-006-0245-5

Zimmerman, K., May, B., Barnes, K., Arynchyna, A., Alford, E. N., Wessinger, C. A., … Rocque, B. G. (2020). Anxiety, depression, fatigue, and headache burden in the pediatric hydrocephalus population. *Journal of Neurosurgery: Pediatrics, 26*(5), 483–489. https://doi.org/10.3171/2020.4.PEDS19697