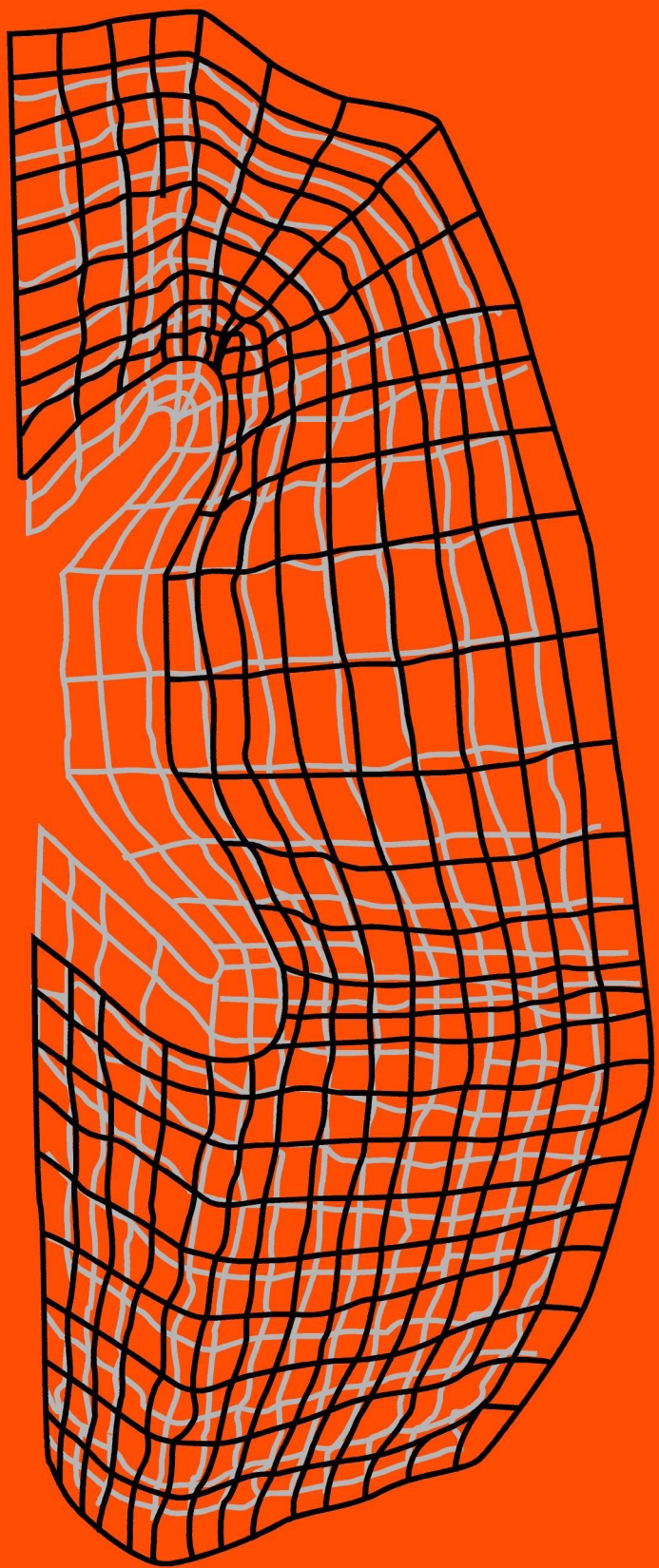

HYDROCEPHALUS IN CHILDREN

Cognition and behaviour

Barbro Lindquist

2007

Göteborg University

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Barbro Lindquist

Department of Pediatrics
Institute of Clinical Sciences

The Sahlgrenska Academy at Göteborg University
Sweden

Göteborg 2007

Surgical treatment of hydrocephalus. Şerefeddin Sabuncuoğlu, 1465.

Abstract

Aims: The main objective of this thesis was to explore the cognitive and behavioural consequences of hydrocephalus in children born at term and preterm, with or without myelomeningocele (MMC) and with or without concomitant neurological impairments, such as cerebral palsy (CP), epilepsy or learning disabilities.

Material and methods: From a population-based cohort of all 107 children with hydrocephalus born in 1989-1993, 73 of the surviving children were assessed with intelligence tests and most of them also using behavioural and autism rating scales. Thirty-six of 47 (77%) children with an IQ of ≥ 70 and eight children with MMC but no hydrocephalus were assessed with a neuropsychological test battery (NIMES) and compared with age- and gender-matched controls.

Results: One-third of the children were normally gifted (IQ > 85), another 30% had a low-average IQ of 70-84 and 37% had learning disabilities (IQ < 70). An IQ of < 70 was found in 42% of children without MMC and in 29% of those with MMC. Children born preterm had a lower IQ than those born at term. Children with CP and/or epilepsy had significantly lower IQ scores than those without these impairments. Parents rated 67% and teachers 39% of the children as having behavioural problems. Learning disabilities increased the risk significantly. Almost all the children with CP and/or epilepsy had behavioural problems. Learning disabilities, CP and epilepsy significantly increased the risk of autistic symptoms, which were present in 13 %, in 4 % of those with MMC and in 20 % of those without MMC. Children with hydrocephalus both with and without MMC and with an IQ of > 70 performed significantly less well than controls on learning, memory and executive functions but not on registration skills. There were no differences between children with hydrocephalus in combination with MMC and those without MMC, whereas children with MMC but no hydrocephalus and normal intelligence performed as well as controls on all the neuropsychological functions.

Conclusions: The majority of children with hydrocephalus had learning disabilities or a low-average IQ, as well as behavioural problems, and some had autistic symptoms. Despite average or slightly below average intelligence, children with hydrocephalus had major difficulties with learning and memory and with executive functions, regardless of the aetiology of the hydrocephalus. Only MMC did not appear to influence cognitive and neuropsychological outcome as much as the brain lesion causing or caused by the hydrocephalus.

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List of publications

I. Lindquist, B., Carlsson G., Persson, E-K., Uvebrant, P.
Learning disabilities in a population-based group of children with hydrocephalus.
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Behavioural problems and autism in children with hydrocephalus – a population-based study.
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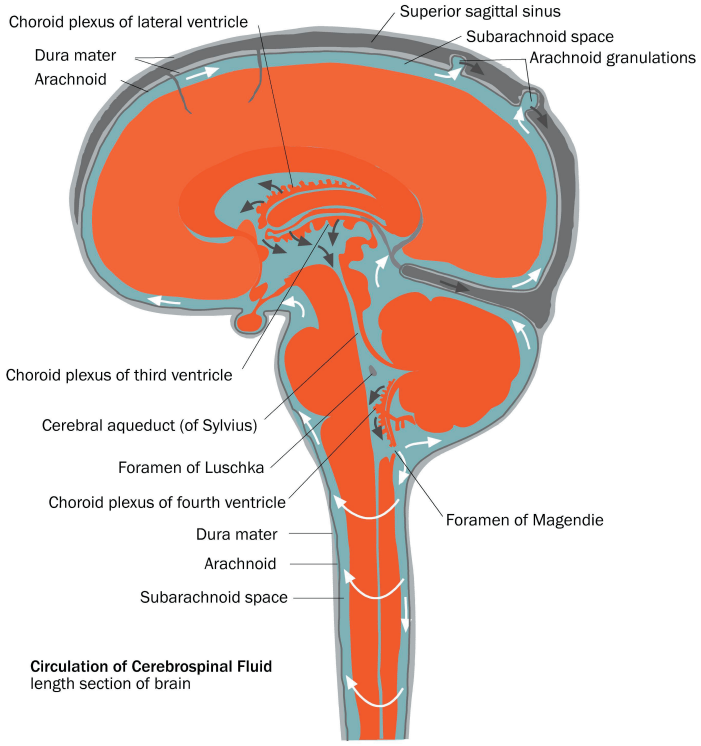
Abbreviations

ADHD	Attention Deficit Hyperactivity Disorder
ASD	Autism Spectrum Disorder
CARS	Childhood Autism Rating Scale
CNS	Central nervous system
CP	Cerebral palsy
CSF	Cerebrospinal fluid
CORSI	Corsi Block Test
DSM	Diagnostic and Statistical Manual of Mental Disorders
FSIQ	Full scale IQ
HOQ	Hydrocephalus Outcome Questionnaire
HC	Hydrocephalus
IH	Infantile hydrocephalus
IQ	Intelligence Quotient
MMC	Myelomeningocele
NIMES	Neuropsykologiska utredningsmetoder för inlärning, minne och exekutiva funktioner för barn i skolåldern
PIQ	Performance IQ
PVL	Periventricular leukomalacia
RAVLT	Rey Auditory-Verbal Learning Test
ROCF	Rey Oesterreich Complex Figure
SD	Standard deviation
TOLT	Tower of London Test
VIQ	Verbal IQ
WISC	Wechsler Intelligence Scale for Children
WPPSI	Wechsler Preschool and Primary Scale of Intelligence

Introduction

Medical background

Hydrocephalus generally refers to an increase in cerebrospinal fluid (CSF) volume, caused by a disturbance in the production, circulation or resorption of the CSF that is secondary to some pathological event or structural brain anomaly. It is accompanied by the enlargement of the cerebral ventricles, which has an impact on the structure and function of the brain. Hydrocephalus is a fairly common disorder of childhood, with a prevalence of 0.8 per 1,000 live births in a recent population-based study (Persson et al., 2005). In children born at term, prenatal aetiology dominates (70%), most often in the form of malformations and infections. In children born preterm, perinatal causes dominate (60%), mainly in the form of intraventricular haemorrhage (Fernell et al., 1987). Another congenital cause of hydrocephalus is myelomeningocele (MMC). In MMC, the neural tube fails to close completely in the very early development of the foetus, which results in the destruction of nerves and the medulla, which in turn causes paralysis in the lower limbs and impairs bladder control and bowel function. About 80% of children with MMC have an associated Arnold Chiari malformation including the brainstem and the cerebellum that introduces a barrier to CSF outflow from the ventricular system to the subarachnoid space.



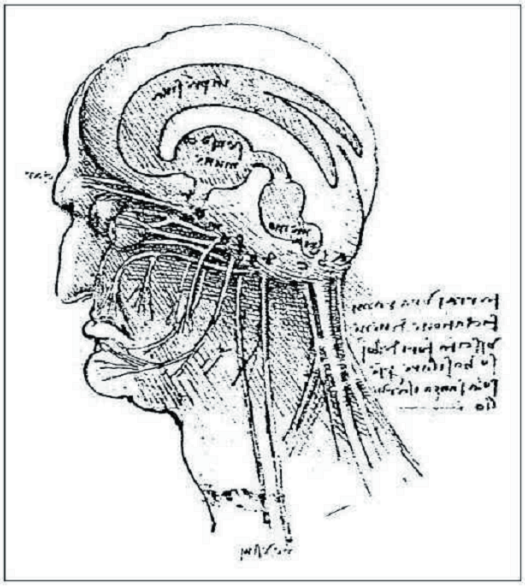
Circulation of Cerebrospinal Fluid
length section of brain

History of hydrocephalus and its treatment

Hydrocephalus was first described by Hippocrates (466-377 BC), as a liquefaction of the brain caused by epileptic seizures. In ancient times, it referred to fluid collections surrounding the brain. The first accurate description of the ventricular anatomy and the CSF was made by Claudius Galen of Pergamon (130-200 AD), who obtained his knowledge from animal dissection, but he still described hydrocephalus as fluid collections outside the brain.

In western literature, Leonardo da Vinci published the first illustration of the ventricular system in 1510, but, in 1551, Andreas Vesalius (1514-1564) was the first to understand and describe the fact that the water was collected in the ventricles. One century later, Thomas Willis (1621-1675) postulated that CFS must flow into the venous system (Schulze 1968; Aschoff et al., 1999).

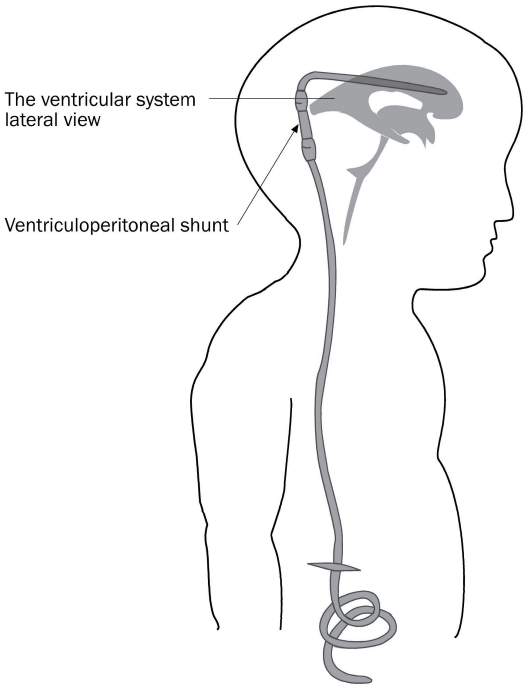
In 1465, the Turkish pioneer surgeon Şerefeddin Sabuncuoğluin published a book entitled *Imperial surgery*, in which he described and illustrated surgical techniques. He recommended the treatment of hydrocephalus by decompression of the ventricular system using a scalpel and he used olive oil and wine for antisepsis (Elmaci, 2000). This work was ignored for a long time as it was written in Turkish when Arabic was the scientific language and it was not known of in the western world until the middle of the twentieth century, while the recommended treatment of hydrocephalus in the eighteenth and nineteenth centuries was bandaging and dehydrating drugs. At the turn of the century, occasional attempts were made to implant shunts of different kinds to drain the ventricles. In 1949, the first functional shunt valve was constructed and implanted and the breakthrough in effective treatment came in 1955-1960, when silicone was also introduced as a construction material.



The ventricular system illustrated by Leonardo Da Vinci 1510.

Treatment of hydrocephalus today

The most frequent treatment of hydrocephalus is still the so-called shunt operation, in which a plastic tube placed in the ventricle leads the CSF to another body space, usually the peritoneal cavity or atrium of the heart. The CSF is resorbed there by the omentum and the peritoneum. A valve is connected to prevent the over-draining of the ventricles. Before the introduction of this method, in the 1960s, mortality in children born with hydrocephalus was very high, about 50% (Hagberg & Sjögren, 1966). In the beginning, there were many complications, but, since the 1970s, the methods have become more effective, with a survival rate of 90%, in many cases without disabilities. Despite fairly frequent postoperative complications, shunting has been shown to reduce intracranial pressure and improve neurological and cognitive function (Mataro et al., 2001). An alternative treatment is ventriculostomy, which has been in use since the 1990s (Hopf et al., 1999). With this method, a hole is made in the bottom of the third ventricle, whereby the circulation of the CSF can be reconstructed. Lately, attempts have also been made to minimise the consequences of hydrocephalus by performing prenatal surgery by shunt-operating foetuses between 24 and 32 weeks of gestation. In a follow-up of prenatally operated children, the IQ was about the same as in children operated on after birth (Cavalheiro et al., 2003). There is also experience of foetal surgery for MMC. No functional improvements have been found in the lower extremities or bladder and bowel functions, but the need for shunt operations is reported to be significantly reduced. As this surgery has so far also led to an increased rate of early delivery and mortality, it is not generally recommended (Bruner et al., 1999; Farmer et al., 2003).



Cerebral development

Knowledge of the biological processes and timing of CNS maturation offers an opportunity to understand the impact of disruption on cognitive performance. The CNS begins to develop around day 40 of embryonic life and, at around 100 days of gestation, it is recognisable in its mature form. Most research suggests that there is a hierarchical progression within the CNS, with cerebellar/brain stem areas maturing first, followed by posterior areas and then anterior regions, particularly the frontal cortex. Prefrontal regions are the last to mature. The cells generate early in gestation in the neural tube and then, at around six weeks of gestation, they migrate from there to predetermined locations within the nervous system. Most of the migration is completed by the 16th week of gestation, but the process is not fully completed until five months after birth. Once neurons have migrated, they begin the process of differentiation in which the nerve-cell bodies develop and the axons and the dendrites grow and form synaptic connections. In this process, there is also selected cell death, about 50% in some areas. It is thought that the neurons that do not make appropriate connections are eliminated. The myelination of the axons, which facilitates nerve conduction, begins around the 14th week of gestation, but most myelination occurs postnatally, especially during the first three years of life, and then at a slower rate until the peri-pubertal stage where there is an increase, possibly associated with hormonal changes within the brain. Myelination is essential in providing effective interhemispheric communication and the input-output of sensory and motor information.

Risk factors

Risk factors for disturbed brain development can be pre-, peri- or postnatal. Genetics and maldevelopments are important etiological factors causing MMC or learning disabilities in children, for example. Maternal health (infections, stress), alcohol and drug addiction, smoking and malnutrition are other examples of prenatal risks. Perinatal factors such as preterm birth, birth complications and postnatal cerebral infection, nutritional problems and trauma may cause anomalies in the CNS. Interruptions in normal myelination can lead to impaired information processing, reduced speed and reduced attention. Psychosocial factors such as maternal deprivation or other disturbances in the mother-child relationship, as well as a disadvantaged environment as a whole, may also have an impact on development (Anderson, 2001).

Neuropsychological background

Neuropsychology is defined as the study of the relationship between brain function and behaviour (Kolb & Wishaw, 1989). This involves behaviour related to the structure and function of the brain, including cognitive functions such as learning, memory, attention and executive functions, as well as behavioural consequences traditionally referred to as neuropsychiatry (regulation of activity, conduct disorders and autism spectrum disorders).

The evolution of applied clinical neuropsychology has been tremendous during the last few decades. Our modern knowledge about the localisation of functions in the brain, and of understanding the origins of behaviour, stems from Alexander Luria's

screening and diagnosis of impaired servicemen during the First World War (Luria, 1973). These adult models may form the basis of our knowledge of neurological disorders in children as well, but they are insufficient in the challenging attempt to understand the effects of injuries to the developing brain (Anderson, 2001). Child neuropsychology has developed by studying behaviour and learning disabilities, for example. For a long time now, there have been observations and descriptions of special characteristics in children with different kinds of brain injury and the development and use of neuropsychological assessment has contributed valuable knowledge about the way different background factors manifest themselves in abilities of learning and behaviour. The parallel development of neuroimaging techniques has also contributed to a stronger connection between brain injury and its consequences. These techniques per se are, however, instruments that are too rough to explain cognitive and behavioural outcome, as the same picture of injury can appear differently in different individuals. It is a challenge to try to understand how the interactions between organic, social, developmental and psychological factors affect the child and lead to the observed outcome. Child neuropsychology as a science is useful for this purpose and the development of knowledge and methods is ongoing.

Intelligence

In 1905, Alfred Binet constructed the first test of intelligence, a commission from the French government, with the main purpose of identifying children at school with learning disabilities. His theoretical concept of intelligence included the abilities of planning, strategies and reflection. His tests were still used, in revised forms, until the 1960s. The methods that are used most frequently today to measure intelligence are the Wechsler scales. They are translated and used all over the world, which makes it possible to compare results in a way that is valid and reliable. David Wechsler developed the original scale for adults in the late 1930s and the first intelligence scale for children in 1949 (Wechsler, 1949). It has been subsequently adapted over the years and the norms have been periodically updated, which is essential as, when outdated norms are used, a child's IQ score will generally be higher than it is when current norms are used. Wechsler viewed intelligence as "the capacity of the individual to act purposefully, to think rationally and to deal effectively with his or her environment". The subtests of the WISC-III (Wechsler, 1992) have thus been selected to tap many different mental abilities, which taken as a whole indicate a child's general intellectual ability. This ability is expressed in the Full Scale IQ (FSIQ), which is subdivided into Verbal IQ (VIQ) and Performance IQ (PIQ). The WISC-III is used for children between six and 16 years of age. In 1967, Wechsler constructed a similar scale for children between three and seven years of age, the WPPSI (Wechsler Preschool and Primary Scale of Intelligence) (Wechsler, 1967), which is also divided into VIQ, PIQ and FSIQ. This makes it possible to compare the children's abilities over time.

For younger children, developmental scales based on parental interviews and observations of children are used when evaluating cognitive abilities; they include the Bayley Scales of Infant Development (Bayley, 1993), or the Griffiths' Developmental Scale (Griffiths', 1980).

Another attempt to define intelligence was made by Howard Gardner (Gardner, 1993) who described eight "signs" of intelligence that he defined as "the capacity to solve problems or to fashion products that are valued in one or more cultural setting". His theories have been accepted and applied by educators and educational theorists, but, in clinical practice and research, the Wechsler Scales are predominant.

Hydrocephalus and neuropsychology

It has been shown that early-onset hydrocephalus (i.e. congenital or developing during the first year of life) is frequently associated with deficiencies in intellectual and/or behavioural development. (Dennis et al., 1981; Fernell et al., 1987; Riva et al., 1994; Lumenta et al., 1995; Kao et al., 2001). The intraventricular pressure produced by hydrocephalus causes the expansion of the ventricles and the displacement of adjacent brain structures. With obstruction, the ventricles expand in a posterior-to-anterior direction. If left untreated, the ventricular expansion causes cerebral oedema, which initially affects cerebral white matter and eventually extends to the grey matter. Also in treated hydrocephalus, the posterior regions of the brain are more susceptible to damage because of the posterior-to-anterior progression (Del Bigio, 1993; Mataro et al., 2001).

It has been shown that regional variations in brain tissue composition in children with shunted hydrocephalus correlate with a variety of cognitive and visuomotor functions. In children with early-onset hydrocephalus, measurable reductions were found in the size of two cerebral white-matter structures, the corpus callosum and the internal capsule, which correlated positively and significantly with non-verbal cognitive measures (Fletcher et al., 1992; Hannay 2000). Another study comparing cognitive measures, motor and executive function in children with shunt-treated hydrocephalus and spontaneously arrested hydrocephalus revealed that the thickness of the corpus callosum was strongly correlated with various non-verbal measures and also with fine motor co-ordination. In that study, there was no connection with executive functions and only a weak connection with verbal measures. The highest correlation with performance IQ was found in the shunt-treated group (Fletcher et al., 1996).

Also in children born preterm with hydrocephalus both hydrocephalus per se and the common complication of haemorrhage and leukomalacia can cause corpus callosum anomalies and the loss of cerebral white matter. Hydrocephalus – independent of aetiology – therefore appears to be associated with greater impairment of non-verbal cognitive skills than verbal skills (Dennis et al. 1981; Fletcher et al., 1992, 1996). Moreover, the underlying insult (infection, haemorrhage or malformation) often causes not only hydrocephalus but also other lesions that cause or worsen the impairment.

The elevated incidence of left-handedness in children with MMC and hydrocephalus (about 30% compared with 10% in the normal population) has been explained by white-matter loss in the corpus callosum (Wassing et al., 1993), or other developmental disorders which cause delayed or incomplete lateralisation with left- or mixed-handedness as a consequence.

Intelligence

In a study from 1962 of children with hydrocephalus who were not surgically treated, only 46% survived infancy and the survival to adult life was only 20-23%. At the time of follow-up, only 38% of the children achieved IQ scores in the average range (Laurence & Coates, 1962). Intellectual performance in children with hydrocephalus has since been investigated continuously and the overall IQ has been reported to be in the range of low average or below. About 30-40% of the children have been reported to have an IQ of < 70, which implies learning disability and the need for special schools (Lumenta et al., 1995; Hoppe-Hirsch et al., 1998; Heinsbergen et al., 2002). Children with hydrocephalus and MMC have been found to have higher IQ scores than children with hydrocephalus without MMC (Hoppe-Hirsch et al., 1998; Kao et al., 2001).

Verbal functions

It is frequently documented that children with hydrocephalus generally have a characteristic test profile in which verbal intelligence exceeds non-verbal intelligence (Dennis et al., 1981; Donders et al., 1991; Riva et al., 1994; Brookshire et al., 1995; Lumenta et al., 1995). It can therefore be assumed that language is generally well preserved in children with hydrocephalus. Back in the 1960s, however, specific language deficits were described; they included the "cocktail party syndrome", characterised by hyperverbal behaviour and the use of advanced vocabulary that did not always correspond to understanding the meaning of words or the context in which they should be used (Taylor, 1961; Hagberg & Sjögren, 1966). Later findings have confirmed and further elaborated these early results. Children with hydrocephalus often have difficulty with pragmatics and discourse, verbal fluency and the comprehension of complex grammatical structures (Donders et al., 1991; Dennis & Barnes, 1993; Dennis et al., 1994; Brookshire et al., 1995; Vachha & Adams, 2003). Barnes et al. (2001) found that children with hydrocephalus were as good as controls at reading words, but this decoding speed did not contribute to reading comprehension. They suggested that the reason for poor reading comprehension could be attributed to anomalies of the corpus callosum, which prevent the co-operation between the brain hemispheres necessary for this function. A further study by Barnes et al. (2004) revealed that children with hydrocephalus had difficulty integrating information from previously read sentences in a text in order to understand the next sentence, which reflect deficits in both long-term memory and working memory.

Non-verbal functions

Non-verbal deficits are explained as being related to the loss of white matter in the posterior regions of the brain and also to the abnormalities of the corpus callosum that are frequently found (Fletcher et al., 1992; Fletcher et al., 1996; Hannay, 2000). The loss of periventricular white matter also causes visual perception problems as in children born preterm with periventricular leukomalacia (PVL) (Jacobson et al., 1996) and in children with congenital hemiplegia (Carlsson et al., 1994). Non-verbal problems manifest themselves as poor performance on visual recognition/discrimination tasks, eye-hand co-ordination, visuo-construction, visuo-orientation

and recognition of faces (prosopagnosia) (Houliston et al., 1999). The enlargement of the ventricles is likely to cause damage to the optic nerves and oculomotor pathways, which also helps to explain the children's visual impairments (Houliston et al., 1999). In children with MMC and hydrocephalus, it was previously hypothesised that upper-limb dysfunction could help to explain their eye-hand co-ordination and visuo-constructional difficulties (Wills et al., 1990). Fletcher et al. (1992) found that impairments in co-ordinate perception were not due to impaired movement, as children with MMC had difficulties not only in drawing tasks that require both visual perception and motor control, but also on tasks with limited motor components. Dennis et al. (2002) reported that children with MMC and hydrocephalus have relative strengths on visual perception tasks involving categorical relationships, such as recognising objects and faces, and relative difficulty with objects in motion and figure-ground tasks.

Memory

Only a few studies have been performed on memory function in children with hydrocephalus. Scott et al. (1998) found that, in children with an IQ of > 70, those with shunted hydrocephalus performed less well on both verbal and non-verbal memory tasks than children with arrested or no hydrocephalus. Likewise, Fletcher et al. (1992) reported that children with hydrocephalus were impaired in relation to controls on both verbal and non-verbal memory measures. Yeates et al. (1995) explored verbal learning and memory in children with MMC and hydrocephalus and reported deficits in recall but assets in the recognition of word lists. These findings were confirmed in a study of implicit and explicit memory (learning without and with conscious recollection respectively), where children with MMC and hydrocephalus had a relatively intact implicit memory and a poor explicit memory, in both perceptual and verbal tasks (Yeates & Enrile, 2005). It could be expected that children with MMC and hydrocephalus with their abnormalities in the posterior cortex and reported visuo-perceptual difficulties would under-achieve in implicit perceptual memory. However, the hypothesis suggested by Casey et al. (2000), that implicit memory is mediated by more distributed brain systems in children appears to be confirmed here.

Executive functions

"The executive functions consist of those capacities that enable a person to engage successfully in independent, purposive, self-serving behaviour" (Lezak, 2004). Executive dysfunction means having difficulty with planning and organisation and using strategies, inability to use feedback and rigid or concrete thought process. The few existing studies of executive function in children with hydrocephalus generally illustrate single, or just a few, aspects.

Fletcher et al. (1996) reported poorer results on planning tasks performed by children with hydrocephalus, but they were not discussed in terms of executive dysfunction, but as a result of a generalised spatial problem-solving deficit due to the loss of white matter. Anderson et al. (2002) identified significant symptoms of executive dysfunction in children with hydrocephalus in both auditive-verbal and visuo-per-

ceptual tests. Vachha & Adams (2005) found that children with MMC and hydrocephalus had ineffective meta-cognitive strategies when trying to learn word lists.

Behaviour and attention

Children with physical disabilities due to brain lesions have been shown to have more behavioural and emotional disorders than the general population (Rutter, 1981; Seidel et al., 1975). These disorders may be unrelated to the lesion, they may be a consequence of it, or secondary to the dysfunction when it comes to the way the children are able to cope socially and emotionally (Herzberg & Herzberg 1977; Connell & McConnell 1981). As many as about 40% of children with hydrocephalus have been reported to have behavioural problems (Connell & McConell 1981; Fernell et al., 1991; Fletcher et al., 1995; Williams & Lyttle, 1998). While parents and teachers often report behavioural deficits, the children themselves seldom perceive these problems (Williams & Lyttle. 1998). The aetiology of the hydrocephalus does not appear to affect the rate of behavioural problems, at least not in children with hydrocephalus without MMC, as much as the co-existence of other neuroimpairments such as learning disabilities, which multiply the risk (Fernell et al., 1991).

There are few studies focusing directly on attentional processes in children with hydrocephalus. Fletcher et al. (1996) showed that children with hydrocephalus solved fewer problems on tests of focused attention and selective attention. They argued, however, that the results possibly did not reflect poorer frontal lobe control but difficulty sustaining attention and greater distractibility, i.e. functions that are assumed to be mediated by posterior white-matter regions of the brain. When comparing attention processes in children with shunted hydrocephalus and children with ADHD (Attention Deficit-Hyperactivity Disorder) and normal controls, children with hydrocephalus displayed an inability to focus and shift attention compared with controls (Brewer et al., 2001). Burmeister et al. (2005) found that 31% of a group with MMC and hydrocephalus had ADHD, mostly the inattentive type (23%), using a parent rating scale.

Autism

The core symptoms of autism spectrum disorders (ASD) are impaired social interaction and communication, a lack of flexibility in behaviour and an inability to use the imagination (DSM-IV, 1994). In the general population, it has been suggested that the current rate of ASD is between 30 and 60 cases per 10,000, about a quarter of which meet the full criteria for autism (Chakrabarti et al. 2001; Rutter, 2005). Several reports have described a connection between autism and the severity of brain dysfunction (Chakrabarti et al., 2001; Fernell et al., 1991; Fombonne 1999; Kielinen et al., 2004). The prevalence of autism in children with hydrocephalus has, however, rarely been investigated. Fernell et al. (1991) reported autistic symptoms in 23% of children with hydrocephalus without MMC.

Psychological and social issues

Children with hydrocephalus with or without MMC represent a wide variety of outcomes, where some children after treatment have spared cognitive or motor

functions, while others are affected, with severely disabling impairments of motor and intellectual functioning and additional problems of epilepsy and cerebral palsy. However, the majority appear to have problems that put them at risk of learning difficulties at school, cognitive and social inferiority to peers and, secondarily, low self-esteem. Connell (Connell & Connell, 1981) found a high rate of neurotic disturbance in children of primary-school age. A study of disability and quality of life in children with MMC and hydrocephalus revealed 1 SD lower quality of life for these children than for children with MMC only and isolated hydrocephalus without MMC (Cate et al., 2002). There is a need for further studies of the psychological and social consequences of hydrocephalus in children in order to give the most appropriate support to children and parents. Knowledge of the kind of motor, cognitive and neuropsychological impairments that generally occur, help to ask the right questions and choose the appropriate methods when examining the children are all important, but we need better instruments to meet the need for advice and care. Kulkarni et al. (2004) constructed a Hydrocephalus Outcome Questionnaire (HOQ) for measuring the health status in the children, as well as a questionnaire for assessing parents' concerns about their child with hydrocephalus (Kulkarni, 2006), which could be useful for this purpose.

Aims

Neurosurgical treatment has developed during the last few decades and there has been an increase in the survival of children with early-onset hydrocephalus. Moreover, the aetiological panorama has changed, with an increase in the survival of children born very preterm, with a high risk of developing post-haemorrhagic hydrocephalus (Fernell et al., 1990), as well as a reduction in the birth of children with neural tube defects (Shurtleff & Lemire, 1995).
It is therefore important to reconsider, update and extend earlier research in the fields of cognition, neuropsychological functioning and behaviour, in order to be able correctly to inform parents about the prognosis and provide the child with the optimal support.

Aims

- To assess cognitive functions and investigate the prevalence of behavioural problems and symptoms of autism in a population-based group of children treated surgically for hydrocephalus during the first year of life
- To determine whether the cognitive and behavioural outcome differed between children with neural tube defects associated with the hydrocephalus and those without these defects
- To compare cognitive outcome in children born at term with those born at earlier gestational ages and to see whether preterm birth per se added the risk of subsequent behavioural problems or autistic symptoms
- To see whether additional impairments in the form of cerebral palsy and/or epilepsy increased the risk of cognitive and behavioural problems and autistic symptoms
- To investigate the profile of intelligence regarding verbal and performance IQ in children with hydrocephalus
- To explore the extent to which the neuropsychological functions of learning, memory and executive abilities are impaired in children with hydrocephalus with an IQ of ≥ 70 and to find out whether children with hydrocephalus without MMC differ from those with MMC and compare their results with those of controls

Material

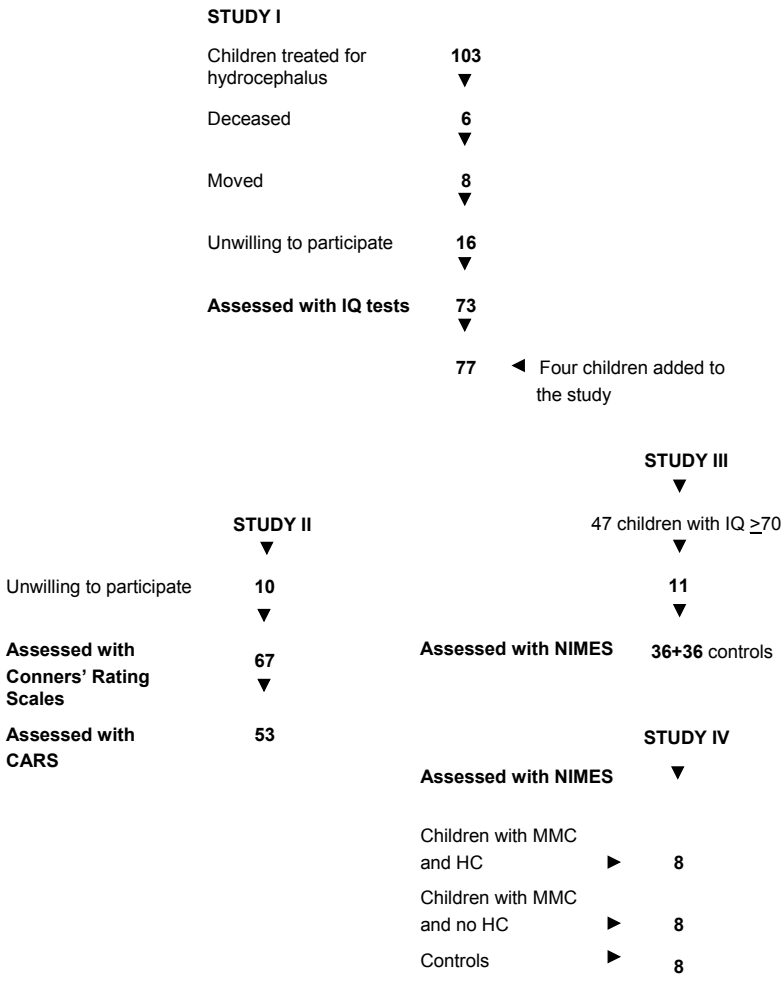


Figure 1. Overview of the study groups participating in cognitive testing, behavioural assessment (Conners' Rating Scales), assessment for autism (CARS) and assessment for neuropsychological functions (NIMES).

Subjects and methods

Study I:
Learning disabilities in a population-based group of children with hydrocephalus
The children who were studied were drawn from an ongoing epidemiological study, comprising all 206 children born during the period 1989 to 1998 and treated for hydrocephalus in the western Swedish health-care region (Persson et al., 2005). The age group suitable for psychological assessment comprised the 103 children born during the five-year period 1989-1993. Six of the children had died and eight had moved out of the region and were therefore excluded from the study. Of the remaining 89 children, 16 were unwilling to participate, leaving 73 as the study group. The 30 children who had died or were lost to follow-up did not differ from the study group in terms of aetiology, associated MMC, rate of preterm birth or whether they were born with hydrocephalus or developed it later during the first year of life (Table 1).

	Lost to follow-up	Study group
Hydrocephalus/MMC	11 (37%)	28 (38%)
Infantile hydrocephalus	19 (63%)	45 (62%)
Born at full term (> 36 weeks of gestation)	23 (77%)	52 (71%)
Born preterm (< 37 weeks of gestation)	7 (23%)	21 (29%)
Hydrocephalus at birth	12 (40%)	28 (38%)

Table 1. Aetiological characteristics in a studied group of 73 children with hydrocephalus compared with 30 children lost to follow-up.

Of the 73 children studied, five (7%) had cerebral palsy, eight (11%) epilepsy and nine (12%) both cerebral palsy and epilepsy. Among the children born preterm, the single most common cause of hydrocephalus was cerebral haemorrhage at birth (9 of 21) and, among children born at term, MMC was the most common cause (23 of 52) (Fig. 2).

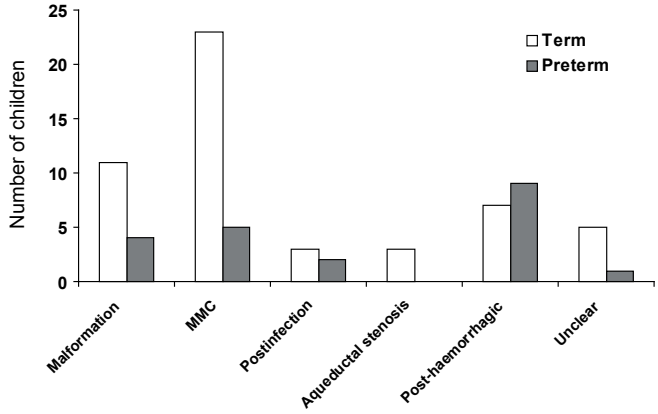


Figure 2. Aetiology of hydrocephalus in 73 children born in 1989-1993.

Study II:
Behavioural problems and autism in children with hydrocephalus – a population-based study

In connection with the psychological testing, the parents were asked to rate their children's behaviour using the Conners' Parents Rating Scales (Conners, 1989) and they were also asked for their permission to allow the children's teachers to use Conners' Teachers Rating Scales (Conners, 1989). The parents of 65 children agreed (parents of 64 children filled in the Parents' Scale and teachers of 55 children were allowed to use the Teachers' Scale). After the data collection was finished, at the time at which the collected material was evaluated, another four children who were treated for hydrocephalus between 1989 and 1993 were found. One of them had moved out of the region, while the other three were asked to participate in the study of behavioural problems and autism. Two agreed. So, from the population of 107 children, 67 were rated with the Conners' Scales; 66 on the Parents' Scales and 57 on the Teachers' Scales. Twenty-six children had hydrocephalus in combination with MMC. Seventeen children were born preterm and 50 were born at full term. The children were between five and twelve years old (mean 8 years 4 months) at the time of rating.

There were no significant differences between the study group and the 25 (27%) unwilling to participate, in terms of gender, learning disabilities, percentage of MMC or preterm birth.

Study III:
Learning, memory and executive functions in children with hydrocephalus

Forty-six children participating in study one and one of the two added children from study two had an IQ of ≥ 70, which was the inclusion criterion for participating in study three. Thirty-six agreed (77%), 23 boys and 13 girls, aged eight to 13 years, with a median IQ of 84 (range 70-112). Sixteen children had hydrocephalus in combination with MMC (median IQ = 78; range 71-109) and 20 children had infantile hydrocephalus (median IQ = 89; range 70-112). Six children with infantile hydrocephalus had a post-haemorrhagic hydrocephalus and four children a post-infectious hydrocephalus. In six, the hydrocephalus was caused by a malformation and one child had a stenosis of the aqueduct. In three children, the aetiology was unknown. The eleven (23%) children who were unwilling to participate did not differ from the study group in terms of gender (50% vs. 60% boys), aetiology (40% in both groups had MMC) or IQ (84 vs. 93; p=0.17).

The 36 children in the study groups were compared with an age- and gender-matched control group of 36 healthy children from Swedish mainstream schools, comprising children with an IQ of > 70. The median age in both groups was 11.6 years (range 8-13).

Study IV:
Cognitive functions in children with myelomeningocele without hydrocephalus

Between 1992 and 1999, there were 69 children with MMC among the 188,998 children born in western Sweden, i.e. a live birth prevalence of 3.5 per 10,000. Nine

of these children (15%) did not develop hydrocephalus, corresponding to a prevalence of 0.5 per 10,000 live births. One child had moved out of the country and the eight remaining children and parents all agreed to participate in the study. The children were between eight and 13 years of age (mean age 10.5), five boys and three girls. They were matched for age and gender with eight children with MMC and hydrocephalus (mean age 11.2) and eight controls (mean age 10.7),
The normal controls were selected from mainstream schools and were considered to have an average intelligence. The inclusion criterion for children with hydrocephalus was an IQ of at least 70.

Methods

Measures of intelligence (Studies I and IV)

The 73 children (41 boys and 32 girls) were five to 10 years of age at the psychological examination. Intelligence was assessed in 20 children using the WPPSI-R (Wechsler, 1991) and the WISC-III was used in 42 (Wechsler, 1992). For WISC-III, the English 1992 norm data were used, while the Swedish 1991 norms were used for the WPPSI-R. With four children, it was only possible to use parts of the Wechsler scales and their full-scale IQ was estimated on the basis of incomplete results. The Griffiths' Developmental Scales (Griffiths, 1980) were used in eight children with a developmental age of less than three years. Their development quotients were then converted to IQ equivalents. Three children had communication difficulties in combination with severe learning disabilities, which made it impossible to perform formal testing, and their developmental level was estimated as profound retardation (IQ < 20). The DSM-IV criteria were used to describe the different levels of learning disability. An IQ of below 20 thus meant profound mental retardation, IQ 20-34 severe retardation, IQ 35-49 moderate and IQ 50-69 mild mental retardation. Children with an IQ of between 70 and 84 were considered as low average, and those with IQ >84 were considered as average or above average.
In study IV the eight children with MMC without hydrocephalus were tested with the 1992 version of WISC-III (Wechsler, 1992). Six of the children with MMC and hydrocephalus had been tested in study one with WISC-III and two children with WPPSI-R (Wechsler, 1991).

Measures of behaviour (Study II)

The Conners' Rating Scales for parents and teachers were used (Conners, 1989). The parents' 48 item scales include the sub-scales of conduct problems, learning problems, psychosomatic, impulsive-hyperactive, anxiety and the hyperactive index. The teachers' 28 item scales include the sub-scales of conduct problems, hyperactivity, inattentive-passive and the hyperactivity index. The latter index is frequently used clinically, for example, when evaluating medication in children with hyperactivity problems. It consists of ten items for each scale measuring the extent to which the child performs behaviours that are regarded as indicative of an underlying diagnosis of hyperactivity. The items in the parents' and teachers' scales have four alternative answers; not at all, just a little, pretty much or very much. The cut-off level for problems much above average is set to 1.5 standard deviations (T-score > 65) and, for

problems very much above average, 2 standard deviations (T-score > 70). A standard deviation of 1.5 corresponds to the 95th percentile, which means that only 5% of the children in a normal population are expected to reach that level.
Parents of 66 children and teachers of 57 children agreed to participate. The "missing" teacher ratings are mostly due to the parents' refusal to allow teacher ratings. Ten children were rated only by parents, 56 by both parents and teachers and one child was rated only by a teacher, which means that a total of 67 children were rated using one or both of the scales.

Measures of autism (Study II)
The prevalence of autism was investigated using the Childhood Autism Rating Scale (CARS) (Schopler et al., 1988), a 15-item behavioural rating scale developed to identify children with autism and to distinguish them from developmentally handicapped children without the autistic syndrome. The items are: relating to people (communication abilities), imitation, emotional response, body use (stereotyped body movements, occupied with some peculiar finger- and hand-positions, self-mutilation), object use (how to use objects and toys), adaptation to change, visual response (for example, avoiding eye contact, staring into space, being fascinated by sparkling or rotating objects or looking at objects from a peculiar angle), auditive response (can appear to be deaf or over-react to auditory stimuli), taste, smell, touch and pain response (occupied with smelling, licking and tasting and often indifferent to pain), fear or nervousness, verbal communication, non-verbal communication, activity level, level and consistency of intellectual response and a general clinical impression of autistic behaviour. Each item was rated on a scale of 1-4 with midpoints, forming a scale with 7 classes (1, 1.5, 2, 2.5 and so on). The CARS scores may range from 15 to 60 and children with scores of 30 to 36 are considered to have mild to moderate autistic symptoms, while a score of over 36 defines severe autism.
When screening for signs of autism, eleven children were found to function well in every respect and were therefore not assessed at the second stage. Two children already had a diagnosis of autism and were not re-assessed. The remaining 53 children were all assessed using CARS. Two of the children with borderline scores of between 20 and 30 were subsequently re-assessed.

Measures of neuropsychological functions (Studies III and IV)
The instrument that was used was a Swedish translation of a neuropsychological test battery for children seven to 14 years of age (NIMES – Neuropsykologiska utredningsmetoder för inlärning, minne och exekutiva funktioner hos barn i skolåldern) (Croona & Kihlgren, 2000), the "Neuropsychological assessment of the school-aged child" (Anderson et al., 1997), comprising ten tests of different neuropsychological functions in the domains of auditive-verbal and visuo-spatial learning and memory and executive abilities (Table 2).
Registration skills were assessed with the Corsi block test (Milner, 1971), where the child was asked to tap up to nine blocks spatially distributed on a board in sequences of increasing lengths, and the auditive-verbal Digit span test (Wechsler, 1992), where the task was to repeat sequences of digits forward of increasing length.

FUNCTIONS	AUDITIVE-VERBAL	VISUO-SPATIAL
REGISTRATION SKILLS	Digit Span	Block Span
SHORT-TERM MEMORY	Story Recall	Complex Figure of Rey
		Recall
LEARNING	Rey Auditory-Verbal Learning test	Spatial Learning Test
LONG-TERM MEMORY	Story Recall	Complex Figure of Rey
	Delayed recall	*Delayed recall*
	Rey Auditory-Verbal Learning Test	Spatial Learning
	Delayed recall	*Delayed recall*
EXECUTIVE FUNCTIONS	Verbal Fluency Test	Tower of London
		Trail Making Test
		Complex Figure of Rey
		Organisation

Table 2. Tests in NIMES measuring auditive-verbal and visuo-spatial registration skills, memory and the executive functions of problem-solving and planning and organisation.

Verbal learning and memory was assessed with Rey Auditory-Verbal Learning Test (RAVLT) (Rey, 1964), where the child has five trials to listen to and learn as many words as possible and immediately recall this list after interference , and Story Recall (Christensen, 1979; Anderson & Lajoie, 1996), where the child listens to two short stories and then tries to immediately recall them correctly. Spatial learning and memory was assessed with the Spatial Learning Test (Lhermine & Signoret, 1972; Anderson & Lajoie, 1996). The children were asked to memorise and recall the positions of nine pictures on a wooden board. Another spatial learning and memory task was to copy a complex figure (the Complex Figure of Rey and Oesterreich, ROCF) (Rey, 1941) and then to draw it spontaneously without looking at the original. This is also a measure of visuo-constructional ability.

Long-term memory for auditive-verbal and visuo-spatial material was measured by asking the children to recall the word list (RAVLT) and the stories (Story Recall) after 30 minutes and to place the pictures (Spatial Learning Test) and draw the Complex Figure of Rey (ROCF) as correctly as possible.

Four tests assessed the visual executive functions of planning and problem-solving; the Trail making Test A and B (Spreen & Strauss, 1991), where the children were asked to draw lines between numbers vs. numbers and letters as quickly as possible (a measure of speed and flexibility), an evaluation of the child's organisational ability when drawing the Complex Figure of Rey, and the problem-solving task of the Tower of London Test (Shallice, 1982). In the Tower of London Test, the children were asked to change the position of three wooden balls on sticks, similar to models presented on 12 cards, in a prescribed number of moves. The solution time was also considered. Aspects of language executive function were measured with the Verbal Fluency Test (Gaddes & Crocket, 1975), where the children were asked to find out

as many words as possible in one minute, beginning with F, A and S, respectively.
Two tests of recognition (of spatial and verbal material) were also administered. Recognition abilities are not thought to reflect short-term memory, long-term memory or executive functions. The recall of visuo-spatial and auditive-verbal material by seeing or hearing a clue does not require strategic abilities to organise material to be stored in or retrieved from long-term memory.
Assessments of cognition, behaviour and autistic symptoms were performed by psychologists at six child development centres/habilitative services constituting the study area.

Statistics
Statistical analyses were performed using STATISTICA 7.0 for Windows (Statsoft Inc.).
Continuous and normally distributed variables were assessed with ANOVA and with post hoc analysis using Tukey HSD. Kruskal-Wallis with post hoc analysis using Mann-Whitney was used for variables that were continuous and not normally distributed. Categorical data were analysed using Chi-square and Spearman correlations.
In all analyses, results were judged statistically significant if the type I error was less than 5% (i.e. $p < 0.05$).

Ethics
The studies were approved by the Ethics Committee at the Medical Faculty at the University of Göteborg. Informed consent was given by all the children and their parents.

Results

Study I:
Intelligence

One-third (24 of 73) of the children were normally gifted with an IQ over 85, while another 22 (30%) had a low-average IQ in the range of 70-84. More severe learning disabilities were found in 27 children (37%), who had an IQ of less than 70. No children were found in the IQ interval 20-34. The median IQ for the whole group was 75.

The IQ scores for the 73 children with hydrocephalus were distributed in a manner that was roughly similar to the expected normal curve, but displaced about 20 IQ scores below normal, and with a sub-group at the lower end of the curve, indicating a sub-population with more severe learning disabilities (Fig. 3).

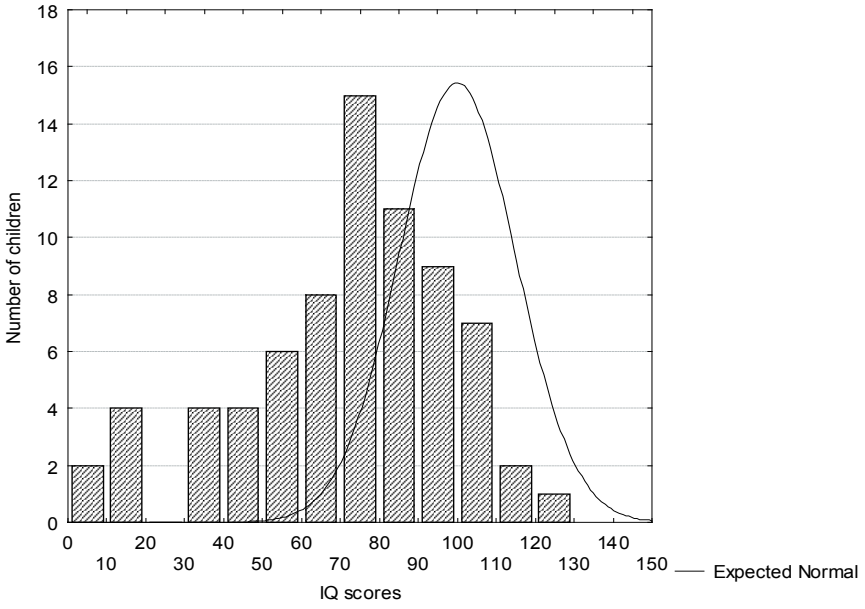


Figure 3. IQ in 73 children with hydrocephalus compared with an expected normal group.

Profiles of verbal and performance IQ

Of the 73 children, 58 were able to complete the WPPSI-R or the WISC-III and thus made it possible to compare the results for verbal IQ (VIQ) with those for performance IQ (PIQ). There was a significant difference ($p < 0.001$) between verbal (median 90) and performance (median 76) IQ. The difference was also significant in the group of 24 children with normal intelligence. Forty-four (76%) of the 58

children had higher VIQ than PIQ scores, 26 (59%) of them significantly higher, i.e. 15 or more IQ scores.

Results on WISC-III subtests
When looking at the results on the 11 subtests for the 40 children tested with WISC-III, the mean scores were below average on Picture arrangement, Object assembly and Coding, which are all performance subtests. On the verbal sub-tests of Information, Similarities and Vocabulary, the mean scores were about average (Table 3). In a normal population, the mean score is 10 and scores of 7-13 comprise the average area, which means that scores of < 7 represent a below-average result.

Scale	Mean score
Information	11.45
Similarities	9.33
Arithmetic	8.05
Vocabulary	10.08
Comprehension	8.63
Digit span	7.15
Picture completion	8.05
Picture arrangement	6.80
Block design	7.40
Object assembly	6.48
Coding	5.56

Table 3. Mean scores for 40 children with hydrocephalus on WISC-III sub-tests, where, in a normal population, M=10 and 7-13 comprise the average area.

IQ in children with and without associated MMC
There were more children (42%) with an IQ of < 70 in the group without MMC than in the group with MMC (29%). Their median IQ was, however, similar, 76 in the group without MMC and 75 in the group with MMC. This is explained by the greater variability in IQ levels of children in the non-MMC group, while the children with MMC were clustered around the IQ range of 70-85. For both the 32 children without MMC and the 26 with MMC, the verbal IQ was significantly higher than the performance IQ; $p < 0.001$ and $p < 0.01$, respectively.

Intelligence in relation to gestational age
The 52 children (23 with MMC and 29 without) born at term had a slightly higher IQ of 76 than the 21 (five with MMC and 16 without) preterm children, who had a medium IQ of 68. The 16 preterm children had a medium IQ of 61 and the nine children in this group with post-haemorrhagic hydrocephalus had the lowest IQ of 57.

Hydrocephalus present at birth
Twenty-eight children (38%), 10 with MMC and 18 without MMC, had large

heads already at birth, while 45 children (18 with MMC and 27 without) developed hydrocephalus during the first year of life. The 18 children without MMC who were born with hydrocephalus had a median IQ of 60, while the 27 children without MMC, whose hydrocephalus developed during the first year of life, had a median IQ of 84, $p < 0.05$ (Table 4).

	Hydrocephalus at birth		Hydrocephalus first year of life	
	n	median IQ	n	median IQ
All children	28	IQ 71	45	IQ 77
MMC	10	IQ 77	18	IQ 73
IH	18	IQ 60	27	IQ 84

Table 4. Median IQ in 28 children born with hydrocephalus compared with 45 who developed hydrocephalus during their first year of life; results for the whole group and for MMC and IH respectively.

Children with cerebral palsy and/or epilepsy

Almost one-third (22) of the 73 children had cerebral palsy and/or epilepsy added to their hydrocephalus. The median IQ of these children was 66, which was significantly lower than the IQ of 78 in those without CP or epilepsy ($p < 0.01$). The nine children with a combination of both cerebral palsy and epilepsy had the lowest IQ of 58.

Study II:

Behavioural problems

The parents of 44 of the 66 children (67%) rated them as having some kind of behavioural problem much above average in one or more of the sub-scales, i. e. a T-score of > 65. Forty of the 44 children, or 61% of all the children, were rated as very much above average, i.e. a T-score of > 70. Eighteen of the 44 children had a T-score of > 65 in one sub-scale, ten children in two and 16 in three or more sub-scales. Almost half the 66 children were reported by their parents to have learning problems and almost one-third to have psychosomatic symptoms. One-third was regarded as hyperactive, according to the hyperactivity index (Table 5).

Behaviour scales	T-score 66-70 n (%)	T-score > 70 n (%)	Total > 65 n (%)
Conduct problem	4 (6)	7 (11)	11 (17)
Learning problem	3 (5)	28 (42)	31 (47)
Psychosomatic	2 (3)	17 (27)	19 (29)
Impulsive-hyperactive	5 (8)	6 (9)	11 (17)
Anxiety	1 (2)	7 (11)	8 (12)
Hyperactivity index	5 (8)	16 (24)	21 (32)

Table 5. Percentages of parent-rated behavioural problems in 66 children with hydrocephalus, much above (T-score 66-70) and very much above (T-score >70) average.

The teachers of 57 children found that 22 (39%) had behavioural problems much above average in some of the four teachers' scales and 15 (26%) had scores very much above average (T-scores > 70). Twelve children had T-scores of > 65 in one sub-scale, one child in two and nine in three or more sub-scales. According to the teachers, attention problems were the most common (30%), especially in the form of inattention-passivity.

Behavioural problems in relation to learning disabilities, cerebral palsy and epilepsy
According to parents' ratings, the 23 children with an average IQ (85-115) had fewer behavioural problems than the 21 with an IQ below average (IQ 70-85) and the 22 with learning disabilities (IQ <70) (Table 5). Twenty of the 22 children with learning disabilities had some kind of behavioural problem, compared with 24 of the 44 (55%) children with an IQ of > 69, p = 0.01 (Table 6).

IQ	n	No behavioural problems n (%)	Conduct problems n (%)	Learning problems n (%)	Psycho-somatic n (%)	Impulsive-hyper-active n (%)	Anxiety n (%)	Hyper-activity index n (%)
< 70	22	2 (9)	6 (27)	17 (77)	7 (32)	6 (27)	4 (18)	13 (59)
70-85	21	5 (24)	2 (10)	11 (52)	6 (29)	2 (10)	1 (5)	4 (19)
> 85	23	15 (65)	3 (13)	3 (13)	6 (26)	3 (13)	2 (9)	4 (17)

Table 6. Behavioural problems much above or very much above average, related to cognitive function in 66 children according to the Conners' Parent Rating Scales.

In addition, the teachers reported a significant correlation between cognitive function and behavioural problems. Seven of 37 (19%) children with an IQ of > 69 had problems of this kind compared with 15 of 20 (75%) with learning disabilities (p = 0.001).
All the 18 children with CP and/or epilepsy added to their hydrocephalus were rated by parents and 16 (89%) of them were found to have problems in one or more behavioural areas, which was significantly more common than 28 of 48 (58%) without CP or epilepsy, p = 0.02. On teachers' scales, there were no significant differences between the groups; seven (44%) of the 16 rated children with CP and/or epilepsy had behavioural problems, as did 15 (37%) of 41 without these additional impairments.

Behavioural problems in children with infantile hydrocephalus and in children with MMC
There was a slight, yet not significant, difference between children with infantile hydrocephalus and those with MMC, when it came to the number of behavioural problems according to parents. Fifteen of 40 (38%) children with infantile hydrocephalus were rated as hyperactive compared with six of 26 (23%) with MMC. The percentage of children regarded as having learning problems was about the same in both groups.
Of the 57 children rated by teachers, five of 36 (14%) with infantile hydrocephalus were reported to have conduct problems compared with only one of 21 children with MMC. Likewise, the hyperactivity index indicated hyperactivity in nine of 36

(25%) children with infantile hydrocephalus compared with three of 21 (14%) with MMC. The differences did not reach significance. Children with MMC were somewhat more frequently considered to be inactive/passive than those with infantile hydrocephalus, seven of 21 (33%) compared with nine of 36 (25%) respectively.

Behavioural problems in relation to gestational age at birth
Seventeen (25%) of the 67 children were born before 37 weeks of gestation. All 17 were rated by parents and no significant differences were found between the children born at full term and those born preterm, although the latter were reported somewhat more frequently to have anxiety problems. Nor did the two groups differ on teachers' ratings (45 of the 57 children were born at term and 12 were born preterm).

Comparison between parents' and teachers' ratings
The results from parents' and teachers' scales that were possible to compare, i.e the hyperactivity index and conduct problems, revealed that there was significant agreement in terms of the hyperactivity index ($r = 0.41$; $p < 0.05$). Parents rated 21 of 66 (32%) children as hyperactive using the hyperactivity index, while teachers rated 12 of 57 (21%) as hyperactive. Conduct problems were reported by parents in eleven children (17%), compared with six (11%) by the teachers.

Autism
Of the 67 children, two already had a diagnosis of autism before the study and, using the CARS assessment, five more obtained scores above the cut-off score of 30 for an autism diagnosis. Two children with scores of between 20 and 30 were subsequently re-assessed and were then found to fulfil the criteria for autism. There were therefore nine of 67 (13%) children with autistic symptoms.
Six (27%) of the 22 children with learning disabilities (IQ < 70) had autistic symptoms, which was significantly more than three of the 45 (7%) without learning disabilities, $p = 0.03$. These symptoms were also significantly more frequent in children with additional impairments in the form of CP and/or epilepsy; six of 18 (33%) compared with three of the 49 (6%) without these impairments, $p < 0.01$.
Of the 26 children with MMC, only one had autistic symptoms compared with eight of 41 (20%) children with infantile hydrocephalus.
Autistic symptoms were as common in children born at term (seven of 50) as in those born preterm (two of 17).

Study III
The two clinical groups, children with infantile hydrocephalus and children with MMC with hydrocephalus, obtained significantly lower results than controls on the four neuropsychological functions of short-term memory ($p<0.001$), long-term memory ($p<0.001$), learning ($p<0.001$) and executive functions ($p<0.001$). When it came to the functions of registration skills, there were no significant differences either between the children with hydrocephalus with or without MMC, or between the clinical groups and controls and all three groups slightly underachieved accord-

ing to norms (Table 7).
In the function of short-term memory, children with hydrocephalus both with and without MMC performed about 1 SD below the test standard mean compared with normal results in the control group.
In long-term memory sub-tests, the two groups with hydrocephalus performed 1-2 SD below the mean for standard norms, with the greatest difficulties noted on Story delayed recall and delayed recall of the Complex Figure of Rey. Controls performed according to norms.
The two sub-tests of learning, both the verbal and the spatial, produced results about 1 SD below the mean for the children with hydrocephalus. Controls performed according to norms on the verbal subtest and 1 SD above the mean on the spatial subtest.
All five sub-tests of executive functions comprised difficulties for the children with hydrocephalus, where those both with and without MMC performed equally, about 1 SD below the standard mean, while controls achieved results 1 SD over the mean

Functions and subtests	IH			MMC			Controls			
	n	Mean	SD	n	Mean	SD	n	Mean	SD	p
Registration skills	20	42.45	9.73	16	42.78	8.36	36	45.69	6.43	ns
Corsi block	20	44.10	11.31	16	45.06	9.74	36	47.94	10.42	ns
Digit span	20	40.80	10.48	16	40.50	10.10	36	43.44	8.15	ns
Short-term memory	19	36.53	11.12	14	33.25	9.68	36	52.19	9.22	***
ROCF recall	19	36.79	15.07	14	33.43	9.78	36	53.89	14.43	***
Story recall	20	36.20	13.39	16	34.50	13.28	36	50.50	8.04	***
Learning	20	40.65	16.18	16	40.78	9.21	36	54.82	7.32	***
Spatial learning	20	42.15	21.79	16	44.44	12.80	36	60.31	7.82	***
RAVLT 1-5	20	39.15	14.84	16	37.13	13.72	36	49.33	12.23	*
Long-term memory	20	37.54	12.76	13	35.31	8.19	36	51.17	8.01	***
Spatial delayed recall	20	39.60	16.29	16	40.63	13.06	36	49.61	9.16	**
ROCF delayed recall	20	36.50	14.38	13	32.77	12.04	36	53.14	15.13	***
Story delayed recall	20	32.60	13.71	16	31.06	9.04	36	47.17	7.59	***
RAVLT delayed recall	20	41.45	17.56	16	36.38	12.96	36	54.75	11.62	**
Executive functions	19	41.68	8.98	14	41.39	8.80	36	54.92	4.41	***
Trail Making Test A	20	39.05	14.28	16	38.63	10.98	36	53.14	6.71	***
Trail Making Test B	20	39.30	12.04	16	40.94	13.97	36	52.00	6.40	***
ROCF organisation	20	47.80	16.41	14	41.36	13.08	36	60.44	13.31	***
Tower of London	19	38.42	10.87	16	40.06	14.38	36	58.56	5.86	***
Verbal fluency	20	42.80	12.59	16	45.19	9.56	36	50.47	11.97	***

*p<0.05, **p<0.01, ***p<0.001

Table 7. Means and standard deviations (SD) for five neuropsychological functions and the constituting sub-tests in NIMES for 36 children with hydrocephalus, 20 with infantile hydrocephalus (IH) and 16 with myelomeningocele (MMC), plus 36 controls.

on the visuo-spatial planning tasks of Tower of London and Complex Figure of Rey and according to norms on the other sub-tests.
When comparing the results in the two domains of auditive-verbal and visuo-spatial abilities, there were no differences between children with hydrocephalus with or without MMC; both groups obtained significantly poorer results ($p<0.001$) than controls in both domains.
The sub-tests of Recognition were not included in the function of long-term memory. There were no differences between the clinical groups and controls in terms of auditive-verbal recognition, where the three groups performed according to norms. However, in the spatial recognition task, the children with hydrocephalus performed significantly below controls ($p<0.01$).

Study IV
Of the eight children with MMC without hydrocephalus, two children had an IQ of < 70, i. e. mild learning disabilities. When comparing this group with the children with MMC and hydrocephalus, the performances of the children with hydrocephalus were significantly poorer on the functions of registration skills ($p<0.05$), long-term memory ($p<0.05$) and executive functions ($p<0.01$). The children with only MMC obtained fairly normal results according to standard test norms, while the children with MMC and hydrocephalus obtained results 1-2 SD below. Children with MMC and hydrocephalus performed less well in the visuo-spatial domain ($p=0.05$) but not in the auditive-verbal domain.
When the two children with an IQ of < 70 were excluded from the group with only MMC, this group significantly exceeded those with MMC and hydrocephalus in all functions and on the majority of the sub-tests, as well as in the auditive-verbal and visuo-spatial domains (Table 8). Only on the three tests of spatial learning, Tower of London and verbal fluency did the groups not differ significantly. The median T-scores did not differ very much, at most 1 SD (on two sub-tests of long-term memory), but the ranges were generally much narrower in the MMC group.

When compared with controls, there were also different results when including or excluding the children with learning disabilities. When included, the group with

Domains	MMC n=8		MMC+HC n=8		Controls n=8		
	Median	Range	Median	Range	Median	Range	p
Auditive-verbal	50.50	30-55	33.75	30-46	51.33	40-62	**
Visuo-spatial	51.11	31-55	37.39	26-43	56.67	47-59	***

**p≤0.01, ***p<0.001

Table 8. Medians and ranges in the auditive-verbal and visuo-spatial domains of neuropsychological functions in six children with myelomeningocele (MMC) without hydrocephalus (HC) and an IQ of ≥ 70 and eight children with MMC and HC with an IQ of ≥ 70.

only MMC differed from controls in two functions; learning ($p<0.05$) and executive abilities ($p<0.05$), and in the visuo-spatial domain ($p<0.05$), but, with the two children excluded, there were no statistically significant differences between the groups in any function or domain.

Children with MMC and hydrocephalus differed significantly from controls on the five functions and in both domains, where short-term memory ($p<0.001$), learning ($p<0.001$) and executive functions ($p<0.001$) constituted the greatest difficulties for the children with MMC, who obtained results 1.5-2 SD below standard test norms.

When the two clinical groups were compared with the controls, it was evident that children with MMC combined with hydrocephalus were those that primarily contributed to the group differences. The children with MMC in combination with hydrocephalus differed significantly in all five tested functions compared with controls (Registration skills; $p=0.01$, Short-term memory; $p<0.001$, Learning; $p<0.001$, Long-term memory; $p<0.05$, Executive functions; ($p<0.001$)). Children with only MMC differed significantly from controls, but to a lesser extent than those with hydrocephalus, on learning ($p=0.05$) and executive functions ($p<0.05$) respectively, but not regarding registration skills or short- or long-term memory. After excluding the two children with an IQ of < 70, their performance was equal to that of controls (Figure 4).

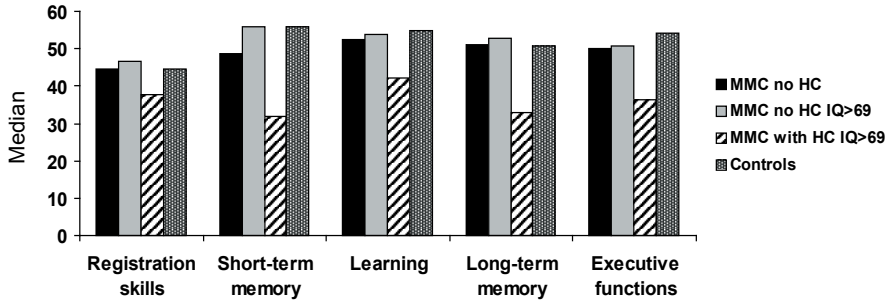


Figure 4. Medians of five neuropsychological functions for eight children with myelomeningocele (MMC) without hydrocephalus (HC), six children with MMC without HC and an IQ of ≥ 70, eight children with myelomeningocele and hydrocephalus and eight controls.

Discussion

The general aim of this study was to explore and describe a population-based group of children treated for hydrocephalus during their first year of life, in terms of intellectual, neuropsychological and behavioural functions and the presence of autistic symptoms. It was also of interest to analyse differences between sub-groups with or without MMC and to see whether preterm birth, epilepsy or cerebral palsy influenced cognitive or behavioural outcome.
These findings should also make it possible to further discuss and better understand the role of hydrocephalus per se in creating the characteristic assets and deficits of these children.

Methodological considerations
Since the study included all the children aged five to ten in the region and, as the children lost to follow-up did not differ in characteristics from the ones being studied, the results could be regarded as representative and thus relevant to clinical practice. The last study, which focused on cognitive functions in children with MMC without hydrocephalus, recruited children born later than the children in Studies I-III, in order to obtain the appropriate ages.
The methods for measuring intelligence, the Wechsler and Griffiths scales, are well known and have also been frequently used in studies of children with hydrocephalus. The WISC-III has a documented inter-scorer agreement (Wechsler, 1992), which was important, as many psychologists were involved in the test procedure. The Conners' Behaviour Rating Scales, especially the Hyperactivity Index, are well established. However, one weakness was the fact that the parents' and teachers' scales were not fully compatible. The instrument used for assessing autism, CARS, would not be the instrument of choice today. However, at the start of the study, this was the most frequently used instrument for this purpose in the region and one with which the experienced participating psychologists were familiar. The fact that the participating psychologists were experienced and that the children and parents did not have to travel far or to meet too many new persons should also outweigh the risk of weak test reliability.
The NIMES neuropsychological test battery consists of well-known, often used tests, gathered and validated both in Australia and Sweden.
It would have been valuable if we had also gathered information about the lateralisation of linguistic superiority and the functional integrity of the cerebral hemispheres, by using a dichotic listening test. Additionally, it would have been interesting to supplement the study with a measure of quality of life and psychological health.

Intelligence
This study revealed that one-third of the children with hydrocephalus were normally gifted, slightly less than one-third had a low-average intelligence and slightly more than one-third were learning disabled. When comparing the studied group of children with a normal population, the distribution of intelligence was similar but with

a displacement of 20-30 IQ scores toward lower levels and with a group of children with very low scores.
The characteristic better verbal than performance IQ was noted even before it became common to treat hydrocephalus surgically (Hagberg & Sjögren, 1966; Tew et al. 1979) and it has frequently been described over the years (Dennis et al., 1981; Donders et al., 1991; Riva et al., 1994; Brookshire et al., 1995; Lumenta et al., 1995). In this study, this difference was present in children with and without learning disabilities, with and without MMC and regardless of whether they were born with hydrocephalus or developed it during their first year of life. A significant difference (in any direction) between verbal and performance IQ is found in 24% of a normal population of children, according to the WISC-III manual. The more than double prevalence of significantly better VIQ than PIQ in this population-based study convincingly suggests that this is characteristic of children with hydrocephalus.

Neuropsychological functions
The 36 children with hydrocephalus and an IQ of ≥ 70 performed significantly less well compared with controls in both the auditive-verbal and visuo-spatial domains, as well as on the functions of short- and long-term memory, learning and executive abilities. However, their registration skills and ability for verbal recognition were average and the same as those of controls.

Behaviour
The Isle of Wight study of behavioural problems in children in general (Rutter et al., 1976) revealed that about seven per cent of the children had what was then labelled psychiatric disorders, with emotional and conduct disorders as the most common types. Furthermore, three to five per cent of all school-age children have been reported to have clinically significant attention deficit/hyperactivity disorder, as defined in DSM-IV.
When studying the prevalence of behavioural problems in this population of children with hydrocephalus, the parents of as many as two-thirds of the children considered that their children had problems in at least one area on the Conners' scales. The corresponding rate for teachers was about 40%. In both parents' and teachers' ratings, the majority of the children obtained scores *very* much above average.
Parents also reported that one-third of the children had psychosomatic symptoms, but, as the questions relating to psychosomatic symptoms include headache, stomach-ache and nausea, they may in this population also reflect somatic symptoms related to the ventriculo-peritoneal shunt and not necessarily psychological symptoms.

Autism
The prevalence of autistic symptoms in this study was 13%, much higher than the 30 to 60 cases per 10,000 (0.03-0.06%) in the general population (Chakrabarti et al., 2001, Rutter, 2005).

Children with infantile hydrocephalus compared with children with MMC

The children with MMC displayed a fairly homogeneous pattern in full-scale IQ, with intelligence clustering around the 70-85 IQ interval, whereas children without MMC appeared to be more heterogeneous, with both very low and fairly high IQ scores. The median IQ was, however, similar in both groups. The better verbal IQ compared with performance IQ was significant for children both with and without MMC, as has been shown earlier (Dennis et al., 1981; Riva et al., 1992; Hoppe-Hirsch et al., 1998).

When assessing neuropsychological functions in children with an IQ ≥ 70, there were no differences in any function or domain between children with or without MMC. Children with or without MMC have traditionally most often been studied separately and each group has revealed impaired memory and executive functions (Dalen et al., 2005; Vachha & Adams 2005; Yeates & Enrile, 2005). However, performing the same tests with almost identical results, it can be assumed that the difference in the aetiology of the hydrocephalus was not that important for the children's outcome.

As the inclusion criterion was an IQ of 70 or more, there was a range in IQ of between 70 and 112 and, to estimate the impact of IQ level on their results, the children were divided into two groups, one with average intelligence and one with low-average intelligence. The groups did not differ in terms of learning and long-term memory but differed significantly in terms of short-term memory and executive functions.

When comparing children with infantile hydrocephalus and those with hydrocephalus associated with MMC, the former were found to have behavioural problems and hyperactivity in particular, slightly more often. Hyperactivity was also reported more frequently by parents than teachers, which could be explained by the fact that activities at school are structured and help the children to concentrate more effectively. From the teachers' ratings, a pattern emerged in which children with infantile hydrocephalus were hyperactive and those with MMC were more inactive-passive. This was also reported in a recent study of temperament patterns in children with MMC, where this group differed significantly from a standardised population by being less attentive (Vachha & Adams 2005).

In this study, only one child with MMC had symptoms of autism. When this child was excluded, eight of 41 (20%) of the children with infantile hydrocephalus had autistic symptoms, a similar percentage to the 23% reported in an earlier study of children with hydrocephalus without MMC (Fernell et al., 1991).

Learning disabilities related to behaviour

There was a significant difference in parents' ratings between children with learning disabilities and those with normal or nearly normal cognitive function. About 90% of the children with learning disabilities (IQ < 70) had some behavioural problems compared with about half of those with an IQ over 69 and 35% of those with an IQ of > 85. Furthermore, the teachers reported that children with hydrocephalus and learning disabilities manifested problems significantly more often than the normally gifted children. Of those with an IQ below 70, as many as 75% were considered

to have behavioural problems compared with about one in five of those who were normally or nearly normally gifted. Hydrocephalus in association with learning disabilities therefore appeared strongly to increase the risk of behavioural problems, which has also been shown previously (Fernell et al., 1991).

Gestational age and debut of hydrocephalus related to intelligence and behaviour
In this study, children born preterm, especially those with post-haemorrhagic hydrocephalus, had a lower IQ than those born at term, which was in accordance with earlier reports that these children fared less well than children with congenital hydrocephalus (Brookshire et al., 1995; Fletcher at al., 1997; Heinsbergen et al., 2002).
Somewhat unexpectedly, children born preterm did not have behavioural problems more frequently than those born at term. This is in contrast to a large Swedish follow-up study of children born extremely preterm (Stjernqvist & Svenningsen, 1999), in which it was found that these children had behavioural problems significantly more often (32%) than those born at term (10%). It may be that the factor of hydrocephalus with its underlying brain lesions and learning disabilities in half of the children born preterm in this study outweighed the smaller contribution that just being born preterm makes to behavioural problems.
In children with MMC, there was no difference in median IQ (77 and 73 respectively) between those who were born with hydrocephalus and those who developed hydrocephalus at a later stage. In the IH group, the 27 who developed hydrocephalus during the first year of life had an IQ in the low-average area, while the 18 (40%) children born with hydrocephalus had a median IQ of < 70. This is in agreement with an earlier population-based study of hydrocephalus overt at birth, where 45% of the children had learning disabilities (Fernell et al., 1987).

Additional impairments of cerebral palsy and epilepsy related to intelligence, behaviour and autism
Significantly lower IQ scores were found in the one-third of the children who had cerebral palsy and epilepsy added to their hydrocephalus.
Since only five children in the study of neuropsychological functions had cerebral palsy and/or epilepsy, no comparisons were made between children with and without these impairments to neuropsychological abilities.
Additional impairments significantly increased the risk of behavioural problems, as almost all children with cerebral palsy and /or epilepsy had problems in one or more behavioural areas, according to their parents. In this study, as in earlier reports (Chakrabarti et al., 2001; Fernell et al., 1991; Fombonne, 1999; Kielinen et al., 2004), autistic symptoms were also much more common among children with more severe brain dysfunction. These symptoms were thus found to be five times more common in children with signs of a more widespread brain lesion resulting in epilepsy or cerebral palsy (33%) compared with six per cent of children without these impairments.

The impact of hydrocephalus on cognition and neuropsychological functions

In current research on hydrocephalus, the group of children with hydrocephalus in combination with MMC are the most studied, for natural reasons. It is homogeneous according to aetiology and outcome and it represents a fairly large group of congenital childhood impairments. However, in this study, an attempt was made to explore how hydrocephalus contributes to the cognitive and behavioural problems, by studying these aspects in all children with hydrocephalus and also by studying intelligence and neuropsychological functions in children with MMC without hydrocephalus.

The children with hydrocephalus and MMC constituted a homogeneous group on intelligence measures, while the children with isolated hydrocephalus represented a wide range on the IQ scale. The early neural tube defect and malformation of the brain in children with MMC can certainly result in outcomes of varying severity, but, compared with the widespread aetiology (malformations, infections, haemorrhages) in isolated hydrocephalus, the latter group comprise a much more heterogeneous group.

Children with hydrocephalus both with and without MMC obtained significantly better results in verbal IQ than performance IQ. These non-verbal learning problems have been described as being caused by the loss of cerebral white matter (Fletcher, 1992), which can in turn be a consequence of early maldevelopment, as well as high pressure caused by hydrocephalus that destroys both white-matter tissue and cortical neurons (Del Bigio, 1993). In this study, the mean sub-scale scores of the 40 children tested with WISC-III therefore produced results under average on visuo-perceptual performance tasks. As the loss of periventricular white matter generally also causes visual perception problems in children, as children born preterm with periventricular leukomalacia (PVL) (Jacobson et al., 1996), children with both preterm birth and hydrocephalus can be expected to run a very high risk of developing non-verbal cognitive impairment.

Six of the eight children with MMC without hydrocephalus displayed no difference between verbal and performance IQ, while the two children with learning disabilities had a significantly poorer performance IQ compared with verbal IQ. Although the group is small, it can be presumed that the two children with learning disabilities had a more severe early malformation causing their poorer outcome in overall terms and that the other six children did not display the characteristic cognitive profile because of the absence of hydrocephalus.

When examining neuropsychological functions in children with hydrocephalus and referring to IQ profiles, these children could be expected to obtain better results in auditive-verbal than visuo-spatial domains, which was not the case. It has been shown that the relatively strong verbal abilities as measured by intelligence tests such as the WISC-III and WPPSI-R are of limited value for predicting good language function. These tests include single word knowledge, which is mostly very good in children with hydrocephalus but does not measure abilities in pragmatics or discourse where these children have great difficulty (Dennis & Barnes 1993; Barnes & Dennis 1998; Barnes et al. 2001; Vachha & Adams 2003). The Wechsler scales are

therefore not sufficient when examining language function in children with hydrocephalus.

When it came to both short-term and long-term memory, the children with hydrocephalus had great difficulty in both the auditive-verbal and visuo-spatial domains, but, on immediate memory tasks (registration skills of digit span and Corsi block), their results were equal to those of controls. The results confirm that implicit memory appears to be relatively unaffected in children with early brain disorders. The immediate recall of numbers and imitating pointing at blocks do not require conceptual analysis but are perceptually driven (implicit memory). Memorising and recalling a story, even if it is very short, on the other hand, requires this kind of analysis (explicit memory). In a study that compared children with MMC, traumatic brain injuries and orthopaedic injuries, Yeates et al. (2005) found that those with MMC did not differ from the other groups in terms of implicit memory but obtained significantly poorer results for explicit memory when compared with non-brain-injured children. This finding may be explained by the hypothesis that implicit memory is mediated by less specialised brain systems in children than in adults and is therefore less vulnerable (Yeates & Enrile, 2005). Some earlier studies of children with hydrocephalus did not reveal problems with memory (Cull & Wyke 1984; Donders et al., 1991), but subsequent research has changed this picture.

Significant verbal and non-verbal memory deficits have been reported in children with both MMC and infantile hydrocephalus (Vachha & Adams 2005; Ralph et al., 2000; Scott et al., 1998; Yeates & Enrile, 2005). This study revealed that children with hydrocephalus had significant difficulty recalling short stories, both immediately and after 30 minutes. This explicit memory task may partly be a consequence of discourse problems, but it also contains a working memory component. In a study of meaning construction and comprehension, Barnes et al. (Barnes et al., 2004) found that children with hydrocephalus failed in the comprehension of meaning due to deficits in both long-term memory and working memory. They had problems remembering and integrating information from one sentence to another, although they had no difficulty understanding single words and sentences. Deficits in working memory, as well as in information-processing speed, in children with MMC were also found in a recent study (Boyer et al., 2006).
In this study, visuo-spatial memory tasks also presented great difficulty to all the children with hydrocephalus compared with controls. The visuo-constructional task of copying and remembering the Complex Figure of Rey appeared to be more difficult than the spatial memory task in which the children were asked memorise the position of pictures. The character of memory problems in children with hydrocephalus are currently well documented, but the neural correlates have not been studied (Dennis, 2006). It is argued that changes in the ventricular system associated with hydrocephalus can lead to the compression of temporal lobes, hippocampal structures and other subcortical structures involved in memory (Scott, 1998).
Another aspect of memory is recognition, where the children in this study performed as well as controls on the verbal recognition task. Although they had great problems

retrieving words from a word list, they were able to recognise the words in a text. This phenomenon was explained by Yeates et al. (Yeates et al., 1995) as a lack of strategies for the retrieval of stored information, but that this information could be retrieved with a cue.

Strategic and planning abilities were studied in terms of executive functions, where the children with hydrocephalus had significant difficulty compared with controls. There were no differences between children with or without MMC, which indicates that the visuo-motor tests (Trail Making Test and Tower of London Test) also reflect deficits in planning and speed and not deficits in motor functions in children with MMC. Deficits in visual and visuo-spatial functions can be partly explained by the thinning of the posterior cortex, which is often found as a secondary CNS insult in children with hydrocephalus (Dennis et al., 1981, Fletcher et al., 1996). In spite of the fact that children with hydrocephalus have fluent spontaneous language in everyday life, they had difficulty performing the word-finding task in this study. Fletcher et al. (1992, 1996) have shown that the motor and spatial deficits displayed by children with early hydrocephalus are strongly related to the integrity of the corpus callosum.

Inter-hemispheric communication is also essential for language function. In a study of children with MMC and hydrocephalus, a relationship was found between idiom comprehension deficits and congenital agenesis of the corpus callosum (Huber-Okrainec et al., 2005). In the present study, idiom comprehension was not studied, but the memory problems could be partly explained by the children's difficulty finding meaning and their lack of ability to organise and structure the information, which makes retrieval difficult. Fletcher et al. (2004) found that only four per cent of children with MMC were found to have a normal corpus callosum and 52% were missing one or more corpus callosum structures. In children with MMC, these structural abnormalities may be an expression of the neural tube midline closure defect which may include agenesis of the corpus callosum, but destructive effects of the hydrocephalus by stretching and thinning the corpus callosum must also be considered (Del Bigio, 1993).

Although the studied group of children with MMC without hydrocephalus was small, only eight children, it should be possible to obtain an indication of the effect of hydrocephalus on cognitive development by matching them with both children with MMC in combination with hydrocephalus and controls. The children with only MMC differed significantly from controls on the functions of learning and executive abilities and in the visuo-spatial domain, but, when the two children with an IQ of < 70 were excluded, there were no differences in any neuropsychological function or domain between children with MMC and controls. This suggests that MMC alone is not a major determinant of neuropsychological functions. However, the two children with MMC and an IQ of < 70 illustrate that there is also variability among children with MMC without hydrocephalus in terms of cognitive outcome. Children with only MMC performed significantly better than those with hydrocephalus

on registration skills, long-term memory and executive functions, even when the two children with an IQ of < 70 were included, and, when the groups were compared without these two, the differences were significant for all neuropsychological functions and for both the auditive-verbal and the visuo-spatial domains.

The similar results on neuropsychological functions, when comparing children with MMC without hydrocephalus with controls, and the similar results between children with hydrocephalus with or without MMC indicate that, despite congenital malformations causing brain anomalies in children with MMC, the alterations to brain structures caused by hydrocephalus may be responsible for the majority of cognitive and neuropsychological problems in children with average and low-average intelligence.

Conclusions

- Hydrocephalus may result from diverse aetiologies, may be associated with various additional neuroimpairments and the severity of disability may vary, but the majority of affected children had problems with cognition and behaviour and many had autistic symptoms.
- There was no difference in median IQ between children with and without MMC, but children with infantile hydrocephalus were represented in all IQ areas, while children with MMC were clustered around the IQ range of 70-85. Children with infantile hydrocephalus had slightly more behavioural problems compared with those with MMC. Both groups had problems with the regulation of activity; however, children with MMC tended to be more passive and children with infantile hydrocephalus had more problems with hyperactivity.
- Children born at full term had a slightly but not significantly higher IQ than those born preterm. There were no differences between children born preterm or at full term regarding the frequency of behavioural problems or autistic symptoms.
- The additional impairments of cerebral palsy and epilepsy significantly heightened the risk of developing cognitive and behavioural problems, as well as autistic symptoms.
- Children with hydrocephalus irrespective of aetiology characteristically had better verbal IQ than performance IQ as measured with intelligence tests. However, they had considerable problems with learning, memory and executive functions on both auditive-verbal and visuo-spatial do mains. An examination with standard intelligence tests is therefore not enough in order to understand and describe language functions in these children but should be supplemented with neuropsychological measures.
- When examining children with hydrocephalus and low-average to average intelligence, there was no difference between children with or without MMC when it came to the neuropsychological functions of learning, memory and executive abilities. Children with hydrocephalus had deficits in short-term and long-term memory, as well as in planning and organisation. They had assets in immediate recall and verbal recognition. Given a clue, they had a chance to recall what they had learned.
- Children with MMC without hydrocephalus and without learning disabilities did not display a better verbal IQ than performance IQ, like children with MMC and hydrocephalus. They also appeared to perform normally on tests of learning, memory and executive functions.

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Sammanfattning

Syfte: Syftet med studien var att undersöka begåvning och beteende hos barn med hydrocefalus; födda med eller utan ryggmärgsbråck (MMC), födda i normal tid eller för tidigt, och med eller utan funktionshinder som cerebral pares (CP), epilepsi eller utvecklingsstörning.

Material och metod: Av alla 107 barn som behandlats för hydrocefalus 1989-1993 i västra sjukvårdsområdet, undersöktes 73 med begåvningstest, 67 med skattningsskalor för beteende (Conners) och 54 för autism (CARS). Trettiosex av de 47 barn som hade IQ ≥ 70 samt åtta barn med MMC utan hydrocefalus undersöktes med ett neuropsykologiskt testbatteri (NIMES) och jämfördes med ålders- och könsmatchade kontroller.

Resultat: En tredjedel av barnen hade normal begåvning (IQ >85), ytterligare 30% hade låg begåvning inom normalområdet (IQ 70-84) medan 37% hade en utvecklingsstörning (IQ < 70). Av barnen med MMC hade 29% IQ < 70. Motsvarande andel av barnen med hydrocefalus utan MMC var 42%. För tidigt födda barn hade lägre IQ än fullgångna. Barn med CP och/eller epilepsi hade signifikant lägre IQ än de utan dessa funktionsstörningar.
Baserat på föräldrarnas skattning hade 67% av barnen beteendeproblem jämfört med 39% enligt lärarna.. Utvecklingsstörning ökade risken för beteendeproblem betydligt och nästan alla barn med CP och/eller epilepsi hade beteendeproblem. Autistiska symtom fanns hos 13% av barnen, hos fyra procent av dem med MMC och hos 20% av dem utan MMC. Utvecklingsstörning, CP och epilepsi ökade risken för autism signifikant.
Barn med hydrocefalus, både med och utan MMC och med IQ > 70 presterade signifikant sämre än kontrollerna gällande inlärning, minne och exekutiva funktioner men inte avseende omedelbart minne. Det fanns ingen skillnad mellan normalbegåvade barn med hydrocefalus med och utan MMC, medan normalbegåvade barn med MMC utan hydrocefalus fick resultat jämförbara med kontrollerna på alla neuropsykologiska test.

Slutsats: En majoritet av barnen med hydrocefalus hade en begåvning inom det nedre normalområdet eller utvecklingsstörning samt beteendeproblem. Många hade även autisitiska symtom. Trots genomsnittlig eller låg genomsnittlig begåvning hade barnen stora svårigheter med inlärning, minne och exekutiva funktioner, oberoende av den bakomliggande orsaken till hydrocefalus. Enbart MMC utan samtidig hydrocefalus tycktes inte påverka kognitiva och neuropsykologiska funktioner i samma utsträckning som den hjärnskada som orsakade eller orsakades av hydrocefalus.

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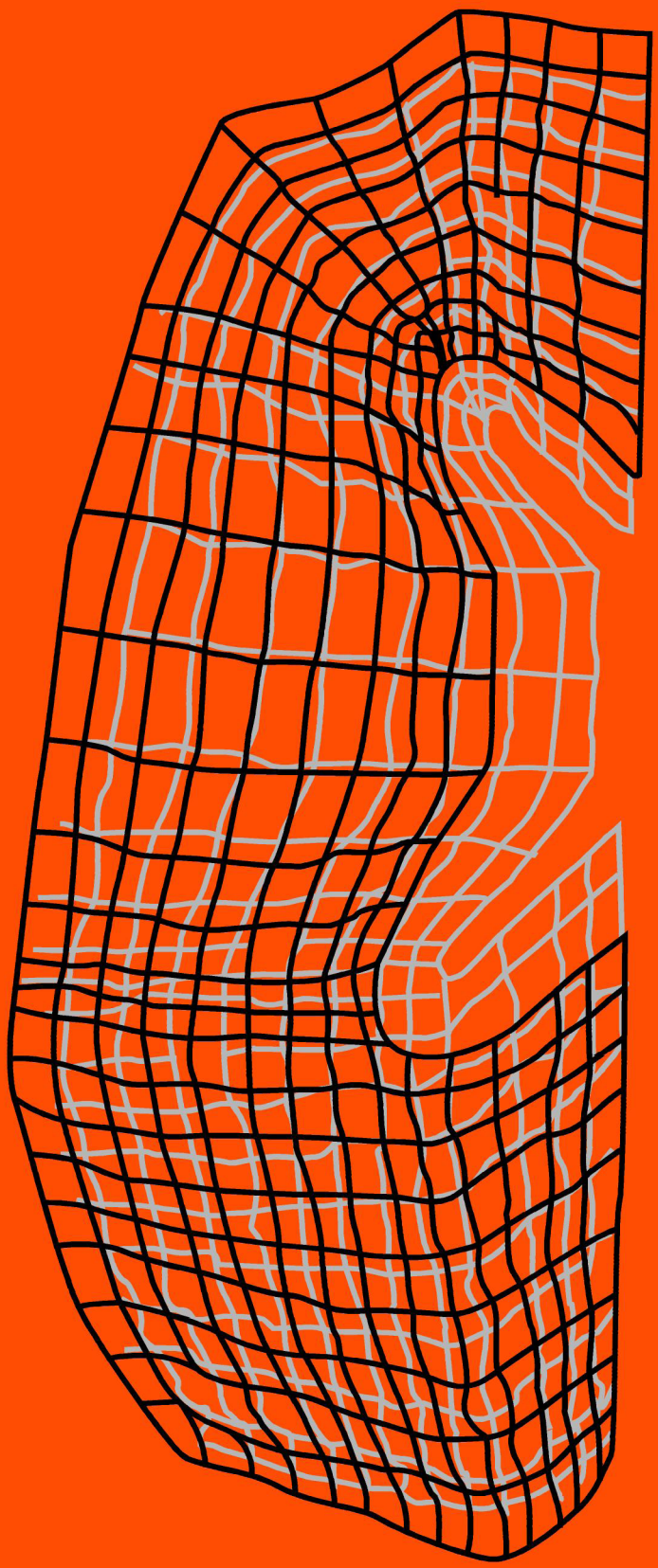

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