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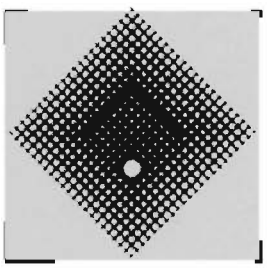

ATTENTION, PROCESSING SPEED AND EXECUTIVE FUNCTIONS IN CHILDREN WITH OBSTRUCTIVE AND COMMUNICATING HYDROCEPHALUS

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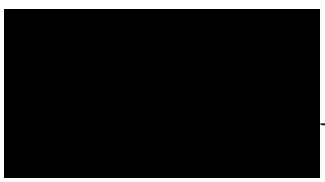
This research thesis was submitted in partial fulfilment of the requirements for the Doctor of Psychology (Clinical Neuropsychology), 2004.

Statement of Authorship

Except where reference is made in the text, this thesis contains no material published elsewhere. All the work is that of the author except where referenced in the text of this thesis.

Signed 

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Ethics Committee Approval

The original study, that involved the data collection phase, received ethics approval from the Royal Children's Hospital Ethics in Human Research Committee on 18th of February 2000 (see Appendix A). The current archival study received ethics approval from the Victoria University Human Research Ethics committee on 3rd of June 2003 (see Appendix B).

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The current study is an archival study. The original study was developed and conducted by staff from the RCH. A research assistant recruited participants and collected the data. The original study included a large test battery mentioned in the procedure section. The present author was involved after the completion of the data collection phase. A portion of the test scoring, some data entry as well as data collation, organisation and analysis, were completed by the author. The author formulated the thesis topic, aims and hypotheses, in conjunction with her supervisors.

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Abstract

Hydrocephalus (HC) is a condition involving an increased volume of cerebral spinal fluid (CSF) in the ventricular system, caused by a variety of prenatal and postnatal aetiologies. In obstructive HC there is an obstruction to the flow of CSF and in communicating HC there is an interference with the reabsorption of the CSF while the ventricular pathways remain functional. The compressive effects of HC may lead to thinning and stretching of the corpus callosum, and also damage projection fibres, optic tracts and olfactory pathways. Consistent with this neuropathology, progressive HC in early childhood may result in neuropsychological impairment such as gross and fine motor deficits, spatial perceptual problems, and slowed processing speed. To date there is a paucity of research that compares the neuropsychological functioning of children with obstructive and communicating HC. This study aimed to compare attention, processing speed and executive functions in children with obstructive and communicating HC. In total 116 children (age range 7 to 16 years) were included. Forty-three participants had obstructive HC (mean age = 10.07 years *SD* = 2.45 years), 31 participants had communicating HC (mean age = 10.65 years *SD* = 2.40 years) and 42 participants were sibling controls (mean age = 10.62 years *SD* = 2.44 years). Subtests from the Test of Everyday Attention for Children were administered to assess selective, sustained and divided attention. The Rey Complex Figure (RCF), Contingency Naming Test, Tower of London, Digits Backwards and the Behavioural Rating Inventory of Executive Function were administered to assess executive functions. When compared to controls, both the obstructive and communicating HC groups exhibited problems with attentional and executive functions. Subtle differences between the two HC groups were also observed, however with the exception of RCF these differences failed to reach significance. Qualitative analyses of MRI scans revealed that the HC groups had a similar incidence, type and degree of white matter and corpus callosum abnormalities, but differed in the location of white matter loss.

CHAPTER 1: ATTENTION, PROCESSING SPEED AND EXECUTIVE FUNCTIONS IN CHILDREN WITH OBSTRUCTIVE AND COMMUNICATING HYDROCEPHALUS

1.1 Overview

The literature consistently reports that children with hydrocephalus (HC) have a variety of cognitive and behavioural problems. It is unclear if children who have different aetiologies or types of HC have a distinctly different cognitive profile. The current study focuses on two types of HC, obstructive and communicating, and investigates if these sub-groups can be differentiated in terms of attention, processing speed and/or executive functions.

1.2 Aetiology and Neuroanatomy

HC literally means "water brain" and refers to an increase of fluid within the skull. The term HC is applied to any condition in which enlargement of the ventricular system occurs as a result of an imbalance between the production and absorption of the cerebrospinal fluid (CSF) (Baron & Goldberger, 1993). HC is not a disease entity rather it is the final common pathway of several conditions that have the same effect on the brain (Barkovich, 1995; Mori, 2000). Blockages in one, or more, of the following locations are most often the cause of HC: narrowed sections of the ventricular system (generally part of the third ventricle or the aqueduct of the fourth ventricle), the subarachnoid space or the arachnoid granulations (Bannister, Russell, Rimmer & Arora, 2000). To relieve raised intracranial pressure (ICP), which is very destructive on brain

tissue, HC is usually treated by diverting the CSF fluid through a ventriculo-peritoneal (VP) shunt (Barkovich, 1990).

CSF is mainly produced in the choroid plexus, located in the floor of the lateral, third, and fourth ventricles (Victor & Ropper, 2001). Under normal circumstances, from its site of formation in the lateral ventricles, CSF flows downward through the third ventricle, aqueduct, fourth ventricle, and foramens Magendie and Luschka to the subarachnoid spaces surrounding the brain and spinal cord. Finally it arrives in the lateral and superior surfaces of the cerebral hemispheres, where most of it is absorbed into the venous circulation by the arachnoid villi (CSF flow is illustrated in *Figure 1*). The primary function of the CSF is to provide protection for the brain and spinal cord from impact to the spinal column and skull and acute changes in venous pressure. It may also be important in the removal of waste products from the CNS (Menkes, 1990; Victor & Ropper, 2001).

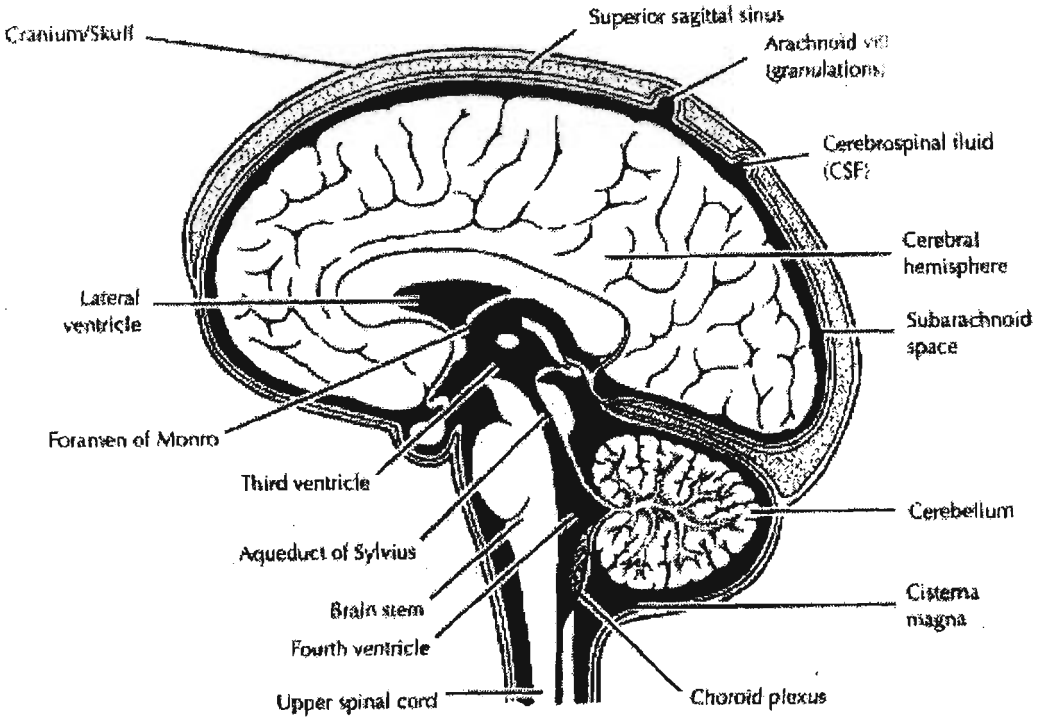

Figure 1. Cross –section of the brain showing cerebral spinal fluid circulation Taken from Hydrocephalus, Madeline Foundation (1995) and Anderson, Northam, Hendy & Wrennall (2001).

In terms of the developmental stages HC is more likely in infancy, where it has been estimated as affecting 6 in every 10,000 or 0.06% (Hommet, Billard, Gillet, Barthez, Lourmiere, Santini, de Toffol, Corcia & Autret, 1999). The current project focuses on early-onset HC, defined as HC, which develops before 12 months of age, often within the first days of life (Fletcher, Dennis & Northrup, 2000).

HC is commonly classified into type (communicating, obstructive), aetiology (congenital, acquired) and/or whether the condition is complicated or uncomplicated (Fletcher, Brookshire, Bohan, Brandt & Davidson, 1995). In 1914 Dandy and Blackfan

introduced the terms *communicating* and *non-communicating (obstructive)* hydrocephalus to distinguish the type of HC in which there is a circulatory abnormality within the ventricular system (Victor & Ropper, 2001). Obstructive (non-communicating) HC occurs when, as the name suggests, there is obstruction to the CSF flow (Barkovich, 1995; refer to *section 1.21* for more detail). In communicating HC there is a free flow of CSF within the ventricular system but the absorption of CSF is interrupted (Johnston, 1995; refer to *section 1.22* for more detail).

The second classification of HC is aetiology. Congenital HC usually refers to the condition being present early in development, where there is a disturbance in the development of the pathways of CSF circulation. Congenital and perinatal forms of HC are usually detected before, at, or shortly after birth (Barkovich, 1995). In contrast, acquired HC refers to the fact that HC develops secondary to some event such as a tumour, haemorrhage or infection, which blocks the channels of circulation (Victor & Ropper, 2001). Fletcher, Brookshire, Bohan, et al. (1995) reported that 70% of cases of HC are of the obstructive congenital form and the remaining 30% are the acquired, usually communicating form. The third classification uncomplicated/complicated refers to whether HC occurs alone (uncomplicated) or in association with other clinical problems (complicated) (Fletcher, Brookshire, Bohan, et al., 1995).

To address the difficulties with HC classification, Raimondi (1988) proposed a grading system for HC in general: Grade 1 includes slit ventricle syndrome, Grade 2 represents ventricles equal in volume to the cerebral parenchyma, Grade 3 identifies ventricles of a greater volume than the cerebral parenchyma, and Grade 4 where the ventricles are so dilated the parenchyma is barely identifiable. A further clarification of

HC was suggested (Raimondi, 1994) to include intraparenchymal (cerebral oedema) and extraparenchymal types, subclassified into subarachnoid, cisternal, and intraventricular forms. However, the traditional basis of grouping HC (i.e. congenital and acquired) is still the most widely utilised in research. Critics of this traditional classification system argue that it cannot always be determined if HC is due to an acquired or congenital aetiology, as in the case of congenital versus acquired obstruction of the aqueduct. Usually the type of obstruction is classified as acquired or congenital depending on the microscopic features of the lesion (Friede, 1989). However, the histopathologic classification of congenital and acquired lesions is controversial (Friede, 1989). Reviewing the literature Drachman and Richardson (1961) found disagreement in the criteria used for classification. Experimental data showed that one of the "congenital lesions" could be induced by viral infection of ependyma. Friede (1989) suggested that the difference between "congenital" and "acquired" obstruction, is probably due to the timing of the disease rather than any other factor. To avoid this conundrum some authors classify HC as either obstructive or communicating HC. Though this classification system is not without problems as in some cases of HC both communicating and obstructive forms can occur simultaneously (Friede, 1989).

Children with obstructive HC have usually developed it secondary to congenital brain malformations that occur in the first trimester of gestation. HC is usually detected shortly after birth because of rapid expansion of the head size to accommodate the increase in CSF. In contrast communicating HC occurs secondary to an insult to the brain peri or postnatally. HC is usually identified through brain ultrasonography routinely conducted on infants who are at high risk. It has been suggested that the distinction

between obstructive and communicating HC is interesting from developmental perspective because obstructive HC tends to develop early in gestation it is associated with a "snowballing" effect on subsequent brain development, whereas communicating HC occurs as the result of peri or postnatal injury to a brain that has developed normally (Wills, 1993). This has been suggested that the two groups will therefore differ neuropsychologically with the latter being less impaired (Wills, 1993). Due to the differences in the timing of onset children with obstructive HC have usually had the destructive causes of HC for a longer duration than children with communicating HC and children who have prolonged periods of increased ICP have higher morbidity rates and high mortality rates (Erickson et al., 2001).

1.21 Obstructive Hydrocephalus

Most frequently obstructive HC is associated with congenital brain malformations such as spina bifida, Arnold Chiari Malformations, Dandy Walker Syndrome and aqueductal stenosis (Brewer, Fletcher, Hiscock & Davidson, 2001). The obstructive form of HC is a condition in which there is an obstruction to the flow of CSF causing it to accumulate within the ventricles, enlarging and expanding the cerebral hemispheres (Victor & Ropper, 2001). There are several sites where obstruction to the flow of CSF may occur. The foramen of Monro, which is the pathway from the lateral ventricles to the third ventricle, may be blocked by a tumour or by the horizontal shift that results from a large unilateral hemispheral mass, resulting in expansion of one lateral ventricle or a portion of one. Large tumours of the third ventricle may block both foramina of Monro, leading to dilation of both lateral ventricles.

Other sites of obstruction of the CSF pathways are the foramina Luschka and

Magendie. Foramina Luschka and Magendie are the three apertures of the fourth ventricle and as such allow the ventricular system to communicate with the subarachnoid space (Nolte, 1999). Obstruction occurs when there is a failure of the opening of the foramina or most commonly in the subarachnoid space around the brainstem due to post inflammatory or post haemorrhagic fibrosing meningitis. These latter forms of obstruction result in enlargement of the entire ventricular system, including the fourth ventricle (Victor & Ropper, 2001).

1.211 Aqueduct Stenosis

Aqueduct Stenosis (AS) accounts for approximately 20% of congenital HC cases. Children with AS develop HC because of congenital narrowing of the aqueduct of Sylvius (Barkovich, 1995). This leads to a blockage of the aqueduct, which ordinarily allows the circulation of CSF out of the lateral ventricles, through the third ventricle and into the fourth ventricle. As a result of the blockage CSF accumulates in the ventricles, leading to ventricular enlargement and consequently great distension throughout the ventricular system and in severe cases, compression of the cortex and grey matter (Bannister et al., 2000). Agenesis of the corpus callosum is common in AS, but other brain malformations are rare (Barkovich, 1990).

1.212 Cysts

Cysts can arise in any part of the brain and may be of sufficient size to create an obstruction in the usual CSF pathway resulting in HC. Commonly, an arachnoid/epidermoid cyst in the lateral ventricles or posterior fossa is the causal factor (Fewell, Levy, & McComb, 1996). Arachnoid cysts are primary congenital lesions within the arachnoid membrane that expand by CSF secretion. They are lined with

arachnoid membranes and filled with CSF (Bannister, Russell, Rimmer & Mowle, 1999). There are two types, those with open necks and free access of CSF into their interiors, and those with narrow necks that trap CSF within them. These cysts are a result of developmental abnormality, or follow inflammation or trauma to the arachnoid space. The cysts lie anywhere within the intracranial cavity or spinal canal. Postnatally, narrow necked cysts are identified because they expand to such a size that they raise intracranial pressure, obstruct the ventricular system and cause HC (Bannister et al., 1999).

Although cysts may occur secondary to other HC aetiologies, and while they may not pose an immediate threat to CSF circulation, they may eventually grow to a size sufficient to give rise to excess intracranial pressure (ICP) and/or cause another point of blockage. In some cases, it is possible to safely and effectively excise (surgically remove) or fenestrate (remove via a syringe inserted into the cyst) the entire cyst and reduce ICP. Treating the HC therefore becomes redundant; the complications of shunting devices therefore do not necessitate consideration, and the focus switches to possible damage done (in the acute stages of HC) and cyst biopsy (to estimate chance of recurrence).

It may not be possible to totally remove the cyst, in which case the remaining parts (which may regrow or continue growing) are operated on for removal at another point in time. In other cases it may not be feasible to safely remove or fenestrate any of the cyst and in this situation a shunt (cysto-peritoneal shunt) may be inserted directly into the cyst to continually drain it, relieving the raised ICP (Ciricillo, Cogen, Harsh, & Edwards, 1991). However, the cysto-peritoneal shunt may not be sufficient to halt the progression of the HC, in which case a VP shunt is also inserted (Almeida, Matushita,

Mattosinho-Franca, & Shibata, 1990; Golden, Rourke, & Bruce, 1987).

1.213 Dandy Walker Syndrome

Dandy Walker Syndrome (DWS) is relatively rare occurring in about 1 per 30,000 births. The clinical triad of the DWS (which has been observed in experimental rat models to occur as early as 12 days of gestational age; Oi, Yamada, Sato, & Matsumoto, 1996) describes a disorder involving the formation of fluid-filled cystic fourth ventricle with partial to complete agenesis of the cerebellar vermis (Leech & Goldstein, 1991). Specifically, as Dandy-Walker malformations occur early in gestation (Oi et al., 1996), the resulting cyst causes enlargement of the posterior fossa, substantial dilation of the fourth ventricle and the cerebellum and lateral venous sinuses of the skull to be pushed upward and laterally. About 70-80% of children with DWS develop HC often due to pressure on the surrounding tissues by the enlarging cyst (Chuang, 1986).

1.214 Spina Bifida

Spina Bifida (SB) is the most common form of neural tube defect. However, the incidence of SB has markedly declined in the past decade due to folic acid being taken before the 28th day of pregnancy (Victor & Ropper, 2001). SB is a condition in which there is a congenital malformation of the CNS such that the infant's vertebral column fails to fuse properly and the vertebrae do not develop correctly. Consequently an opening in the spine occurs (Welch & Lorenzo, 1991). There may be no external sign of the opening or there may be a herniation of the meninges of the spinal column out through the opening. This herniation usually appears as a bulge or sac on the infant's back. The opening can occur anywhere along the spine but most commonly occurs in the lower back region. There are two types of SB; Cystica and Occulta. In the latter and

milder form, the cord remains inside the canal and there is no external sac (Victor & Ropper, 2001). SB Oculta is usually asymptomatic and detected by accident. SB Cystica has more serious sequelae and is subcategorised into two groups, Meningomyelocele and Meningoceles, which are defined by the contents of the sac (Bryan, 1994).

Ninety percent of children with SB have the Meningomyelocele form, which occurs at the rate of one in every 1,000 births. In this form the protruding sac contains the spinal cord as well as CSF, meninges and skin. Furthermore, malformations of the brain are often present, the most common being the Arnold-Chiari malformation (Bryan, 1994).

The second form of SB Cystica is a Meningocele, which is named after the characteristic meningoceles that are usually found in the lumbosacral region. This form is characterised by protrusion of the meninges, CSF and skin through the midline spinal defect. The spinal cord is usually normal and, with the exception of increased risk of HC, the prognosis is usually positive (Welch & Lorenzo, 1991). Children with SB Cystica usually develop HC because of the Chiari II malformation of the cerebellum and brainstem whereas children with aqueductal stenosis develop HC because of a congenital narrowing of the Aqueduct of Sylvius (Barkovich, 1995).

1.215 Chiari Malformations

The Chiari Malformations describe congenital anomalies of the base of the brain. Characteristic morphological features include an elongated medulla and pons, extension of the cerebellar tongue and a narrowed aqueduct. The displaced medulla and cerebellar tissue occludes the foramen magnum. The foramens of Luschka and Magendie open into the cervical canal and the arachnoidal tissue around the herniated brainstem and

cerebellum is fibrotic. All of these factors are considered to be the cause of HC (Victor & Ropper, 2001). Meningoceles are often found when a kink in the spinal cord is present below the herniated tail of the cerebellum, which is pushed posteriorly by the lower end of the fourth ventricle (Victor & Ropper, 2001). The cranial nerves have a long upward pathway and thus may be compressed or stretched. Chiari recognized four types of abnormality. In recent years the term has been restricted to Chiari's type I and II, which refer to cerebellomedullary malformation without and with a meningomyelocele, respectively (Victor & Ropper, 2001).

1.22 Communicating Hydrocephalus

The second form of HC is the communicating type, in which there is interference with the reabsorption of CSF while the pathways within the ventricular system remain functional (Baron & Goldberger, 1993). The site of obstruction in communicating HC is within the subarachnoid space, around the base of the brain, or over the surface of the cerebral hemispheres. It may also be at the arachnoid villi (Johnston, 1995).

Whereas most obstructive forms of HC are of congenital aetiology, communicating HC is often the result of perinatal or postnatal insults to a brain that has otherwise developed normally and therefore is most commonly of acquired aetiology (Fletcher, Brookshire, Bohan, et al., 1995). Occasionally, communicating HC may be congenital because of a developmental failure in the subarachnoid space (Johnston, 1995).

1.221 Intraventricular Haemorrhage

The most common cause of communicating HC is intraventricular haemorrhage (IVH) (Fletcher, Brookshire, Bohan, et al., 1995). Typically IVH occurs in premature or

very low birth weight infants (less than 1500 grams) where inadequately developed blood vessels rupture and bleed into the ventricles. The site of haemorrhage is typically the germinal matrix (Fletcher, Bohan, Brandt, Brookshire, Beaver, Francis, Davidson, Thompson & Miner, 1992). This mix of blood and CSF interferes with the balance of production and absorption of CSF (Fletcher et al., 1992). It is recognised that infants with haemorrhages confined to the germinal matrix (grade 1) and ventricular system without ventricular dilation (grade 2) are at less risk of neurological sequelae than infants with more diffuse bleeding and dilated lateral ventricles (grade 3) or bleeding involving the ventricles and neuronal cell death (grade 4) (Hoon & Melham, 2000; Ment, Vohr, Allan, Katz, Schneider, Westerveld, Duncan & Makuch, 1999).

Of infants with grade 3 to 4 IVH, approximately 50% will have static or transient ventriculomegaly and 50% will require treatment for post-haemorrhagic HC. Outcome depends predominantly on the grade of the IVH, severity of post-haemorrhagic HC and promptness of its neurosurgical management (Ment et al., 1999; Rahman, Murshid, Jamjoom & Jamjoom, 1993).

Of preterm babies with IVH, 13% develop HC with 6% of these children requiring shunting (Fletcher, Landry, Bohan, Davidson, Brookshire, Lachar, Kramer & Francis, 1997). It is not uncommon for premature babies who have grade 3 or 4 haemorrhages to also have periventricular leukomalacia (PVL) (Roth, Baudin, McCormick, Edwards, Townsend, Stewart & Reynolds, 1993).

1.222 Inflammatory Diseases

Inflammatory diseases such as meningitis can result in obliteration of subarachnoid space by stimulating fibrous tissue formation (an obliterative arachnoiditis)

(Leech & Goldstein, 1991). The inflammation and debris block the drainage pathways and may therefore lead to HC. Bacterial meningitis, an inflammation of the membranes that cover the brain and spinal cord, affects about 4 out of every 10,000 children under the age of 4 years (Lieberth, 1994). Thomas (1992) reported that between 1979 and 1989, in Australia, 2.8% of paediatric bacterial meningitis patients developed obstructive HC. In the acute phase HC is caused by 'clumping' of the purulent fluid in the CSF pathways (Barkovich, 1995). Another component may be the inflammation of the arachnoid granulations (Gilles & Davidson, 1971).

Bacterial meningitis is most common in the first month of life but also has a high incidence up to 2 years of age (Victor & Ropper, 2001). The most common cause of bacterial meningitis, in children under 5 years old, is Hemophilus Influenzae type B (Hib) disease. However, since the introduction of the Hib vaccination the incidence of Hib disease, and consequently bacterial meningitis, has markedly declined. In neonatal meningitis, death occurs in 50% to 70% of cases (Gascon & Leech, 1991). Of those who survive, ventricular dilation, intracerebral cysts or cysts communicating with the ventricles as a result of repeated ventricular taps through the fontanelle, commonly occur. Bacterial meningitis is also a significant predictor of shunt malfunction. When the shunt malfunctions, the catheter may become infected and the meningitis spreads, subsequently altering CSF dynamics (Tuli, Drake, Lawless, Wigg, & Lamberti-Pasculli, 2000; Filka, Huttova, Tuharsky, Sagat, Kralinsky, & Krcmeery, 1999).

1.223 Head Injury

HC can be a result of severe head injury complication (Victor & Ropper, 2001). The incidence of post-traumatic HC has been reported as low as 0.7% by some studies

and as high as 29% in others (Guyot & Michael, 2000). Blocking of the aqueduct and fourth ventricle by blood clot and basilar meningeal fibrosis may be the responsible mechanism. In some cases of head injury, raised ICP may cause shearing and tearing of brain tissue, rupturing the blood vessels and causing haemorrhage into the subarachnoid space. Thus the CSF dynamics are disrupted and HC results (Leech & Goldstein, 1991). The pattern of CSF circulation may change dramatically after cranioplasty/craniotomy (Czosnyka, Copeman, Czosnyka, McConnell, Dickinson, & Pickard, 2000), whereby ICP is relieved and the HC arrested. In other cases HC may require active treatment. Traumatic head injury can also cause meningeal cysts (Leech & Goldstein, 1991). In such cases a noncommunicating HC may develop.

1.224 Cerebral Tumours

Cerebral tumours, like cysts, may cause compression and swelling of the brain resulting in inadequate CSF drainage (Oka, Yamamoto, Nagasaka & Tomonaga, 1994). Cerebral tumours in the subarachnoid space can block CSF circulation toward the arachnoid granulations. When this occurs the resulting HC is usually of the communicating form (Oka et al., 1994). Even after excision of the tumour, ICP often remains high due to cerebellar swelling, occlusion of CSF flow, or bleeding into the tumour bed (Imielinski, Kloc, Wasilewski, Liczbik, Puzyrewski, & Karwacki, 1998). Approximately 30-40% of children with cerebral tumours have required a permanent shunt catheter (Bonner & Siegel, 1988; Lee, Wisoff, Abbot, Freed & Epstein, 1994).

1.225 Normal Pressure Hydrocephalus

Raised ICP is not always a feature of HC (Leech & Goldstein, 1991). A form of HC occurs even though CSF pressure is within the normal range and is consequently

termed Normal Pressure Hydrocephalus (NPH). In order to maintain normal pressure CSF formation equilibrates with absorption, which increases in proportion to CSF pressure (Tullberg, Jensen, Ekholm& Wikkelsø, 2001). The decrease in the production of CSF is possibly because of compression of the choroid plexus. Once equilibrium is attained ICP decreases, though it still maintains a slightly higher gradient between ventricle and brain parenchyma (Barkovich, 1990). Finally, a stage is reached where the CSF is at a high normal pressure and the patient presents with cerebral effects of the hydrocephalic state (Victor & Ropper, 2001). The most common cause of NPH is communicating HC with incomplete arachnoid obstruction to CSF drainage. NPH may occur secondary to IVH, intracranial trauma, infections or surgery (Barkovich, 1990).

Characteristic of NPH is the triad of clinical features of a slowly progressive gait disorder, impairment of mental function and sphincter dysfunction (Tullberg et al., 2001; Victor & Ropper, 2001). This form of HC is slowly progressive and does not result in an enlarged cranium. The specific aetiology of NPH is not known (Victor & Ropper, 2001). However, the symptoms and pattern of deterioration reported are similar to the sequelae of the combination of both cerebrovascular and pressure effects on the periventricular white matter (PVWM) and deep grey matter structures (Corkhill & Cadoux-Hudson, 1999; Bradley, Whittemore, Watanabe, Davis, Teresi & Homyak, 1991).

1.3 Pathophysiology

Advances in neuroimaging have made it possible to further investigate the neuroanatomical consequences of HC (Anderson, Northam, Hendy & Wrennall, 2001). Neuroimaging techniques that have been used in the diagnosis, research and management

of HC include air encephalograms, ultrasonic imaging, computerised tomography (CT) and magnetic resonance imaging (MRI).

Air encephalograms were used in early HC studies but have now been replaced by more modern imaging techniques such as CT and MRI scans. CT scans offer a number of advantages over MRI, in that they require shorter examination times and are less expensive to conduct. However, MRI is a popular technique because it provides high-resolution images of brain structures, and it allows the pathophysiological changes in HC to be studied more precisely than with other imaging techniques (Victor & Ropper, 2001). MRI has also proved helpful in early diagnosis of HC, which is made when the ventricles are enlarged in the absence of atrophy (Barkovich, 1990). The advantage ultrasonic imaging provides over other imaging techniques, is that it enables high resolution imaging of the human foetal CNS at approximately the 16^{th} week of gestation. For the examination of the brains of neonates and foetuses ultrasonic imaging is employed.

Although imaging techniques have advanced significantly, it is still difficult in many cases to distinguish between the brain pathology directly attributable to HC and that related to the predisposing structural defect of the pathological condition (Welch & Lorenzo, 1991). One of the challenges of modern neuroimaging is to differentiate ventricular enlargement secondary to HC and that resulting from atrophic ventromegaly (Segev, Metser, Beni-Adani, Elran, Reider-Groswasser & Constantini, 2001). This differentiation is important because it guides the clinician whether to initiate therapy at all, and when surgery is necessary, whether to insert a VP shunt or conduct an endoscopic third ventriculostomy (Segev et al., 2001).

It is widely recognised that early HC may have detrimental effects on the developing brain (Hannay, 2000). The amount of damage to the brain depends upon both the cause and the degree of HC as well as the age of the patient at the time when the HC develops (Barkovich, 1995). The compressive effects of HC are similar across aetiologies, leading to major changes in the midbrain that are influenced by the severity of the condition (Raimondi, 1994). The pathophysiology of HC is multi-factorial in nature and many alterations associated with ventriculomegaly are secondary consequences derived from one or more primary mechanisms (Ding, Lai, McAllister & Canady, 2001). The primary mechanisms associated with HC include the mass effects of compression and stretching of white matter tracts, as well as oedema and ischaemia (Del Bigio & Mc Allister, 1999, cited in Ding et al., 2001).

Ventriculomegaly, disruption of the periventricular ependyma, periventricular oedema, axonal destruction, secondary myelin degeneration and finally a reactive astrocytosis has been described as the sequence of HC (Rubin, Hochwald, Tiell, Liwincz & Epstein, 1975, cited in Anderson, Northam, et al., 2001). As the ventricles dilate, usually in a posterior to anterior direction, pressure is exerted on the surrounding structures and neural fibres. The stretching of neural structures compromises the projection fibres and the optic and olfactory tracts may be also damaged (Del Bigio, 1993; Dennis, Fitz, Netley, Sugar, Harwood-Nash, Hendrick, Hoffman & Humphreys, 1981; Hannay, 2000). This process may also damage the anterior cerebral artery and thus reduce cerebral blood flow leading to ischaemia (Brewer et al., 2001).

Another consequence of ventricular enlargement is the stretching and damaging of corpus callosum fibres. Partial agenesis of the corpus callosum is common in all

congenital forms of HC (Klaas, Hannay, Caroselli & Fletcher, 1999). Even when all portions are present this structure is often thinned and stretched. The extent of the damage to the corpus callosum varies across cases, with the destructive effects being most often apparent in the mid-portion of the corpus callosum (Barkovich, 1995; Barkovich & Newton, 1988). However, dorsal flattening and thinning of the posterior body of the corpus callosum has also been reported (Jenkins, 1991).

If HC persists, distension of the cerebral hemispheres will cause injury to neurons and there is thinning of the cortical mantle throughout the brain (Barkovich, 1990). Thinning or stretching of the cortical mantle in children with HC is often asymmetric in an anteroposterior direction (Del Bigio, 1993; Hannay, 2000). The vertex and occipital lobe have been shown to have greater mantle thinning than frontal regions on autopsy and CT scans of HC children (Dennis et al., 1981). Due to the frontal horns being more rigid than those in the occipital horn region, selective rather than global thinning occurs, because the portion of the brain that atrophies most is that which presses against an inflexible wall (Dennis et al., 1981). In addition, injuries to the cerebellum and posterior fossa, especially in children with DWS and SB meningomyelocele, have been reported (Klaas et al., 1999).

HC also has a detrimental affect on myelination (Rubin, et al., 1975). As axons in the PVWM become elongated, disruption to the surrounding myelin occurs. Other studies have reported that myelination of the corpus callosum is delayed, either from direct interference with myelinogenesis or a secondary destructive effect after myelination has commenced (Hannay, 2000). The effect of raised ICP and CSF volume on myelination has been investigated (Hanlo, Gooskens, van Shoonveld, Tulleken, van

der Knaap, Famer & Willemse, 1997). Hanlo et al. (1997) found a significant correlation between anterior fontanelle pressure (AFP) and degree of myelination present on MRI, however, the volume of CSF correlated poorly with the degree of myelination and mean AFP. Interestingly, most of the children showed some recovery of myelination following CSF drainage, however, this depended on the duration of raised ICP and secondary parenchymal damage (Hanlo et al., 1997). Consistent with Hanlo et al.'s findings, Van der Knaap and Valk (1990) also found a negative relationship between CSF volume and myelination.

It is debated whether cell death plays an important role in the pathophysiology of HC (McAllister & Chovan, 1998). The relatively recent refinement of the mechanism of cell death into two categories, apoptosis (programmed cell death) and necrosis, has renewed the debate about whether dark neurons found in untreated HC brains could fully recover after appropriately timed shunting (Mc Allister, 2000).

1.4 Treatment

The development of ventricular shunt tubing with one-way valves, approximately 50 years ago, marked the beginning of successful treatment of HC (Victor & Ropper, 2001; Wills, 1993). Shunting aims to restore equilibrium between cerebrospinal fluid production and absorption, and it remains the principal effective treatment for HC (Baron & Goldberger, 1993). A shunt or a piece of silastic tubing is surgically inserted usually into either the frontal or occipital horn of the lateral ventricle through a small hole in the calvarium (Barkovich, 1990). In general, the tip of the ventriculostomy tube lies within the lateral ventricle in the region of the foramen of Munro (Barkovich, 1990) in order to

drain the excess CSF from the ventricles into another cavity, usually the abdominal or chest cavity. The tube has a one-way valve so the CSF is directed away from the brain (Johnston, 1995).

Even though shunts have proven to be extremely effective, especially since the development of flexible and relatively inert silicone tubing, they are subject to complications, such as blockage, disconnection, breakage, calcification, infection and haemorrhage (Gilkes, Steers & Minns, 1999; Martinez-Lage, Lopez, Poza, & Hernandez, 1998). Shunt malfunctions are clinically manifested by symptoms of raised ICP and excessive rate of head growth in the infant. The most common malfunction is due to occlusion of the ventricular catheter by choroid plexus or glial tissue, which grows into the lumen (Barkovich, 1990). This is usually shown on imaging studies by correct placement of the shunt but enlarging ventricles. Another form of shunt malfunction is due to disconnection. This can occur at any point within the shunt system and most commonly occurs where the various components are joined together (Barkovich, 1990).

The risk of infection resulting from ventricular shunting has been estimated to be between 1% and 30% (Ammirati & Raimondi, 1987; Barkovich, 1990). In a sample of 431 children, Ammirati and Raimondi (1987) found that those who had a shunt inserted at a younger age had more infections than older children. The major clinical symptom of shunt infection is fever. Symptoms such as anaemia, dehydration, hepatosplenomegaly and stiffness of the neck muscles may also occur. Other complications arising from ventriculo-peritoneal shunting are rare such as subdural haematomas. If they are present, they are usually in children older than three years after overly vigorous drainage of markedly enlarged ventricles (Barkovich, 1990). There may also be complications within

the abdomen and these include ascites, pseudocysts, perforation of the abdominal wall and intestinal obstruction (Barkovich, 1990).

Infrequently, patients may develop granulomatous reactions adjacent to the shunt tube either within or near the ventricle. Some shunted HC patients may develop symptoms of shunt failure in the absence of ventricular enlargement. This has been termed "slit ventricle syndrome" and the cause is not known. In fact, there is disagreement about what clinical and/or pathological findings define this syndrome (Ammirati & Raimondi, 1987; Barkovich, 1990).

Finally, very rarely when the ventricular end of the shunt becomes blocked, CSF may track along the shunt and enter the interstices of the centrum semiovale. Oedema occurs in the area around the shunt and eventually a cyst may form in the white matter surrounding the ventricular catheter. The oedema and cyst resolve with shunt revision (Barkovich, 1990). Shunt malfunctions, such as blockages and disconnections, usually require surgical replacement of the shunt or replacement of malfunctioning parts (Barkovich, 1990). The surgery itself, as does any neurosurgical technique, has risks associated.

Higher incidences of seizures have been noted in children with shunted HC than the general population, varying between 6 to 59%. A study of factors predicting epilepsy in children with HC, found that abnormalities in radiological scans, shunt complications and episodes of raised ICP were all predictors (Bourgeois, Sainte-Rose, Cinalli, Maixner, Malucci, Zerah, Pierre-Kahn, Renier, Hoppe-Hirsche & Aicardi, 1999).

In the past decade third ventriculostomy, another treatment option, has become more popular (Kestle, Cochrane & Alisharan, 2000). This procedure involves making a

hole in the floor or sometimes the wall of the third ventricle (approximately 1-10mm in diameter) so that the CSF can drain into the subarachnoid space at the base of the brain (Johnston, 1995; Kestle et al., 2000). The hole is made usually by ventriculoscope, which is less invasive than other procedures (Johnston, 1995).

The rise in third ventriculostomies, as the initial treatment of HC, is due to the availability of good endoscopic equipment (Kestle, et al., 2000). One study found that the proportion of patients who were treated initially with this procedure ranged markedly (0-100%) between the 43 surgeons that were questioned. The incidence of third ventriculostomies was not related to the surgeons' training or years of experience, however, if the patient had AS, a tectal tumour or a thinned, ballooned floor of the third ventricle, it raised the chance of this procedure being conducted (Kestle, et al., 2000).

1.5 Neuropsychology of Hydrocephalus

Considerable research has focussed on the neuropsychological sequelae of HC and studies have been increasingly directed toward understanding specific deficit patterns, and correlating function with neuropathology (Erickson, Baron & Fantie, 2001). The majority of studies reviewed consisted of heterogenous samples of children with HC.

1.51 Intelligence Quotients

Despite the numerous studies over the past three decades investigating intellectual abilities in early onset HC, the precise nature of the effects of HC on intelligence is still unknown (Erickson, et al., 2001). Before HC was treated by surgical intervention, only 25% of children with HC survived to adulthood (Anderson, 1975; Anderson, Northam, et al., 2001). Less than 40% of the surviving children were within the *Average* range on

tests of intelligence and the majority were severely impaired (Laurence, 1969). In more recent years, samples of children with HC are reported to have full scale IQs in the *Low Average* (Bier, Morales, Liebling, Geddes & Kim, 1997; Dennis et al., 1981; Donders, Rourke & Canady, 1991; Friedrich, Lovejoy, Shaffer, Shurtleff & Beilke, 1991) or *Average* range (Barnes & Dennis, 1998; Brewer et al., 2001; Dennis et al., 1981; Donders et al., 1991; Hetherington & Dennis, 1991; Klaas et al., 1999). Children with HC are now more likely to have normal FIQs than in the previous decades because of improved and earlier diagnosis, better treatment and reduced incidence of shunt complications (Erickson et al., 2001). However, it is important to bear in mind that most studies exclude participants with IQs less than 70, therefore the population mean is likely to be even further below that of the general population.

Early shunting has led to improved survival rates, though arguably not better intellectual outcomes (Hommet et al., 1999). Children who have HC severe enough to require insertion of a shunt have significantly lower IQ scores than children with a history of mild, arrested and unshunted HC (Fletcher, Brookshire, Landry, Bohan, Davidson, Francis, Levin, Kramer & Morris, 1996). Hanlo et al. (1997) reported that successful shunt surgery benefits measured IQ across the life span. Whilst Thompson, Fletcher, Chapieski and Landry (1982) postulate that shunting does not necessarily result in improved cognitive ability rather it halts a decline.

Empirical attempts to document the associations between the number of shunt revisions and neuropsychological outcome have produced inconsistent findings (Anderson, Northam, et al. (2001). Whilst some studies show no relationship between number of shunt revisions and test scores (Dennis et al., 1981; Fletcher, Brookshire, et

al., 1996; Ralph, Moylan, Canady & Simmons, 2000), others have shown a decline in IQ scores with increasing number of shunt revisions (Bier et al., 1997; Menkes, 1990). What has been consistently found is that immediate treatment of HC is crucial as longer durations of untreated HC are associated with poorer IQ outcomes (Dennis et al., 1981; Young, Nulsen & Weiss, 1973). Similarly, a history of shunt infections or bleeding has been correlated with lower IQs (Hunt & Homes, 1976; Wills, Holmbeck, Dillon & McLone, 1990).

Few studies have examined the longitudinal variability of IQ scores in children with HC and findings to date are inconsistent (Anderson, Northam, et al., 2001). Whilst one study has found stability in IQ over time (Tew & Laurance, 1983), others have found a decline in scores with increasing age (Jacobs, Northam & Anderson, 2001; Wills et al, 1990). Brookshire, Fletcher, Landry, Davidson & Francis (1995b) found general instability in IQ scores across a five-year period and substantial fluctuations in scores were evident over the follow-up. They suggested that this instability may be due to several factors such as attention and the effects of repeated exposure to the tasks. In a cross-sectional analysis Dennis et al. (1981) found no significant relationship between age at testing and IQ score. This group concluded that, provided the child was treated for HC before 12 months of age, HC does not progressively debilitate intelligence throughout childhood.

In general, timing of brain insult has significant consequences on intellectual outcome (Ewing- Cobbs, Thompson, Miner & Fletcher, 1994). In the case of congenital versus acquired HC, congenital aetiologies may be characterized by more severe cognitive deficits than acquired HC given the earlier timing of the brain abnormality

(Dennis et al., 1981; Hetherington & Dennis, 1999). However, the site of obstruction in HC (intraventricular or extraventricular) has not been found to be associated with IQ (Dennis et al., 1981; Fletcher, Brookshire, et al., 1995; Riva, Milani, Giorgi, Pantaleoni, Zorzi, & Devoti, 1994).

The influence of medical variables on the outcome of HC is an understudied domain (Anderson, Northam, et al., 2001). In general, poorer outcomes have been noted for children who have HC and other co-morbid conditions (Donders, et al., 1991; Hunt & Homes, 1976; Shurtleff, Foltz & Loeser, 1973; Ralph, Moylan, Canady & Simmons, 2000; Young, et al., 1973). Lower IQ scores, particularly Performance IQ (PIQ), have been found in children who have HC and cerebral palsy, significant visual impairments, or seizures (Dennis et al., 1981; Laurance, 1969; McCullough & Balzer-Martin, 1982; Ralph, et al., 2000). Children with uncomplicated HC, as found in AS, tend to have higher IQs than those with HC in combination with other CNS malformations such as spina bifida or when HC occurs secondary to infections (Dennis et al., 1981). One study of 106 children with SB found that the level of the lesion was most strongly associated with IQ (measured by the Kaufman Brief Intelligence Test) (Bier et al.,1997).

Studies have attempted to correlate intellectual functioning with various neuroantomical features (thickness of cortical mantle, brain mass) (Hunt & Holmes, 1976; Shurtleff et al., 1973; Young, et al., 1973). Research to date indicates that cortical mantle size does not directly relate to intellectual potential and children with very thin cortical mantles can have normal intelligence (Lonton, Barrington, & Lorber, 1975, cited in Erickson et al., 2001; Lorber, 1980). However, prior to shunt surgery reduced mantle thickness (calculated by brain mass) is usually associated with a low FIQ (Choudhury,

1995; Jackson & Lorber, 1984; Tew & Laurence, 1983; Thompson, Eisenberg & Levin, 1982). Whilst a positive relationship may exist between cortical mantle size and intelligence in group studies, exceptions may occur at the individual level (Erickson et al., 2001).

The comparison of ventricular size after shunting and long-term IQ impairment has provided mixed results. Some studies reported the two variables to be correlated, but only if the distension is above a certain threshold level (Foltz & Shurtleff, 1963, Renier, Sainte-Rose, Pierre-Kahn, & Hirsch, 1998; Thompson et al., 1982; Young et al., 1973;). Other research has found no association between post-shunt ventricular size and IQ (Hanlo et al., 1997; van der Knaap, Valk, Bakker, Schooneveld, Faber, Willemse & Gooskens, 1991). With regard to corpus callosum, one study found that children with an intact corpus callosum tended to have higher IQ scores than children with agenesis of the corpus callosum (Fletcher et al., 1992). The degree of myelination determined by MRI has been found to be weakly correlated with performance on Bayley Scales in infants. However as children mature, gray matter volumes are more strongly related to IQ than overall white matter volumes (Fletcher, Bohan, Brandt, Kramer, Brookshire, Thorstad, Davidson, Francis, McCauley, & Baumgartner, 1996).

1.52 Motor Skills

Motor skill deficits are frequently reported among children with HC (Dennis et al., 1981; Fletcher et al., 2000). Both gross motor and fine motor skill difficulties have been described. For example, one study found that performances across a variety of motor skill tasks were poor and fell between one and three standard deviations below the mean for normally developing children (Hetherington & Dennis, 1999). These deficits,

according to Hetherington and Dennis (1999), are of no surprise because the neuropathology often involves brain regions implicated in motor control, such as the cerebellum and brainstem, corpus callosum, PVWM tracts (including the cerebrospinal tract), basal ganglia and the cerebral cortex.

Most research examining gross motor function in children with HC has been restricted to SB aetiology (Hetherington & Dennis, 1999). Certainly, gross motor deficits would be expected in many children with SB because lower limb dysfunction is common in these children, due to lesions of the spinal cord (Hetherington & Dennis, 1999). In fact in one study gait abnormalities were reported in 92% of a group of children with SB and HC (Fletcher et al., 1995). Demonstrating that gross motor abnormalities are not unique to SB, gait abnormalities were also present in 44% of children with HC as a result of IVH and 36% of a group with AS (Fletcher, Levin & Butler, 1995). Likewise, another study found that both ambulatory and non-ambulatory children with HC, secondary to various aetiologies, demonstrated significant impairment on a test of gross motor function (Hetherington & Dennis, 1999).

Significant research has been aimed at characterising the fine motor skills of children with HC, which are essential for handwriting, drawing and other academically relevant skills (Hetherington & Dennis, 1999). Regardless of gross motor skill level, deficits in upper limb and hand function including fine motor skill and psychomotor speed are frequently reported (Hetherington & Dennis, 1999). Of note, a large study of 187 children with HC arising from various aetiologies, found that 74% of children with shunted HC had abnormalities in fine motor coordination (Fletcher, Levin, et al., 1995).

Even though current research suggests that regardless of aetiology, children with

HC demonstrate some motor deficits, there is limited literature on differences in motor function profiles across the various aetiologies. A recent study compared motor function in 42 children with HC arising from four aetiologies; two aetiologies were congenital (SB meningomyelocele and AS) and two were acquired (IVH) and infections/adhesions of the brain). Four motor domains were studied: persistence and smooth motor control, strength, fine motor skills, and balance, gait and posture. On 8 of the 10 motor subtests the congenital HC group performed more poorly than the acquired HC group (Hetherington & Dennis, 1999). Multivariate analysis of variance showed that the test of gross motor skill (standing balance) contributed most to this difference (Hetherington & Dennis, 1999). Further evaluation of differences in motor function as a function of aetiology of HC requires larger samples than have been utilised to date (Fletcher et al., 2000).

Various socio-demographic and medical factors have been shown to be correlated with motor outcome (Wills, 1993). Hetherington and Dennis (1999) found that handedness, gender, and seizures were weakly related to some motor functions. However, due to a small sample size, independent effects of these variables were not investigated. It has been reported that HC children are more prone to mixed handedness and that there is an increased incidence of left handedness (Dennis et al., 1981).

Fletcher, Bohan, et al. (1996) found that the cross-sectional area of the corpus callosum was correlated with fine motor skills. In particular, when hypoplasia was included in the analyses, corpus callosum abnormalities always involved the posterior body and splenium (Fletcher, Bohan, et al., 1996).

1.53 Non-Verbal, Visual Spatial Skills

Relatively consistent is the finding that children with HC have problems with visual-motor and visual spatial tasks (Dennis et al., 1981; Fletcher et al., 1992; Fletcher, Brookshire, et al., 1995; Fletcher et al., 2000), both on tasks that require constructional performance and on tests that minimize motor demands, such as judgement of line orientation (Lindgren & Benton, 1980). Many studies report poorer PIQ than VIQ scores on WISC-R and WISC-III (Brookshire, Fletcher, Bohan, Landry, Davidson & Francis, 1995b; Dennis et al., 1981; Donders et al., 1991; Fletcher et al., 1992; Holler, Fennell, Crosson, Boggs & Mickle, 1995; Shaffer, Friedrich, Shurtleff, Shurtleff & Wolf, 1986; Tew & Laurance, 1975). In fact, Wills et al. (1990) reported that a significant difference between PIQ and VIQ occurred at double the rate found in the normative population. Notwithstanding this, not all studies have found a difference in PIQ and VIQ scores (Friedrich et al., 1991; Hommet, Cottier, Billard, Perrier, Gillet, De Toffol, Sirinelli, Bertrand & Autret, 2002). However, even if there is not a significant difference between PIQ and VIQ, the latter will often be marginally higher. For example, in the Hommet et al. (2002) study the mean VIQ was almost five points greater than the mean PIQ.

One explanation for observed PIQ-VIQ differences is that they are due to shunt surgery and placement. Most shunts are inserted in the right parietal region and such surgery damages brain tissue, leading to a greater impairment of the functions subserved by the right hemisphere or intact connections to this region (Fletcher, Brookshire, et al., 1996). This interpretation is supported by Fletcher and colleagues (1996) observation that the PIQ-VIQ differences were only evident in premature children with shunts in situ. Alternatively, Anderson, Northam, et al. (2001) stated that this association might reflect

the fact that those children with more severe HC have received treatment.

Motor problems, even of a mild degree, have been reported to limit the development of non-verbal intelligence (Dennis et al., 1981; Thompson, Fletcher, Chapieski, Landry, Miner & Bixby, 1991). Consistent with this interpretation, Dennis et al. (1981) found that motor deficits were strongly associated with lower PIQ scores but had no effect on verbal intelligence. Slower processing speed has also been associated with a lower PIQ. Supporting this view, Thompson et al. (1991) found no difference in VIQ and PIQ on an un-timed test of general cognitive ability. Nevertheless, it has been argued that reduced movement does not fully explain non-verbal deficits because mobile and non-mobile children with HC have been found to have similarly low PIQs (Abercrombie, 1968). Furthermore, not all studies have been able to attribute poorer PIQs to the motor demands of the task (Brookshire et al., 1995b). Anderson, Northam, et al. (2001) suggested that children with HC may have impaired higher-order motor planning skills and spatial problem-solving skills that in turn compromise their PIQ. Many of the traditional "performance" tests have inherent demands on executive functions and as the item difficulty increases there is often a corresponding increase in the demand of executive functions in the test items. It has been suggested that executive function deficits may impede children with HC's performance on non-verbal reasoning tasks (Anderson, Northam et al. 2001, Erickson, et al., 2001). Consequently, reported lower PIQ's are likely to be due to a combination of factors such as visual-spatial perceptual deficits, impaired motor skills, slowed processing speed and executive function deficits.

Ocular motor function deficits have been found in children with HC (Houliston,

Taguri, Dutton, Hajivassilou & Young, 1999). Deficits such as abnormalities in gaze and movement, and/or refraction and accommodation, are commonly reported (Blakemore & Eggers, 1978, cited in Dennis et al., 1981; Hein, 1979). It has been hypothesised that certain visual problems of HC limit the child's visual experience, which then alters the structure and function of the visual cortex, itself being abnormally developed (Dennis et al., 1981). These deficits have a negative effect on visual acuity, visually guided behaviour and non-verbal intelligence (Dennis et al., 1981). A study that examined visual-processing skills in children with HC, identified deficits in shape recognition, simultaneous perception, perception of movement, colour perception, orientation, object recognition and face recognition (Houliston et al., 1999).

Children with AS may be most vulnerable to oculomotor problems because of the pressure on midbrain collicular structures, affecting the cranial nerve nuclei, causing third (oculomotor) and sixth (abducens) nerve palsy. Cranial nerve III innervates four of the six extrinsic eye muscles and the levator muscle on the upper eyelid, whilst cranial nerve VI innervates the lateral rectus, one of the six extrinsic eye muscles (Tortora & Grabowski, 1993). Visual conditions like strabismus (a condition where the axes of the eyes differ so they do not focus on the same object) chronically reduce concordant binocular vision and limit visual experience, which, in turn, will disrupt the development of the visual cortex (Dennis et al., 1981; Tortora & Grabowski, 1993).

Reported differences in visual motor and spatial abilities between the separate HC aetiologies vary. Congenital aetiologies, in the Dennis et al. (1981) study, demonstrated the greatest discrepancies in their WISC-R PIQ-VIQ scores. Post-natal aetiologies, on the other hand, demonstrated comparable VIQ-PIQ scores (Dennis et al., 1981). Fletcher,

Bohan, et al. (1992) postulated that non-verbal skills might be selectively impaired in children with obstructive HC due to the relative greater loss of cerebral white matter in the right hemisphere. Alternatively, deficits may be due to callosal agenesis, which limits access to right hemisphere functions. They noted that children with acquired HC related to prematurity may have fewer CNS anomalies because the brain was developing normally but acquired damage, rather than with congenital HC, where there is an abnormality in the early development of the brain structures i.e. AS. Not all studies have found aetiological differences in PIQ-VIQ. Fletcher, Brookshire, Bohan, et al. (1995) compared children with SB, AS and IVH on the different subtests of the Test of Visual-Perceptual Skills and found no significant difference in performances across aetiologies.

A history of seizures has been related to poor development of non-verbal intelligence (Dennis et al., 1981; Schiottz-Christiansen & Bruhn, 1973). Dennis et al. (1981) found that IQ measures did not differ between the seizure forms (motor-sensory, neocortical, rhinencephalic, tonic-adversive, absence, automatism, generalised or unknown) or seizure frequency. However, the presence of EEG seizure focus and the severity of the EEG disturbance determined from EEG reports (not analysis of the EEG protocols) influenced the level of non-verbal intelligence. Importantly, in the children who had a seizure focus for five out of the six children it was in the right hemisphere. Therefore it could be argued that the presence of right hemisphere focused seizures limited the development of nonverbal intelligence, rather than seizures in general.

Neuroanatomical correlates of lower PIQ scores have been investigated (Dennis et al., 1981; Donders et al., 1991; Fletcher, Bohan, et al., 1992; Fletcher, Bohan, et al., 1996). Such studies have found that children with lower PIQ scores than VIQ showed

proportionately greater brain thinning in posterior brain regions relative to anterior regions (Dennis et al., 1981). MRI studies evaluating areas of the corpus callosum, internal capsules and centrum semiovale, have found that the cross-sectional area of the corpus callosum is more strongly correlated with PIQ than with VIQ (Fletcher, Bohan et al., 1992; Fletcher, Bohan et al., 1996). In addition, non-verbal measures were also correlated with the volume of the right ventricle and with the area of the right and left internal capsules (Fletcher, Bohan et al., 1992).

Donders et al. (1991) speculated that differences in PIQ and VIQ were a reflection of persistent dysfunction in the posterior regions of the right cerebral hemisphere. Furthermore they suggested that there is more white matter interregional integration in the right hemisphere than the left and HC tends to be particularly destructive of white matter. Visual-spatial tasks often require processing of novel or complex stimuli and this is thought to be a function of the white matter interregional connections (Rourke, 1987; 1989). Thus, the relatively inefficiency in dealing with novel or complex visual-spatial stimuli was related to dysfunction of white matter within the posterior regions of the right cerebral hemisphere (Donders et al., 1991).

1.54 Verbal Skills

As discussed, several studies of children with early onset HC have documented discrepancies in their performance on traditional measures of intelligence, with better verbal skills compared to non-verbal skills (Brookshire, Fletcher, Bohan, Landry, Davidson & Francis, 1995a). Intact use of grammar, and relatively good performance on formal tests of vocabulary, comprehension and syntax have been reported (Horn, Lorch, Lorch & Culatta, 1985), as have the intact word recognition skills, use of morphology

and understanding of grammar (Dennis, Hedrick, Hoffman, & Humphreys, 1987). However, children with HC are not without language difficulties (Brookshire et al., 1995a).

The pattern of fluent, well articulated speech that is syntactically correct but characterized by perseveration in responses, excessive use of social phrases, inappropriate familiarity in manner, and a tendency to introduce personal experiences into conversation inappropriately or out of context has been observed in HC children (Barnes & Dennis, 1997; Hadenius, Hagberg, Hyttnas-Bensch & Sjorgren, 1962; Tew, 1979) and has been termed the "cocktail party syndrome" (CPS) (Tew, 1979). CPS refers to the fact that their speech is fluent but deficient in content, contains several references to personal experiences (that are not shared with the person they are speaking with) and reveals a confident, perhaps overly familiar, social demeanour (Barnes & Dennis, 1997).

Although other authors have found evidence of aspects of CPS in children with HC, the syndrome itself is controversial. Dennis, Hedrick, Hoffman & Humphreys (1987) argue that CPS is not a true syndrome rather it describes the pattern of superficial, perseverative language observed in some children with HC. Supporting this view, Tew (1979) noted that these behaviours appeared to decline with age in many children and that CPS was more strongly associated with lower IQ and impaired mobility than with HC. Perhaps the strongest argument for CPS being a symptom of a broader pattern of cognitive impairment is provided by Rourke (1989) who has included CPS as one of the features of Non-Verbal Learning Disorder (NLD) (Anderson, Northam, et al., 2001). In a recent study, Yeates, Loss, Colvin & Enrile (2003) found about 50% of children with HC in their sample had a pattern of deficits consistent with NLD. However, their sample was

restricted to children with SB.

Whilst Yeates et al. (2003) findings indicated that half of the children with myelomeningocele and shunted HC demonstrated a pattern of assets and deficits consistent with the NLD syndrome, the same proportion of these children did not. In general, these children demonstrated more variability in their in their strengths and weaknesses than their siblings, that is more suggestive of substantial individual differences in neuropsychological functioning that are characteristic of children with SB and HC (Fletcher et al., 2000; Yeates et al., 2003). Thus the authors caution against making generalizations of applying the NLD model to children with SB and early-onset HC. Even though the NLD model may not always be present in children with HC, language deficits are commonly reported (Barnes & Dennis, 1998; Brookshire et al., 1995a; Dennis et al. 1987)

Horn, Lorch, Lorch and Culatta (1985) postulated that the hyperverbal behaviour evident in some children with HC was due to a deficit in selective attention rather than a deficit in language abilities. Children with HC have difficulty focussing their attention on the pertinent information and ignoring distractors. Baron and Goldberger (1989) explained hyperverbal behaviour in terms of a failure of executive control, rather than a fluency/word retrieval problem. They speculated that children with HC have difficulty initiating, inhibiting and monitoring their speech output for logic and relevance (Anderson, Northam, et al., 2001).

Other language deficits have been described in HC children. Poor metalinguistic skills (the ability to explain and reflect on or monitor language) have been reported (Dennis, et al., 1987). In addition, the speech of children with HC has been reported as

lacking in appropriate content and their verbal fluency is unusually high (Dennis et al., 1987). Further deficits have been noted in their phonological awareness, in the ability to describe word meaning and the comprehension of complex grammatical structures (Dennis et al., 1987; Brookshire et al., 1995a; Prigatano, Zeiner, Pollay & Kaplan, 1983).

Thus, the language and speech abilities of HC children are neither globally impaired nor globally proficient (Dennis et al., 1987). These children often produce single words in isolation better than they use the same words in context. They are slower than their peers to produce content words appropriate to semantic-contextual cues (Barnes & Dennis, 1998). In contrast, they are within age expectations on producing the same words in referential naming tasks and recognising words (Dennis et al., 1987). However, they do not have a comparable level of comprehension of written discourse (Barnes & Dennis, 1998).

More recently, research has focussed on the narrative language abilities of children with early onset HC (Brookshire et al., 1995a). Dennis, Jacenik and Barnes (1993) analysed the narratives of 49 children aged 6 to 15 years. They found that children with HC produced narratives that conveyed less of the pertinent content, included more ambiguous material and were more convoluted and verbose (Dennis et al., 1993). Dennis et al. (1987) considered these problems with narrative discourse to reflect deficits in pragmatic language that were a product of problems with language processing rather than social language deficits.

In a companion study Dennis and Barnes (1993) found problems in establishing alternate meanings for ambiguous sentences, understanding figurative expression, making bridging inferences and producing speech acts on the Test of Language Competence-

Expanded Edition (TLC-E). They explained these language deficits as indicative of difficulties with conveying textual language rather than problems at the word and sentence level (Dennis & Barnes, 1993). That is children with HC exhibit a disorder of language usage rather than language content (Anderson, Northam, et al., 2001).

The discrepancy in their apparent language ability and the actual communicative content of what children with HC say may lead others to over-estimate their true abilities (Anderson, Northam, et al., 2001). This can be to the children's disadvantage because some deficits may go unnoticed and inappropriately high expectations may be placed on the child. In summary, difficulties in metalinguistics awareness and understanding of inferential language, combined with slowed comprehension of more complex grammatical structures, may impede the child from understanding information aimed at an age expected level, which has serious consequences especially in the academic domain.

Little is known about whether and how language development can be predicted from the medical history of children with HC (Dennis et al., 1987). It is also unclear if some degree of difficulty with speech and language functioning is characteristic of all forms of HC or early HC or whether it occurs only in certain aetiologies. Prenatal aetiologies of HC have been associated with poorer language skills than peri and postnatal causes (Brookshire, et al., 1995a; Dennis et al., 1987). Brookshire et al. (1995a) studied phonological awareness skills, semantic language ability, fluency and automaticity, and word finding abilities in children with HC due to SB, IVH and AS. Children with SB performed more poorly than those with IVH, and the children with AS performed within normal expectations on all tasks except for rapid automatized naming.

Fletcher et al. (2000) suggested that the deficit in the rapid retrieval of words from semantic memory might be a consistent feature of HC regardless of aetiology, especially considering the problems many children with HC have conveying information in an economical manner.

With regard to differences in language abilities between types of HC, Dennis et al. (1987) found children with communicating HC had preserved higher-order language functions but disrupted speech fluency, suggestive of subtentorial regions being involved in speech fluency. In contrast, children with obstructive HC had fluent speech but disrupted lexical access, sequencing, and grammatical comprehension and these deficits were especially evident in SB.

It has been suggested that disruption of cortical association fibres that occurs secondary to HC may result in hyperverbal speech (Donders, Canady & Rourke, 1990). Brookshire, et al. (1995a) interpreted the word retrieval difficulties as a problem with intra or interhemispheric transfer of information due to corpus callosum anomalies as the task is thought to require the transfer of visual information from visual-perceptual centres in the posterior cortex to speech centres in the anterior left hemisphere (Brookshire et al., 1995a). Fletcher and colleagues (1992; 1996) were unable to correlate verbal abilities with lateral ventricle volumes, area measurements of the internal capsules, semiovale or corpus callosum area. Instead this word retrieval deficit may reflect slower information processing speed, which is often found in children with HC, rather than a language problem per se (Anderson, Northam, et al., 2001). Furthermore, visual problems commonly associated with HC may compromise performance on naming and sequencing tasks (Anderson, Northam, et al., 2001).

1.55 Attention

Attention is not a unitary process; rather it is a multidimensional construct (Fletcher, 1998). Mirsky (1996) conceptualised four components of attention; focus-execute, sustain, encode and shift. Despite parents and teachers reporting attention problems, empirical studies documenting the nature and extent of the attentional impairments of HC children are limited and mainly address children with HC secondary to SB (Anderson, Northam, et al., 2001; Brewer et al., 2001; Erickson et al., 2001). In general, it is unclear whether the attention problems documented are a consequence of faulty attentional system, or of perceptual motor demands of specific tests. However, the most recent evidence suggests that even when motor demands are minimized children with HC have attentional problems (Brewer et al., 2001).

Research tends to centre on aspects of focussed and sustained attentional processes, and to a lesser extent shifting attention. Deficits in focussed attention, the capacity to respond discretely to specific stimuli within an array of stimuli, have been reported in children with HC (Brewer et al., 2001; Fletcher, Brookshire, Bohan, et al., 1995; Fletcher, Brookshire, et al., 1996; Loss et al., 1998; Tew, Laurance & Richards, 1980). Neuroanatomically, according to Mirsky, Anthony, Duncan, Ahearn and Kellam (1991) and Posner and Peterson (1990), focussed attention is subserved by the superior temporal and inferior parietal lobes. Fletcher, Brookshire, Landry, Bohan, Kramer and Morris (1996) argued that poorer focussed attention probably reflected motor and speed of processing deficits ubiquitous in HC. In fact, when the motor component was reduced, children with HC performed as well as control subjects on focussed attention tasks (Fletcher, Brookshire, et al., 1996). Furthermore, a meta-analytic study of the literature

found that although simple focussed attention was intact in children with HC, difficulties may be evident on more complex tasks that demand active scanning (Wills, 1993).

Horn et al. (1985) found that children with HC secondary to SB had poorer focussed attention abilities than normal controls on a computerized vocabulary test. A limitation of this study was that attention tests were given in the visual modality and generalizations were made to the auditory modality (Brewer et al., 2001). According to Brewer et al. (2001), given current knowledge about the neuroanatomic substrates of attention this assumption is inappropriate.

Sustained attention requires the active maintenance of a certain response under conditions with low environmental support (Manly, Anderson, Nimmo-Smith, Turner, Watson & Robertson, 2001). This skill has been anatomically linked to parietal lobe functioning (Posner, Walker, Friedrich & Rafal, 1984) and rostral midbrain structures are thought to be involved in sustained attention to environmental events (Mirsky, 1987). Studies of sustained attention abilities (Fennell, Eisenstadt, Bodiford, Redeiss, & Mickle, 1987; Loller, 1990; Tew et al., 1980) in children with HC have consistently reported deficits. However the studies are mainly confined to samples of children with SB.

Commonly, focussed and sustained attention abilities are assessed using the Continuous Performance Test (CPT) or cancellation tasks. Cooley and Morris (1990) argued that these tasks have been shown to be more sensitive to focussed attention abilities and also require rapid motor responses, which are often impaired in children with HC. Supporting the latter interpretation Fletcher, Brookshire, et al. (1996) found that poor performance on a pencil-and-paper cancellation task could be explained by the task requirement of rapid marking of stimuli. Furthermore, Tew et al. (1980) found that when

no time limit was imposed on cancellation tasks, the accuracy of children with HC was adequate. Similarly, using a CPT, Lollar (1990) found no difference in ability to sustain visual attention between SB children and controls. In contrast to Lollar's (1990) findings, children with HC made more omission and commission errors on a computer-based CPT (Fennell, et al., 1987). These errors were considered to be suggestive of both inattention and impulsivity (Fennell et al., 1987).

Another component of attention, shifting attention, has been studied in HC children. Shifting attention refers to the ability to switch attentional focus in a flexible and adaptive manner (Mirksy et al., 1991). Shifting attention may also be conceptualised as the ability to divide attention. For example, when there is a need to deal with multiple attentional demands in a relatively simultaneous fashion it requires efficient shifting of attentional focus. The prefrontal cortex, medial frontal cortex and cingulate cortex are involved in this ability (Mirsky, et al., 1991). Fletcher, Brookshire, et al. (1996) found that children with HC made more perseverative errors on the Wisconsin Card Sorting Task (WCST), suggesting deficits in shifting attention. Brewer et al. (2001) also found that children with HC exhibited deficits switching attention on the WCST. They pointed out that deficits in shifting attention might depend on the extent to which the midbrain has been damaged by HC.

According to Brewer et al. (2001) there is a need for more extensive study of attentional processes in children with early HC, particularly as anecdotal reports of focussed and sustained attentional deficiencies are common and many children with shunted HC are prescribed stimulant medication for these problems (Brewer et al., 2001).

To date no known study has correlated MRI data with performance on attentional

tasks in children with HC. Notwithstanding this, several authors have postulated the neuroanatomical regions affected by HC on the basis of test performance and neuropsychological theory. Brewer et al. (2001) interpreted difficulties with disengaging attention as consequence of posterior parietal area dysfunction. They suggested that this finding is consistent with other neuropsychological findings as well as the pathophysiology, i.e. greater thinning in posterior cortex than the anterior cortex (Brewer et al., 2001). Posterior brain regions are also implicated because of deficits noted in the ability to focus and shift attention (Fletcher et al., 2000). In contrast to these findings Wills (1993) concluded that deficits in focussed, divided and shifting attention, as well as those in planning, mental tracking or the inhibition of over learned responses, in children with HC, were consistent with anterior brain dysfunction.

1.56 Memory and Learning

As Anderson, Northam, et al. (2001) pointed out, one would expect memory deficits in children with HC because of the pathophysiology of HC. Ventricular enlargement is likely to damage the hippocampal and subcortical structures, which are important to learning and retrieval from long-term memory, respectively (Delis, Kramer, Kaplan & Ober, 1991). Despite this, studies of memory and learning in children with HC have produced inconsistent findings.

Verbal learning abilities in children with HC secondary to SB were investigated by Yeates, Enrile, Loss, Blumenstein and Delis (1995). Children with HC did not differ on first trial learning, however, they did acquire additional words more slowly across trials and consequently their total recall was lower. In addition, pronounced recency effect was demonstrated by poor delayed recall of the original list (Yeates, Enrile, Loss,

Blumenstein & Deis, 1995). Other studies have also found deficits in word list and paired associated learning (Cull & Wyke, 1984; Donders et al., 1991; Fletcher et al., 1992; Prigatano et al., 1983; Scott, Fletcher, Brookshire, Davidson, Landry, Bohan, Kramer, Brandt & Francis, 1998).

Immediate recall of sentences has been found to be poorer in children with HC than controls (Dennis, et al., 1987; Fennell et al., 1987; Fletcher, Brookshire, Bohan, et al., 1995), as has literal text recall of stories (Barnes & Dennis, 1998; Fletcher et al., 1992). In contrast, essentially normal verbal memory abilities on a selective reminding test (Donders et al., 1990) and immediate recall of short story elements have also been reported (Cull & Wyke, 1984; Donders et al., 1990).

Assessment of non-verbal memory abilities, in children with HC, has provided mixed findings. Prigatano et al. (1983) found deficits in tactile learning on the Fuld-Object Recognition Test. Furthermore, there have been findings of poor visual-spatial recall on non-verbal memory tests (Prigatano et al., 1983; Scott et al., 1998). However, visual-spatial memory tests are often pencil and paper tests, which require the type of motor skills that are often impaired in children with HC. One limitation of this study, however, is that they only included 10 children with HC and consequently the findings cannot be generalised.

Relatively consistent is the finding that children with HC have deficits in delayed recall and spontaneous retrieval of information (Cull & Wyke, 1984; Donders et al., 1991; Fletcher et al., 1992; Scott et al., 1998; Yeates et al., 1995). Generally, children with HC have been found to perform comparable to controls on recognition measures (Yeates et al., 1995). For example, intact recognition for nonsense shapes and faces has

been reported (Cull & Wyke, 1984), as has performance on non-verbal memory tests when given multiple-choice cues (Prigatano et al., 1983). However, Scott, Fletcher, Brookshire, Davidson, Landry, Bohan, Kramer, Brandt & Francis (1998) found non-verbal recognition was poorer in children with HC.

The impact of motor deficits on visual memory tests has been studied (Donders et al., 1991). Donders et al. (1991) used a motor free facial recognition task and found no deficits. Combined with Prigatano et al.'s (1983) finding that immediate memory for a copying task was impaired, it could be concluded that motor difficulties often present in HC explain poor performances on visual memory tasks. However, Scott et al. (1998) also found problems with HC children on both motor-free and design copying visual memory test, suggesting that motor problems do not solely explain visual memory deficits.

According to Anderson, Northam, et al. (2001) this pattern of relatively intact immediate recall and recognition of information but impaired spontaneous retrieval suggests difficulties in two domains. Firstly, children with HC have impaired development of organisational strategies required for efficient storage and retrieval over time as well as an over-reliance on rote recall. Secondly, children with HC have difficulty with spontaneous retrieval of information and these retrieval problems are related to the severity of subcortical white matter damage (Anderson, Northam, et al., 2001; Delis et al., 1991).

As well as organisational problems suggested by Anderson, Northam, et al. (2001), Cull and Wyke (1984) suggested that there was a lack of ability to use appropriate semantic strategies at the level of encoding. Supporting this interpretation,

other studies have found that children with HC perform more poorly on tasks of encoding and selective reminding (Buschke, 1974; Scott et al., 1998).

Findings have been inconsistent with regard to evidence for better verbal memory than visual memory. Fletcher, Francis, et al. (1992) found poorer non-verbal memory than verbal memory scores in children with HC. However, one year later the children were followed up and no difference between verbal and non-verbal memory abilities were found. Similarly, Scott et al. (1998) found no difference between verbal and spatial learning on a serial learning task.

Aetiological differences in memory abilities were examined by Scott et al., (1998) in children with shunted HC due to SB meningomyelocele, IVH and AS. Across a variety of memory tests no differences related to aetiology were found (Scott et al., 1998).

Fletcher et al. (2000) concluded the relationships of neuropathology to specific memory processes are relatively unexplored. To address this issue, they suggested that neuroimaging techniques combined with assessment of memory and learning would help. Furthermore, the impact of motor difficulties on visual learning and memory is still debated and studies are inconclusive with regard to evidence for non-verbal specific memory deficit (Anderson, Northam, et al., 2001; Fletcher et al., 2000).

1.57 Processing Speed

Despite the lack of specific research into processing speed in HC, several investigators have reported low scores on timed tasks that require visual tracking, eye-hand coordination skills and the physical manipulation of test stimuli (Anderson, Northam et al., 2001). Dennis et al. (1981) also reported reduced response speed on non-

motor tasks such as word-finding and slowed comprehension of complex grammar. Similarly, Anderson, Anderson, Northam, Jacobs and Mikiewicz (2002) found that children with HC were slower on a naming task (CNT), than controls. In another, non-motor task, Fletcher, Brookshire, et al. (1996) found HC groups were slower on three trials of the Stroop test, indicative of some slowing for automatic responses.

Klaas and colleagues (1999) studied interhemispheric transfer (IT) of visual, auditory, tactile and visuomotor information in a group of 13 children with HC and partial agenesis of the corpus callosum associated with AS or SB. Their results suggested that children with HC had a slower IT time, but they found neither the IT task nor the dichotic listening and tactile naming tasks provided clear evidence of difficulties in callosal transfer (Klaas et al., 1999). In contrast to Klaas et al.'s (1991) findings, Anderson, Northam, et al. (2001) suggested that cross-modal tasks that require transfer of visual information from posterior brain regions to naming areas in the left frontal regions, would be expected to be performed poorly by HC children because these tasks are reliant on the corpus callosum, which is abnormal in many HC children.

1.58 Executive Functions

Executive function is a difficult construct to define because it is closely related to attentional and memory skills (Erickson et al. 2001). Furthermore, researchers have long debated what executive functions comprise, although generally it is agreed to encompass the skills necessary for goal-directed behaviour such as the ability to anticipate, establish goals, plan, monitor results and use feedback (Stuss & Benson, 1986; Anderson, 1998). Several authors have postulated models of executive functions, the present study had adopted the model proposed by Anderson, Anderson, Northam, et al., 2001, which

followed from others' definitions (Lezak, 1993; Luria, 1973; Neisser, 1967; Shallice, 1990; Stuss, 1992; Walsh, 1978). Anderson, Northam, et al. (2001) conceptualised executive functions as:

> *Comprising three separable, but integrated, components: (a) attentional control-selective attention and sustained attention; (b) cognitive flexibility – working memory, attentional shift, self-monitoring, and conceptual transfer; and (c) goal setting – initiating, planning, problem solving and strategic behaviour* (Anderson, Northam, et al., 2001, p. 306).

The prefrontal cortex is thought to, at least in part, mediate executive functions (Erickson et al., 2001). Over the past couple of decades researchers have argued that although frontal lobes may play a vital role, the integrity of the entire brain is essential for efficient executive function (Anderson, 1998). This is due to the fact that the prefrontal region has extensive connections to all areas of the neocortex via cortico-cortical projections, as well as connections to the limbic system and subcortical structures (Anderson, Anderson, Northam, Jacobs & Catroppa, 2001). Lesions to any of these areas may effect the efferent or afferent connections to the prefrontal cortex thereby influencing executive functioning (Anderson, Anderson, et al., 2001). Deficits in executive skills in children with HC are likely to be due to damaged white matter tracts that carry information to and from the prefrontal cortex (Fletcher, Brookshire, et al., 1996).

Behavioural descriptions of children with HC commonly include references to increased distractibility, lack of judgement and reasoning, poor adaptive and goal-directed behaviour and poor follow-through skills (Fernell, Gillberg & von Wendt, 1991).

Despite these common anecdotal suggestions that children with HC have executive deficits, compared to other neuropsychological domains, relatively few studies have directly addressed the area (Anderson et al., 2002; Dise & Lohr, 1998; Fletcher, Brookshire, et al., 1996; Landry, Jordan & Fletcher, 1994; Mahone, Zabel, Levey, Verda & Kinsman, 2002; Snow, Prince, Souheaver, Ashcroft, Stefans & Edmonds, 1994).

Goal directed behaviour was investigated in young children with SB and HC, and IQ matched controls by Landry, Jordan and Fletcher (1994). They found that children with HC spent less time in goal directed behaviour and had impaired ability to sustain this behaviour to reach a goal. Landry et al. (1994) concluded that they were unable to distinguish between several possible reasons for this impaired performance. The explanations raised included problems in sequencing, attentional control, planning or a combination of these factors, or possibly another aspect of executive functioning altogether. Alternatively, these children's decreased mobility may have impaired their exploratory behaviour and cause and effect interactions with the environment, which are thought to be important to the development of goal directedness.

Deficits have also been found in the problem solving abilities of children with HC Dise & Lohr; 1998; Fletcher, Brookshire, et al., 1996). In the Fletcher, Brookshire and colleagues' (1996) study, children with HC solved fewer problems on the Tower of London (TOL), particularly on the first trial. However, there were no significant differences on planning time or rule breaks and with enough trials they were able to achieve solutions comparable to the controls (Fletcher, Brookshire, et al., 1996). Likewise, Anderson et al. (2002) found no significant differences between HC children and controls on time taken and extra attempts on TOL. Contrary to Fletcher, Brookshire

et al., Anderson et al. (2002) also found no significant difference in the number of correct items (Anderson et al., 2002). Brewer et al. (2001) postulated that difficulties, observed in their HC sample, on the WCST were due to deficits in novel problem solving. However, they also suggested these difficulties could be due to problems with disengaging attention or with learning procedures, so that they may require longer time to determine the sorting rule.

Fletcher, Brookshire et al. (1996) argue that although children with HC demonstrated problem-solving deficits on measures of executive function (TOL and WCST), their performance was not consistent with frontal lobe impairment of executive functions. Their interpretation was that deficits in problem solving were directly related to impaired sustained attention. Consistent with this explanation Goldberg and Costa (1981) and Rourke (1995) hypothesized that children with white matter disease have problems with complex integrative tasks and novel problem solving activities, that are dependent on the integrity of right posterior systems.

Another domain of executive functioning, abstraction abilities, was researched by Snow, Prince, Souheaver, Ashcraft, Stefans and Edmonds (1994). They compared this domain in different subgroups of children with SB and found mild deficits. They concluded that deficits were indicative of possible anterior system dysfunction.

Dise and Lohr (1998) examined conceptual reasoning, problem solving, mental flexibility and mental efficiency in 36 adolescents with SB, terming these skills 'higher mental functions.' Despite having IQ within normal limits, these SB children tended to exhibit difficulty in at least one component of these 'higher mental functions'. Similarly, using the Contingency Naming Test (CNT) Anderson et al. (2002) found HC children to

be slower but to make more self-corrections, suggesting intact self-regulation but decreased mental efficiency.

Recently, two studies have examined behavioural aspects of executive functions utilising the Behavioural Rating Inventory of Executive Function (BRIEF) (Anderson et al., 2002; Mahone et al., 2002). The Anderson et al. (2002) HC sample consisted of 45 children with either congenital or acquired aetiologies. On composites of 'metacognition', 'working memory' and 'initiate', HC children's scores were significantly higher than those of control subjects, indicating poorer functioning. Mahone et al. (2002) employed a different methodology to study behavioural aspects of executive functions. They compared parents' and children's reports on the BRIEF. Their results indicated that children considered themselves to have significantly more problems than their parents did on the scales comprising the behavioural regulation index. On the other hand, parents rated their children to have more problems on the 'metacognition index' compared to the 'behavioural regulation index.'

1.6 Research Limitations and the Current Study

Early research in children with HC, as with many other neurological disorders, focussed on general domains of neuropsychological function such as intelligence, verbal and visual-spatial skills (Fletcher et al., 2000). However, because HC is not a single disease entity and neuropathology can vary, neuropsychological outcomes can also differ within the clinical population. Although there may be some characteristic features of HC, it is unlikely a single profile or unique pattern of neuropsychological strengths and weaknesses will sufficiently describe a child with HC (Anderson, Northam, et al., 2001).

For this reason, Fletcher et al. (2000) advocate that it is useful to study neuropsychological function in HC in terms of a set of outcome domains. However, the variability within these domains should be considered in relation to sampling and to factors that are characteristic of the hydrocephalic condition such as the aetiology, neuropathology, treatment and medical sequelae of HC (Fletcher et al., 2000).

The literature is limited, in most part, to children with HC secondary to SB (Anderson, Northam, et al., 2001; Fletcher et al., 2000; Jacobs et al., 2001;Yeates et al., 2003). Children with SB are reported to have more severe cognitive deficits as a consequence of the major CNS abnormalities associated with the disorder (Anderson, Northam, et al., 2001; Erickson et al, 2001; Wills, 1993). Furthermore, most studies involve small samples (Cull & Wyke, 1984; Hommet et al., 1999; Friedrich et al., 1991; Jacobs et al., 2001; Klass et al., 1999). Studies involving relatively large HC samples (Brookshire et al., 1995; Brewer et al., 2001; Dennis et al., 1981, 1986; Donders et al., 1991; Fletcher, Brookshire, et al., 1996; Fletcher, Bohan, et al.; 1996) have included children with arrested and unshunted HC, and the total number of children with shunted HC is often less than 20. Group numbers are further reduced when the HC sample is divided into aetiological groups for analysis (Fletcher et al., 1992; Hetherington & Dennis, 1999) or into age groups (Dennis et al., 1987). These small group sizes restrict the power of statistical comparisons and increase the probability of Type II errors (the likelihood of concluding that there is no significant difference between the groups when in fact there is) (Schmidt, 1996).

According to Baron and Goldberger (1993), further limitations of the research have been selection of tests with inappropriate norms, unbalanced control and

experimental groups and failures to dissociate the requirements inherent to successful mastery of task. Conclusions are therefore limited by inconsistent findings, difficulty in interpreting test performances and methodological problems (Fletcher et al., 2000).

The literature is also limited by the paucity of current research about the cognitive sequelae of obstructive and communicating HC. Although the literature suggests that the site of obstruction (intraventricular or extraventricular) does not affect intellectual outcome in any systematic way (Dennis et al., 1981; Fletcher, Bohan, et al., 1992; Riva et al., 1994), congenital aetiologies have been associated with more severe cognitive deficits than acquired HC. However no known studies to date have specifically addressed attentional and executive functions, or processing speed in the different types of HC.

In general, studies of heterogeneous samples of HC have reported deficits in attention and executive functions (Anderson et al., 2002; Brewer et al., 2001; Dise & Lohr, 1998; Fletcher, Brookshire, et al., 1996; Landry et al., 1994; Mahone et al., 2002; Snow et al., 1994). However, conclusions are impeded by disagreement about the neuropsychological explanation for these deficits. It is not surprising that several authors have suggested that more systematic studies of attention, processing speed and executive functions of children with HC are required, incorporating large samples and MRI techniques to examine how specific deficits are related to neuropathology (Anderson, Northam, et al., 2001; Fletcher et al., 2001; Erickson et al., 2001).

Numerous questions are still to be answered due to the mentioned limitations of previous literature. Some of these questions are: What specific attention, processing speed and executive functions deficits do children with HC exhibit? Do children with obstructive and communicating differ in terms of their attention, processing speed and

executive function? Do children with obstructive and communicating HC differ in terms of the degree and nature of neuropathology?

This study was designed to address some of these unanswered questions and limitations of previous literature with a broad aim to investigate the neuropsychological functioning of children with early treated HC. The sample included children aged 7 to 15 years recruited over a two-year period (2000-2001).

The theoretical implications of this study are that it may provide better understanding about brain behaviour relationships. For example, if the two HC groups have distinctly different cognitive profiles, it may suggest that this is a consequence of different pathophysiology. Alternatively, no difference between the groups may suggest that regardless of the mechanism of HC, the cognitive outcome and pathophysiology are similar across groups. The clinical implications of this study are that it may provide more prognostic information about the attention, processing speed and executive functions of children with HC. These findings will assist parents, teachers and other clinicians in their management of children with HC.

1.8 Aims and Hypotheses

The current study aims to extend the current research in attention, processing speed and executive functions in children with early treated HC. In addition, it aims to document the cognitive profiles of children with early treated communicating and obstructive hydrocephalus and utilize qualitative analysis of neuroradiological data to determine if the site of neurological injury is related to any pattern of deficits in the above cognitive domains.

It is apparent that no study has examined neuropathological differences between obstructive and communicating HC. Due to the timing and mechanism involved in obstructive HC it is hypothesized that this group will display more damage to midline structures such as dysgensis of the corpus callosum, which is reported to be more common in congenital aetiologies (Hannay, 2000). Given this assumption it is hypothesised that:

i. The obstructive HC group will perform more poorly on tasks that measure focussed, sustained and divided/shifting attention, than the communicating HC group.

ii. The obstructive HC group will be significantly slower than the communicating HC group on tasks of processing speed.

iii. The obstructive HC group will do more poorly on executive function measures (planning, organisation, problem solving, mental flexibility, self-regulation) than the communicating HC group.

iv. The obstructive HC and communicating HC groups will differ on qualitative reports of the degree of white matter injury and dysgenesis of the corpus callosum, such that the obstructive HC group will display more pronounced injury.

CHAPTER 2: METHOD

2.1 Participants

The current archival study included 116 children with a mean age of 10.42 years (*SD* = 2.43 years). The sample was comprised of 60 males and 56 females, divided across two clinical groups (obstructive and communicating HC) and a control group.

2.11 Clinical Participants

2.111 Obstructive HC

Forty-three participants (mean age =10.07 years, *SD* =2.45 years) had the obstructive form of HC of whom 25 were male and 18 were female. The obstructive HC group was comprised of 24 participants with AS, one participant with Arnold Chiari Malformation, three participants with DWS and 14 participants with a cyst. Of the participants with cysts their location was as follows; seven participants had posterior fossa cysts, two participants had lateral ventricle cysts, one participant had a third ventricle cysts and four participants had cysts in other locations.

2.112 Communicating HC

Thirty-one children had the communicating form of HC. This group (mean age = 10.65 years, *SD* = 2.40 years) consisted of 18 males and 13 females. The communicating HC group was entirely comprised of children with IVH.

2.12 Inclusion Criteria for Clinical Participants

Inclusion in the clinical groups required that the child was diagnosed with HC, based on magnetic resonance imaging (MRI), medical record review and neurological examination, and treated during their first year of life at the Royal Children's Hospital

(RCH) Melbourne, Victoria, or at the Sydney Children's Hospital (SCH), New South Wales, Australia. The children with HC were grouped into communicating or obstructive HC. Most children had been classified into these HC subtypes at the time of diagnosis, however this classification was confirmed by an independent neurosurgeon who viewed the child's medical records and MRI scans. Children with primary neurological conditions not necessarily associated with HC, eg, stroke, trauma, tumour or a history of cerebral infection such as meningitis, encephalitis, spina bifida or an uncontrolled seizure disorder, were excluded. Five participants were excluded because their WISC-III full-scale intelligence quotients were below a standard score of 70. Four children were excluded because they were less than seven years old. Fifty clinical participants (31 obstructive HC and 19 communicating HC) had a MRI scan conducted at the RCH or the SCH.

2.13 Control Participants

The control group was comprised of 42 (17 males and 25 females) sibling controls (mean age = 10.62 years, *SD* = 2.44 years). Control participants were siblings of the children in the HC groups. Inclusion criteria were that the child had no history of cerebral nervous system infection, disease or trauma, or significant illness.

2.2 Materials

A battery of neuropsychological tests assessing general intellectual functioning, visual perception, attention, processing speed and problem solving/executive functioning skills (organization, mental flexibility, planning and concept formation) was administered. Parents or guardians were asked to complete a questionnaire regarding

demographics, family background, medical history, parental concerns, developmental milestones, and educational history (see Appendix C).

2.21 Intellectual Functioning

The Australian adaptation of the Wechsler Intelligence Scale for Children - 3rd Edition (WISC-III; Wechsler, 1991) was used to assess intellectual functioning. All of the participants were administered 11 of the subtests (full version plus one optional test, Digit Span). As well as the Full Scale Intelligence Quotient (FIQ), the Verbal (VIQ) and Performance Intelligence Quotient (PIQ) were calculated according to test protocol (Wechsler, 1991).

2.22 Attention

Attention is not a single construct and is comprised of several components. The attentional processes that were assessed in the current study were focussed, sustained and divided attention. Selected subtests from the Test of Everyday Attention for Children (TEA-Ch; Manly, Robertson, Anderson & Nimmo-Smith, 1999) were employed to assess these attentional constructs. The TEA-Ch consists of nine subtests measuring the following domains of attention: visual selective, sustained auditory, shifting, or divided (auditory and visual). The SkySearch, Score! and SkySearch DT subtests were utilised.

2.221 Focussed Attention

SkySearch assesses visual focussed attention. In this subtest children were shown a picture in a cue book of identical and non-identical paired 'spaceships', and instructed that another sheet would follow which also displays identical (target items) and non-identical pairs (distractor items) of spaceships. Next the children were given a laminated A3 sheet with rows of paired spaceships, mostly distractor items (20 target 'identical'

pairs and 108 'non-identical' pairs). The children were then instructed to circle the identical pairs of spaceships as quickly as possible and when they had finished to tick a box located at the lower right corner. The number of pairs correctly circled was recorded, as was the number of errors, and the time taken to completion. The overall score is the number of pairs correctly circled per second. For this study the number of targets identified was converted to a standard score and employed as the measure of selective attention. Prior to completing the SkySearch main test the children were given a practice A4 sheet to ensure they had comprehended the instructions.

To control for differences that are attributable to motor speed rather than visual selection, the children were all administered a motor control version of SkySearch. SkySearch-Motor Control requires the child to circle all of the identical pairs of spaceships as quickly as they can on an A3 sheet filled only with identical pairs (Manly et al., 1999). The score attained is then used in a formula described in the manual.

2.222 Sustained Attention

Score! is designed to test auditory sustained attention. The test required the child to listen to an audiotape over 10 different trial items, with the child instructed to silently count the number of tones (without using their fingers) for each trial. Initially, a bell sounds, followed by a number of tones, until another bell sounds. At the end of the trial the child was required to verbalise the number of tones between the first and second bells. In each trial item, between 9 and 15 identical tones of 345ms are presented, separated by interstimulus intervals of variable duration (between 500 and 5000ms). Two practice trials were given. The number of correct, incorrect and omitted responses were recorded. The score used was the number of correct target responses and this was converted to a

standard score.

2.223 Shifting/Divided Attention

SkySearch Dual Task (DT) is a test of divided attention skills. It requires the child to complete a task similar to that of SkySearch, which only differed in the location of the targets, and to simultaneously and silently count the number of tones presented within each item of an auditory counting task. In the auditory counting task the tones are the same as those in Score! however, this time the tones were regularly paced at one per second. The child was given practice items. The test ended and timing stopped when the child completed the visual search component. The numbers of correct, incorrect and omitted responses for each of the two elements of this task were recorded, and a final DT score was calculated from an equation described in the manual (Manly et al., 1999).

2.23 Processing Speed

The Rapid Automatised Naming Test (RAN; Denckla & Rudel, 1974) is a measure of processing speed. The RAN employed in the current study consisted of the RAN colour chart. This chart was presented on a 27.5cm X 35.5cm white paper with colours arranged in five rows of 10 items (Neuhaus, Foorman, Francis & Carlson, 2001).

Before the task commenced it was ensured that the child could identify the stimuli accurately. The child was instructed to name each colour as quickly as they could. Each test was scored for the time it took to name each of the colours on the chart from left-to-right and top-to-bottom. The time it took to complete the chart was recorded and the child's responses including any uncorrected errors were noted. The spontaneous self-corrections were also recorded. This provided three parameters; time taken, errors and

self-corrections. Raw scores of time taken for the RAN colour subtest were converted into z scores using Wiig, Zureich and Chan's(2000) norms.

Coding is one of the performance scale subtests of the WISC-III. The child is required to copy simple symbols that are paired with simple geometric shapes or numbers. Using this key the child copies the symbol in its corresponding shape or under its number. The child has to complete as many symbols as he/she can in a 120-second time limit. The raw score is the number of symbols the child completed in 120 seconds. The raw score was changed into an age scaled score, which was then converted into a z score.

The Contingency Naming Test (CNT) trial 1 and 2 time raw scores were combined to give a measure of rapid word retrieval and information processing time (Trial 1 and 2 of the CNT are described in detail in *section 2.234*). The raw score was converted to a z score using Anderson, Anderson, Northam and Taylor (2000) norms.

2.24 Executive Functions

Measures of executive function were chosen to assess two of the executive function subdomains (goal setting and cognitive flexibility). The third and fourth subdomain attentional control and information processing are incorporated in the attention and information processing speed section. In addition, a behavioural measure of executive function was used.

2.241 Cognitive Flexibility, Self-Monitoring/Regulation

The CNT is a measure of speed of name retrieval and mental flexibility (mental set shifting), however it is also thought to capture elements of self-regulation, inhibition, and processing speed (Anderson, Anderson, et al., 2001; Anderson et al., 2000). The task

involves the child pointing to a series of coloured shapes, each containing a smaller shape inside. The inside and the outside shapes are the same for some of the stimuli and different for others (Taylor, Hack & Klein, 1998).

The child was instructed to name only the colours in Trial 1 and only the outside shapes in Trial 2. In Trial 3, a simple switching task, the naming rule (contingency) required the child to name the colour if the outside and inside shapes match, and to name the shape if they do not match. In trial 4, a complex switching task, the child was told to apply the same contingency as trial 3 except when a backward arrow appears above the shape. In this case the child is told to reverse the contingency and name the colour instead of the shape and vice versa (Taylor et al., 1998). In each of the trials the child is given up to five training trials to learn the rule. When an errorless performance on a training trial occurred or five training trials given, the child was administered the test trial. Variables include the time taken, number of errors, and number of self-corrections recorded in the simple and complex switching tasks (Anderson et al., 2002).

The current study utilized two scores obtained from Trial 4: time taken to complete the task and efficiency score. The efficiency score measures speed and accuracy, taking into account both time and errors made. The formula for the efficiency score is *efficiency* = [(1/time) / SQRT(errors+1)] x 100 (Anderson et al., 2000). Trial 4 was used because it is the most cognitively demanding trial and is considered to be a measure of mental/cognitive flexibility (Anderson et al., 2000). Raw scores were converted to z-scores using Anderson et al.'s (2000) norms.

2.242 Organization

The Rey Complex Figure Test (RCFT) (Rey, 1964) is a measure of sensory-

motor, perceptual, planning and organizational abilities (Anderson, Anderson & Garth, 2001). The children were timed whilst copying the RCF as accurately as possible. The figure is a complex geometric design consisting of a large rectangle, vertical and horizontal centrelines, two diagonals, and various external attachments and internal sections of the large rectangle. The accuracy score (score range: 0 to 36) is a reflection of the number and accuracy of elements of the figure copied (according to scoring protocol of Osterreith, 1944).

The Rey Complex Figure - Organisational Strategy Score (RCF-OSS) developed by Anderson, Anderson & Garth (2001) was also used in the current study. RCF-OSS reflects the child's level of organisational and strategic development where level 1 is unrecognisable, level 2 is poor organization, level 3 random organization, level 4 piecemeal/fragmented organization, level 5 part configural organization, level 6 conceptual organization and level 7 excellent organization. In the current study level 1, 2 and 3 were collapsed into one variable labelled 'poor organization,' level 4 was labelled 'fragmented organization', level 5 was 'part configural', and level 6 and 7 were collapsed into a 'conceptual' variable.

2.243 Planning and Problem Solving

The Tower of London (TOL) (Anderson, Anderson & Lajoie, 1995; Shallice, 1982) is primarily a test of planning ability, but it is also considered to be sensitive to impairments in working memory, problem solving and mental flexibility (Lezak, 1995). In this study the TOL was administered and scored according to the protocol outlined by Anderson et al. (1995). The task required the children to move three coloured balls on three posts of differing heights, in a specific number of moves, so that they match the

configuration presented on a stimulus card. The TOL comprises 12 items of increasing difficulty with the number of moves allowed to solve the problem varying from 2 to 5 depending on the difficulty. The children were allowed as many attempts as possible within a 60 second time limit and failed attempts were penalised in the overall summary score (Anderson et al., 1995). The raw score was utilized which incorporates the following parameters: number of failed attempts and total time taken in seconds to complete the trials. As standard scores were not available for children over 13 years of age, the raw score was employed.

2.244 Working Memory

The backwards component of the Digits Span subtest is thought to be a measure of working memory (Lezak, 1995). Digits backwards is the second trial of the Digit Span subtest. A string of digits (ranging from 2 to 8 digits in length) was read to the child at a rate of one digit per second. The child was instructed to repeat the sequence of digits in the reverse order (i.e. 6-3-8-1 would become 1-8-3-6). Each digit sequence is presented in two trials and the test was discontinued when the child failed both trials or repeated the maximum (eight) digits backwards. The raw score for the number of correct trials backwards was converted to a z score using Gardner's (1981) norms.

2.245 Behavioural Aspects of Executive Function

The Behaviour Rating Inventory of Executive Functioning (BRIEF; Gioia, et al., 2000) is an 86-item questionnaire that assesses behavioural aspects of executive function. In this study the parent version was administered. The 86 items provide eight theoretically and empirically derived scales (inhibit, shift, emotional control, initiation, working memory, planning/organization, organization of materials and behavioral

monitoring) two indices (Metacognitive and Behavioral Regulation) and one composite score (Global Executive Composite) (Anderson et al., 2002). The Behavioral Regulation Index measures a child's capacity to regulate behavioural and emotional responses and to adapt behaviour according to the situation. The Metacognition Index provides an indication of the child's ability to initiate, plan, organize, hold information in working memory and sustain actions necessary to complete a problem in a variety of daily settings. The summary measure, the Global Executive Composite, incorporates all 8 clinical scales. For each clinical scale raw scores were converted to T scores ($M = 50$, $SD = 10$). Scores more than 1.5 standard deviations above the mean were considered to be abnormally elevated.

2.25 Socio-Economic Status

The socio-economic status (SES) of the participants was assessed using Daniel's Scale of Occupational Prestige (Daniel, 1983). It is a self-report measure of the parental occupation that consists of a 7-point rating scale in which 1 reflects high SES and 7 refers to low SES.

2.26 Magnetic Resonance Imaging (MRI)

MRI scans were conducted on 51 clinical participants and interpreted by two independent radiologists at the RCH to quantify the pathology. A standard protocol was followed for rating the MRI scans and involved the assessment of ventricular size, cortical mantle, midline structures, and white matter abnormalities. The following MRI sequences were used; T1 weighted 3D volumetric dataset in the case of the ventricles, and T2 weighted dual echo in the case of grey/white matter ratio.

Two radiologists completed a MRI report checklist that included the following

items: site of the shunt, presence of calvarian thickening (yes or no), ventricular size (normal, dilated, slit or entrapped), subdural collection (yes or no), corpus callosum signal (normal, focal abnormality, diffuse abnormality), rostrum, genu, body, and splenium (normal, thick, thin), white matter loss (deep or subcortical) severity of white matter loss (mild, moderate, severe). Refer to Appendix C for an example.

There was no systematic reason for the 23 children not having MRI imaging data. There were several reasons that MRI data was not able to be included in the current study such as families declining MRI, difficulty in arranging MRI, and movement artefact precluding analysis. Of those who did not have an MRI a similar proportion of participants were from each HC group, 12 from the obstructive HC group and 11 from the communicating HC group. On simple analysis there was no difference between their IQ and the HC children who had MRI scans conducted.

2.3 Procedure

Neurosurgical records were accessed at the RCH and SCH to obtain the names of children who had been diagnosed and treated for HC in the first year of life between 1985 and 1994, inclusive. From these patients, those between the ages of 7 and 15 (during March 2000 and October 2001) were considered for inclusion. The medical records of these children were then requested and reviewed for recruitment according to selection criteria. Letters of invitation were sent to families of the eligible children, followed by a phone call by the project coordinator. Families who consented to participate in the project, and who had a sibling in the eligible age range, were asked if this child was also willing to take part in the study.

Written informed consent for the children and the parent(s) to participate in the study was obtained. The parent(s) were then asked to complete a general questionnaire (Appendix D) and the BRIEF. The assessment phase was conducted over a two-year period between 1999 and 2000. Each assessment took approximately three to four hours; including one or two 15-30 minute breaks. The tests utilized in the current study were embedded in a larger neuropsychological test battery. The order of battery administration was as follows: WISC-III, Wide Range Achievement Task- 3rd Edition, Speed of Handwriting test, Tea-Ch subtests SkySearch, SkySearch Motor control, Score! and Sky Search DT, California Verbal Learning Test- Children's Version (CVLT-C), Visual Memory Index, Matrix Reasoning, CVLT-delay, RAN, RCFT Copy, Controlled Oral Word Association Test, CNT, TOL, RCFT delayed recall.

When it was possible children in the clinical group had a MRI scan conducted on the same day as their neuropsychological testing. In some cases MRI scans could not be conducted on the same day as neuropsychological testing so these children had a scan within a month of the their cognitive assessment.

A neuropsychological feedback report was provided for each child. A letter of thanks was also sent to each family, including a letter requesting permission to place a copy of their child's results in the medical records for future reference. Raw data was collated and entered into the statistical computer program SPSS version 11.0 for Windows.

2.4 Statistical Analyses

All metric data scores from the TEA-Ch, RAN, CNT, TOL, RCF, WISC-III coding and digit span were converted to z scores using the most appropriate norms available. WISC-III intelligence quotients remained as standardised scores. The BRIEF indices were converted to T-scores.

Between group (obstructive HC, communicating HC and control) differences on WISC-III IQ Scales, TOL component scores, SES ratings, and age of participants were analysed using Analysis of Variance (ANOVA). Between group differences, on each cognitive domain (attention, processing speed and executive functions), were assessed with Multivariate Analysis of Variance (MANOVA) (see Table 1).

Pillai's Trace criterion was employed as the statistic to assess the significance of the main effects and interactions. Pillai's Trace is more robust than Wilks' Lambda, Hotellings Trace and Troys's gcr criterion to violations of the assumptions of MANOVA (Tabachnik & Fidell, 1996). Post-hoc analysis of group comparisons were conducted using Tukey's Honestly Significant Difference (HSD) procedure to control for the number of pairwise comparisons, keeping the error rate at an overall alpha level at 0.05.

Group differences for two categorical variables were explored using Chi-Square analysis. Categorical data was obtained for RCF-OSS, errors and self-corrections made on the RAN-colours subtest, and the qualitative analysis of the MRI data (see Table 1). Differences between gender and medical variables were also assessed using Chi-square analyses. An alpha level of 0.05 was used for all tests.

Table 1 displays the cognitive constructs that that were analysed using MANOVA. It also shows the dependent variables included in each MANOVA.

Table 1

Cognitive Domains and Test Variables Analysed Using MANOVA

Cognitive Construct	*Test Variable*
Attention	
Focussed	TEA-Ch SkySearch
Sustained	TEA-Ch Score!
Shifting/Divided	TEA-Ch SkySearchDT
Processing Speed	
	RAN-Colours Subtests
	WISC-III Coding
	CNT-Trial 1 & 2 time taken
Executive Function	
Cognitive Flexibility	CNT-Trial 4 Efficiency
Problem Solving	TOL-Summary Score
Planning	RCF-Accuracy Score
Working Memory	Digits Backwards
Behavioural Aspects of Executive Function	BRIEF Scales

CHAPTER 3: RESULTS

3.1 Demographic Characteristics of Participants

Table 2 presents the number of males and females in each participant group. It also displays the mean (and standard deviation) age and socio-economic status (SES) ranking, for the obstructive HC, communicating HC and control groups.

Table 2

Demographic Variables for Clinical and Control Participants

Demographics	Participant Group		
	Obstructive Hydrocephalus	Communicating Hydrocephalus	Control
n	43	31	42
Gender			
Male, *n* (%)	25 (58.1)	18 (58.1)	17 (40.5)
Female, *n* (%)	18 (41.9)	13 (41.9)	25 (59.5)
Age (Years)			
M	10.07	10.65	10.62
SD	2.45	2.40	2.44
SES			
M	4.23	4.56	3.99
SD	1.27	1.19	1.20

Note: Means with the same superscript are significantly different at $p<0.05$

Separate ANOVAs revealed no significant difference between the obstructive HC, communicating HC and control groups on mean age [F (2,113) = 0.72, p>0.05] or SES [F (2, 111) =1.85 , p>0.05]. Despite a higher proportion of females in the control group, Chi-square analysis found no significant group differences for gender [χ^2 (2, N = 116) = 3.34, p>0.05].

3.2 Medical Variables of Clinical Participants

Table 3 shows the proportion of children in the HC groups with co-morbid conditions.

Table 3

Co-morbid Conditions in the Obstructive and Communicating Hydrocephalus Groups

	Obstructive Hydrocephalus (n = 43) (%)	Communicating Hydrocephalus (n = 34) (%)
Cerebral Palsy	11.60	19.4
Seizures	23.30	35.50
PVL	9.30	19.4

Note: Means with the same superscript are significantly different at p<0.05

PVL= Periventricular White Matter Leukomalacia

Chi-square analysis revealed that the obstructive HC and communicating HC group did not significantly differ in the incidence of cerebral palsy [χ^2 (1, N = 74) = 0.85, p>0.05], seizures [χ^2 (1, N = 74) = 1.32, p>0.05], or PVL [χ^2 (1, N = 74) = 1.56, p>0.05]

3.21 Shunt Locations

Table 4 shows the frequency of shunt location for both the obstructive and communicating HC groups.

Table 4

Frequency of Shunt Location in the Hydrocephalic Groups

Shunt Location	Obstructive Hydrocephalus ($n = 43$)	Communicating Hydrocephalus ($n = 31$)
Right Ventricle, *n (%)*	28 (65.11)	28 (90.32)
Left Ventricle, *n (%)*	4 (9.30)	2 (4.65)
Bilateral, *n (%)*	4 (9.30)	1 (3.23)
Right Ventricle and Posterior Fossa, *n (%)*	5 (11.63)	0 (0.00)
Left Ventricle and Posterior Fossa, *n (%)*	1 (2.33)	0 (0.00)
ETV only, *n (%)*	1 (2.33)	0 (0.00)

All except one participant had been treated by shunt implantation, three participants had a shunt inserted followed by an endoscopic third ventriculostomy (ETV), and one child had only undergone ETV (see Table 4).

Table 4 shows that shunt location was most commonly in the right ventricle for both HC subtypes (28 in the obstructive HC group and 28 in the communicating HC group). Four children in the obstructive group had left ventricle shunt location, four children had bilateral, five children had right ventricle and posterior fossa, one child had left ventricle and posterior fossa, and one had an ETV only. Of the 31 children in the

communicating group, only two participants had a left ventricle shunt, and one participant had a bilateral shunt placement (see Table 4).

3.22 Shunt Revisions and Complications

Table 5 demonstrates the frequency of shunt revisions and complications in the obstructive HC and communicating HC groups.

Table 5

Frequency and Type of Shunt Revisions and Complications

		Obstructive Hydrocephalus (n = 43)	Communicating Hydrocephalus (n = 31)
Shunt Revision			
	Routine	0	1
	Infection	2	1
	Blockage	16	11
Infection and Blockage		4	1
Infection and Displacement		0	1
Disconnection		3	1
Disconnection and Blockage		1	2
Other		1	0
Slit Ventricle		0	2
No Shunt Revisions or Complications		16	11

Note: Means with the same superscript are significantly different at $p<0.05$

Chi-square analysis revealed that the obstructive HC and communicating HC groups did not significantly differ in the frequency of shunt revisions and complications [$\chi^2(10, N = 74) = 9.10, p>0.05$]. The obstructive HC group 18 participants underwent shunt revisions, four participants had shunt infections and blockages, and four participants had a shunt disconnection and/or blockage. Sixteen participants had not required a shunt revision or experienced any complications.

In the communicating HC group 13 participants had shunt revisions (12 due to blockage or infection and one was routine). One participant in this group had acquired an infection and blockage of their shunt and four participants' shunts had become disconnected and/or blocked. Eleven participants had not had a shunt revision or complication (see Table 5).

3.3 General Intellectual Ability

Table 6 presents means and standard deviations of the WISC-III scales for the obstructive HC, communicating HC and control groups

Table 6

Means and Standard Deviations of WISC-III Scales for Clinical and Control Participants

		Participant Group		
		Obstructive Hydrocephalus	Communicating Hydrocephalus	Control
WISC-III Scale		(n = 43)	(n = 31)	(n = 42)
FIQ				
	M	94.42[a]	97.42[b]	106.90[a,b]
	SD	10.72	13.99	10.42
VIQ				
	M	97.16[a]	101.94	106.48[a]
	SD	11.42	12.99	11.81
PIQ				
	M	92.09[a]	93.55[b]	106.43[a,b]
	SD	11.91	18.34	9.96

Note: Means with the same superscript are significantly different at $p<0.05$

ANOVAs revealed significant group differences in FIQ [F (2, 113) = 13.20, $p<0.001$], VIQ [F (2, 113) = 6.41, $p<0.01$] and PIQ [F (2, 113) = 14.29, $p<0.001$]. Post hoc comparisons found that the obstructive HC and communicating HC groups had

significantly lower mean FIQ's than the control group ($p<0.001$ and $p<0.01$, respectively). Similarly, post hoc analyses revealed that the control group had a significantly higher mean PIQ than the obstructive HC and communicating HC groups ($p<0.001$). With regard to VIQ, the control group had a significantly higher mean VIQ than the obstructive HC. However, the control group and the communicating HC group did not differ on mean VIQ. The obstructive HC and communicating HC groups did not differ from each other on any of these variables.

3.4 Attention

Table 7 presents *z* scores (means and standard deviations) of the TEA-Ch subtests SkySearch, Score! and SkySearch DT, for the obstructive HC, communicating HC and control groups.

Table 7

Z Score Means and Standard Deviations of TEA-Ch subtests for Clinical and Control Participants

	Participant Group					
	Obstructive Hydrocephalus (n = 42)		Communicating Hydrocephalus (n = 31)		Control (n = 42)	
	M	*SD*	*M*	*SD*	*M*	*SD*
Focussed						
SkySearch Targets	0.07[a]	1.09	0.24	1.16	0.76[a]	0.56
Sustained						
Score!	-0.64[a]	1.10	-0.73[b]	1.10	-0.02[a,b]	0.92
Shifting/Divided						
SkySearch DT	-1.53[a]	1.18	-1.57[b]	1.28	-0.94[a,b]	0.83

Note: Means with the same superscript are significantly different at $p<0.05$

The group main effect on the attention MANOVA was significant [Pillai's Trace = 0.16, $F(6,222) = 3.24$, $p<0.01$, $\eta^2 = 0.08$]. Follow up univariate tests revealed that the

groups differed on SkySearch targets [F (2, 112) = 5.87, p<0.01, η^2 =0.10], Score! [F (2, 112) = 5.50, p<0.01, η^2 = 0.09] and SkySearch DT [F (2, 112) = 4.17, p<0.05 η^2 = 0.07].

Post hoc analyses revealed that the obstructive HC group identified significantly fewer targets on SkySearch than the control group (p<0.05). On the Score! subtest the control group performed significantly better than the obstructive HC and communicating HC groups (p<0.05). The control group performed significantly better on SkySearch DT than both HC groups (p<0.05). No other group differences were found.

In summary, both HC groups had more difficulty than controls on tasks of sustained (Score!) and divided (SkySearch DT) attention. Only the obstructive HC group performed more poorly than controls on a focussed attention task. The HC groups performed similarly across the domains with no between group differences found.

3.5 Processing Speed

Table 8 presents the *z* score means and standard deviations of the WISC-III Coding subtest, the time taken in seconds for RAN-Colours subtest, and time taken on trials 1 and 2 combined on the CNT subtest, for each participant group.

Table 8

Z Score Means and Standard Deviations of Coding, RAN – Colours Subtest and CNT Trial 1 and 2 for Clinical and Control Groups

	Participant Group					
	Obstructive Hydrocephalus ($n = 42$)		Communicating Hydrocephalus ($n = 31$)		Control ($n = 42$)	
	M	*SD*	*M*	*SD*	*M*	*SD*
Coding	-0.60[a]	1.10	-0.38[b]	1.02	0.44[a,b]	1.08
RAN -Colours	1.87	1.34	2.15[b]	1.50	1.27[a]	1.14
CNT- Trials 1 and 2	0.21[a]	1.32	0.40[b]	1.35	-0.47[a,b]	1.11

Note: Means with the same superscript are significantly different at $p<0.05$

A MANOVA revealed a significant difference in the performance of the groups on tests of processing speed [Pillai's Trace = 0 .20, F (6, 222) = 4.10, $p<0.01$, $\eta^2 = 0.10$].

Follow up univariate tests revealed that Coding [F (2, 112) = 11.18, $p<0.01$, $\eta^2 = 0.17$], RAN-Colours [F (2, 112) = 4.33, $p<0.05$, $\eta^2 = 0.07$], and time taken on CNT trial 1 and 2 [F (2, 112) = 5.09, $p<0.01$, $\eta^2 = 0.08$] differed significantly between the groups.

Post hoc comparisons found a significant difference between the control and obstructive HC group ($p<0.001$) and between the control and communicating HC groups ($p < 0.01$) on Coding. It was also revealed that the control group was faster at completing the RAN-colours subtest than the communicating HC group ($p<0.05$), but not than the obstructive HC group ($p>0.05$). In addition, post hoc analyses found that the control group took less time to complete the first two trials on the CNT than both HC groups *($p<0.05$)*. No other group differences were significant.

Table 9 presents the percentage of errors and self-corrections made by the obstructive HC, communicating HC and the control group on the RAN colours subtest.

Table 9

Percentage of Errors and Self-Corrections on the RAN Colours Subtest for Clinical and Control Groups

	Participant Group		
	Obstructive Hydrocephalus	Communicating Hydrocephalus	Control
RAN Colours	(%)	(%)	(%)
Errors			
No Errors	86.0	93.5	83.3
< 3 Errors	100.0	100.0	97.6
>2 Errors	0.0	0.0	2.4
Self Corrections			
Yes	51.2	71.0	66.7
No	48.8	29.0	33.3

Note: Means with the same superscript are significantly different at $p<0.05$

Even though the communicating HC group was slower than the control group at completing the RAN colours subtest (see Table 6), chi-square analysis found no group differences for the number of participants that made errors on RAN-colours [χ^2 (2, N = 116) = 1.72, p>0.05]. Similarly, there was no significant group differences for the number of participants that made self-corrections [χ^2 (2, N = 116) = 3.59, p>0.05].

In summary, the HC groups exhibited processing speed deficits on tasks of psychomotor speed and rapid naming. Although the HC groups were slower on the RAN they were not less accurate. While no group significant differences were identified between the HC groups, the communicating HC group had lower mean scores on rapid word retrieval tasks (RAN and CNT), whereas the obstructive HC group had slightly poorer scores on a visuo-motor skill task (Coding).

3.6 Executive Function

3.61 Planning, Mental Flexibility, Working Memory and Self Regulation

Table 10 displays the *z* score means and standard deviations of the of TOL summary score, the CNT (trial 4) efficiency score and WISC-III digits backwards, for the obstructive HC, communicating HC and control groups.

Table 10

Z Score Means and Standard Deviations of the CNT Trial 4 Efficiency Score, the TOL Summary Score and Digits Backwards for the Clinical and Control Groups

	Participant Group					
	Obstructive Hydrocephalus ($n = 37$)		Communicating Hydrocephalus ($n = 24$)		Control ($n = 41$)	
	M	SD	M	SD	M	SD
CNT Efficiency Score	-0.52[a]	0.96	-0.65[b]	0.90	0.05[a,b]	0.68
TOL Summary Score	-1.24[a]	1.53	-1.34[b]	1.81	0.13[a,b]	0.91
RCF-Accuracy Score	-0.63	0.96	-1.43[b]	2.11	-0.26[b]	0.72
Digits Backwards	-0.24	1.03	-0.01	1.10	0.26	0.92

Note: Means with the same superscript are significantly different at $p<0.05$

A between groups MANOVA indicated a significant difference in the performance on tests of executive function of the three groups [Pillai's Trace = 0.36, F (8, 194) = 5.33, $p<0.001$, $\eta^2 = 0.18$]. Univariate tests showed a significant difference

between groups on CNT trial 4 efficiency score [F (8,99) = 6.93, $p<0.01$, $\eta^2 = 0.13$], TOL summary score [F (2,99) = 12.60, $p = 0.001$, $\eta^2 = 0.20$] and RCF-accuracy score [F (2,99) = 6.53, $p < 0.01$, $\eta^2 = 0.12$], but not on digits backwards [F (2,99) = 2.45, $p>0.05$, $\eta^2 = 0.05$].

Post hoc analyses revealed that the control group had significantly higher mean CNT trial 4 efficiency score than both HC groups ($p<0.01$). Post hoc analysis also found a significant difference in the TOL summary score between the control and obstructive HC groups ($p<0.001$) and between the control and communicating HC groups ($p<0.001$). The only significant difference on the RCF-accuracy score was between the control and communicating HC groups ($p<0.01$). No other group differences were found.

The TOL summary score incorporates the number of failed attempts and the time taken to complete the 12 trials. The components of the TOL, namely the number of failed attempts, time taken, corrections and repetitions were analysed in separate ANOVAs to help determine what factors, if any, were influencing performance.

Table 11 presents *z* scores of the means and standard deviations of these TOL component scores for the obstructive HC, communicating HC and control groups.

Table 11

Z score Means and Standard Deviations of the TOL Component Scores for Clinical and Control Participants.

	Participant Group					
	Obstructive Hydrocephalus (n = 42)		Communicating Hydrocephalus (n = 30)		Control (n = 42)	
	M	*SD*	*M*	*SD*	*M*	*SD*
Time Taken	0.67[a]	1.48	0.64[b]	1.42	-0.27[a, b]	0.86
Failed Attempts	0.24	1.52	0.62[b]	1.58	-0.39[b]	1.11
Correct Items	-1.10[a]	3.14	-1.00[b]	2.15	0.51[a, b]	1.25

Note: Means with the same superscript are significantly different at p<0.05

Separate one way ANOVAs revealed that the groups differed significantly on time taken [F (2, 113) = 7.26, $p < 0.01$], failed attempts [F (2, 113) = 4.95, $p < 0.01$], and number of correct items [F (2, 113) = 6.07, $p < 0.01$].

Post hoc analyses revealed that control group took significantly less time to complete the TOL than the HC groups ($p< 0.01$). The communicating HC group made more failed attempts than the control group ($p< 0.01$) but not than the obstructive HC group. Both HC groups performed similarly and both achieved significantly fewer correct

items than the control group ($p< 0.05$). No other group differences were identified.

3.62 Organization

The communicating HC group was found to be less accurate at copying the RCF than the other two groups. Because this task involves visual-perceptual skills, planning and organisation, organisational strategies were analysed to determine if they contributed significantly to RCF accuracy. Table 12 displays the type of RCF organizational strategies employed by the three participant groups (obstructive HC, communicating HC and control).

Table 12

Organizational Strategies Utilised by the Clinical and Control Groups

	Rey Complex Figure Organizational Strategies			
	Poor	Fragmented	Part Conceptual	Conceptual
Participant Group	(%)	(%)	(%)	(%)
Obstructive HC [a]	27.9	16.3	48.8	7.0
Communicating HC [a, b]	45.2	9.7	25.8	19.4
Control [b]	14.3	19.0	47.6	19.0
N	116			

Note: Means with the same superscript are significantly different at p<0.05

Chi-Square analysis revealed that the groups used different strategies to complete the RCF (χ^2 (6, N =120) = 14.54, p<0.05). As can be seen in Table 12 children in the communicating HC group were more likely to utilize poor strategies (45.2%) than

children in the obstructive HC group (27.9%) and control participants (16.3%). Whereas, almost half of the participants in the control and obstructive groups employed a part conceptual approach (47.6% and 48.8%, respectively), only 23.5% of communicating HC participants used this strategy. The trend was that more of the children from the communicating HC group did poorly on the RCF-OSS than the control and obstructive HC groups and the higher organizational scores (part-conceptual and conceptual) were achieved by the control group.

With regard to cognitive aspects of executive function, both HC groups exhibited deficits in mental flexibility, self-regulation, problem solving and planning tasks. However, the HC groups had similar working memory abilities to those of controls. The HC groups did not differ significantly in any area of executive function. Compared to controls the communicating HC group made more failed attempts on TOL, which suggests impulsivity. The communicating HC group also performed more poorly than controls on tasks of planning and organisation, whereas the obstructive HC group did not differ to controls on these domains.

3.63 Behavioural Aspects of Executive Function

Table 13 displays means and standard deviations of the each of the BRIEF scales for the obstructive HC, communicating HC and control groups.

Table 13

T-Score Means and Standard Deviations of the BRIEF Scales for the Clinical and Control Participants.

	Obstructive Hydrocephalus (n =36)		Communicating Hydrocephalus (n =26)		Control (n = 35)	
	M	*SD*	*M*	*SD*	*M*	*SD*
Inhibit	52.69[a]	12.16	52.19	10.56	46.29[a]	10.00
Shift	54.33[a]	13.93	51.50	12.14	44.54[a]	8.92
Emotional Control	54.67	14.98	55.88[b]	13.30	47.49[b]	10.60
Initiation	57.97[a]	12.77	55.65[b]	12.34	45.54[a,b]	9.92
Working Memory	57.64[a]	13.87	53.92[b]	11.66	45.86[a, b]	9.70
Plan/Organize	52.17	11.72	52.35	9.77	48.94	10.65
Materials Organization	54.39[a]	12.70	53.77[b]	13.05	43.91[a, b]	9.24
Monitor	55.44[a]	14.34	53.65	12.13	45.54[a]	9.92

Note: Means with the same superscript are significantly different at $p<0.05$

A between groups MANOVA revealed a significant group differences on BRIEF Scales [Pillai's Trace = 0.28 F (16, 176) = 1.82, $p<0.05$, $\eta^2 = 0.14$]. Follow up univariate

tests yielded a significant difference between the groups on the BRIEF indices shift [F (2,94) = 6.34, p<0.01, η^2 = 0.11], inhibit [F (2,94) = 3.57, p<0.05, η^2 = 0.07], initiate [F (2, 94) = 11.02, p<0.001, η^2 = 0.19], working memory [F (2, 94) = 8.98, p<0.001, η^2 = 0.16], organization of materials [F (2, 94) = 8.59, p < 0.001, η^2 = 0.16], monitor [F (2, 94) = 5.58, p<0.01, η^2 = 0.11], and emotional control [F(2, 94) = 3.93, p<0.05, η^2 = 0.08]. However, there was no significant difference on the BRIEF index plan and organize [F (2, 97) = 1.34, p>0.05, η^2= 0.03].

Post hoc analyses revealed the control group initiated more activities (p<0.05), were better at organizing their materials (p<0.05) and had better working memory abilities (p<0.05) than both HC groups. Moreover, the obstructive HC group was significantly poorer at shifting their attention, and inhibiting and monitoring their behaviour. On the other hand, the communicating HC group was poorer at controlling their emotions than the control group. No other group differences were found.

In summary, parents of both HC groups reported problems with several behavioural aspects of executive function (such as working memory, initiating activities, inhibiting behaviour, and organisation of materials) than did the parents of both groups of HC children. In addition to these deficits, children with obstructive HC were rated to have problems with inhibition, shifting attention, and monitoring their behaviour, whereas children with communicating HC had difficulties with controlling their emotions.

3.7 Summary of Attention, Processing Speed and Executive Function Test Results

Table 14 demonstrates significant differences between the three participant

groups on test variables.

Table 14

Significant differences between the two HC groups and the Control Participants on Test Variables

Cognitive Domain	*Test Variable*	*Obstructive-Communicating*	*Control-Obstructive*	*Control-Communicating*
IQ			***	**
VIQ			*	
PIQ			***	***
Attention				
	SkySearch		*	*
	Score!		*	*
	SkySearchDT		*	*
Processing Speed				
	RAN-Colours			*
	WISC-III Coding		***	**
	CNT Trials 1 & 2		*	*
Executive Function				
	CNT-Trial 4		**	**
	TOL		***	***
	RCF			**
	RCF-OSS	*		*
	Digits Backwards			
BRIEF				
	Inhibiting Behaviour		*	
	Shifting		**	
	Emotional Control			*
	Initiation		***	**
	Working Memory		***	*
	Plan/Organise			
	Material Organisation		**	**
	Monitoring Behaviour		*	

* indicates a significant difference at the $p<0.05$ level
** indicates significant difference at the $p<0.01$ level
*** indicates as significant difference at the $p<0.001$ level

3.8 Magnetic Resonance Imaging Data

3.81 White Matter Abnormalities

Table 15 presents the type and location of white matter abnormalities in the obstructive and communicating HC participants demonstrated on MRI scans.

Table 15

Percentages of White Matter Abnormalities in Participants with Obstructive and Communicating Hydrocephalus Demonstrated on MRI

	Obstructive Hydrocephalus (%)	Communicating Hydrocephalus (%)
Degree of White Matter Loss		
None	24.1	0.0
Mild	34.5	63.2
Moderate	24.1	26.3
Severe	17.2	10.5
Location of White Matter Loss		
Deep	75.9[a]	82.4[a]
Deep and Subcortical	0.0[a]	17.6[a]

Note: Means with the same superscript are significantly different at $p<0.05$

Chi-square Analysis revealed no significant relationship between type of HC and degree of white matter loss [χ^2 (3, N = 46) = 7.02, $p>0.05$]. Notably the entire

communicating HC group had some degree of white matter loss, whilst approximately one-quarter of the obstructive HC participants did not have any white matter loss. There was a relationship between type of HC and location of white matter loss [χ^2 (2, N = 46) = 9.28, p<0.05]. The location of white matter loss in the obstructive HC group was restricted to deep white matter (79.5%), whereas the communicating group had both deep white matter loss (82.4%) deep and subcortical white matter loss (17.6%).

3.82 Corpus Callosum Abnormalities

Table 16 presents the percentages of the type and location of corpus callosum abnormalities demonstrated in the participants with obstructive HC and communicating HC on MRI scans.

Table 16

Corpus Callosum Abnormalities in Participants with Hydrocephalus Demonstrated on MRI

Corpus Callosum	Obstructive Hydrocephalus (%)	Communicating Hydrocephalus (%)
Rostrum		
Normal	82.8	90.0
Thick	3.4	0.0
Thin	13.8	10.0
Genu		
Normal	80.0	80.0
Thick	6.7	0.0
Thin	13.3	20.0
Body		
Normal	53.3	45.0
Thick	3.3	10.0
Thin	43.3	45.0
Splenium		
Normal	51.7	60.0
Thick	6.9	5.0
Thin	37.9	35.0
Absent	3.4	2.0

Note: Means with the same superscript are significantly different at $p<0.05$

Chi-square analysis of rostrum abnormalities revealed no significant group differences [χ^2 (2, N = 49) = 0.90, p>0.05], with the majority of participants having a normal rostrum (82.8% obstructive HC and 90% communicating HC). Further analysis yielded no significant relationship between genu abnormalities and HC type [χ^2 (2, N = 48) = 1.67, p>0.05] or between the corpus callosum body abnormalities and type of HC [χ^2 (2, N = 48) = 0.96, p>0.05]. With regard to the genu the majority of each HC group (80%) had no abnormalities in this region. Of the communicating HC group 20% had a thin genu, whilst 13.3% of the obstructive HC group had a thin genu. The remaining obstructive HC group had a thick genu (6.7%).

Almost half the participants in the HC groups had normal corpus callosum bodies (45 and 50%, respectively). The remaining obstructive HC group had thin (46.7%) or thick corpus callosum bodies (3.3%). The remaining communicating HC participant had thin (45%) or thick (10%) corpus callosum bodies.

Splenium abnormalities did not differ significantly between HC groups [χ^2(3, N = 47) = 1.19, p>0.05]. Of note, only about half of the HC groups exhibited problems in these regions. The majority of splenium abnormalities were due to thinning (37.9% of obstructive HC and 35% of communicating HC). However, thick spleniums were also evident (see Table 15). Absence of the splenium was uncommon in both groups, only occurring in 3.4% of the obstructive HC group and 2% of the communicating HC group.

Not surprisingly the majority of the two HC groups had white matter loss. The degree did not differ between the groups, however the location did (obstructive HC group

had deep white matter loss and communicating HC had deep and deep and subcortical white matter loss). The HC groups were very similar on the presence, location and proportion of corpus callosum abnormalities. With considerable proportion of each group having a normal corpus callsoum. Interestingly, the most common location of abnormalities was in the body and the splenium.

CHAPTER 4: DISCUSSION

4.1 Hypotheses

The hypotheses that the obstructive HC group would exhibit more profound deficits in attention, processing speed and executive functions, compared to children with communicating HC were not supported. Similarly, the hypothesis that the obstructive HC and communicating HC groups would differ in the degree of white matter injury and dysgenesis of the corpus callosum was not supported. Notwithstanding the lack of support for these hypotheses, cognitive and neuropathological differences between the two HC groups were observed.

4.2 Intellectual Functioning

In general, children with HC had a lower FIQ than control participants and this discrepancy cannot be explained by SES differences. The clinical and control groups performed within the *Average* range on FIQ, and consistent with expectations the obstructive HC group obtained the lowest FIQ. Several other studies have also found children with HC perform within the *Average* range on FIQ (Barnes & Dennis, 1998; Brewer et al., 2001; Dennis et al., 1981; Donders et al., 1991; Hetherington & Dennis, 1991; Klaas et al., 1999). Erickson et al. (2001) reported that children with HC are now more likely to have FIQs in the *Average* range than in the previous decades because of improved and earlier diagnosis, better treatment and reduced incidence of shunt complications (Erickson et al., 2001).

However, the current finding is in contrast to previous research that found children with HC perform within the *Low Average* range on measures of general

intelligence (Bier et al., 1997; Donders et al., 1990; Fletcher, Bohan et al., 1992; Fletcher et al., 1997; Friedrich et al., 1991; Jacobs et al., 2001; Tew & Laurance, 1975). Superior FIQ for this HC group could be explained by the fact that children with SB were excluded from the study. Children with SB are consistently reported to have lower IQ scores than other aetiological groups, as a consequence of the major cerebral anomalies associated with the disorder (Anderson, Northam, et al., 2001; Brookshire, Landry, et al., 1995; Fletcher, Bohan et al., 1992; Fletcher, Brookshire et al., 1995; Fletcher, Brookshire et al., 1996). Studies that included SB children and report an *Average* IQ for their HC sample, also include children with other aetiologies who have higher IQs than children with SB, thereby increasing the mean FIQ of the HC sample (Barnes & Dennis, 1998; Dennis et al., 1981; Hetherington & Dennis, 1991). When compared to other studies that have separated aetiological subgroups, similar mean FIQ's were observed to that of Brewer et al.'s congenital HC group (2001), Dennis et al.'s postnatal aetiology group (1981), Donders et al.'s HC group (1991) and Fletcher, Brookshire, et al.'s (1996) premature HC group.

Similar to previous research, children with HC had lower Performance IQ's than control participants (Brookshire et al., 1995; Dennis et al., 1981; Donders et al., 1990; Fennell et al., 1987 Fletcher, Bohan, et al., 1992; Fletcher et al., 1997; Friedrich et al., 1991; Snow et al., 1994; Tew & Laurance, 1975). In HC, visual-perceptual and visual-motor deficits, which load heavily on PIQ, are thought to be a result of thinning in the posterior brain regions (Brookshire et al., 1995a; Dennis et al., 1981; Fletcher et al., 1997; Friedrich et al., 1991). The compression that causes occipital horn enlargement may also lead to visual tract abnormalities and ocular coordination problems,

compromising visual perceptual abilities. These visual disturbances chronically limit the visual experience, which in turn disrupts the development of the visual cortex and leads to further deficits in visual-perceptual functioning. As a consequence of these deficits non-verbal intelligence is often affected (Dennis et al., 1981). Another reason for depressed PIQ is because white matter interregional connections are often damaged by HC, and these neuroantomical substrates are crucial for processing novel or complex stimuli on visual spatial tasks (Rourke, 1987).

It is important to note that all of the communicating HC group and 86% of the obstructive HC group had a VP shunt inserted in their right parietal lobe, a neuroantomical region that is thought to subserve visual-spatial functions. Some authors have postulated that injury to the right parietal lobe caused by shunt surgery may be the cause of visual-spatial problems in children with HC (Fletcher, Brookshire, et al., 1996). Others have suggested that shunt surgery would imply more severe HC and consequently the neuropathology and neuropsychological consequences are more severe (Anderson, Northam, et al., 2001). Given that visual-spatial abilities are thought to be mediated by the right parietal lobe, and children with HC in this study displayed deficits in this domain, it may be postulated that the HC sample had significant injury to white matter in the right posterior hemisphere, either as a consequence of HC or the shunt surgery or both.

With regard to verbal cognitive abilities, only the obstructive HC group had a significantly lower mean VIQ than controls. This is consistent with previous findings that obstructive (intraventricular) HC was associated with poorer language skills than communicating (extraventricular) HC (Dennis et al., 1987). Dennis et al. (1987)

suggested that in obstructive HC the intraventricular obstruction of CSF causes pressure within the ventricular system, resulting in children displaying deficits in the central aspects of language, those involving core elements of linguistic content and form. In contrast, children with communicating HC demonstrate difficulties in peripheral language abilities, such that they have preserved language abilities but impaired fluency (Dennis et al., 1987). The finding that obstructive HC results in preserved fluency but impaired semantic and visual word finding as well as poor grammatical comprehension, suggests that intraventricular pressure present in obstructive HC damages the left temporal lobe as well as corpus callosum fibres (Dennis et al., 1987). The left temporal lobe mediates comprehension of language and word finding abilities, whereas the corpus callosum is important in transferring visual information to naming centres in the left hemisphere (Lezak, 1995). Supporting this explanation, previous research has reported corpus callosum anomalies in HC (Hannay, 2000) and posterior brain regions are reported to be more affected than anterior ones (Dennis et al., 1981). On the other hand, reduced verbal fluency observed in communicating HC would suggest that sub-tentorial regions might be might be important for this function (Dennis et al., 1987).

4.3 Attention

Overall, children with obstructive HC and communicating HC exhibited deficits in attentional skills compared to the control group. Contrary to expectations the two HC groups did not differ significantly on any of the attentional components.

Compared to control participants the obstructive HC group demonstrated difficulties selecting target information from an array of distractor items. Focussed

attention difficulties were also noted in the communicating HC group, who performed half a standard deviation below controls, however this finding was not significant (p=0.057). An increased number of participants in the communicating HC group may have increased the power of the analyses to detect the smaller effect size between communicating HC and control groups. No significant differences were found between the HC groups, who performed within 0.2 of standard deviation of each other.

The deficits in focussed attention displayed by the obstructive HC group, and to a lesser extent the communicating HC, are consistent with previous research (Brewer et al., 2001; Fletcher, Brookshire et al., 1996; Loss et al., 1995; Tew et al., 1980). Similar to these studies, the current study used a cancellation-type task to assess focussed attention. It has been argued that motor problems and slow processing speed ubiquitous in HC may contribute to the focussed attention difficulties in children with HC (Fletcher, Brookshire, et al., 1996; Loss et al., 1998). It should be noted that motor difficulties and slowed processing speed were taken in to account when analysing the data. Hence the number of targets identified, rather than the time per target, was examined so any slowness in processing and motoric difficulties would not be as influential.

It is possible that visual scanning deficits reported in children with HC may be a reason for their poorer performance (Dennis et al., 1981). Certainly, in the current sample, children with obstructive HC and communicating HC had a much higher incidence (approximately 50% in each group) of visual problems as compared to the control group (only 2.5% had visual problems). Consistent with this finding, deficits of gaze and eye movement and /or refraction and accommodation have been reported in children with HC. Altered pressure in the midbrain collicular structures, affecting the

third cranial nerve nuclei, causes third and sixth cranial nerve palsies and paralyses of the upward gaze (Dennis et al., 1981). Moreover, HC can result in compression that causes occipital horn enlargement and may also lead to visual tract abnormalities and ocular coordination (Riva et al., 1994). These visual motor and visual perceptual difficulties would impede a child's performance on any task with visually based stimuli.

Deficits in focussed attention have been related to dysfunction of the posterior attention system (Erickson, et al., 2001). More specifically, right inferior parietal lobe and superior temporal lobe functioning has been associated with focussed attention (Mesulam, 1987). In addition, sub-cortical brain structures in the posterior right hemisphere have also been implicated in deficient focussed attention abilities (Fletcher, Brookshire, et al., 1996). In children with HC, focussed attention deficits may be the consequence of damage to the posterior parietal regions caused by compression and stretching of white matter tracts and oedema (Fletcher, McCauley et al., 1996; Ito, Saijo, Araki, Tanaka, Tasaki, Cho & Miyamoto, 1997).

In comparison to control participants, both HC groups had difficulty sustaining their concentration on an auditory task, supporting previous research (Brewer et a., 2001; Fennell et al., 1987; Loss et al., 1998; Tew et al., 1980). Contrary expectations the two HC groups had similar sustained attention abilities. Whereas previous research has mainly used a CPT to assess sustained attention (Fennell et al., 1987; Brewer et al., 2001; Tew et al., 1980), the current study utilised an auditory sustained attention task from the TEA-Ch. Unlike the CPT, Score! does not involve motor or visual-spatial skills, therefore deficits in sustained attention exhibited by the children with HC cannot be explained in terms of difficulties with motor or visual-spatial demands of the task.

Mirsky et al. (1991) suggested that deficits in sustained attention reflect damage to the rostral midbrain structures (Mirsky et al., 1991). In children with HC the midbrain may develop abnormally due to congenital disorders, as is the case of Chiari Malformation, or may be damaged by the compressive effects of HC (Del Bigio, 1993; Raimondi, 1994).

Obstructive HC and communicating HC children demonstrated deficits in their ability to divide/shift their attention between two competing stimuli. Contrary to expectations the two HC groups had very similar divided attention abilities. To date there is a paucity of research investigating this domain of attention. Furthermore, comparisons with other studies are problematic due to different methodologies and conceptualisations of this attentional construct. Current findings are consistent with Wills (1993), who reported that children with HC have difficulty with tasks that demand more active mental tracking and higher-level attentional skills such as shifting attention. Other studies have found children with HC have difficulties shifting attention on the WCST (Brewer et al., 2001; Fletcher, Brookshire et al., 1996).

One such study that employed the WCST was Fletcher, Brookshire, et al. (1996). They found that children with HC had difficulties on several domains, in particular the perseverative errors and number of correct categories, which Mirsky (1996) suggested was a measure of shifting attention. However, they did not interpret their results as a deficit in shifting attention. Instead they hypothesized that the pattern of performance on the WCST and other tests, reflected problems with arousal and inattention. They suggested that children with HC were more distractible and never "got into the task." These types of attention deficits implicate impairment to the subcortical structures in the right posterior hemisphere that represent part of an arousal-activation brain system

producing problems with intention (Fletcher, Brookshire et al., 1996; Heilman, Watson & Valenstein, 1993). In contrast to Fletcher, Brookshire et al. (1996) and Brewer et al.'s (2001) conclusions, Wills (1993) suggests that the deficits in shifting attention were due to mental tracking and planning difficulties, a pattern consistent with damage to anterior brain regions. In the present study Children with HC had difficulties dividing their attention. Due the combination of cognitive deficits displayed by children with HC differentiating whether these attention difficulties are the result of anterior system or posterior and subcortical attentional system dysfunction is not possible.

The current study employed a task that required the children to divide their attentional resources between auditory and visual stimuli. The ability to divide one's attention means dealing with multiple attentional demands and being able to switch efficiently between different activities. Neuroantomically, the anterior cingulate and the prefrontal cortex have been associated with this ability (Mirsky et al., 1991). In contrast, Brewer et al., (2001) suggested that in children with HC impairment in shifting attention depended on the extent to which the midbrain was damaged. If the ability to shift attention was mediated by the midbrain, as suggested by Brewer et al. (2001), children with congenital aetiologies, in particular those children with DWS and Chiari Malformation, would be expected to perform more poorly than the children with IVH. However, the obstructive HC and communicating HC groups performed similarly on this task suggesting other contributing factors to these shifting attention difficulties.

4.4 Processing Speed

Children with obstructive HC demonstrated deficits compared to control

participants in processing speed on both a psychomotor task and on rapid naming tasks. Children with communicating HC also demonstrated processing speed deficits on tasks of psychomotor speed and rapid naming of colours and shapes. Contrary to predictions the two HC groups did not significantly differ on this cognitive domain. However, there were some subtle differences noted between the HC groups. The obstructive HC group had lower scores on the psychomotor speed task than the communicating HC group, whilst the communicating HC group performed more poorly on tasks that required rapid naming of colours and shapes.

Though there are few empirical studies of processing speed in children with HC, it has been reported that these children are generally slower at timed tasks that require visual tracking, eye-hand co-ordination skills and physical manipulation of test materials (Anderson, Northam, et al., 2001). Consistent with the literature, in the current study children with HC did more poorly than controls on a task of processing speed that required pencil and paper responses (Fletcher, Brookshire et al.1996) but also on two non-motor based processing speed tasks (CNT-trial 1 and 2, and RAN-colours). Despite their slowness, the HC children were not less accurate than the control group on the RAN. The finding was consistent with Fletcher, Brookshire et al. (1995) who also found that children with HC performed more poorly on the RAN.

Information processing deficits are not surprising given the high incidence of corpus callosum abnormalities in children with HC (Fletcher, Brookshire et al., 1996; Hannay, 2000; Klaas et al., 1999). Tasks that measure processing speed, such as the RAN and CNT, require the transfer of visual information from posterior brain regions to naming centres in the anterior regions of the left hemisphere via the corpus callosum.

4.4 Executive Function

The current study found that children with obstructive HC and communicating HC had executive function deficits when compared to controls. With the exception of organisational skills, the obstructive HC and communicating HC children did not significantly differ on any of the executive function domains.

The obstructive HC group demonstrated problem solving and planning deficits on a novel task (TOL). Further analysis revealed they took longer to complete each item and achieved fewer correct items than the controls, but did not fail more attempts. The communicating HC group also demonstrated deficits in problem solving and planning. As a group, the communicating HC children were slower to complete items, completed fewer correct items and made more failed attempts, than controls. More failed attempts would imply that children with communicating HC were more impulsive than controls. Interestingly, the communicating HC group also had planning difficulties on another task of visual-spatial task (RCF), whereas the obstructive HC group did not. Perhaps the communicating HC group's performance on the RCF was affected by an impulsive approach.

Deficits in problem solving and planning abilities are consistent with previous research (Fletcher, Brookshire et al., 1996). However, discrepancies between the current findings and previous research are evident on two TOL variables; time taken to solve a problem (Anderson et al., 2002; Fletcher, Brookshire, et al., 1996) and the number of correct items (Anderson et al., 2002).

Unlike the current study, Fletcher, Brookshire et al. (1996) found that children

with HC were comparable to controls on time taken to complete the TOL. This discrepancy in time taken can be explained by the fact that the Fletcher, Brookshire et al. (1996) only analysed trial 1, whereas the current study analysed the time taken for the entire task (12 trials). Differences in processing speed may become more evident as the task becomes more complex.

Consistent with Fletcher, Brookshire, et al. (1996), Anderson et al. (2002) also found that children with HC were comparable to controls on time taken to complete the TOL. The differences between the current study and the Anderson et al. (2001) study could be explained by differences in IQ. The difference in the IQ of the control group and HC group in the Anderson et al. (2002) study (control = 102.92 to HC = 97.33) was considerably less than in the current study (control = 106.90, obstructive HC = 94.42 and communicating HC = 97.42), which may account for the discrepancy. Supporting this view, a comparison of raw scores found that the control group in the current study performed on average 30 seconds faster and achieved more correct items than the control group in the Anderson et al. (2001) study. Conversely, both HC groups in the current study were slower and completed fewer correct items.

With regard to organisational skills, the obstructive HC group performed similarly to the controls, whilst a large proportion of the communicating HC group demonstrated poor conceptual organizational strategies. The obstructive HC group and the communicating HC group differed significantly on organisational abilities, however contrary to expectations the communicating HC group displayed more difficulty. Although the communicating HC group had more difficulties than controls on their planning and copying of the RCF, the obstructive HC group performed similarly to

controls.

Neither of the HC groups differed to the control group on working memory abilities. However, both HC groups performed approximately half a standard deviation below the control group, which would suggest difficulties at a clinical level. These non-significant findings may be explained by a lack of statistical power. For example, power analyses revealed that there was only a 50% chance of detecting the small-moderate effect size present (η^2=0.05).

Children with HC displayed deficiencies in mental flexibility, but contrary to expectations, the obstructive HC and communicating HC groups performed similarly on this domain. Findings of reduced self-regulation and mental flexibility in the HC children are consistent with previous research (Anderson et al., 2002; Dise & Lohr (1998). Mental flexibility has been associated with the frontal-striatal system, implicating the prefrontal cortex and basal ganglia (Eslinger & Grattan, 1993). In children with HC, these skill deficits may reflect damage to subcortical regions and white matter tracts carrying information to and from the prefrontal cortex. For example, grade 4 IVH is associated with intraparenchymal bleeding, which can penetrate the head and body of the caudate nucleus. Similarly damage to the basal ganglia may be caused by axonal injury and disturbance of myelination in the periventricular area (Hetherington & Dennis, 1987). Subcortical damage may also occur secondary to increased pressure on the midline structures (Fletcher, Brookshire, et al., 1996).

As well as using standardised cognitive measures to assess executive functions, the current study examined behavioural aspects of executive functions using the parent form of the BRIEF. The HC groups exhibited deficits in both the metacognition and

behavioural regulation indices of the BRIEF. According to parents, the obstructive HC group display problems with working memory, organizing their materials, inhibiting and monitoring behaviour, and shifting attention. The communicating HC group were also reported to have difficulties with working memory and organizing their materials, additionally they experience problems with controlling their emotions.

Two other studies have administered the BRIEF to HC samples (Anderson et al., 2002; Mahone et al., 2002). Similar to Anderson et al. (2002) and Mahone et al.'s (2002) findings, parents of HC children identified they had problems on the BRIEF metacognition index. Further comparisons to the Mahone et al. (2002) are not possible because of different methodologies. Whilst the Mahone et al. (2002) study compared parent reports to the adolescent reports, the current study utilised only the parent report. Noteworthy is that the current study's HC group performed similarly to the HC group in the Anderson et al. (2002) study. In the present research children with HC exhibited behavioural problems in several domains of the BRIEF, which suggests a generalized pattern of behavioural problems.

The current study's findings suggest that children with HC have difficulties with many aspects executive function. Providing an explanation for these deficits is an area of contention amongst the literature. Some authors propose that HC children do not have executive function problems per se, rather they had generalised visual-spatial problems and difficulties sustaining attention, interpreted as indicative of right posterior attentional system dysfunction rather than frontal lobe dysfunction (Fletcher, Brookshire et al., 1996). Moreover, Fletcher, Brookshire, et al. (1996) suggested that children with HC are underaroused, more distractible and never 'get into the task,' referring to this pattern of

deficits as an *intentional* system problem. Supporting this view, Donders et al., (1990) have reported that children with HC have greater difficulty processing complex or novel stimuli, particularly in the visual-spatial domain. Fletcher, Brookshire, et al. (1996) suggested that observational studies may provide more information about whether children are more distractible, and their underarousal and inability to 'get into' the task is the reason for their problems with executive functions. An alternative hypothesis for executive function deficits in HC was suggested by Snow et al. (1994). They interpreted difficulties that SB children had with cognitive abstraction skills was due to anterior cortical damage.

It is difficult to interpret the current findings as supporting either the Fletcher, Brookshire, et al. (1996) posterior attention system deficit hypothesis or the Snow et al. (1994) anterior cortical hypothesis. Instead support was found for both interpretations. For example, the present study found that children with HC had difficulty sustaining attention, a depressed PIQ, performed poorly on tasks that required problem solving and planning abilities in the visual-spatial domain, and were noted to have problems with 'initiating behaviour' on the BRIEF, consistent with Fletcher, Brookshire, et al.'s *intentional* system hypothesis (1996). However supporting Snow et al.'s (1994) interpretation, children with HC demonstrated problems on executive function tasks that are not as reliant on visual-spatial skills (mental flexibility, self-regulation, behavioural aspects of executive function). It is possible that the observed deficits in attentional control, mental flexibility and self-regulation may associated with deficits in focussed and sustained attention in this population. However, on the basis of the present findings, disruption to the neural connections carrying information to and from the prefrontal

cortex cannot be disregarded.

4.5 Magnetic Resonance Imaging Data

Consistent with the findings that children with obstructive HC and communicating HC have deficits in their attentional skills, processing speed and executive functions, the majority of the two HC groups demonstrated white matter loss on MRI. Whilst 75% of the obstructive HC group displayed white matter abnormalities, the entire communicating HC group had some degree of white matter loss. Greater white matter loss in the obstructive HC group was predicted, as it is mainly comprised of children with congenital aetiologies. Congenital causes of HC have been associated with a "snowballing effect on subsequent brain development" (dismigration and dysgenesis anomalies), whereas the communicating HC group consists of children who had postnatal insults to a brain that has developed normally (Wills, 1993, p. 251). Despite these distinct differences in timing, mechanism and aetiology of HC, in nearly a quarter of children with obstructive HC there was no significant white matter injury. The reason for this is difficult to ascertain. One explanation is that it may reflect the different disease processes of the separate aetiologies. The communicating HC group consisted entirely of children with IVH secondary to prematurity or low birth weight, which has been associated with white matter injury and PVL in the absence of HC. Perhaps another reason is because a proportion of children with obstructive HC had only mild ventriculomegaly that was not sufficient to cause white matter loss. The differences are unlikely to be related to the duration raised ICP as children with HC secondary to IVH have routine cranial ultrasounds and it would be expected that ventriculomegaly would be

detected in the first few weeks of life. Children with congenital abnormalities may not have HC detected until neurological signs and increased skull circumference were present. It is possible that the failure to find white matter injury in approximately 25% of the obstructive HC group may be a function of the sensitivity of the neuroimaging procedures used.

The location of white matter loss differed between the two groups, with the obstructive HC group having deep white matter loss and the communicating HC group having both deep and subcortical white matter loss. Previous research has also found that children with grade 4 IVH can damage the caudate nucleus, a subcortical structure (Hetherington & Dennis, 1999). With regard to the corpus callosum, most participants in both HC groups displayed normal rostrums and genus. About half the HC groups had thin corpus callosum body or splenium. Given the previous research has consistently found corpus callosum abnormalities in the HC population, the current finding was expected (Barkovich, 1990; Dennis et al., 1981; Hannay, 2000).

The incidence of co-morbid disorders such as seizures, cerebral palsy and PVL did not differ between the obstructive HC and communicating HC groups. They were also very similar on variables such as the location of shunts and the frequency of shunt revisions, infections and blockages. More compromise of cognitive skills, especially PIQ, has been noted in children who have co-morbid conditions with seizures reported to have the most detrimental effects (Dennis et al., 1981).

As the proportion of participants with co-morbid conditions did not differ between the two HC groups, the impact of co-morbidities is not clear. Inspection of mean scores on the tests administered revealed that, overall children in both HC groups

who had seizures performed more poorly than those who had not suffered a seizure. For example a comparison of children with obstructive HC and seizures (FIQ: M =88.90, SD = 10.79, PIQ: M =85.00, SD = 11.25, VIQ: M = 94.20, SD = 10.90) and those with obstructive HC without seizures (FIQ: M =96.09, SD = 10.28, PIQ: M = 94.24, SD = 11.41, VIQ M = 98.06, SD = 11.58) found there was difference of seven points in FIQ, nine points on PIQ and three points on VIQ. The communicating HC children who had seizures (FIQ: M = 91.36, SD = 12.71, PIQ: M =87.45, SD = 15.79, VIQ: M = 96.91, SD = 11.02) compared to the communicating HC children who did not have seizures (FIQ: M =100.75, SD = 13.82, PIQ: M =96.90, SD = 19.15, VIQ: M = 104.70, SD = 13.33) were more than nine points poorer on FIQ and PIQ and almost eight points poorer on VIQ.

Preliminary analysis suggests that children who had seizures performed more poorly than those who did not have seizures, which is consistent with Dennis et al. (1981) findings that even a single seizure has a negative effect on cognitive outcome. The presence of seizures may indicate a greater degree of neuropathology. Consequently a greater degree of neuropsychological impairment would most likely be present.

These differences between means on IQ measures of the two groups require appropriate statistical analyses, which is beyond the scope of the current research. Furthermore, the small number of participants in each HC group with seizures (approximately 10) mean it is unlikely there would be sufficient power to detect a significant difference with effect sizes of this magnitude. However, thorough statistical analyses of these variables with larger samples may help to determine the contribution of medical variables.

4.6 Clinical and Theoretical Implications

In summary, children with obstructive HC exhibited deficits in their verbal and non-verbal intellectual abilities compared with controls. Deficits were also evident in focussed, sustained and shifting attention skills. As a group they were slower to process information than controls, and particular difficulties were evident on tasks involving psychomotor skills. As well as attention and processing speed deficits, impaired executive functions in the domains of problem solving, mental flexibility, planning and self-regulation were also evident. Behaviourally, the obstructive HC group was reported to have difficulty in several aspects of executive function (planning and organisation of materials, behavioural regulation, inhibition, shifting attention, and initiating activities).

Although the communicating HC group also had a mean FIQ and PIQ below that of normal healthy controls, VIQ was age appropriate. Similar to the obstructive HC group the communicating HC group demonstrated significant attentional difficulties, however, deficits were only noted in two components of attention; sustained and divided. The communicating HC group were also slower to process information, but their deficits were more pronounced in the rapid naming tasks rather than the psychomotor speed task. Executive functions were also impoverished in the communicating HC group. Similar to the obstructive HC group deficits in problem solving, planning, mental flexibility and self-regulation were observed, in addition to these deficits organisational problems were also evident. Behaviourally, they were also found to have problems with working memory and organising their materials, as well as controlling their emotions.

The finding that children with HC have poorer intellectual abilities, attention,

processing speed and executive functions have profound implications for them in their everyday life, in particular their educational progress. For example, children with HC in this study demonstrated difficulties in focussing their attention on important information, filtering out distracting information and sustaining their concentration. In the classroom these cognitive deficits would translate to distractibility and inattention. Children with HC may also take longer to understand concepts and information, and take more time to complete tasks. Given the combination of the mentioned deficits and the deficits in other cognitive domains, such as language, it is not surprising that children with HC are reported to have academic difficulties (Anderson, Northam, et al., 2001; Erickson, et al., 2001).

Perhaps as a consequence of their cognitive difficulties, and increased awareness of limitations, children with HC are reported to have social, emotional and behaviour problems (Fletcher, Brookshire, Landy, et al., 1995). Some authors have suggested that deficits in non-verbal cue comprehension and inappropriate familiarity described in some children with HC may be partly responsible for social skills problems (Tew, 1979). Moreover, as peers become more important during school age and the social demands become more complex, children with HC are reported to display poorer adaptive behaviour (Fletcher, Brookshire, Landy, et al., 1995), socialisation (Holler et al., 1995) and lower self-esteem (Erickson, 200; Fernell et al., 1991).

Whereas studies of children with localised brain injuries allow the inference of relationships between neuroanatomy and specific cognitive functions (Erickson et al., 2001), the current research assists in the understanding of how diffuse neuropathology, as occurs in HC, can affect particular cognitive functions. Theoretically, these results are

surprising because it was expected that the obstructive HC group would have more profound neuroantomical damage due to differences in the timing and aetiology of the pathology. As a consequence of the greater neuronal injury, it was expected that children with obstructive HC would exhibit more profound deficits in attention, processing speed and executive functions, but this hypothesis was not supported by the study's findings. These results also have significant theoretical implications for the understanding of neuropathology in HC. On the basis of the current findings, children with obstructive HC and communicating HC exhibit similar deficits in attention, processing speed and executive functions, which would appear to be the result of white matter loss and corpus callosum abnormalities, noted in both HC groups.

The lack of significant differences between the two HC groups on most cognitive and neuroantomical measures, suggest that regardless of aetiology the compressive and stretching effects of HC on the brain are similar. Certainly, one study has commented that regardless of aetiology when ICP reaches a certain threshold the detrimental neurological effects are similar (Raimondi, 1994). Another explanation may be that neuropathological and neuropsychological differences in the types of HC are evident closer to time of onset but become similar as acute effects of HC are not as prominent. It is also important to note that children with obstructive and communicating HC did not differ on shunt malfunction variables. This may be one reason for the lack of significant difference on neuropsychological outcomes between the two HC groups.

Importantly, this study adds to the paucity of recent research on attention, processing speed and executive functions in children with HC children. Clinically, this information may be relevant to neuropsychologists when giving feedback to parents

about these deficits. On the basis of this study, children with obstructive HC and children with communicating HC have several similarities in their prognosis with regard to cognitive outcomes. Understanding the types of cognitive difficulties children with HC experience is essential for developing rehabilitation measures that target these skills (Erickson et al., 2001).

It is apparent that a study of this magnitude has not been previously conducted with an Australian sample. Therefore, this study makes a significant contribution to the literature on children with HC in Australia. Given the potential differences in how medical centres internationally treat and manage children with HC, these findings may be particularly useful for clinicians working with children with HC in Australia.

This study provides recent cognitive and neuropathological information about children with HC. This is important because several landmark studies were conducted more than 20 years ago and consisted mainly of children with HC secondary to SB, the incidence of which has markedly declined over the past decade. Moreover, advances in treatment of HC have lead to an increased survival rate and better cognitive outcomes that are not reflected in older research (Anderson, Northam, et al., 2001).

4.7 Limitations

A methodological limitation of the current study was that the MRI data was qualitatively analysed. Volumetric quantitative analysis is preferable because it is more objective and precise. The findings were further limited by the fact that only 70% of the HC children had MRI scans. If more children had MRI scans conducted this may have increased the power to detect differences of smaller magnitude between the groups.

However, it may be argued that smaller differences, that cannot be easily detected, are of less clinical importance and do not warrant further investigation.

Small sample sizes have been a criticism of the previous studies of children with HC. The current study employed a relatively large sample (116 children) and most of the analyses revealed a moderate to large effect sizes and greater power to detect differences between the HC and control groups. However, where the effect size was smaller, such as it was between the two HC groups, increased numbers would have benefited the analyses. Referring to Cohen (1984), to have an 80% chance of detecting a small to moderate effect size (η^2= 0.01-0.06) a sample of more than 64 children in each of the HC groups would have been required.

Other studies (Brewer, et al., 2001; Fletcher, Brookshire, et al., 1996) have suggested that using attention tasks that minimise the motor-skill demands would be more informative about children with HC's attentional skills. In the current study the focussed attention task from the TEA-Ch required an element of motor control and visual scanning, often impaired in HC, and it is possible that this contributed to the findings that obstructive HC children had deficits in this area.

Another limitation of the current research is that it did not examine visual-motor and graphomotor abilities in the HC groups. Given that several studies have reported that these skills influence performance on tasks requiring manipulating of test materials, copying, writing, and visual scanning abilities, this may have assisted in the interpretation of test performance. In particular, it may have assisted in the interpretation of the obstructive HC group's performance on the selective attention task. If visual scanning problems were identified, difficulties on SkySearch may have been due to these visual

deficits rather than a problem with focussing attention.

The RAN tests were used as one measure of processing speed in the current study. Its validity as a measure of processing speed might be debated in this population because it relies on naming colours. Word- retrieval deficits that may affect performance on the RAN have been reported in children with HC in some studies (Brookshire et al., 1995b; Fletcher, Brookshire, Bohan, eta l., 1995) but not others (Dennis et al., 1987; Donders et al., 1991).

Inherent in the study of attention, processing speed and executive function is the difficulty of separating these constructs in assessment. Furthermore, the difficulty in defining and separating constructs makes it challenging to appropriately assess them. This difficulty has been noted by Erickson et al. (2001) who stated that executive functions are closely related to attention and memory functions and because of this overlap in skills, deficits in one area will impact on other cognitive domains. Due to these factors, differentiating the contribution of each cognitive construct to a child's neuropsychological profile is difficult.

This particular study did not specifically address inhibitory control a construct that has been proposed by some authors to be an important aspect of executive functions (Denckla, 1996). It was beyond the scope of this study to examine all aspects of executive functions. It is acknowledged that this may be a variable that the two groups differed on and consequently it is an important area for further study.

Given that it is difficult to determine aetiology, some children could have been 'misdiagnosed' and thus entered in the incorrect group for analysis. It should be mentioned that this issue was given careful consideration. Regardless of whether or not

the child had previously been diagnosed with obstructive HC or communicating HC, neuroradiology reports and an independent neurosurgeon confirmed the diagnosis.

Determining the effects of the type of HC is problematic. The communicating HC consisted entirely of children with IVH, secondary to low birth weight or prematurity. Consequently, these children are at greater risk of white matter injury regardless of whether or not they have HC. Therefore it is difficult to disentangle the cognitive deficits due to the white matter injury caused by communicating HC and that attributed to other factors of the condition.

The current study's sample reduces representativeness of HC because it omitted children with SB. However, children with SB have other neuropathology that may be difficulty to determine what the unique contribution is of obstructive and communicating HC on neuropsychological function.

The age range of this sample was large and developmental factors may have been impacting on the child's performance. This study does not provide sufficient information about the impact of HC at a particular age, instead it provides information about children within the 7 to 15 year age group, and consequently developmental differences are not obvious. For example, if children do more poorly on certain tasks as they get older and the chronic effects of HC are more evident, combining all the age groups together may mask this fact. It may appear as though between the age of 7 and 15 years old children with HC will have deficits in a particular domain, when in fact it is only children older than 12 years.

Though some researchers may argue that the lower IQ in both HC groups explains the poorer performances on many tasks, Fletcher, Brookshire, et al. (1996) found weak to

moderate correlations between IQ and measures of attention and executive functions. These weak correlations suggest that attention and executive tests measure variability in outcome that is not accounted for by differences in IQ. Similarly, Brewer et al. (2001) found that IQ did not significantly contribute to performance on the tests of attention. They covaried for FIQ and found that it did not alter the pattern of their results and argued that IQ scores are weak constructs as causal variables in studies of children with brain disorders. They suggested that IQ scores were better conceptualised as outcome measures, and that the processes that lead to poor performances on cognitive measures also lead to poorer performances on IQ. For example, poor performance on the TOL may reflect the fact that children with HC have difficulty with novel problem solving tasks. Alternatively, it may also be due to children with HC having difficulty learning procedures or slow to process information.

Within each domain Tukey's Honestly significant difference was used to control for the number of pairwise comparisons. It may be argued that there were a number of neuropsychological variables compared across domains, in all 24, may increase the risk of Type 1 error. Therefore, another limitation of the current study is that it did not control for Type 1 error.

4.8 Future Research

Prospective and longitudinal studies that commence close to the age of HC onset are required to determine the effects of different types and aetiologies of HC at various ages. It may also help to determine if the effect of aetiology, timing of onset, and mechanism of HC are most evident at a particular age, and how they influence cognitive

development.

Developmentally, little is known about the emergence of neuropsychological functions in children with HC, especially before school age (Erickson et al., 2001). Longitudinal studies that examine the effects of HC on cognition at different ages, commencing in the preschool years, would provide important information about the developmental course of HC. This type of study is also pertinent to developing prognostic information about the various HC aetiologies. Understanding early neuropsychological abilities may allow predictions of future challenges for the school years and lead to appropriate interventions and management (Erickson et al., 2001). Tracking the development of children with HC is important because studies have reported that as children with HC get older they fall further behind their peers cognitively, academically, socially and emotionally (Dennis et al., 1987; Erickson et al., 2001).

Developmental research would also be beneficial in examining social, emotional and behavioural difficulties and the impact of cognitive factors on these domains in children with HC. Such studies might allow insight into later emotional and social behaviour problems (Erickson, et al, 2001). For example, particular deficits in infancy may lead to more serious behavioural issues in later childhood. Early detection and rehabilitation of these social and emotional behavioural problems may avoid greater problems at a later stage. This is important because behavioural problems have implications for socialization and school placement, such that challenging behaviours in a child may preclude them from attending a mainstream school even if they have sufficient intellectual abilities to manage in a mainstream school (Erickson et al., 2001).

Another area of behaviour that has been reported to be deficient in children with

HC is adaptive living skills (Fletcher, Brookshire, et al.,1995; Hurley, Dorman, Laatsch, Bell & D'Avignon, 1990). The fact that children with HC have long-term monitoring and invasive neurosurgical treatment, often have academic difficulties and can be a burden on their family are factors for the development of adjustment difficulties (Fletcher & Levin, 1988). Despite this there are few studies that have examined this issue in children with HC. Therefore, future research on behavioural outcomes in children with HC is much needed. On possible area of study would be to examine adaptive skills in children with obstructive and communicating HC to see if they can be differentiate on this domain. Instruments such as the Vineland Adaptive Behavioural Scale could be utilised to provide quantitative assessment of these skills.

This study only identified subtle differences between the two types of HC, future research could examine if cognitive differences are related to co-morbid conditions or medical complications such as seizures or PVL. Investigating these factors may provide some explanation for the variability observed in the cognitive profile of children with HC.

An important research question raised by previous research is that of the cause of executive function deficits in children with HC. Determining whether they are due to posterior attention system dysfunction, disruption in the connections to the prefrontal cortex, a combination of both, or another reason altogether, may assist with our understanding of the pathophysiology of HC. Functional MRI studies, combined with cognitive assessment, would identify the neuroantomical regions utilised by children with HC whilst completing executive function tasks. To study the nature of attentional deficits and their impact on their cognition and behaviour, observational assessments of

behaviour in children with brain injury have been suggested. It has been argued that this type of dynamic assessment has greater ecological validity than psychometric tests, and give a clearer picture (Barkley, 1994; Fletcher, Brookshire et al., 1996). The relationship between IQ and other measures may be an interesting area for future research.

Future research would assist understanding the effects of HC and the associated aetiologies on the brain, and how they contribute to the various neuropsychological profiles evident. To examine this issue, research should incorporate cognitive measures and quantitative MRI-measurements of different brain structures and patterns of changes in gray and white matter (Fletcher, Brookshire, et al., 1996). It may be beneficial for future research to include techniques that are more sensitive to white matter injury such as diffusion tensor imaging. The use of these more sensitive neuroimaging procedures might reveal a greater degree of white matter injury in the children with HC that on less sensitive procedures is undetectable.

Though the current findings have added to the limited research on attention and executive functions in children with HC it is still unclear how these cognitive constructs influence the neuropsychological function in this population. Consequently, further empirical research into the nature of these skill deficits is required (Anderson, Northam, et al., 2001; Brewer et al., 2001; Erickson et al., 2001; Fletcher, Brookshire, et al., 1996; Fletcher, et al., 2000).

REFERENCES

Abercrombie, J.L.J. (1968). Some notes on disability: Movement, intelligence quotient and attentiveness. *Developmental Medical Child Neurology, 10,* 206-213.

Almeida, G.M., Matushita, H., Mattosinho-Franca, L.C., & Shibata, M.K. (1990). Dandy-Walker syndrome: posterior fossa craniectomy and cyst fenestration after several shunt revisions. *Child's Nervous System, 6,* 335-337.

Ammirati, M., & Raimondi, A. (1987). Cerebrospinal fluid shunt infections in children. A study on the relationship between the etiology of hydrocephalus, age at the time of shunt placement, and infection rate. *Child's Nervous System, 3,* 106-109.

Anderson, E.M. (1975). Cognitive deficits in children with spina bifida and hydrocephalus: A review of the literature. *British Journal of Educational Psychology, 45,* 257-268.

Anderson, V. (1998). Assessing executive functions in children: Biological, physiological, and developmental considerations. *Neuropsychological Rehabilitation, 8,* 319-350.

Anderson, P., Anderson, V., & Garth, J. (2001). Assessment and development of organizational ability: The Rey complex figure organizational strategy score (RCF-OSS). *The Clinical Neuropsychologist, 15(1),* 81-94.

Anderson, P., Anderson, V., & Lajoie, G. (1995). The tower of London test: Validation and standardization for pediatric populations. *The Clinical Neuropsychologist, 10,* 54-65.

Anderson, V., Anderson, P., Northam, E., Jacobs, R., & Mikiewicz, O. (2002). Relationships between cognitive and behavioural measures of executive function in children with brain disease. *Child Neuropsychology, 8(4),* 231-240.

Anderson, V., Anderson, P., Northam, E., Jacobs, R., & Catroppa, C. (2001). Development of executive functions through late childhood and adolescence in an Australian sample. *Developmental Neuropsychology, 20(1),* 385-406.

Anderson, P., Anderson, V., Northam, E., & Taylor, H.G. (2000). Standardization of the Contingency Naming Test (CNT) for school-aged children: A measure of reactive flexibility. *Clinical Neuropsychological Assessment, 1,* 385-406.

Anderson, V., Lajoie, G., & Bell, R. (1996). *Neuropsychological Assessment of the School-Aged Child.*

Anderson, V., Northam, E., Hendy, J., & Wrennall, J. (2001). *Developmental Neuropsychology: A Clinical Approach.* East Sussex: Psychology Press.

Bannister, C.M., Russell, S.A., Rimmer, S., & Mowke, D.H. (1999). Fetal arachnoid cysts: Their site, progress, prognosis, and differential diagnosis. *European Journal of Pediatric Surgery, 9(1),* 27-28.

Bannister, C.M., Russell, S.A., Rimmer, S., & Arora, A. (2000). Pre natal ventriculomegaly and hydrocephalus. *Neurological Research, 22,* 37-42.

Barkovich, A. J. (1990). *Paediatric Neuroimaging.* New York: Raven Press.

Barkovich, A. J. (1995). *Paediatric Neuroimaging (Volume 2).* New York: Raven Press.

Barkovich, A.J., & Newton, T.H. (1988). Anomalies of the corpus callosum: Correlation with further anomalies of the brain. *Journal of Neuroradiology, 15,* 171-179.

Barkovich, A.J., & Newton, T.H. (1989). MR of aqueductal stenosis: Evidence of a broad spectrum of tectal distortion. *American Journal of Neuroradiology, 10,* 471-476.

Barnes, M.A., & Dennis, M. (1998). Discourse after early-onset hydrocephalus: Core deficits in children of average intelligence. *Brain and Language, 61,* 309-334.

Baron, I.S., & Goldberger, E. (1993). Neuropsychological disturbances of hydrocephalic children with implications for special education and rehabilitation. *Neuropsychological Rehabilitation, 3(4),* 389-410.

Bier, J., Morales, Y., Liebling, J., Geddes, L., & Kim, E. (1997). Medical and social factors associated with cognitive outcome in individuals with myelomeningocele. *Developmental Medicine and Child Neurology, 39,* 263-266.

Blakemore, C., & Eggers, H.N. (1978). Animal models and human visual development. In S.G. Cool & L. Smith (Eds.), *Frontiers in Visual Science.* New York: Springer Verlag.

Bourgeois, M., Sainte-Rose, C., Cinalli, G., Maixner, W., Malucci, C., Zerah, M., Pierre-Kahn, A., Renier, D., Hoppe-Hirsch, E., Aicardi, J. (1999). Epilepsy in children with shunted hydrocephalus. *Journal of Neurosurgery, 90* (2), 274-281.

Bradley, W. G., Whittemore, A.R., Watanabe, A.S., Davis, S.J., Teresi, L.M., & Homyak, M. (1991). Association of deep white matter infarction with chronic communicating hydrocephalus. Implications regarding the possible origin of normal-pressure hydrocephalus. *American Journal of Neuroradiology, 12,* 31-39.

Brewer, V.R., Fletcher, J.M., Hiscock, M., & Davidson, K.C. (2001). Attention

processes in children with shunted hydrocephalus versus attention deficit-hyperactivity disorder. *Neuropsychology, 15*(2), 185-198.

Brookshire, B., Fletcher, J.M., Francis, D.J., Thompson, N., Bohan, T.P., Landry, S.H., Davidson, K.C., & Milner, M.E. (1993). Language functions in children with hydrocephalus. *Journal of Clinical and Experimental Neuropsychology 15,* 58–72.

Brookshire, B. L., Fletcher, J.M., Bohan, T.P., Landry, S.H., Davidson, K.C., & Francis, D.J. (1995a). Specific language deficiencies in children with early onset hydrocephalus. *Child Neuropsychology, 1(2),* 106-117.

Brookshire, B. L., Fletcher, J.M., Bohan, T.P., Landry, S.H., Davidson, K.C., & Francis, D.J. (1995b). Verbal and nonverbal discrepancies in children with hydrocephalus: A five-year longitudinal follow-up. *Journal of Pediatric Psychology 20(6),* 785-800.

Bryan, D. (1994). Neural tube defects. In M.J. Robinson & D. M. Robertson (Eds.), *Practical Paediatrics.* Melbourne: Churchill-Livingstone.

Buschke, H. (1974). Components of verbal learning in children: Analysis by selective reminding. *Journal of Experimental Child Psychology, 18,* 488-496.

Choudhury, A.R. (1995). Infantile hydrocephalus: Management using CT assessment. *Child's Nervous System, 11,* 220-226.

Chuang, S. (1986). Perinatal and neonatal hydrocephalus: Part 1. Incidence and aetiology. *Perinatal Neonatology, 10,* 8-19.

Circillo, S. F., Coegen, P.H., Harsh, G.R. & Edwards, M.S.B. (1991). Intracranial arachnoid cysts in children: A comparison of the effects of fenestration and

shunting. *Journal of Neurosurgery 74,* 230-235.

Cooley, E.L., & Morris, R.D. (1990). Attention in children: A neuropsychologically based model of assessment. *Developmental Neuropsychology, 6,* 239-274.

Corkhill, R. G., & Cadoux-Hudson, T.A.D. (1999). Normal pressure hydrocephalus: developments in determining surgical prognosis. *Current Opinion in Neurology. 12(6),* 671-677.

Cull, C., & Wyke, M. (1984). Memory function of children with spina bifida and shunted hydrocephalus. *Developmental Medicine and Child Neurology, 26,* 177-183.

Czosnyka, M., Copeman, J., Czosnyka, Z., McConnell, R.S., Dickinson, C., & Pickard J.D. (2000). Post-traumatic hydrocephalus: influence of craniectomy on the CSF circulation. *Journal of Neurology, Neurosurgery and Psychiatry. 68(2),* 246-248.

Daniel, A. (1983). *Power, Privilege, and Prestige: Occupations in Australia.* Melbourne: Longman-Chesire.

Del Bigio, M. (1993). Neuropathological changes caused by hydrocephalus. *Acta Neuropathologica, 85,* 573-585.

Denckla, M. B., & Rudel, R. (1974). Rapid "automatized" naming of pictured objects, colors, letters and numbers by normal children. *Cortex, 10,* 186-202.

Dennis, M., & Barnes, M.A. (1993). Oral discourse after early-onset hydrocephalus: Linguistic ambiguity, figurative language, speech acts, and script-based inferences. *Journal of Pediatric Psychology, 18(5),* 639-652.

Dennis, M., Fitz, C.R., Netley, C.T., Sugar, J., Harwood-Nash, D.C.F., Hendrick, E.B., Hoffman, H.J., & Humphreys, R.P. (1981). The intelligence of hydrocephalic

children and adolescents. *Archives of Neurology, 38,* 607-615.

Dennis, M., Hendrick, E.B., Hoffman, H.J., & Humphreys, R.P. (1987). Language of hydrocephalic children and adolscents. *Journal of Clinical and Experimental Neuropsychology, 9,* 593-621.

Dennis, M., Jacennik, B., & Barnes, M.A. (1994). The content of narrative discourse in children with adolescents after early-onset hydrocephalus and in normally developing peers. *Brain and Langauge, 46,* 129-165.

Ding, Y., Lai, Q., McAllister, J.P., & Canady, A.I. (2001). Impaired motor learning in children with hydrocephalus. *Pediatric Neurosurgery, 3,* 182-189.

Dise, J.E., & Lohr, M.E. (1998). Examination of deficits in conceptual reasoning abilties associated with spina bifida. *American Journal of Physical Medicine and Rehabilitation, 77,* 247-251.

Donders, J., Canady, A.I., & Rourke, B.P. (1990). Psychometric intelligence after infantile hydrocephalus. *Child's Nervous System, 6,* 148-154.

Donders, J., Rourke, B.P., & Canady, A. (1991). Neuropsychological functioning of hydrocephalic children. *Journal of Clinical and Experimental Neuropsychology, 13* (4), 607-613.

Drachman, D.A., & Richardson, E.P. Jr. (1961). Aqueductal narrowing, congenital and acquired. A critical review of the histological criteria. *Archives of Neurology, 5,* 552-559.

Eslinger, P., & Grattan, L. (1993). Frontal lobe and frontal-striatal substrates for different forms of developmental changes in human infants. *Behavioural Neuroscience, 10 (3),* 526-537.

Erickson, K., Baron, I.S., & Fantie, B.D. (2001). Neuropsychological functioning in early hydrocephalus: Review from a developmental perspective. *Child Neuropsychology, 7(4),* 199-229.

Ewing-Cobbs, L., Thompson, N.M., Miner, M.E., & Fletcher, J.M. (1994). Gunshot wounds to the brain in children and adolescents: Age and neurobehavioural development. *Neurosurgery, 35,* 225-233.

Fennell, E.B., Eisenstadt, T., Bodiford, C., Redeiss, S., & Mickle, J. (1987). The assessment of neuropsychological dysfunction in children shunted for hydrocephalus. *Journal of Clinical and Experimental Neuropsychology, 9,* 25-26.

Fernell, E., Gillberg, C., & von Wendt, L. (1991). Behavioural problems in children with infantile hydrocephalus. *Developmental Medicine and Child Neurology, 33,* 388-395.

Fewell, M. E., Levy, M.L., & McComb, J.G. (1996). Surgical treatment of 95 children with 102 intracranial arachnoid cysts. *Pediatric Neurosurgery, 25,* 165-173.

Filka, J., Huttova, M., Tuharsky, J., Sagat, T., Kralinsky, K., & Krcmeery Jnr, V. (1999). Nosocomial meningitis in children after ventriculoperitoneal shunt insertion. *Acta Paediatrica, 88,* 576-578.

Fletcher, J. M. (1998). Attention in children: Conceptual and methodological issues. *Child Neuropsychology, 4(1),* 81-86.

Fletcher, J., Bohan, T.P., Brandt, M.E., Brookshire, B.L., Beaver, S.R., Francis, D.J., Davidson, K.C., Thompson, N.M., & Miner, M.E. (1992). Cerebral white matter and cognition in hydrocephalic children. *Archives of Neurology, 49,* 818-824.

Fletcher, J., Bohan, T.P., Brandt, M.E., Kramer, L.A., Brookshire, B.L., Thorstad, K.,

Davidson, K.C., Francis, D.J., McCauley, S.R., & Baumgartner, J.E. (1996). Morphometric evaluation of the hydrocephalic brain: relationships with cognitive development. *Child's Nervous System, 12,*192-199.

Fletcher, J., Brookshire, B.L., Bohan, T.P., Brandt, M.E., & Davidson, K.C. (1995). Early Hydrocephalus. In B.P.Rourke.(Ed.), *Syndrome of Nonverbal Learning Disabilities: Neurodevelopmental Manifestations* (pp 25-46). New York: Guilford Press.

Fletcher, J., Brookshire, B.L., Landry, S.H., Bohan, T.P., Davidson, K., Francis, C., Thompson, N.M., & Miner, M.E. (1995). Behavioural adjustment of hydrocephalic children: Relationships with etiology, neuropsychological, and family status. *Journal of Pediatric Psychology, 20,* 765-781.

Fletcher, J.M., Brookshire, B.L., Landry, S.H., Bohan, T.P., Kramer, L.A., & Morris, R.D. (1996). Attentional skills and executive functions in children with early treated hydrocephalus. *Developmental Neuropsychology, 12(1),* 53-76.

Fletcher, J.M., Dennis, M., & Northrup, H. (2000). Hydrocephalus. In K.O Yeates, M.D., Ris, & G. Taylor (Eds.), *Paediatric Neuropsychology: Research, Theory and Practice.* New York: Guilford Press.

Fletcher, J. M., Francis, D.J., Thompson, N.M., Brookshire, B.L., Bohan, T.P., Landry, S.H., Davidson, K.C., & Miner, M.E. (1992). Verbal and nonverbal skill discrepancies in hydrocephalic children. *Journal of Clinical and Experimental Neuropsychology,14(4),* 593-609.

Fletcher, J. M., Landry, S.H., Bohan, T.P., Davidson, K.C., Brookshire, B.L., Lachar, D., Kramer, L.A., & Francis, D.J. (1997). Effects of intraventricular hemorrhage and

hydrocephalus on the long-term neurobehavioural development of preterm very-low-birthweight infants. *Developmental Medicine and Child Neurology, 39,* 596-606.

Fletcher, J. M., Levin, H.S., & Butler, I.J. (1995). Neurobehavioral effects of brain injury on children: Hydrocephalus, traumatic brain injury, and cerebral palsy. In, M.C. Roberts (Ed.), Handbook *of Pediatric Psychology* (2nd ed., pp. 362-383).

Fletcher, J. M., McCauley, S.R., Brandt, M.E., Bohan, T.P., Kramer, L.A., Francis, D.J., Thorstad, K., & Brookshire, B.L. (1996). Regional brain tissue composition in children with hydrocephalus: Relationships with cognitive development. *Archives of Neurology 53,* 549-557.

Foltz, E.L., & Shurtleff, D.B. (1963). Five-year comparative study of hydrocephalus in children with and without operation. *Journal of Neurosurgery, 65,* 211-216.

Friede, R.L. (1989). *Developmental Neuropathology.* Berlin, Springer-Verlag.

Friedrich, W.N., Lovejoy, M.C., Shaffer, J., Shurtleff, D.B., & Beilke, R.L. (1991). Cognitive abilities and achievement status of children with myelomeningocele: A contemporary sample. *Journal of Pediatric Psychology, 16(4),* 423-428.

Gardner, M.F. (1988). *Test of Visual –Perceptual Skills.* San Francisco: Health.

Gascon, G. G., & Leech, R.W. (1991). Clinical Presentation. *Hydrocephalus: Current Clinical Concepts.* St. Louis.

Gilles, F.H., & Davidson, R.I. (1971). Communicating Hydrocephalus associated with deficient dysplastic parasagittal arachnoid granulations. *Journal of Neurosurgery, 35,* 421.

Gilkes, C.E., Steers, A.J.W., & Minns, R.A. (1999). A classification of CSF shunt

malfunction. *European Journal of Pediatric Surgery, 9*(Suppl. 1), 19-22.

Gioia, G.A., Isquith, P.K., Retzlaff, P.D., & Pratt, B.M. (2000). *The Behavioural Rating of Executive Function.* Odessa, Florida: PAR.

Golberg, R., & Costa, L. (1981). Hemisphere differences in the acquisition and use of descriptive systems. *Brain and Language, 14,* 144-173.

Golden, J.A., Rourke, L.B., & Bruce, D.A. (1987). Dandy-Walker syndrome and associated anomalies. *Pediatric Neuroscience, 13*, 38-44.

Guyot, L.L., & Michael, D.B. (2000). Post-traumatic hydrocephalus. *Neurology Research, 22,* 25-28.

Hadenius, A., Hagberg, B., Hyttnas-Bensch, K., & Sjorgen, I. (1962). The natural prognosis of infantile hydrocephalus. *Acta Paediatrica, 51,* 117-118.

Hannay, J. (2000). Functioning of the corpus callosum in children with early treated hydrocephalus. *Journal of the International Neuropsychological Society, 6*, 351-361.

Hanlo, P.W., Gooskens, R.J., van Schooneveld, M., Tulleken, C.A., van der Knaap, M.S., Faber, J.A., & Willemse, J. (1997). The effect of intracranial pressure on myelination and the relationship with neurodevelopment in infantile hydrocephalus. *Developmental Medicine and Child Neurology, 39,* 286-291.

Heilman, K.M., Watson, R.T., & Valenstein, E. (1993). Neglect and related disorders. In K.M. Heilman & E. Valenstein (Eds.), *Clinical Neuropsychology.* (3rd ed, pp 279-336). New York: Oxford University Press.

Hein, A. (1978). Eye motility and visual motor development. In S.J Cool. & E.L. Smith (Eds.), *Frontiers in Visual Science.* New York: Springer Verlag.

Hetherington, R., & Dennis, M. (1999). Motor function profile in children with early onset hydrocephalus. *Developmental Neuropsychology, 15*(1), 25-51.

Holler, K.A., Fennell, E.B., Crosson, B., Boggs, S.R., & Mickle, J.P. (1995). Neuropsychological and adaptive functioning in younger versus older children shunted for early hydrocephalus. *Child Neuropsychology, 1,* 63-73.

Hommet, C., Billard, C., Gillet, P., & Barthez, M.A. (1999). Neuropsychologic and adaptive functioning in adolescents and young adults shunted for congenital hydrocephalus. *Journal of Child Neurology 14(3),* 144-150.

Hommet, C., Cottier, J.P., Billard, C., Perrier, D., Gillet, P., De Toffol, B., Sirinelli, D., Bertrand, P., & Autret, A. (2002). MRI morphometric study and correlation with cognitive functions in young adults shunted in congenital hydrocephalus related to spina bifida. *European Neurology, 47*(3), 169-174.

Hoon, A., & Melham, E.R. (2000). Neuroimaging: Applications in disorders of early brain development. *Developmental and Behavioural Pediatrics. 21*(4), 291-302.

Horn, D.G., Lorch, E.P., Lorch, R.F., Jr., & Culatta, B. (1985). Distractibility and vocabulary deficits in children with spina bifida and hydrocephalus. *Developmental Medicine and Child Neurology, 27,* 713-720.

Houliston, M.J., Taguri, A.H., Dutton, G.N., Hajivassilious, C., & Young, D.G. (1999). Evidence of cognitive visual problems in children with hydrocephalus: a structured clinical history-taking strategy. *Developmental Medicine and Child Neurology, 4,* 298-306.

Hunt, GM. & Holmes, A.E. (1976). Factors relating to intelligence in treated cases of spina bifida cystica. *American Journal of Diseases of Children, 130,* 823-827.

Imielinski, B. L., Kloc, W., Wasilewski, W., Liczbik, W., Puzyrewski, R., Karwacki, Z. (1998). Posterior fossa tumors in children - indications for ventricular drainage and for V-P shunting. *Child's Nervous System 14,* 227-229.

Jackson, P.H., & Lorber, J. (1984). Brain and ventricular volume in hydrocephalus. *Zeitshrift fur Kinderchirargie, 39,* 91-93.

Jacobs, R., Northam, E., & Anderson, V. (2001). Cognitive outcome in children with myelomeningocele and perinatal hydrocephalus: A longitudinal perspective. *Journal of Developmental and Physical Disabilities, 13(4),* 389-405.

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

Kestle, J., Cochrane, D., & Alisharan, R. (2000). The initial treatment of hydrocephalus: an assessment of surgeons' preference between third ventriculostomy and shunt insertion. *Neurological Research, 22*(1): 65-68.

Johnston, I. (1995). *Hydrocephalus: A Handbook for Parents and Patients.* Australia: Rawson Graphics.

Klaas, P.A., Hannay, J., Caroselli, J.S., & Fletcher, J.M. (1999). Interhemispheric transfer of visual, auditory, tactile, and visuomotor information in children with hydrocephalus and partial agenesis of the corpus callosum. *Journal of Clinical and Experimental Neuropsychology, 21(6),* 837-850.

Landry, S.H., Jordan, T., & Fletcher, J.M. (1994). Developmental outcomes for children with spina bifida. In M.G. Tramontana, & S.R. Hooper. (Eds.), *Advances in Neuropsychology (Volume 2).* New York: Springer Verlag.

Laurance, K.M. (1969). Neurological and intellectual sequelae of hydrocephalus. *Archives of Neurology, 20,* 73-81.

Lee, M., Wisoff, J.H., Abbott, R., Freed, D., & Epstein, F.J. (1994). Management of hydrocephalus in children with medulloblastoma: prognostic factors for shunting. *Paediatric Neurosurgery, 20(4),* 240-7.

Leech, R. A., & Goldstein, E. (1991). Classification and mechanisms. *Hydrocephalus: Current clinical concepts.* R. W. L. R. A. Brumback. St. Louis, MO, Mosby - Year Book: 45-70.

Lezak, M.D. (1995). Neuropsychological Assessment. (3rd ed.). New York: Oxford University Press.

Lindgren, S.D., & Benton A.L. (1980). Developmental patterns of visuospatial judgment. *Journal of Pediatric Psychology, 5,* 217-225.

Lollar, D.J. (1990). Learning patterns among spina bifida children. *Zeitschrift fur Kinderchirugie, 45* (Supp. 1), 39.

Lonton, A.P., Barrington, N.A., & Lorber, J. (1975). Lacunar skull deformity related to intelligence in children with myelomeningocele and hydrocephalus. *Developmental Medicine and Child Neurology, 35,* 58.

Lorber, J. (1980). Is your brain really necessary? *Science, 4,* 7-20.

Mahone,.E.M., Zabel, A.T., Levey, E., Verda, M., & Kinsman, S. (2002). Parent and self-report ratings of executive function in adolescents with myelomeningocele. *Child Neuropsychology, 8* (4), 258-270.

Manly, T., Robertson, I.H., Anderson, V., & Nimmo-Smith, I. (1999). *The Test of Everyday Attention for Children Manual.* Suffolk: Thames Valley Test Company.

Manly, T., Anderson, V., Nimmo-Smith, I., Turner, A., Watson, P., & Robertson, I.H., (2001). The differential assessment of children's attention: The Test of Everyday Attention for Children (TEA-Ch), normative sample and ADHD performance. *Journal of Child Psychology and Psychiatry, 42* (8), 1065-1081.

Martinez-Lage, J.F., Lopez, F., Poza, M., & Hernadez, M. (1998). Prevention of intraventricular haemorrhage during CSF shunt revisions by means of a flexible coagulating electrode. *Child's Nervous System, 14,* 203-206.

McAllister, J.P. (2000). Hydrocephalus enters the new millennium: An overview. *Neurological Research, 22,* 2-3.

McAllister, J., & Chovan, P. (1998). Neonatal hydrocephalus. Mechanisms and consequences. *Neurosurgical Clinics of North America, 9, 73-83.*

McCullough, D.C., & Balzer-Martin, L.A. (1982). Current prognosis in overt neo-natal hydrocephalus. *Journal of Neurosurgery, 57,* 378-383.

Menkes, K.J. (1990). *Textbook of Child Neurology* (4th ed.). Philadelphia: Lea & Febinger.

Ment, L., Vohr, B., Allan, W., Katz, K.H; Schneider, K.C; Westerveld, M.; Duncan, C.C., & Makuch, R.W. (1999). The etiology and outcome of cerebral ventriculomegaly at term in very low birthweight preterm infants. *Pediatrics 104,* 243-248.

Mirsky, A.F. (1987). Behavioural and psychophysiological markers of disordered attention. *Environmental Health Perspectives, 74,* 191-199.

Mirsky, A.F. (1996). Disorders of attention: A neuropsychological perspective. In R.G. Lyon & N.A. Krasnegor (Eds.), *Attention, Memory, and Executive Functions* (71-

96). Baltimore: Brookes.

Mirsky, A.F., Anthony, B.J., Duncan, C.C., Ahearn, M.B., & Kellam, S.B. (1991). Analysis of the elements of attention : A neuropsychological approach. *Neuropsychological Review, 2,* 109-145.

Mori, K., (2000). Actualities in hydrocephalic classification and management possibilities. *Neuropsychological Research, 22,* 127-130.

Neuhaus, G., Forrman, B.R., Francis, D.J., & Carlson, C.D. (2001). Measures of information processing in rapid automatized naming (RAN) and their relation to reading. *Journal of Experimental Child Psychology, 78,* 359-373.

Nolte, J. (1999). *The Human Brain. An Introduction to Its Functional Neuroanatomy.* (4th ed.). St Louis: Mosby Inc.

Oi, S., Yamada, H., Sato, O., & Matsumoto, S. (1996). Experimental models of congenital hydrocephalus and comparable clinical problems in the fetal and neonatal periods. *Child's Nervous System, 12,* 292-302.

Oka, Y, Yamamoto, M., Nagasaka, S., & Tomonaga, M. (1994). Endoneurosurgical treatment for hydrocephalus caused by intraventricular tumors. *Child's Nervous System, 10,* 162-166.

Osterreith, P. (1944). Le test de copi d'lune figure complexe. *Archives de Psychologie,* 30, 206-356.

Osterreith, P. (1993). The complex figure copy test. *The Clinical Neuropsychologist, 7,* 3-21.

Posner, M.I. & Deheane, S. (1994). Attentional networks. *Trends in Neuroscience, 17,* 75-79.

Posner, M.I. & Peterson, (1990). The attention system of the human brain. *Annual Review of Neuroscience, 13,* 25-42.

Posner, M.I., Walker, J.A., Friedrich, F.J. & Rafal, R.D. (1984). Effects of parietal lobe injury on covert orienting of visual attention. *Journal of Neuroscience, 4,* 1863-1874.

Prigatano, G.P., Zeiner, H.K., Pollay, M., & Kaplan, R.J. (1983). Neuropsychological functioning in children with shunted and uncomplicated hydrocephalus. *Child's Brain, 10,* 112-120.

Rahman, N., Murshid, W.R., Jamjoom, Z.A., & Jamjoom, A. (1993). Neurosurgical management of intraventricular haemorrhage in preterm infants. Journal of the Pakistan Medical Association, 43*(10), 195-200.*

Raimondi, A.J. (1994). A unifying theory for the definition and classification of hydrocephalus. Children's Nervous System, 10, *2-12.*

Ralph, K., Moylan, P., Canady, A., & Simmons, S. (2000). The effects of multiple shunt revisions neuropsychological functioning and memory. Neuropsychological Research, 22, 131-136.

Reiner, D., Sainte-Rose, C., Pierre-Kahn, A., & Hirsch, J.F. (1988). Prenatal hydrocephalus: Outcome and prognosis. Child's Nervous System, 14, 10-14.

Rey, A. (1964). L'examine psycholgique dans les cas d'encephalopathie tramatique. Archives de Psychologique, 28, 286-340.

Riva, D., Milani, N., Giorgi, C., Panatleoni, C., Zorzi, C., & Devoti, M. (1994). Intelligence outcomes in children with shunted hydrocephalus of different etiology. Child's Nervous System, 10, 70-73.

Roth, S. C., Baudin, J., McCormick, D.C., Edwards, A.D., Stewart, A.L., & Reynolds, E.O. (1993). Relation between ultrasound appearance of the brain of very preterm infants and neurodevelopmental impairment at eight years. Developmental Medicine and Child Neurology, 35, 755-768.

Rourke, B.P. (1987). Syndrome of non-verbal learning disabilities: The final common pathway of white matter disease/dysfunction. The Clinical Neuropsychologist, 1, *209-234.*

Rourke, B.P. (1989). Nonverbal learning disability. The syndrome and the model. *New York: Guilford Press.*

Rourke, B.P. (1995). Syndrome of Non-Verbal Learning disabilities. *New York: Guilford Press.*

Rubin, R., Hochwald, G., Tiell Liwnicz, B., & Epstein, B. (1975). Reconstitution of the cerebral cortical mantle in shunt-corrected hydrocephalus. *Developmental Medicine and Child Neurology, 17,* 151-156.

Schiottz-Christiensen, E., & Bruhn, P. (1973). Intelligence, behaviour and scholastic achievement subsequent to febrile convulsions: An analysis of discordant twin pairs. *Developmental Medicine and Child Neurology, 15,* 565-575.

Schmidt, F.L. (1996). Statistically significant testing and cumulative knowledge in psychology: Implications for training or researchers. *Psychological Methods, 1,* 115-129.

Scott, M., Fletcher, J.M., Brookshire, B.J., Davidson, K.C., Landry, S.H., Bohan, T.P., Kramer, L., Brandt, M., & Francis, D.J. (1998). Memory functions in children with early hydrocephalus. *Neuropsychology, 12,* 578-589.

Segev, Y., Metser, U., Beni-Adani, L., Elran, C., Reider-Grosswater, I., & Constantini, S. (2001). Morphometric study of eh midsagittal MR imaging plane in cases of hydrocephalus and atrophy in normal brains. *American Journal of Neuroradiology, 22,* 1674-1679.

Shaffer, J., Wolfe, L., Friedrich, W., Shurtleff, H., Shurtleff, D., & Fay, G. (1986). Developmental expectations: Intelligence and fine motor skills. In D. Shurtfleff (Ed.), *Myelodysplasias & Extrophies: Significance, Prevention and Treatment.* (359-373). New York: Grune & Stratton.

Shallice, T. (1983). Specific impairments of planning. *Philosophical Transcripts of the Royal Society of London, 298*, 199-299.

Shurtleff, A.B., Foltz, E.L., & Loeser, J.B. (1973). Hydrocephalus: A definition of the progression and relationship to intellectual function. A*merican Journal of Diseases of Children, 125,* 688-693.

Snow, J.H., Prince, M., Souheaver, G., Ashcraft, E., Stefans, V., & Edmonds, J. (1994). Neuropsychological patterns of adolescents and young adults of spina bifida. *Archives of Clinical Neuropsychology, 9,* 227-287.

Stuss, D.T., & Benson, D.F. (1986). *The Frontal Lobes.* New York: Raven Press.

Tew, B. (1979). The "Cocktail Party Syndrome" in children with hydrocephalus and Spina Bifida. *British Journal of Disorders of Communication, 14(2)*, 89-101.

Taylor, H.G., Hack, M., & Klein, N. (1998). Attention deficits in children with <750gm birthweight. *Child Neuropsychology, 4,* 21-34.

Tew, B., & Laurence, K.M. (1975). The effects of hydrocephalus on intelligence, visual perception and school attainment. *Developmental Medicine and Child Neurology,*

17, (35), 129-134.

Tew, B., & Laurence, K.M. (1983). The relationship between spina bifida children's intelligence test scores on school entry and at school leaving: A preliminary report. *Child Health Care and Development, 9,* 13-17.

Tew, B., Laurance, K., & Richards, A. (1980). Inattention among children with hydrocephalus and spina bifida. *Zeitschrift fur Kinderchirurgie, 31,* 381-385.

Thomas, D.G. (1992). Outcome of paediatric bacterial meningitis 1979-1989. M*edical Journal of Australia, 19, 519-520.*

Thompson, M.G., Eisenberg, H.M., & Levin, H.S. (1982). Hydrocephalic infants: Developmental assessment and computed tomography. *Child's Brain, 9,* 400-410.

Thompson, N.M., Fletcher, J.M., Chapieski, L., & Landry, H. (1991). Cognitive motor abilities in preschool hydrocephalics. *Journal of Clinical and Experimental Neuropsychology, 13(2),* 245-258.

Tortora, G.J., & Grabowski, S.R. (1993). Principles of Anatomy and Physiology (7^{th} ed.). New York: HarperCollins.

Tuli, S., Drake, J., Lawless, J., Wigg, M., & Lamberti-Pasculli, M. (2000). Risk factors for repeated cerebrospinal shunt failures in pediatric patients with hydrocephalus. *Journal of Neurosurgery 92,* 31-38.

Tullberg, M., Jensen, C., Ekholm, S., & Wikkelsø, C. (2001). Normal pressure hydrocephalus: Vascular white matter changes on MR images must not exclude patients from shunt surgery. *American Journal of Neuroradiology, 22,* 1665-1673.

van der Knaap, M.S., & Valk, J. (1990). MR Imaging of the various stages of normal myelination during the first year of life. *Neuroradiology, 31,* 459-470.

van der Knaap, M.S., Valk, J., Bakker, C.J., Schooneveld, M., Faber, J.A.J., Willemse, J., & Gooskens, R.H.J.M. (1991). Myelination as an expression of the functional maturity of the brain. *Developmental Medicine and Child Neurology, 33,* 849-857.

Victor, M., & Ropper, A.H. (2001). *Adam's and Victor's Principles of Neurology* (7th ed.). USA: McGraw-Hill.

Welch, K., & Lorenzo, A. (1991). Pathology of hydrocephalus. In Bannister, C., & Tew, B. (Eds.), *Current Concepts in Spina Bifida and Hydrocephalus* (pp. 55-82). New York: Cambridge University Press.

Wechsler, D (1991). *Manual of the Wechsler Scale of Children's Intelligence-III.* San Antonio, TX: Psychological Corporation.

Wiig, E. H., Zureich, P., & Chan, H.H. (2000). A clinical rationale for assessing rapid automatized naming in children with language disorders. *Journal of Learning Disabilities. 33*(4), 359-374.

Wills, K. (1993). Cognitive functioning in children with spina bifida and/or hydrocephalus. *Journal of Clinical Child Psychology, 22,* 247-265.

Wills, K.E., Holmbeck, G., Dillon, K., & McLone, D. (1990). Intelligence and achievement in children with myelomeningocele. *Journal of Pediatric Psychology, 15,* 161-167.

Yeates, K.O., Enrile, B., Loss, N., Blumenstein, E., & Delis, D.C. (1995). Verbal leaning and memory in children with meningomyelocele. *Journal of Pediatric*

Psychology, 20, 801-812.

Yeates, K.O, Loss, N., Colvin, A.N., & Enrile, B.G. (2003). Do children with myelomeningocele and hydrocephalus display nonverbal learning disabilities? An empirical approach to classification. *Journal of the International Neuropsychological Society, 9*, 653-662.

Young, H.F., Nulsen, F.E., & Weiss, M.H. (1973). The relationship of intelligence and cerebral mantle in treated infantile hydrocephalus. *Paediatrics, 52*, 54-60.

List of Appendices

Appendix A:

RCH Notification of Human Research Ethics Approval

Melbourne, Victoria

Department **ETHICS IN HUMAN RESEARCH COMMITTEE**

Flemington Road,
Parkville, Victoria, 3052
Australia.
Telephone: (03) 9345 5522
Facsimile: (03) 9345 5789

APPROVAL

EHRC REF. No: **98052 B**

PROJECT TITLE: Neurological and neuropathological correlates of early-treated hydrocephalus

INVESTIGATOR(S): E Northam, P Anderson, J Rosenfeld

DATE OF MODIFICATION APPROVAL: **18 February 2000**

DURATION: 24 months

SIGNED: [redacted]
COMMITTEE REPRESENTATIVE

18/2/2000
DATE

CONDITIONS

ALL PROJECTS

1. Any proposed change in protocol and the reasons for that change, together with an indication of ethical implications (if any), must be submitted to the Ethics in Human Research Committee for approval.
2. The Principal Investigator must notify the Secretary of the Ethics in Human Research Committee of:
   - Actual starting date of project.
   - Any adverse effects of the study on participants and steps taken to deal with them.
   - Any unforeseen events.
3. A progress report must be submitted annually and at the conclusion of the project, with special emphasis on ethical matters.

DRUG TRIALS

4. The investigators must maintain all records relating to the study for a period of 23 years.
5. The investigator(s) must report to the Sponsor and the Ethics in Human Research Committee within 24 hours of becoming aware of any serious adverse event experienced by any subject during the trial.

Appendix B:

Victoria University Notification of Human Research Ethics Approval

Victoria University
Department of Psychology
Human Research Ethics Committee
Co-Chairs: Heather Gridley
Anthoula Kapsalakis

PO Box 14428
MELBOURNE CITY MC VIC 8001
Australia

Telephone:
(03) 9365 2405

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VICTORIA UNIVERSITY OF TECHNOLOGY

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TO WHOM IT MAY CONCERN

Tuesday, 3rd June, 2003

The project titled "Neuropsychological profiles of children with acquired and congenital hydrocephalus" conducted by Ms. Nerissa Cordy under the supervision of Dr. Alan Tucker, received ethics approval from the Department of Psychology's Human Research Ethics Committee on the 03/06/03.

This project is been undertaken as part of the Doctor of Psychology (Neuropsychology) program conducted at Victoria University.

Yours Sincerely
Anthoula Kapsalakis

[redacted]
Co- Chair
Department of Psychology
Human Research Ethics Committee

Appendix C:

MRI Qualitative Data Checklist for Neuroradiologists

HYDROCEPHALUS PROJECT

PATIENT NAME:
UR:
DOB:
DOS:

VP SHUNT: Y / N
SITE:

CALVARIAL THICKENING: Y / N

VENTRICULAR SIZE:

NORMAL	DILATED	SLIT	ENTRAPPED

SUBDURAL COLLECTION Y / N

CORPUS CALLOSUM
SIGNAL: NORMAL
FOCAL ABNORMALITY
DIFFUSE ABNORMALITY

SIZE	**ROSTRUM**	**GENU**	**BODY**	**SPLENIUM**
NORMAL				
THICK				
THIN				
SIGNAL				
NORMAL				
FOCAL ABN				
DIFFUSE ABN				

WHITE MATTER
NORMAL Y / N
WHITE MATTER LOSS: DEEP / SUBCORTICAL
MILD MODERATE SEVERE

CAUSE FOR WM LOSS EVIDENT ON SCAN:

SHUNT TRACT SCARRING:

OTHER ABNORMALITIES/ COMMENTS:

Appendix D:

Parent General Information Questionnaire

General Information

Child's Name: _____

Gender: M / F Date of Birth: ___/___/ 19___

Name of School: _____

Name of School Teacher: _____

Grade/Year: _____ Handedness: Right ☐ Left ☐ Both ☐

Contact Name: _____

Contact Address: _____

Phone Number: (H) _____(W)_____

Family Data

Father: Name: _____

Occupation: _____

Highest level of Education: _____

Mother: Name: _____

Occupation: _____

Highest level of Education: _____

Number of siblings: _____ First Language: _____

Medical History

Has your child experienced any of the following?

Head injury? Yes / No If Yes, please describe: _____

_____

Seizures? Yes / No If Yes, please describe: _____

_____

Hospitalization? Yes / No If Yes, please describe: _____

_____

Medical Questionnaire

Subject #: _____ Consultant: _____

Name: _____

U.R.: _____

D.O.B. (Age): / /19 (____:____)

Age band: 1 ☐ 2 ☐

Date shunt inserted (age-months): ___/_19____ (_____)

Gender: M ☐ F ☐

Type of HC: Congenital obstructive ☐ Acquired communicating ☐

Shunt Type: VP ☐ VA ☐ CP ☐

Shunt location: Right hemisphere ☐ Left hemisphere ☐

Bilateral ☐

Co-morbid conditions: seizures ☐ periventricular leukomalacia ☐

cerebral palsy ☐ history of neonatal hypoxia ☐

other: _____

Surgical Complications: shunt revisions ☐ Due to: Infection ☐

Blockage ☐

other: _____

Neurological Screen Details: _____

Radiological Scale: Mild ☐ Moderate ☐ Severe ☐

Other details: _____

Hearing Problems? Yes / No If Yes, please describe: _____

_____

Visual Problems? Yes / No If Yes, please describe: _____

_____

Current Parental Concern

Have you felt any concerns regarding your child's:

Educational development? Yes / No If Yes, please describe: _____

_____

_____

Intellectual development? Yes / No If Yes, please describe: _____

_____

_____

Behavioural development? Yes / No If Yes, please describe: _____

_____

_____

Physical development? Yes / No If Yes, please describe: _____

_____

_____

Developmental History

What was your impression of your child's language development?

Early ☐ About Right ☐ Delayed ☐

Do you notice anything unusual about your child's language? If Yes, please describe: _____

_____

When did your child first begin to:

Crawl: _____

Walk: _____

Speak first words: _____

Put 2-3 words together: _____

Education

Has your child:

Repeated a school year or grade? Yes / No If Yes, which? _____

Missed a lot of school? Yes / No Approx no. of days:_____

Received any educational assistance? Yes / No