1988

Psychological sequelae of infantile hydrocephalus.

Jacobus. Donders

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PSYCHOLOGICAL SEQUELAE OF INFANTILE HYDROCEPHALUS

by

Jacobus Donders

A Dissertation
Submitted to the
Faculty of Graduate Studies and Research
through the Department of Psychology
in Partial Fulfillment of the
Requirements for the Degree of
Doctor of Philosophy
at the
University of Windsor

Windsor, Ontario, Canada

1988
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ABSTRACT

The intellectual, neuropsychological, and emotional sequelae of infantile hydrocephalus were assessed in a sample of 48 five- to eight-year-old children who had been surgically treated for hydrocephalus in the first year of life. Children were given a comprehensive battery of psychological tests. Their parents completed questionnaires pertaining to the psychological adjustment of their child and to their own distress. The significance of a variety of medical history variables with regard to psychometric intelligence was assessed with stepwise discriminant analyses. This revealed that many medical history variables were neutral with regard to intellectual outcome. The presence of additional medical problems in infancy, as well as current ocular defects, were the most significant variables that were associated with a high likelihood of mental retardation. Cluster analyses were used to identify subtypes of hydrocephalic children on the basis of their performance on a group of neuropsychological measures. Two clusters were found, which differed from each other mainly in level of performance. In general, all hydrocephalic children tended to perform relatively poorly on measures of complex visual-spatial processing. Cluster analyses on the results of the child personality questionnaires also revealed two clusters. One of these clusters included mainly children that were unimpaired, whereas the other cluster
included children with many cognitive and physical deficits. Discriminant and regression analyses revealed that there were no strong relationships between distress of the parents and a variety of physical and social variables. It was concluded that a comprehensive evaluation of the complete medical history of hydrocephalic children can help to identify children that are at risk for impaired intellectual development. However, the cognitive and behavioral outcome of infantile hydrocephalus spanned a wide range: from severely impaired to superior. The possibility of true neuropsychological or emotional subtypes could not be determined on the basis of this study. The possibility that early impairments of visual-spatial processing may represent a risk factor with regard to cognitive or behavioral dysfunctions at a later age was discussed.
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CHAPTER I
INTRODUCTION

1. General aspects of hydrocephalus.

1.1. Definition and classification.

Hydrocephalus refers to a group of conditions associated with ventricular enlargement, which in the majority of cases is secondary to obstruction of the normal egress and flow of cerebrospinal fluid at some point between the sites of formation and the sites of absorption (Bell & McCormick, 1978). This definition excludes cases in which ventricular enlargement is due to primary atrophic or hypoplastic cerebral conditions.

Hydrocephalus may rarely result from overproduction of the cerebrospinal fluid (CSF) seen in choroid plexus papillomas. This is the only exception to the general rule that all hydrocephalic conditions are obstructive.

Hydrocephalus is neither a disease nor a syndrome. As Gabriel (1980) has pointed out, it is a morbid entity with multiple causes. In all cases, however, the normal relationships of pressure and volume movement within the interconnected compartments of the CSF are altered (Etheridge, 1983). The result is enlargement of the ventricles with progressive cranial distention when hydrocephalus begins prenatally, in the newborn period, or in early childhood, but this does not occur when the onset is in later childhood or the adult years (Bell & McCormick,
By convention, obstructive hydrocephalus is divided into intraventricular obstructive, or noncommunicating, hydrocephalus (IVOH), and extraventricular obstructive, or communicating, hydrocephalus (EVOH). This classification is based on the ability of the CSF to leave the fourth ventricle and enter the subarachnoid spaces, i.e., with the foramen of Majendie as the cut-off point for obstruction (Roach, 1981). Thus, in IVOH the obstructive site is within the ventricular cavity, including the outlet foramina of the fourth ventricle, and in EVOH the obstruction occurs distally to the fourth ventricle foramina, in the cisterns or cerebral subarachnoid space itself (Gabriel, 1980).

In the older literature one may find the terms 'internal hydrocephalus' as a referent for IVOH and 'external hydrocephalus' as representing EVOH. The term 'hydrocephalus ex vacuo' refers to ventricular dilatation secondary to cerebral atrophy or hypoplasia, which is excluded from our definition of hydrocephalus. ' Arrested' hydrocephalus is the result of spontaneous or surgical termination of hydrocephalus of any cause. 'Normal pressure' hydrocephalus is a condition in which there is chronic hydrocephalus associated with neurological and mental symptoms (including apraxia of gait, bladder incontinence and dementia), but in which the CSF pressure has returned to the normal range.

IVOH is most likely to occur due to structural lesions where the caliber of the pathways of CSF are most narrow, including the
aqueduct of Sylvius, the fourth ventricle, and the foramen of Monro (Bell & McCormick, 1978). The major cause of EVOH is obliterative arachnoiditis with obstruction at the level of the basal cisterns, tentorial hiatus, or over the cerebral convexities (Roach, 1981). Table 1. (adapted from Bell & McCormick, 1978) provides an overview of the most important conditions associated with the various forms of hydrocephalus. The specific forms of pathology will be discussed in a later section.

Both IVOH and EVOH can occur as congenital and acquired conditions. Although the pathological aspects of congenital forms of hydrocephalus are fairly well established, the etiological factors responsible are for the most part obscure. Factors that may play a role include maternal malnutrition, exposure to radiation, intrauterine infections, and, rarely, an inheritable (X-linked) form of hydrocephalus. This is in contrast to what is known about cases of acquired hydrocephalus, where the etiology is often apparent and is known to include meningitis, intracranial bleeding, and central nervous system tumors (Hammock & Milhorat, 1981).

1.2. Incidence.

Hydrocephalus is a common disorder. The incidence of congenital hydrocephalus has been estimated at 3-4 per 1000 births, of which half are associated with spinal neural tube defects (Milhorat, 1978). The frequency of acquired forms is probably three times as great. With first pregnancies and with increasing age of the mother, the incidence is increased (Hammock
Table 1.

Classification and types of obstructive hydrocephalus.

A. Intraventricular obstructive (noncommunicating)
hydrocephalus.

1. Maldevelopments of the aqueduct (stenosis, atresia,
gliosis).
2. Obstruction due to mass lesions (neoplasm, cyst,
 hematoma, aneurysm of the vein of Galen).
3. Obstruction secondary to exudate, hemorrhage,
 parasites, or acquired aqueductal septum.
4. Obstruction of the fourth ventricle outlet foramina
(Arnold-Chiari malformation, Dandy-Walker syndrome,
arachnoiditis).

B. Extraventricular obstructive (communicating)
hydrocephalus.

1. Post-infectious, post-hemorrhagic, or developmental
 adhesions of basilar cisterns or surface subarachnoid
 space.
2. Arachnoid villi obstruction by erythrocytes.
3. Communicating hydrocephalus with Arnold-Chiari
 malformation.
4. Developmental failure of arachnoid villi.
and Milhorat, 1981). Cases of EVOH represent approximately 30 percent of all childhood hydrocephalus patients (Gabriel, 1980). By far the most common type of congenital hydrocephalus is that associated with other major central nervous system anomalies, including various forms of spina bifida. In some forms of spina bifida (especially lumbosacral myelomeningoceles) 90 percent of the cases have accompanying hydrocephalus (Gabriel, 1980). Because of the frequency of combined spina bifida and hydrocephalus we shall briefly discuss the most important forms of spina bifida and related defects.

1.3. Hydrocephalus and spina bifida.

Spina bifida results from a failure in fusion of the vertebral column (dysraphism). The result is a bony cleft through which varying quantities of brain or spinal cord tissue protrude (Gabriel, 1980). The precise term used to describe this herniation depends upon the type of tissue protruding.

In meningocele, the meninges herniate and form a cystic sac filled with spinal fluid. The spinal cord does not participate in the herniation and may or may not be abnormal. In myelomeningocele, the spinal cord is herniated into the sac and ends there or may continue in an abnormal way further downward. 'Spina bifida cystica' is a general term that is used to describe both conditions. Of all the defects collectively termed spina bifida cystica, about 75 percent are myelomeningoceles and about 25 percent meningoceles.
The majority of myelomeningoceles and meningoceles are located in the caudal parts of the spinal cord, especially the lumbar and lumbosacral regions. The reason for this is that the posterior neuropore (the last portion of the neural tube that closes) is located there and the primary defect in spina bifida is a failure in neural tube closure (Gabriel, 1980). Physical deficits that may result from a lesion at such a location include paraplegia and para-anesthesia of the lower extremities and bladder and bowel incontinence.

According to Gabriel (1980), thoracic and cervical myelomeningoceles are usually not associated with hydrocephalus and, in contrast, 90 percent of lumbosacral myelomeningoceles are complicated by hydrocephalus. This is, however, contradicted by a number of studies that report a higher incidence of hydrocephalus with higher levels of myelomeningocele (e.g., Hunt, Lewin, Gleave, & Gairdner, 1973; Lorber, 1971; Lorber 1972; Smith & Smith, 1973).

1.4. Cerebrospinal fluid formation and absorption.

The following discussion of cerebrospinal fluid (CSF) formation and absorption is mainly based on the discussions by Bell and McCormick (1978), Etheridge (1983) and Gabriel (1980). CSF is secreted, at least in part, by the choroid plexi in the lateral ventricles. The biochemistry of CSF secretion is still unclear. The total production rate is approximately 500 ml per day. Total CSF volume ranges from 50 ml in newborns to 150 ml in adults. This indicates that there must be continuous substantial
absorption of CSF. The capacity for drainage is two to four times the normal rate of production.

Due to arterial pulsations of choroid plexi pump mechanisms the CSF leaves the lateral ventricles through the foramina of Monro and enters the third ventricle from whence it moves via the aqueduct of Sylvis into the fourth ventricle. The CSF then enters the basal cisterns and spinal subarachnoid space via the foramina of Luschka and Magendie. Approximately 80 percent of CSF flows into the cerebral subarachnoid space and the remaining 20 percent circulates into the spinal subarachnoid space.

CSF absorption occurs in many regions, including across the ependyma within the ventricles, via arachnoid granulations into major venous channels, and through spinal perineural spaces into the lymphatic system. The major pathway is drainage into the venous system by way of the arachnoid villi that are located in the granulations of Pacchioni along the sinus sagittalis superior and other major venous sinuses. CSF drainage is controlled by CSF pressure, pressure within the cerebral venous system, and resistance of the arachnoid villi to CSF flow. In the case of hydrocephalus, alternative but usually inadequate methods of CSF absorption may be developed, such as accumulation of fluid in the periventricular white matter.

1.5. Pathogenesis of hydrocephalus.

Any obstruction of the flow of CSF along the pathway between the foramen of Monro and the granulations of Pacchioni will result in more CSF under increased pressure with subsequent dilatation of
the ventricular and subarachnoid spaces. The lateral ventricles
tend to enlarge first and to the greatest degree (Etheridge,
1983). The arterial pulse pressure that is responsible for pumping
the CSF through its normal pathways will increase with higher mean
CSF pressure. Regardless of the site of obstruction, progressive
hydrocephalus will then result in progressive attenuation and
eventual atrophy of the cerebral white matter (Bell & McCormick,
1978). According to Gabriel (1980) this is the result of the fact
that the increased intraventricular pressure leads to rupture of
the ventricular ependymal lining. This allows free and continuing
transependymal flow of CSF into white matter, producing spongy,
atrophic, and edematous dissolution of nerve fibers. This is a
major cause of the thin cerebral mantle that is so characteristic
of the brains of hydrocephalic children. Considerable
reconstruction of the thinned cerebral mantle can occur after

The atrophic changes in the cerebral white matter involve a
primary destruction of axons, a secondary loss of myelin, and
chronic astrogliosis (Hammock & Milhorat, 1981). The cerebral
cortex and deep nuclear grey matter are relatively spared, a
phenomenon that has been attributed to the fact that the
metabolically more active grey matter is more redundantly supplied

Pathologic effects of hydrocephalus depend considerably on
the age of the patient, the expansibility of the skull, and the
type and duration of hydrocephalus (Hammock & Milhorat, 1981). In
infants, the skull is still readily expandable and therefore there will be marked distention of the head. In older children the skull is no longer able to accommodate the continuous ventricular enlargement and signs of increased intracranial pressure will occur, including headaches, nausea, vomiting, and diplopia. These children have less extensively enlarged ventricles, but more flattened gyri (Etheridge, 1983). Infants with IVH show more rapid progression of head enlargement than do those with EVOH (Bell & McCormick, 1978).

As dilatation of the lateral ventricles proceeds other pathologic effects will follow. The atrophy of the white matter is typically accompanied by spongy edema of the brain surrounding the ventricles. There may be rupture of the septum pellucidum, and thinning and elongation of the interhemispheric commissures may occur (Hammock & Milhorat, 1981).

The parietal lobe and vertex are supposed to be most affected by the ventricular enlargement (Etheridge, 1983). The latter has been offered as an explanation for the increased muscle tone and hyperreflexia in the lower limbs in these patients, which may take the form of spastic paraplegia in the absence of spinal cord defects. The mechanism involved here is that long cortical efferent fibers arising from the leg area of the Rolandic cortex must project around the angle of the lateral ventricle to enter the internal capsule, and these fibers get stretched severely by progressive ventricular dilatation (Etheridge, 1983). For this reason, the motor problems related to the lower extremities are referred to as 'long tract signs' in the literature. The legs are
much more affected than the arms because the fibers that descend
to the upper limbs arise more laterally on the motor strip and
their pathway is more distal to the ventricles (Bell & McCormick,
1978).

Although most of the deficits discussed so far are related to
enlargement of the lateral ventricles, pathologic effects can also
occur due to changes at other locations. Third ventricular
dilatation (not uncommon after aqueduct stenosis), for example,
may lead to atrophy of hypothalamic nuclei (Etheridge, 1983). The
latter may result in significantly abnormal function of the
hypothalamic-pituitary axis associated with a variety of endocrine
changes (Gabriel, 1980).

Perceptuo-motor and visuo-spatial deficits may be related to
a variety of factors (Etheridge, 1983; Gabriel, 1980). Dilatation
of the third ventricle may stretch the optic tract and chiasma and
exert pressure on the mesencephalic tectum. There may also be
stretching of fibers to and from the occipital cortex due to
dilated posterior horns of the lateral ventricles.

If hydrocephalus is associated with other intracranial
abnormalities, such as the Arnold-Chiari malformation (to be
discussed later), there is pressure on, or deformation of, the
lower brain stem and cerebellum. Cerebellar damage may lead to
ataxia and other motor dysfunctions. Deranged lower brain stem
functions may present as pseudobulbar palsy, manifested by
difficulty in sucking, feeding, and phonation, and results in
regurgitation, drooling, and aspiration (Gabriel, 1980).
Although progressive abnormal head enlargement is an important clinical sign of hydrocephalus, significant ventricular enlargement can precede increase in head circumference or a detectable degree of suture separation. Bell and McCormick (1978) have mentioned irritability, recurrent vomiting, feeding poorly, and inadequate weight gain as other common signs of hydrocephalus. Seizures are not unusual with post-meningitic or post-hemorrhagic forms of hydrocephalus. Later signs in hydrocephalic infants include the 'cerebral cry', characterized by its brevity and high-pitched, shrill quality, and ocular signs such as the 'setting sun', which is a constant downward deviation of the eyes.

1.6. Pathology of noncommunicating hydrocephalus (IVOH).

Conditions that may be the cause of IVOH include aqueductal lesions (including atresia, stenosis and gliosis), congenital malformations involving the fourth ventricle (Arnold Chiari malformation and Dandy-Walker syndrome), space-occupying lesions in the posterior fossa, intra- or paraventricular tumors, paraventricular arachnoid cysts, peri- and intra-ventricular hemorrhage, and aneurysms of the vein of Galen (Roach, 1981). In this section we will discuss only the more common of these causes.

1.6.1. Aqueductal lesions.

Obstruction within the aqueduct of Sylvius is the most common site of intraventricular obstruction in infants with congenital hydrocephalus (Bell & McCormick, 1978). This is not surprising given the narrowness of conduit at this place. The next most
Important area of obstruction is the fourth ventricle (Gabriel, 1980).

In aqueductal stenosis the aqueduct is histologically normal but is abnormally reduced in size and caliber. The pathogenesis is often unclear but it is commonly assumed to be secondary to developmental abnormalities or inflammations. It has even been suggested that aqueductal stenosis may be a consequence, rather than a cause of hydrocephalus (Williams, 1973; cited in Bell & McCormick, 1978).

In many instances aqueductal stenosis is accompanied by atresia or forking of the aqueduct, which means that there are nonpatent multiple channels. This is often associated with other CNS abnormalities such as myelomeningocele and Arnold-Chiari malformations (Bell & McCormick, 1978) and with malformations of neighboring structures, including fusion of the corpora quadrigemina and third nerve nuclei (Gabriel, 1980).

Aqueductal gliosis is a post-inflammatory process, usually secondary to infection or hemorrhage (Gabriel, 1980). The aqueduct is narrowed or occluded by overgrowth of subependymal glia while the ependymal lining is replaced by gliosis (Bell & McCormick, 1978). In both stenosis and gliosis of the aqueduct the onset of occlusion is insidious and the course progressive.

1.6.2. **Arnold-Chiari malformation.**

Chiari malformations are cerebellar malformations that, alone or in combination with other anomalies, account for approximately 40 percent of all hydrocephalic children (Gabriel, 1980). Chiari
has described four forms of this defect between 1891 and 1895. The
tform that is most common and that is most often associated with
hydrocephalus is type 2 and is referred to in the literature as
the Arnold-Chiari malformation. Most cases of Arnold-Chiari
malformation have IVOH, but if the major obstruction takes place
at the level of the ambient cisterns EVOH may develop.

The Arnold-Chiari malformation is characterized by caudal,
downward displacement of the cerebellum, medulla, lower pons, and
fourth ventricle through the foramen magnum into the upper
cervical canal, and is associated with cerebellar hypoplasia
(Roach, 1981). The defect is almost always associated with
myelomeningocele and/or hydrocephalus. Arnold-Chiari
malformation is supposed to be an abnormality of brain structures
that develops in concert with spina bifida cystica, not secondary
to it (Bell & McCormick, 1978). The hydrocephalus is most likely
related to stretching and subsequent narrowing of the aqueduct by
the downward displacement of the tentorium and the posterior fossa
Furthermore, both the foramina of Luschka and Magendie and the
basal cisterns are occluded as a result of impaction of the
foramen magnum or atresia of the foramina outlets (Gabriel, 1980).

1.6.3. Dandy-Walker syndrome.

The Dandy-Walker syndrome refers to a congenital obstruction
of the foramina of Luschka and Magendie and associated development
of a large porencephalic cystic distention of the fourth
ventricle. This 'cyst' (which is, in fact, the fourth ventricle;
Bell & McCormick (1978) herniates caudally and displaces the cerebellar hemispheres. The cardinal features are hypoplasia of the cerebellum, flattening of the brain stem, and occlusion or atresia of the fourth ventricle foramina (Etheridge, 1983; Roach, 1981).

This entity must be distinguished from arachnoidal cysts in the posterior fossa that lie above the foramen of Magendie. In the latter cases, there is separation of the cyst from the fourth ventricle and a normally developed cerebellum that is only displaced or deformed by the cyst (Roach, 1981).

The hydrocephalus that is associated with the Dandy-Walker syndrome often does not become clinically manifest until after infancy (Gabriel, 1980). Suggestive, but not conclusive, of the diagnosis are enlargement of the head and a characteristic configuration of the head, with very prominent occipital regions (Bell & McCormick, 1978). Associated features may include nystagmus, ataxia, and cranial nerve deficits.

1.6.4. Peri-ventricular tumors.

According to Etheridge (1983), tumors are common causes of hydrocephalus. Tumors can be causes of IVOH as well as EVOH. The third ventricle can be obstructed by craniopharyngiomas and gliomas; pinealomas, astrocytomas and glioblastomas can compress the aqueduct, and ependymomas, medulloblastomas, and cerebellar astrocytomas may compress the fourth ventricle.

1.7. Pathology of communicating hydrocephalus (EVOH).
BVOH is less common in children than IVOH. As was stated above, the locus of obstruction in BVOH is distal to the fourth ventricle foramina. Some of the possible causes of IVOH (especially the Arnold-Chiari malformation) may result in BVOH. Most forms of normal-pressure hydrocephalus are of the communicating type and involve incomplete arachnoidal obstruction to CSF drainage (Gabriel, 1980). The two most common causes of BVOH in young children are intra-ventricular hemorrhage and meningitis.

1.7.1. Intra-ventricular hemorrhage.

According to Volpe (1983), on whose chapter most of the following discussion is based, intra-ventricular hemorrhage is the most common variety of neonatal intracranial hemorrhage. Most cases are observed in infants less than 32 weeks of gestation. Perinatal hemorrhage can be anoxic or traumatic. In most infants the hemorrhage originates in the matrix at the level of the head of the caudate nucleus and the foramen of Monro. In approximately 80 percent of the cases the hemorrhage ruptures through the ependyma and fills the ventricular system. Hydrocephalus may develop due to obliterative arachnoiditis in the posterior fossa that either blocks the outflow of CSF from the fourth ventricle or the flow of CSF through the posterior fossa to the supratentorial space for reabsorption. A less common sequel is IVOH due to obstruction at the aqueduct of Sylvius. The ventricular dilatation that follows intraventricular hemorrhage shows a disproportionate involvement of the occipital horns.
1.7.2. Meningitis.

Etheridge (1983) designates post-inflammmatory hydrocephalus as the most common variety of EVOH. The mechanisms of production of hydrocephalus in meningitic cases is described by Gabriel (1980) as clumping of purulent fluid in the CSP drainage channels in the acute phase, and organization of exudate and blood, resulting in fibroses of the subarachnoid spaces, in the chronic phase. Bacterial meningitis tends to produce cerebral cortical arachnoiditis, whereas parasitic meningitis produces cisternal obstruction.

1.8. Relationship to age.

Chiari malformations and acqueudctal lesions account for 80 percent of all hydrocephalus in the period from 0 to 2 years and represent 60 percent of all hydrocephalus regardless of age (Gabriel, 1980). Intra-uterine infection, perinatal hemorrhage, and neonatal meningitis are other important causes in this age period. In early to late childhood the most common causes for hydrocephalus are posterior fossa neoplasms and obstructions of the acqueduct. Fungul and parasitic infections are the most common infections that cause hydrocephalus in this age group.

As was outlined above, abnormal head growth is characteristic of infantile cases of hydrocephalus whereas symptoms of increased intracranial pressure and focal signs are more common in children in whom the disorder has a later onset. Premature infants have a normal postnatal progressive head growth that is usually not indicative of hydrocephalus.
1.9. Diagnostic studies.

Before going into the disorders that should be distinguished from hydrocephalus we will briefly discuss the most common techniques that are used in the diagnostic process. Details about these and other procedures can be found in Bell and McCormick (1978) and Gabriel (1980).

Serial measurements of the occipito-frontal circumference of the head are a simple but important technique to detect abnormal rate of head enlargement. Roentgenogram and transillumination of the skull can reveal several skull abnormalities, including posterior fossa abnormalities (such as associated with the Dandy-Walker syndrome) and lacunar skull deformities (LSD, craniolacunae, or Luckenschadel). The latter refers to a webbed appearance of the skull that is common in infants with myelomeningocele.

Angiography is of value when hydrocephalus is believed to be caused by vascular abnormalities, such as an aneurysm of the vein of Galen. Pneumoencephalography and isotope cisternography (also called isotope encephalography) are procedures that are more directly aimed at visualizing the ventricles and CSF pathways themselves, by means of the injection of respectively air and radiopharmaceuticals. They are useful to distinguish TVOH from EVOH and may provide critical information in the elderly to differentiate patients with normal-pressure hydrocephalus (who might substantially benefit from a shunt procedure) from those in whom the dementia is due to other factors, such as Alzheimer's disease or multiple infarcts.
Computerized axial tomography (CT scan) is the most important technique for definitive diagnosis, localization of the site of obstruction, and exclusion of mass or compressive lesions. The procedure is far less invasive and presents less risk to the patients than, for example, angiography or pneumoencephalography. A CT scan demonstrates not only the size and position of the ventricles, but also the width of the subarachnoid spaces at the base of the brain and over its convexity. The procedure is also important in evaluating the effects of shunt therapy and in distinguishing acute shunt failure from other causes of neurological signs of abrupt onset in the shunted child.

1.10. Differential diagnosis.

Considerations of syndromes other than hydrocephalus are especially important in the newborn and young infant because the spectrum of causes of abnormal head enlargement is larger and more complex than in older children. In cases of hydrocephalus, differential diagnosis of the underlying cause may have considerable implications for subsequent treatment (e.g., an intracranial neoplasm may be removed whereas congenital anomalies are typically permanent).

We have already mentioned the importance of differentiating the normal accelerated head growth in premature infants from hydrocephalus. Other conditions that have to be ruled out include megalencephaly, skeletal disorders, and cerebral gigantism. These disorders are discussed in detail by Bell and McCormick (1978) on whose chapter most of the following discussion is based.
Megalencephaly refers to the presence of head enlargement because the brain is of excessive weight, either as the result of a congenital malformation or a degenerative disease. CT scan will reveal a normal ventricular system with excessive brain tissue.

There are several skeletal disorders or dwarfism syndromes in which the head is disproportionately large for the body, but in which there is not necessarily hydrocephalus. Acrondoplasia, for example, is characterized by short extremities, short and stubby fingers and macrocephaly, but the ventricular system is often normal.

Children with cerebral gigantism have an abnormally rapid rate of somatic growth and skeletal maturation. They typically have an abnormally large brain with generous-sized ventricles, but not hydrocephalus.

1.11. **Treatment of hydrocephalus.**

Spontaneous and permanent arrest of hydrocephalus, with at least partial restoration of the ventricular size and of the cerebral mantle is rare. Laurence (1974) has indicated that a mortality rate of about 50 percent may be expected in non-treated cases.

The main form of treatment for hydrocephalus involves a shunting procedure in which a surgically installed valve directs the CSF from the ventricular system to another body cavity (usually peritoneal or atrial), from which it may be absorbed. Revisions of the shunt are required periodically with somatic growth. Complicating shunt-induced infections, such as meningitis
and ventriculitis, approximate 5 percent (Gabriel, 1980). Obstruction of the shunt, usually at the distal end, is another major problem that necessitates continued follow-up. Other possible complications of shunt treatment include shunt nephritis, pulmonary thromboembolism and craniosynostosis (Bell & McCormick, 1978).

The mortality and morbidity rates of hydrocephalus have changed considerably over the last three decades. Since the introduction of valve-regulated shunts in the early fifties, the outlook for patients with neonatal hydrocephalus has improved significantly. Lorber (1971) has argued that although the better treatment resulted in a decreased mortality rate, the morbidity rate did not decrease. His argument was that improved therapeutic techniques managed to keep children alive that previously would have died and that those subjects developed severe deficits.

Lorber (1971, 1972) has specified selection criteria to restrict the range of children with neural tube defects and hydrocephalus that should receive treatment. Gross enlargement of the head (exceeding the 90th centile by at least two centimeters) due to hydrocephalus was one of these criteria. Lorber's criteria have received at best mixed support. Stark and Drummond (1973) reported a decrease in the percentages of wheelchair confinements, damage to the urinary tract, and severe mental handicaps when they applied Lorber's criteria but also reported that this application did not abolish the occurrence of severe handicaps. Stein, Schut, and Ames (1975), reporting on a retrospective study, indicated that if they would have used Lorber's criteria they would have
rejected from treatment a considerable proportion (54 percent) of children who had satisfactory outcomes. In fact, Lorber (cited in Lewin, 1980) has provided several cases whose ventricle expansion fills at least 95 percent of the cranium, but still display above average intellectual development.

Most authors agree that not all children with hydrocephalus are necessary candidates for treatment and suggest some consideration of the expected quality of life but refrain from strict criteria such as suggested by Lorber. In general, the consensus in the literature is that the mortality rate for hydrocephalus is greatly reduced and that the morbidity of shunted cases has also declined considerably over the last decade (McCullough & Balzer-Martin, 1982). The 5-year survival rate for an unselected series now approaches 90 percent (Hammock & Milhorat, 1981).

Lister, Zachary, and Brereton (1977) studied the causes of death in a sample of 200 cases of open myelomeningocele and hydrocephalus. The majority of the children that died succumbed during the first three months of life, usually to intracranial hemorrhage or infection. Many of these patients died before there was a possibility of considering treatment of the hydrocephalus.

2. Intellectual development of children with hydrocephalus.

2.1. Methodological aspects.

The vast majority of the studies on the psychological sequelae of hydrocephalus that are reported in the literature have
concentrated on global assessments of intellectual abilities. A substantial number of these studies have attempted to relate the intelligence levels attained by the subjects in their sample to medical history variables. Most of the studies are, however, purely descriptive and many of them do not use adequate statistical analyses. Furthermore, it is often very difficult to compare the results from one study to those from another study due to important differences in methodology. This is particularly disturbing since the literature shows substantial lack of agreement with regard to the relative importance of different parameters of the medical history for subsequent intellectual development in hydrocephalic children.

Raimondi and Soare (1974) have presented an excellent review of the reasons why it is difficult to compare one study to another. They reported five sources of difficulties, including (1) lack of information regarding shunt variables, (2) use of different instruments to assess intelligence, (3) differing methods of data presentation and of definition of "normal", (4) inclusion of varying causes, and (5) sample bias.

2.1.1. Shunt variables.

According to Raimondi and Soare (1974), the most difficult information to obtain from many studies is that regarding the age at which a shunt was first inserted, information regarding the integrity of cerebral structures at the time of shunt insertion, and whether and how the shunting system was kept functioning until the time the child was evaluated. It is very disturbing that some
studies do not provide information about these variables since different studies report conflicting results concerning the predictive validity of some of these variables (e.g., age at shunting, number of shunt infections and revisions) with regard to later intelligence. These discrepancies will be discussed in a later section.

Not only is information frequently lacking with regard to when a shunt was placed and how it was maintained, but the nature of the shunt is also an underreported variable. There appears to be disagreement among medical experts about which type of shunt (especially ventriculo-atrial versus ventriculo-peritoneal) is to be preferred for purely medical reasons (see Bell & McCormick, 1978 versus Etheridge, 1983).

2.1.2. Measures of intelligence.

The use of different instruments to assess intelligence is an issue that was not elaborated on very much by Raimondi and Soare (1974), but which is very important in the context of our research project. The issue is partly related to the problem of sample bias, because different studies use widely varying age populations, which has direct implications for the types of tests that can be administered. On the other hand, some studies include children in their sample that have been assessed with very different intelligence tests.

McCullough and Balzer-Martin (1982), for example, included the following instruments: Bayley Scales of Infant Development (Mental Scale), Wechsler Preschool and Primary Scale of
Intelligence (WPPSI), Merrill-Palmer Scale of Mental Tests, Stanford-Binet Intelligence Scales, and Wechsler Intelligence Scale for Children - Revised (WISC-R). They did not specify, however, how many children were assessed with each test and furthermore grouped and averaged all IQ scores from all different tests together as if they were equivalent. Something similar was done by Tromp, Van den Berg, Jansen, and De Vries (1979). They also used a wide variety of tests across subjects and when more than one test was administered to a subject, his average score was used in the analyses.

In general, intelligence quotients (IQs) from different tests can not be considered to reflect the same functional development because different tests assess different functions. The Stanford-Binet, for example, becomes increasingly more verbal with higher age groups. The use of different intelligence tests is also a problem because only some of these tests provide a differentiation between verbal and performance IQ, and an uneven growth of these two aspects of intelligence (with nonverbal intelligence developing less well than verbal intelligence) is a common outcome of hydrocephalus (Dennis et al., 1981).

Even more disturbing is that some studies administer only selected parts of common intelligence tests without apparent consideration of the validity of this practice. Prigatano, Zeiner, Pollay, and Kaplan (1983) administered only the Vocabulary and Block Design subtests of the WPPSI or WISC-R to their subjects and considered the results to be representative of verbal and performance IQ respectively. They did not offer any justification
for this method of assessment, nor did they comment of the reliability or validity of their index.

Finally, some studies do not even report how intelligence was measured. The most dramatic example of this is the study by Amacher and Wellington (1984). They reported that 63.4 percent of their 5-year survivors of shunted infantile hydrocephalus had a "satisfactory intellectual outcome". At the same time, they did not report how intelligence was assessed, what the criteria for 'satisfactory’ outcome were, and did not even report a single numerical figure with regard to the mean or median IQ of their sample.

2.1.3. Data analysis.

The review by Raimondi and Soare (1974) indicated that most studies cite the percentage of survivors over a certain period of time and then the percentage of patients with IQs above and below a certain level. It was apparent that different authors had significantly different opinions about which IQ range was considered to be 'normal'. Furthermore, many of the studies reviewed did not present a mean IQ and its standard deviation for the sample studied. Methodology in hydrocephalus studies improved somewhat over the last decade, but an adequate statistical analysis of the data is still very often lacking.

The vast majority of the studies on the intellectual sequelae of hydrocephalus are purely descriptive and do not provide any information about whether or not statistical analyses were used. The relatively few number of studies that have used statistical
analyses typically have restricted themselves to contrasting
groups that were selected on the basis of the presence or absence
of specific medical history variables.

For example, in one of the best documented studies on
intellectual sequelae that has been reported in the literature so
far, Dennis et al. (1981) investigated whether there were
differences in IQ scores between subjects with different
demographic, symptomatic, pathologic, and therapeutic
characteristics. Analyses of variance (ANOVA) were used to analyze
the importance of each of these variables separately. Possible
correlations between variables are thus artificially eliminated
and the risk of Type I errors (incorrect rejection of the
Null-hypothesis) is increased. Other authors have used a strategy
similar to the one employed by Dennis et al., using t-tests (e.g.,
Prigatano et al., 1983) or chi square analyses (e.g., McCullough
and Balzer-Martin, 1982).

Raimondi and Soare (1974) presented one of the very few
studies that analyzed the data by means of correlational methods.
However, they provided only a number of independent correlation
coefficients between isolated medical history variables and IQ
and did not analyze the combined predictive power of multiple
variables. Hurley, Laatsch, and Dorman (1983) used multiple
regression analyses, reportedly to determine which of their test
variables discriminated between their patient and control groups.
It was not clear how they did this and why they did not use a
discriminant analysis. Tromp, Van den Burg, Jansen, and De Vries
(1979) reported that they had analyzed their data by means of
multiple regression techniques, but did not report sufficient
details of the results from those analyses. The use of such
techniques is precisely one of the objectives of this
dissertation, and will be outlined in a later section.

The importance of different ways of analyzing the data was
specifically highlighted in the study by Soare and Raimondi
(1977). They found that sensory level at birth appeared not to be
significantly related to IQ if children with only myelomeningocele
and children with myelomeningocele and hydrocephalus were analyzed
separately. When the two groups were combined, however, there was
a significant relationship between sensory level and IQ.

2.1.4. Inclusion of varying causes.

As Raimondi and Soare (1974) have indicated, not all studies
include the same hydrocephalic groups in their analyses. Some
studies employ very strict inclusion criteria, other studies are
much less rigorous. For example, Lorber (1971) reported results
on series of unselected cases of children with myelomeningocele
and hydrocephalus, whereas Dennis et al. (1981) employed a variety
of medical and behavioral criteria to exclude certain subjects
from their study.

Some studies that are reported in the literature appear to
have been set up primarily for the investigation of the outcome of
spina bifida cystica and the hydrocephalus that some of these
cases display is sometimes analyzed separately and sometimes not.
For example, Hunt and Holmes (1975; 1976) specifically
investigated features noted at birth that distinguished the
children who did not require shunts and factors relating specifically to the shunt-treated children. In another study, however, Hunt and colleagues just used hydrocephalus as one of the possible predictive factors (among other variables such as sensory level, neural plaques, etc.) with regard to outcome in terms of mobility, intelligence and continence in cases of myelomeningocele (Hunt, Lewin, Gleave, & Gardner, 1973).

Laurence (1974) commented on the lower intelligence of operated versus non-operated cases of myelomeningocele and suggested that this difference was probably largely due to hydrocephalus, but did not report attempts to analyze this further. On the other hand, Shurtleff, Kronmal, and Poltz (1975) specifically compared the incidence of mental retardation between hydrocephalic patients with and without myelomeningocele.

Studies also differ considerably in the types of hydrocephalus that were the primary object of investigation. For example, Hammock, Milhorat, and Baron (1976) studied eight children with myelomeningocele and normal pressure hydrocephalus. Richardson (1978) limited his sample to children with spontaneously arrested congenital hydrocephalus. Krishnamoorthy, Kuehnle, Todres, and DeLong (1984) reported on the outcome of survivors of posthemorrhagic hydrocephalus.

The range of demographic, pathologic, symptomatic, and therapeutic variables that were included in the analyses also varies considerably across studies. While some studies have specifically investigated the effects of various complications, such as infections, seizures, etc. (e.g., Dennis et al., 1981;
Hunt & Holmes, 1975, 1976; Thompson, Eisenberg, & Levin, 1982) other studies have limited their sample to children with uncomplicated hydrocephalus (e.g., Prigatano et al., 1983). Variables that have been identified by some authors as the best indication of later intelligence are not reported or disputed by other authors. For example, the sensory level of the lesion was concluded to be one of the best predictors of intellectual development in children with myelomeningocele and hydrocephalus by Hunt and Holmes (1975; 1976) but this variable was not included in the study by Dennis et al. (1981), whereas Soare and Raimondi (1977) concluded that knowledge of the sensory level is not a reliable predictor of IQ.

2.1.5. Sample bias.

The problem of sample bias is of course related to the fact that not all studies include the same spina bifida and hydrocephalic groups of subjects in their analyses. The inclusion of varying causes can therefore be seen as one example of sample bias. In general, sample bias appears to be operating before subjects are even selected.

Badell-Ribera, Shulman, and Paddock (1966), for example, performed a retrospective study of children that were referred to a rehabilitation center. They indicated in their introduction that this sample was most likely not representative of the whole population of children with hydrocephalus and spina bifida cystica: More severely affected children are less frequently referred to a rehabilitation center.
Another substantial source of bias concerns the age at which intelligence and other psychological variables were assessed. As Raimondi and Soare (1974) indicated, the older the age group at the time of evaluation, the greater the chance that the more severely affected cases have died. We would like to add to this that it is probably also important in this context to consider when the study was done. As was mentioned above, the mortality and morbidity rates of hydrocephalus are supposed to have declined considerably over the last two decades. Therefore, studies reported in the mid-sixties (e.g., Badell-Ribera et al., 1962; Lorber, 1968) probably deal with very different survivors than studies reported in the late seventies and eighties (e.g., Dennis et al., 1981; Soare & Raimondi, 1977).


The first studies of the general intellectual development that were reported in the literature were of children whose spina bifida and hydrocephalus was untreated (Anderson, 1973). Aggressive treatment for these conditions was not common until the early sixties. Many of the later studies report only on treated hydrocephalus (e.g., Dennis et al., 1981; Shurtleff, Potz, & Loeser, 1973) while some other studies do report that they had cases that were not treated but that they all died before intelligence could be assessed in valid ways (e.g., McCullough & Balzer-Martin, 1982).

Badell-Ribera et al. (1966) reported one of the first studies that compared the performance of spina bifida patients with and
without hydrocephalus. 62 Percent of their 75 spina bifida cystica patients had hydrocephalus. The hydrocephalus was not treated in any of the children and eventually became non-progressive. Hydrocephalus was mainly judged to be present on the basis of a history of rapid head growth in the first two years of life and few patients were subjected to neuroradiological studies. At the time of testing, the ages of the subjects ranged from 5 to 21 years and they were assessed with the WISC or the WAIS. The results indicated that the total subgroup with a history of hydrocephalus scored significantly lower in the verbal (VIQ), performance (PIQ), and full-scale (FSIQ) ratings (analyzed by means of Mann-Whitney U-tests). If groups were stratified according to the degree of motor disability, the children in the more severely disabled groups achieved significantly lower IQs than the mildly disabled groups only when there was a history of hydrocephalus.

In an overview of a series of studies conducted between 1956 and 1972, Laurence (1974) compared the intellectual performance of two series of congenital spina bifida cases: an unoperated series of 290 cases born between 1956 and 1962 (with 54 survivors at follow-up in 1968) and an aggressively treated series of 113 cases born between 1964 and 1966 (with 57 survivors in 1973). Little information was provided concerning how many cases in each group had hydrocephalus, but it was reported that "few" had surgery for hydrocephalus in the non-treated group and that the other subjects had a valve operation "if indicated". The unoperated series was assessed with the Stanford-Binet, the operated group with the WISC. To overcome this variation, the Stanford-Binet IQ results
were transformed into the equivalent Wechsler scores according to a formula published by Weider, Moller, and Schramm (1951). Patients with meningocoele did not show significantly different IQs between groups, but patients with myelomeningocele that was operated performed significantly less well intellectually than those who were not operated upon (analyses by means of \( t \)-tests). Laurence suggested that this was probably largely due to progressive hydrocephalus, but he did not provide data to substantiate this. He also failed to analyze the results for children with and without hydrocephalus separately.

Lorber (1971) has reported on two unselected series of cases of myelomeningocele. In the first series, consisting of 323 infants treated between 1959 and 1963, the Terman-Merrill was administered repeatedly after 5 years of age but only the results obtained on the last occasion were reported. No information was provided concerning the distribution of ages of the subjects at that last assessment or about the number of times the subjects had been assessed. Of the 134 survivors 31 had no hydrocephalus, 29 had hydrocephalus but no shunt, and 75 had hydrocephalus with shunt. Lorber reported that the IQ of the 31 without hydrocephalus and of the 29 whose hydrocephalus required no treatment was the same (87) but did not show results for these two groups separately. On the basis of this he concluded that unoperated hydrocephalus of moderate degree was fully compatible with normal intelligence, but he did not indicate why the subgroup of 28 was not shunted, nor did he compare the results of this group by means of statistical analysis to those of the group with shunts (whose
average IQ was 79). It is very plausible that the children who were shunted had more severe levels of ventricular dilatation than the non-shunted children, so a direct comparison of treated versus non-treated cases would not be valid, but the way Lorber presented his data might give rise to considerable misinterpretation.

The data on the second series in Lorber's paper deal predominantly with his formulation of selection criteria for treatment and these were further elaborated on in a subsequent article (Lorber, 1972). It is important in the context of the current discussion that Lorber found that those children with the most serious degree of ventricular dilatation had the poorest intellectual development. He included gross enlargement of the head (exceeding the 90th percentiled by at least 2 cm) in his criteria against treatment.

Shurtleff, Kronmal, and Foltz (1975) reported on a series of 454 hydrocephalic patients with and without myelomeningocele, and with and without treatment. A wide variety of tests were used to assess intellectual development, including the Peabody Picture Vocabulary Test (PPVT), the Goodenough-Harris Draw-A-Person Test, the Stanford Binet, and the Wechsler scales. No specification was made of how many subjects received each test and no comment was made on the comparability of indices, obtained from these different measures. The groups with treatment were shown to have better survival rates and a smaller proportion of mental retarded cases than the untreated groups, but IQ data or statistical analyses were lacking.
Mapstone, Rekate, Nilsen, Dixon, Glaser, and Jaffe (1984) compared the intellectual performance of three groups of children with myelomeningocele. Group 1 consisted of 18 children never requiring a shunt (the authors did not report how many of these children were hydrocephalic or why they were not shunted); group 2 consisted of 41 children who received a shunt and had no complications; group 3 consisted of 16 children with a shunt and with complications. IQ testing was done with the Wechsler scales (87 percent of the sample) or Stanford Binet. The authors did make a comment that they realized that these tests were not equivalent. Although all children in the sample were above the age of 4, no information was provided concerning how old subgroups of children were at testing. The mean IQ of group 1 was 104, that of group 2 was 91, and that of group 3 70. These IQ differences were significant by means of analysis with Student's t-tests. The authors erroneously concluded that all these IQs represented the lower level of the normal range.

Soare and Raimondi (1977) compared 133 children with myelomeningocele and shunted hydrocephalus to 40 children with myelomeningocele without hydrocephalus. The etiologies of hydrocephalus were not specified and little information was provided regarding complications. The latter is unusual in the context of the criticisms expressed by these authors in an earlier publication concerning the lack of these data in the literature (Raimondi & Soare, 1974). As in many other studies, not all children were assessed with the same intelligence test. Although the authors indicated how often the children were tested, there
appeared to be a very large difference in the mean age of the two groups at the time of the first assessment: the shunted children being 3.11 years on the average and the other children having a mean age of 7.4 years. The significance of their finding that mean IQ of the children with shunted hydrocephalus (87.7) was significantly lower than that of children without hydrocephalus (102.3) is therefore unclear. The authors' conclusion that when myelomeningocele is associated with hydrocephalus, the IQ can be expected to be lower, may be plausible but was at the same time not clearly demonstrated by this study.

Shurtleff, Foltz, and Loeser (1973) compared 22 patients with congenital hydrocephalus without myelomeningocele with 37 patients with congenital hydrocephalus and myelomeningocele. A wide variety of assessment instruments were used, but the criteria for inclusion of subjects in the study were well described. Statistical analyses were unclear. The authors reported that the average IQ of the group without myelomeningocele was significantly higher than that of the group with myelomeningocele.

Brown and McLone (1981) re-analyzed the data from the Soare and Raimondi (1977) study to look more closely at the role of CNS infections. They included in their analyses 39 children who were not shunted, 86 children with shunts but without infections, and 42 children with shunts and infections. The mean IQ of the first group was 102, that of the second group 95 and that of the third group 73. Although the authors did not mention what statistical technique they used, they reported that the third group differed significantly from the other two groups. No data were presented on
a statistical comparison between groups 1 and 2. The authors concluded that hydrocephalus alone does not appear to be a significant limiting factor in ultimate intellectual development of children with myelomeningocele, but that subsequent CNS infection is a significant factor that reduces intelligence.

In summary, it appears that the survival rate of children with treatment of hydrocephalus and/or myelomeningocele is far better than that of children without treatment. Those children that do survive despite lack of treatment may very well have less serious CNS defects than the treated children, which may explain why some studies report higher IQs in children that were not treated. At the same time, these studies have suggested that complications, infections and other medical history variables are very important with regard to subsequent intellectual development.

2.3. Verbal versus performance intelligence.

It has already been mentioned that the type of intelligence test used in different studies varies considerably. Moreover, many studies have not assessed all subjects in their sample with the same technique. Only the Wechsler scales provide the opportunity to derive both verbal and performance measures from the same instrument. At the same time, even studies that have assessed all their cases with the Wechsler scales do not always provide information on all of the intelligence measures, but restrict themselves to presenting the data on FSIQ (e.g., Lenton, Barrington, & Lorber, 1975). On the other hand, several studies
that have investigated both verbal and performance intelligence
often have found discrepancies between VIQ and PIQ in
hydrocephalic children.

Badell-Ribera et al. (1966) found that in a sample of spina
bifida cases that patients that had their lesion in the lumbar or
thoracic region and had untreated hydrocephalus had significantly
lower verbal, performance, and full-scale IQs (as analyzed by
Mann-Whitney U-tests) than those patients with comparable spinal
lesions without hydrocephalus. Although the mean PIQ in the
hydrocephalic patients was quite low (79), the mean VIQ (91) was
in the average range. In patients with spinal lesions in the
sacral region (who were less severely motorically disabled) the
PIQ was significantly lower in cases with hydrocephalus (89) as
compared to cases without hydrocephalus (105). The VIQ did not
differ significantly with the presence or absence of hydrocephalus
in this group (104 versus 109). Although these results suggest
that the PIQ is more depressed in hydrocephalic patients than VIQ,
Badell-Ribera et al. did not make a direct statistical analysis to
contrast both IQs within a hydrocephalic population.

Spain (1974) administered the Performance and Hand-eye
Coordination Scales of the Griffiths Mental Development Test and
the Reynell Developmental Language Scale to a sample of 145
three-year-old children with spina bifida. Although these tests do
not provide dependent measures that are equivalent to the IQs that
can be obtained from the Wechsler scales in older children, the
results are relevant in the context of an uneven growth of verbal
versus performance skills in hydrocephalic children. Spain
reported that all children tended to have higher verbal (Reynell) than non-verbal (Griffiths) scores. He did not subject this to statistical analysis, reportedly because the correlation between the two tests was not known (but which he could of course have calculated for his sample himself). Although Spain reported that children with shunts had lower mean scores than children without shunts, it appears from his results that the difference between the Reynell and the Griffiths scores was evident both in groups with and without shunts for hydrocephalus. Therefore, this study does not provide conclusive information regarding the specific effects of hydrocephalus.

Tew and Laurence (1975b) administered the WPPSI to a group of 59 five-year-old children with spina bifida and to a group of controls, matched for sex, place in family, and social class. The mean FSIQ of their 31 shunt-treated patients was significantly lower than that of the children with spina bifida with no hydrocephalus (analysis by means of t-test). The FSIQ of the total patient sample was significantly lower than that of the controls. In the 31 children with hydrocephalus, there was a statistically significant difference between the mean VIQ (78.1) and the mean PIQ (67.1). Although these results appear to support the notion of a more detrimental effect of hydrocephalus on PIQ than on VIQ, it is important to notice that in a later attempt at replication of these findings with another sample Tew (1977) reported that the mean PIQ on the WISC was not significantly lower than the VIQ.

Tew and Laurence (1984) performed a follow-up of the children from their previous (Tew & Laurence, 1975b) study. At the age of
16, the 51 survivors were administered a Wechsler intelligence test (it was not clear whether this was the WISC-R or the WAIS). Although the authors reported the IQ results for the total sample of spina bifida survivors (with VIQ, PIQ, and FSIQ all being significantly lower in the patient than in the control group), they did not report data on specific analysis of those cases with hydrocephalus.

Lonton (1979) reported on the relationship between the degree of hydrocephalus and intelligence, as assessed by the WISC, in a group of 305 children with spina bifida and hydrocephalus. The statistical analyses were unclear. Lonton reported that dramatically enlarged ventricles were not associated with significantly lower VIQ, but that there was a significant effect on PIQ. It appears that this conclusion was based on the finding that the mean verbal IQ in the shunted group (VIQ = 102) and in the non-shunted group (VIQ = 101) was substantially higher than the performance IQ (84 in the shunted patients and 87 in the patients without valves). According to Lonton, these results were in agreement with those of Tromp et al. (1979) but we were unable to find any report on verbal - performance discrepancies in that publication.

Connell and McConnel (1981) examined 41 children who had been treated surgically for hydrocephalus in the early months of life. These children were assessed with the WPPSI or the WISC between the ages of 5 and 12 years. The 23 boys in this sample had mean verbal and performance scores of 99 and 88 respectively; for the
girls the mean scores were 96 and 36. This verbal-performance discrepancy was not subjected to statistical analysis.

Dennis et al. (1981) have reported on 78 patients with shunted hydrocephalus who were assessed with the WISC or the WISC-R between the ages of 5 and 15 years. These authors employed very strict criteria for selection of subjects in their study. For example, only patients with IQ scores of 70 or greater on either or both of the verbal and performance scales of the Wechsler tests were included in the analyses. The latter criterion was used because the proportion of normal individuals with higher verbal IQ than performance IQ increases with an overall increase in IQ, while the reverse occurs as overall IQ decreases (Matarazzo, 1972; cited in Dennis et al., 1981). This is a property of the test, rather than of any abnormality and the implication is that the VIQ and PIQ are most similar at the average level of intelligence. For this reason, Dennis et al. restricted the permitted IQ range in their sample.

Another important feature from the Dennis et al. study is that they not only reported on VIQ, PIQ, and FSIQ, but also employed a fourth score. This concerned a verbal-performance (V-P) discrepancy score that was derived from the following formula:

\[
\frac{(VIQ - PIQ)}{\text{Mean (VIQ + PIQ)}} \times 100
\]

According to Dennis et al., using this formula, the IQ pattern of two children can be directly compared, even when their scores come from different ends of the IQ distribution. It reflects a relative rather than an absolute difference between the two IQ measures. We have not observed this V-P discrepancy score in any other publications.
Dennis et al. used this measure in their analyses of the role of a
variety of demographic, symptomatic, pathologic, and treatment
variables with regard to later intelligence and these analyses
will be described in a later section.

The common outcome of early hydrocephalus, according to
Dennis et al., was an uneven growth of intelligence during
childhood, with PIQ developing less well than VIQ. Compared by
chi-square analysis to the population frequency distribution of IQ
scores, both FSIQ (mean = 90.80) and VIQ (mean = 96.19) scores
were within the expected range and only the PIQ (mean = 86.6)
distribution differed significantly from normal, with this
distribution being skewed towards the lower IQ values.

Hurley, Laatsch, and Dorman (1983) administered the WISC-R or
WAIS to a sample of 30 patients with spina bifida and
shunt-treated hydrocephalus, aged between 10 and 20 years. A
control group consisted of 30 subjects, matched by age within two
years and by FSIQ within 10 points. The subjects in this control
group had diverse medical histories, including mild mental
retardation of unknown etiology, learning disabilities, and
psychiatric disorders. The hydrocephalic and the control group did
not differ significantly in the absolute level of IQs but differed
in the pattern of intelligence on the Wechsler scales.

Hydrocephalic subjects typically had higher VIQ than PIQ but this
was not true for the control group. The statistical analysis used
in this context was to subject the absolute VIQ minus PIQ
difference to ANOVA, which showed a significant difference between
the two groups.
In summary, it appears that there is support for the suggestion that the verbal abilities, as assessed by the Wechsler scales, are relatively less affected by hydrocephalus than the performance skills on those tests. Few studies have, however, subjected this verbal-performance discrepancy to adequate statistical analyses. At the same time, it is also observed that the actual IQ data that are reported in different studies show considerable variation: the obtained mean PIQ ranges from a low of 67.1 in the Tew and Laurence (1975) study to a high of 88 in the Connel and McConnel (1981) study; for the VIQ, the lowest scores are reported by Tew and Laurence (1975), who found a mean of 78.1, whereas Lenton (1979) reports the highest mean of 102. These differences again underline the importance of differences in sample size, selection criteria, etc. that were discussed above.

2.4. Intelligence in relation to medical history variables.

As we indicated in the section on data analysis (2.3.1.), there are relatively few studies reported in the literature that have included a large number of medical history variables (MHV) in their analysis. Furthermore, the statistical analyses in such studies are often poor. To our knowledge, no study has yet been published that has adequately reported on the results of multiple prediction techniques in the context of the relation of MHV to intellectual outcome in hydrocephalic children. One aspect of this dissertation involves the analysis of the predictive power of a variety of MHV with regard to the intelligence of children with shunted hydrocephalus, using multiple discriminant analyses. The
details of this method will be described in a later section. We will now attempt to summarize the studies reported so far in the literature that have dealt with MHV. We will deal with demographic variables, symptom variables, variables related to formative pathology, and variables related to treatment (a classification suggested by Dennis et al., 1981).

2.4.1. **Demographic variables.**

One of the first studies to systematically report on demographic variables was that by Raimondi and Soare (1974). They studied a sample of 200 hydrocephalic children prospectively. The age of the children at testing varied considerably and, therefore, different tests (Cattell Infant Intelligence Scale and Stanford-Binet) were used to assess intelligence for younger and older children. Five diagnostic groups were studied: (1) Internal hydrocephalus (excluding those cases associated with myelomeningocele), (2) hydrocephalus associated with myelomeningocele, (3) hydrocephalus with porencephaly, (4) Dandy-Walker syndrome, and (5) external hydrocephalus. Hydrocephalics whose disorder was secondary to diagnosed infection or tumor were excluded from this study. Independent variables included in this study were diagnostic group, race, mean age at first shunt, number of shunt revisions, and degree of shunt function.

With regard to demographic variables, Raimondi and Soare reported a significant effect of race. This was analyzed by means of a 2 x 4 ANOVA (two races, black and white; four diagnostic
groups because there were too few children with Dandy-Walker syndrome. White patients had higher IQs than black patients and this was independent of diagnostic group. As the authors indicated, this is not an uncommon finding in the normal population and they found the same results among the normal siblings of their patients. The difference between blacks and whites was the largest in the internal hydrocephalus group, but within this group it also appeared that blacks were shunted later than whites and age at shunting was found to correlate negatively with IQ.

Frew (1977) commented on the role of sex in a series of 58 cases of spina bifida and reported that girls had lower IQs on the WISC than boys, but he did not describe in detail how many of his subjects had hydrocephalus and what statistical analysis he used. Nielsen (1980) also investigated the role of sex with regard to intellectual development in spina bifida. He presented data on serial assessments of 30 myelomeningocele patients (with little information about the prevalence, nature, etc., of hydrocephalus). Up to the age of 3 years, there was a slight, but non-significant sex-difference in the favour of girls on the Cattell but at 6 years, however, the mean FSIQ on the WISC was the same for both sexes. It must be noted that far fewer children were available at the age of 6 and that Nielsen did not indicate what statistical test he used.

Sex was also among the demographic variables studied by Dennis et al. (1981). As was indicated above, this study employed very strict selection criteria for the inclusion of subjects. No
significant differences for any IQ measurement (see 2.3.) were found between boys and girls by means of ANOVA.

Dennis et al. also investigated other demographic variables, such as the age at testing and handedness at testing. No significant relationship between age at testing and any IQ score was found. Similarly, right- and left-handed children did not differ significantly on any IQ measure. Lonton (1976) also reported that the WISC scores of children with spina bifida and hydrocephalus showed no significant differences between left- and right-handers (statistical test not specified), but in this sample significantly more mixed handers were found in the low IQ (< 70) groups.

Socio-economic status (SES) was controlled for in some studies but its effect on the intelligence of hydrocephalic children has not frequently been analyzed. Raimondi and Soare (1974) reported that at the time of their publication they had too little data to make comparisons that included SES, but they did suggest that the before-mentioned IQ differences between black and white groups were indicative of SES differences.

Soare and Raimondi studied 173 children with myelomeningocele, 133 of whom developed hydrocephalus and had shunt procedures. They used a SES index, based on the mother's education and family income and the index scores were split at the median of the total sample. The mean IQ of the 85 patients above the median was significantly higher (as analyzed by t-test) than that of those below the median.
Tromp et al. (1979) studied the effect of SES on the intelligence of 78 5-19 year-old hydrocephalic children on a wide variety of intelligence tests. Etiologies of hydrocephalus were diverse, but cases of tumor cerebri and gross cerebral deformation were excluded. These authors reported that the most important technique of analysis in their study included multiple regression analysis, but they did not report important information such as what combinations of independent variables were studied, what the obtained Beta weights and intercepts were, or how much of the total amount of variance could be explained. They did report that educational level, professional level, and estimated IQ of the parents all showed significant correlations with the IQ of their children. These coefficients ranged from .253 to .320. The authors also reported that educational level was used as a "control variable" when evaluating the influence of other predictor variables, such as etiology, sensory level, etc., but did not indicate how this was done.

Lenton and Sklayne (1980) investigated the relationship between social class and intelligence on the WISC in a sample of nearly 1000 patients with spina bifida with a mean age of 14.8 years. Reportedly, "suitable alternative tests" were administered to those subjects who were outside the age or ability range of the WISC. Social class was rated on the basis of paternal employment at the birth of the affected child. ANOVAs were used for statistical analyses. There appeared to be statistically significant effects of social class for all IQs of the WISC. The authors argued that the "major divide" with respect to VIQ was the
boundary between the skilled non-manual and the skilled and semi-skilled manual workers but they did not provide statistical support for this (for example in the form of post-hoc analyses). The authors further emphasized that the range in mean IQ across social classes was less for PIQ (15 points) than for VIQ (25 points) and argued that that was in agreement with other studies that found that verbal skills were more depressed than nonverbal skills in deprived circumstances. The authors seemed to forget, however, that a statistically significant main effect of social class was also found in the analysis for PIQ, so we question the validity of this 15 versus 25 point difference.

In the study by Lenton (1982) that will be discussed in section 2.4.4. social class (based on paternal occupation) correlated significantly with intelligence. The strength of the correlation (−.15) was equivalent to that found for other variables such as location and extent of the lesion. Although these were significant values in this large sample, the magnitude of these and other correlations at the same time suggests to us that the actual amount of variance explained is minimal.

In summary, race and SES appear to be demographic variables that have an important impact on the intelligence of hydrocephalic children (just as they have on the intelligence of normal children), while handedness, sex, and age at testing appear to be of little relevance. It must be realized that not all of these variables can be readily subjected to prediction research. Sex and race are obvious at birth, but handedness is typically not. Any study that attempts to investigate the predictive power of
different WHV must discriminate between variables that are available at the time of treatment and variables that are only apparent at the time of assessment.

2.4.2. Symptoms.

As was reported in section 1, some symptoms, such as visuomotor abnormalities, are quite common among hydrocephalic children and this has been related to the pathogenesis of the disorder. Other symptoms, such as seizures, are less often reported, and with regard to again other symptoms, such as sensory level at birth, there appears to be considerable disagreement in the literature concerning their predictive value vis-a-vis IQ.

Tew and Laurence (1978) investigated the importance of ocular defects for intellectual, academic, and motor performance in a sample of 55 cases of spina bifida and 55 controls, matched for age. The children were assessed close to their tenth birthday with a variety of tests, including the WISC. 40 percent of the spina bifida cases had ocular defects, especially strabismus. The authors reported that ocular defects were "significantly related to shunt treated hydrocephalus" but it was not clear how many cases had hydrocephalus, what the statistical analysis was, and what the numerical value of this relationship was. Significant differences (again unclear how analyzed) were also reported to occur on all three IQs of the WISC between spina bifida cases with ocular defects and those without defects. Although the authors concluded that ocular defects were an indicator of multiple problems (including academic and motor problems which will be
discussed in later sections) they did not make clear whether or not they considered the low intelligence of the affected cases to be a direct result from the ocular abnormalities. Few and Laurence did suggest that ocular defect seemed to be associated with a "specific weakness in visuo-spatial and visuo-motor functioning", based on the finding that the PIQ was 11 points lower in average than the VIQ in the cases with ocular defects. The relevance of such a discrepancy in this below-normal range (VIQ = 73.3; PIQ = 62.1) is not clear, especially since the VIQ is also significantly below normal.

In the study by Dennis et al. (1981), that was described in section 2.3., four groups of symptom variables were studied: early development, ocular abnormalities, motor status, and seizures. Hydrocephalic children were classified according to the presence or absence of antenatal/labor and neonatal/infancy problems, but ANOVA revealed no significant effect on any IQ score. Ocular abnormalities (involving either gaze and movement defects or refraction and accommodation defects) appeared to be associated with a lower absolute level of PIQ, while no effect on the other measures was found. Motor symptoms were classified as normal motor development, mild motor problems, moderate paraplegia, and severe paraplegia. The three groups with motor impairment all had significantly lower PIQ and FSIQ scores than the group with normal motor development, while no effect was found on VIQ or the V-P discrepancy score. Finally, children with seizures had significantly lower PIQ, FSIQ, and V-P discrepancy scores than children without seizures, although their VIQ scores were similar.
Dennis et al. further looked at the interactions between the three different (ocular, motor and seizure) symptoms that influenced intelligence. A Chi-square test showed no statistical association between the frequencies of these symptoms. Dennis et al. concluded that this indicated that symptoms occur independently of one another, but their effects on intelligence may be additive (more symptoms increasing the risk of problems).

Motor disability was also included in the analyses by Badell-Ribera et al. (1963) of the performance of spina bifida children. The details of this study have also been described in section 2.3. These authors assigned their patients to one of five groups, according to the level of the spinal lesion. A Mann-Whitney comparison was made between the combined three groups with severe motor disability (lesion level L5 or higher) and the combined two groups with mild disability (lesion level S1 or lower). This revealed that children in the severely disabled groups achieved significantly lower scores on the WISC than those in the mildly disabled groups only when there was a history of hydrocephalus. There was no significant difference in the subgroup without hydrocephalus.

Abnormalities of the skull represent a group of symptoms that has been investigated repeatedly in the literature and has given rise to a lot of controversy. Lacunar skull deformity (LSD, craniolacunae, or Luckenschadel) was first suggested as an important predictor by Stein, Schut, and Borns (1974). It relates to abnormalities on skull films that are often only apparent in
the first three months of life. The most characteristic feature is a web-like appearance of the skull.

Stein et al. obtained a Gesell Developmental IQ from 156 children with myelomeningocele. The age range of the children was 3-10 years and it is not clear to us how the Gesell test can be applied to the older (> 6.5 years) children in the sample. Of these children, 122 had hydrocephalus and 76 of these 122 cases had LSD. Stein et al. found significant differences in IQ on Student's t-test between children with and without LSD. The mean in the LSD group was 63.8 and 95.8 in the non-LSD group. These authors concluded that the presence or absence of LSD at birth could be used as an early indicator of intellectual capacity. In a later paper Stein and colleagues have proposed that this should be an important factor in the selection for surgery for newborn babies with myelomeningocele (Stein, Schut, & Ames, 1975).

Lonton, Barrington, and Lorber (1975) have disagreed with this conclusion. They had adequate perinatal radiological assessments to assess the presence or absence of LSD for 131 children with myelomeningocele and hydrocephalus. Lonton et al. further classified LSD into five grades of severity, ranging from normal skull to severe cranial defects. Because the normal and the extreme groups contained too few children they were excluded from the analyses. Thus, whereas Stein et al. compared LSD and non-LSD children (regardless of the level of severity), Lonton et al. compared children with definite but varying degrees of LSD. Although Lonton et al. were not very clear about their statistical analyses, it was clear that the mean FSIQ obtained in each of the
three LSD groups (ranging from 82 in the severe LSD group to 90 in the group where LSD was barely perceptible) was nowhere in the region of 68.3 reported by Stein et al. Although these studies used different tests to assess intelligence, this discrepancy is large.

Lotton et al. further indicated that 82 percent of the children in the group with severe LSD (90 percent of whom had shunts) had IQs above 70 and 13 percent had IQs above 100. They concluded that these findings argued against using an LSD rating as a criterion in the selection protocol of children with myelomeningocele and hydrocephalus.

Hunt and Holmes (1975, 1976) assessed intellectual functioning in 83 survivors of spina bifida cystica, of whom 74 had required shunting for relief of hydrocephalus. The Stanford-Binet was used "in most cases", but no further specifications were provided regarding how many subjects were assessed with specific other tests. The age-range at which the children's intelligence was reviewed varied from 1.1 to 7.8 years. Hunt and Holmes investigated the role of many variables with regard to later intelligence in children who had a shunt but no CNS infection. These variables included sensory level of the lesion, occipito-frontal circumference at birth, timing of operation for insertion of shunt, thickness of pallium at time of shunting, enlargement of the head, functioning of shunt, and craniolacunae (= LSD). The authors reported that in 56 children with shunts but without infection they did not find a significant relationship between craniolacunae and intelligence. However, they
did not provide any quantitative data with regard to this relationship and did not specify their statistical analysis.

Fishman, Palkes, Shackelford, and McAlister (1977) related the severity of craniolacunae at birth with later intellectual performance in a group of 77 children with myelomeningocele. No specifications were given regarding the presence, etiology and other variables relating to hydrocephalus. The ages at which the children were tested were also unclear, although only assessments after two years of age were reported. 67 Children were tested with the Stanford-Binet or the WISC-R, 10 with the Cattell. The authors felt it was justified to combine the IQs from these scales in their analysis because of the "high correlation" between the tests. We strongly disagree with this, not only because of the large age-variation in this sample, but also because the reported correlation of .73 between the Stanford-Binet and the WISC-R means that they have just over 50 percent common variance. Furthermore, as was mentioned in section 2.1.2, these tests assess different functions.

Nevertheless, Fishman et al. reported that LSD was present in 72.7 percent of their sample. An ANOVA revealed that there was a significant effect of the severity of craniolacunae, which appeared primarily due to the difference between the group without LSD and the group with the most severe form of LSD (differences between other groups were not significant). Fishman et al. concluded that these results partly agreed with the findings of Stein et al. (1974), but they also pointed out that the mean IQ of the total population of children with LSD in their sample (79.3)
was more near the scores reported by Lonton et al. (1975) for their definite and severe groups than to the mean (63.8) reported by Stein et al. The authors therefore argued for caution in considering LSD as a selection criterion for treatment.

Villani, Giani, Giovanelli, Tomel, Zavanone, and Motti (1976) investigated the role of another form of skull abnormalities, namely cranial changes following shunting procedures. 76 patients who had been shunted for hydrocephalus (tumor cases were excluded) were assessed "at least once in the course of the follow-up programme extending from 2 to 12 years after surgery" with the Terman-Merrill. The skull X-ray signs that were assessed in relation to shunting included vault changes, cranial floor changes, exaggerated air sinuses, and calcified haematoma. The degree of skull change was rated from moderate to severe. Intellectual performance was classified in three broad categories: IQ > 100, 60 < IQ < 101, and IQ < 61. The authors reported that impaired mental development paralleled the degree of skull alteration, but did not report data on any statistical analysis.

A variety of studies have investigated the predictive value of sensory level at birth with regard to later intelligence in children with hydrocephalus and myelomeningocele. It must be emphasized that when reference is made in the literature to 'lesion level', it is not always clear what this means. Some studies refer to sensory level, others to motor level, and other studies to site of the herniating cystic sac in the spina bifida child. Sensory level frequently differs from both external and
radiological levels of the lesion (Hunt, Lewin, Gleave, and Gardner, 1973). We will discuss studies that relate to each of these forms of 'lesion' levels.

Hunt et al. (1973) were one of the first authors to report on the importance of the sensory level of the lesion in children with myelomeningocele. They provided very little information concerning the presence and characteristics of hydrocephalus in their sample of 80 children, but, nevertheless their results are relevant because of the emphasis on sensory level. Sensory level at birth was assessed by prickling the resting baby with a pin, working from the region of the lowest dermatome upwards, and the level at which general arousal occurred was noted. Three groups were defined as (a) high sensory level (T5-T10), (b) intermediate sensory level (T11-L3), and (c) low sensory level (L4 and below). The authors reported that the group with low sensory levels had significantly better IQs than the other two groups combined, but IQ tests and statistical analyses were not specified, the children appeared to have been assessed at quite different ages, and the number of children in each group was quite different.

Hunt and Holmes (1975; 1976) commented more specifically on the role of sensory level in children with myelomeningocele and hydrocephalus, that was not associated with CMG infection. The details of this study have been discussed above. The authors reported that intelligence was significantly related to sensory level of the lesion recorded at birth. Their results appeared to suggest that relatively more children with higher lesions were
intellectually subnormal (defined as IQ < 80), but both IQ measures and statistical analyses were very poorly described.

Lonton (1979) investigated the relationship between vertebral location of the myelomeningocele sac and subsequent intellectual abilities in a sample of 190 children with myelomeningocele and hydrocephalus. Most of the children were assessed at the ages of five and a half to seven years with the WISC. The nature of the statistical tests used was unclear and the method at least questionable: the mean IQs of groups with specific locations were compared to the mean of the whole group to see if there was a significant difference. An analysis of variance or a correlational analysis would have been more appropriate. In any event, Lonton found that children with thoracolumbosacral (TLS) lesions had significantly lower IQs than the sample mean, while children with sacral lesions had significantly higher IQs. Sac level appeared to have other consequences as well. For example, the majority of the children with TLS lesions were chairbound, while none of the children with sacral lesions were so. On the other hand, there was a very high incidence of urinary incontinence in the group with sacral lesions. Although these latter findings may seem obvious for neurological reasons, they again underline the necessity of considering the relationships between multiple independent variables with regard to their combined effect on specific dependent variables. The relevance of factors such as immobility and incontinence will also be discussed in the section on social-emotional development.
Soare and Raimondi (1977) studied 173 children with myelomeningocele, 133 of whom also had shunted hydrocephalus. The details of this study have been discussed in section 2.2. The value of this article was that the authors tried to determine whether there was a relationship between the level of the myelomeningocele sac, sensory level, and IQ, independent of the effects of hydrocephalus. When children with only myelomeningocele and children with both myelomeningocele and hydrocephalus were analyzed separately, they found that neither sensory level nor level of the sac were significantly related to IQ. When the two groups were combined, however the apparent difference between higher sensory and sac levels (relatively low IQ) and lower levels (relatively higher IQ) reached statistical significance on ANOVA. It would have been preferable if this had also been analyzed by means of correlation. Soare and Raimondi concluded that knowledge of the level of the sac or the sensory level is not a reliable predictor of IQ.

Tromp et al. (1979), whose study was outlined in section 2.4.1., failed to find a significant effect of sensory level on intelligence. This was analyzed by means of a three-factor ANOVA (nature of hydrocephalus x etiology x sensory level within the spina bifida group). Encephaloceles stood out as having a very poor intellectual prognosis and the analysis of the effect of sensory level excluded the encephaloceles from the spina bifida group.

Lorber (1971) was not very clear about what he referred to with "site of lesion", but it appeared to relate to level of the
myelomeningocele sac. Details of this study were discussed in section 2.2. Lorber reported that patients with thoraco-lumbar or thoraco-lumbo-sacral lesions had the most severe hydrocephalus, the most severe paralysis, and the highest mortality rate. Intellectual development was inversely related to the site of the lesion, which Lorber attributed to the association of intelligence with the presence and severity of hydrocephalus. Again, statistical analyses were lacking in this study.

Smith and Smith (1973) assessed the importance of the 'neurological level' of the lesion in a series of 88 children with myelomeningocele, 56 of whom had been shunted for hydrocephalus. This 'neurological level' related to a motor level, and was specified in terms of which muscles were affected, with the presence or absence of an active quadriceps as the distinguishing feature between high and low lesions. The motor level was assessed at birth by means of presence or absence of active movements and of posture of the limbs. These authors reported that they had not found sensory loss useful in determining the 'neurological level' in the newborn period, but did not elaborate on this point. Intelligence was assessed when the children were aged 7 to 11, but the instruments were not specified. It appeared that a larger proportion of the low lesion group (81 percent) than of the high lesion group (63 percent) had a 'normal mental state', which was defined as an IQ of 86 or above. Statistical analyses were lacking.

Mapstone et al. (1984; see section 2.2.) reported that in a series of 75 children treated for myelomeningocele and/or
hydrocephalus the IQ dropped with higher levels of the myelomingocele sac. They also reported that this inverse relationship between IQ and lesion level did not reach statistical significance, but did not specify how this was analyzed.

In summary, the literature on the predictive importance of symptom variables with regard to intellectual outcome in hydrocephalus and myelomingocele is characterized by contradictions. While most authors agree on the preponderance of ocular and motor symptoms in hydrocephalic children and the specific effect this may have on their performance intelligence, there is widespread disagreement regarding the role of other symptoms such as craniolacunae and sensory level at birth. The discrepancies among studies of course relate partially to the methodological inadequacies that were discussed before. On the other hand, the fact that not all studies investigate the same combination of variables is problematic because the effect of different symptoms might very well be additive, even if they are relatively independent from each other.

2.4.3. Formative pathology.

Because hydrocephalus can manifest itself in different forms and can be related to a wide variety of underlying pathologies it is not surprising that researchers have attempted to relate intellectual outcome to the type and pathogenesis of the disorder. Some studies have contrasted children with IVOH and children with EVOH, other studies have looked more specifically at the very nature of pathology or site of obstruction. A discussion of
studies dealing with differences between hydrocephalic patients
with and without spina bifida was presented in section 2.2. (Brown
& McLone, 1981; Mapstone et al., 1984; Soare & Raimondi, 1977;
Shurtleff et al., 1973).

Tromp et al. (1979; see 2.4.1. for details) found that
patients with noncommunicating (IVOH) hydrocephalus had
significantly lower IQs (mean = 73.9) than patients with
communicating hydrocephalus (EVOH; mean = 89.1). This was a
replication of a finding reported earlier by these authors (Tromp
& van den Burg, 1978) in a study with a similar methodology but
with a smaller subject sample. This same finding was not
supported, however, by the results of Dennis et al. (1981; see
2.3. for details) who reported that the type of hydrocephalus
affected neither the level nor the pattern of intelligence.
Children with IVOH and EVOH did not differ significantly on any IQ
measurement. At the same time, Dennis et al. did find that the
site of obstruction within the ventricular system affected
intelligence in the IVOH group. Children with blocks of the
aqueduct or anterior third ventricle had lower PIQ than those with
obstruction at the fourth ventricle or its exits.

Apart from differences in sample selection, IQ measurement,
etc., the contradiction between the Dennis et al. and the Tromp et
al. study with regard to the importance of type of hydrocephalus
may be related to the relatively small number (8) of EVOH cases in
the Dennis et al. study. On the other hand, Dennis et al. appeared
to be more conservative in their way of assigning cases to either
group, since they reported that they could not clearly determine
the type of hydrocephalus in 19 cases, while Tromp et al. apparently had no difficulty with the decision on type of hydrocephalus.

Tromp et al. and Dennis et al. also disagreed with regard to the importance of etiological variables. Tromp et al. reported that children with encephalocele (only three cases) performed particularly poorly, but the differences between the other etiological groups (trauma, meningitis, congenital, and associated with spina bifida) were not statistically significant. Dennis et al., however, concluded that the etiology of hydrocephalus affected the pattern of intelligence. Specifically, this related to a significant difference between congenital etiologies and postnatal etiologies. Children with aqueduct stenosis had a significantly greater V-P discrepancy score than children with hemorrhages and hematomas or with infections and adhesions. It is important to realize that this means that there was a difference in pattern of intelligence: the FSIQ, VIQ, and PIQ measurements did not differ significantly. This again illustrates that the way the data are quantified and analyzed has important consequences for the findings.

Shurtleff et al. (1973; see 2.2. for details) discussed pathologic variables such as infections, hemorrhage, and prematurity, but they did not discriminate whether these variables were a cause or a complication of the hydrocephalus. If children with infections with and without myelomeningocele were compared to children with congenital hydrocephalus with and without myelomeningocele, there was a significantly higher percentage of
cases with mental retardation (defined as IQ < 80) in the former group (analysis by means of Chi-square).

Raimondi and Soare (1974; see 2.1. for details) found significant differences in mean IQ among their diagnostic groups. The group with Dandy-Walker syndrome was excluded from this analysis because it had too few cases. A 2 x 4 ANOVA showed a significant effect of race (see 2.4.1.) and of diagnostic groups, but no interaction. The group with myelomeningocele was significantly brighter than any of the three other groups. Of these latter three, the groups with internal hydrocephalus and external hydrocephalus had significantly higher IQs than the group with porencephaly.

McCullough and Balzer-Martin (1982) assessed the intelligence of 37 children with overt congenital hydrocephalus after an average 6-year follow-up period. The tests that were used to assess intelligence were very variable, and, apparently, so was the age at which the children were assessed. The selection criteria (e.g., premature babies, infants with hydrancephaly, and tumor cases were excluded) and the data concerning the etiology or site of obstruction were well described. The authors reported a "clear relationship between cause and outcome" but failed to provide statistical support. The mean IQ of the children with communicating hydrocephalus (109) or Arnold-Chiari malformation (110) was quite different from that of children with aqueductal atresia (71) or Dandy-Walker syndrome (45). It must be noted, however, that the number of cases in some groups was quite small.
(e.g., 3 in the Dandy-Walker group; 4 in the Arnold-Chiari group) and that the standard deviations were generally very large.

Upadhyaya, Bhargava, Dube, Sundaram, and Ochaney (1982) have reported on the intellectual performance in 110 Indian children shunted for hydrocephalus. Most of the children belonged to poor SES strata and were late referrals. Of these cases, 58 were congenital and 52 post-meningitic in origin. Intelligence measurements included Indian versions of the Gesell and Stanford-Binet. The authors provided data for subgroups assessed at different intervals between the age at operation and the age of testing. Analysis by means of multiple independent t-tests failed to reveal significant differences in mental performance between congenital and post-meningitic groups.

In summary, disagreement prevails with regard to the impact of pathologic variables on intellectual development in hydrocephalus. As was noted already, many of the observed discrepancies relate to methodological issues and especially to the way the data are analyzed. Dennis et al. have indicated that the pattern of intelligence may reveal significant differences between diagnostic groups where the absolute levels do not. It is therefore unfortunate that none of the other studies discriminate between verbal and performance intelligence.

2.4.4. Treatment variables.

Studies dealing with comparisons between treated versus not treated hydrocephalic children were discussed in section 2.2. The following discussion will focus on variables that are related to
the children that have been shunted. Although some of these variables (especially measures of the degree of ventricular enlargement and of regional cortical thinning) might also be considered as symptoms, they are discussed in this section because they are variables that are typically assessed at the time of treatment. Furthermore, other authors (e.g., Dennis et al., 1981) also discuss them under treatment variables.

Because of the fact that rapidly progressive hydrocephalus can be fatal if not treated immediately it is not surprising that some authors have investigated the importance of the age of the children at operation. In children with myelomeningocele, the spinal defect is usually treated first. Prompt closure of the sac decreases the rate of infection and permits an early placement of a shunt (Selker, Steward, Cairns, & Chalub, 1973).

Raimondi and Soare (1974; see 2.1. for details) found that intelligence was related to the age at initial shunt placement in their subgroup with 'internal' hydrocephalus (defined in this study as all IVOH and EVOH not due to subdural hematoma and effusion secondary to trauma or infection, and excluding cases with myelomeningocele). In these 50 cases a correlation of -.26 was found between the age at first shunt and IQ (significant at .05 level). The correlation for the group with myelomeningocele was not significant (100 cases; \( r = .13 \)). These authors also reported that if the analysis was broken down into those shunted before six months and those shunted after that, a \( t \)-test indicated that the former group was significantly brighter than the latter.
The authors did not appear to realize, however, that this finding was strongly influenced by the variable diagnostic group.

As was reported in section 2.4.3., they had already found that children with myelomeningocele were significantly brighter than any of the other three groups. From their tables, it was also apparent that this group was in general operated on much earlier than some of the other groups. For example, the mean age of the myelomeningocele children at the first operation was 2.4 months and that of the group with internal hydrocephalus 20.4 months. Therefore, an analysis that breaks the total subject population up into cases operated before and after six months is strongly influenced by diagnostic group. It is therefore not surprising that a significant correlation between age at shunting and intelligence was only found in the 'internal' group, with such a high mean and even higher standard deviation.

In the study by Hunt and Holmes (1975, 1976; see 2.4.2) 90 percent of the shunted cases were operated within three months after birth. The authors reported that the correlation between early operation and possible subnormal intelligence was not significant, but did not quantify this statement. Furthermore, they appeared to have analyzed their data only in terms of the proportion of normal (defined as IQ > 80) versus subnormal children, and did not actually compute correlations coefficients.

Tromp et al. (1979; see 2.4.1.) also failed to find a significant effect of age at the first shunt operation. They also provided other important information. Analysis of variance revealed that cases of IVOH were generally operated upon much
earlier than those of EVOH. This is important in the context of their earlier reported finding that children with IVOH had lower IQs than children with EVOH. This combination of findings may suggest that children with IVOH had generally more severe hydrocephalus than those with EVOH and therefore received more rapid treatment, but this was not analyzed by these authors. At the same time, this entanglement of variables again illustrates the need for a type of analysis that considers the predictive power of specific medical history variables in concert with other medical history variables.

Tromp et al. also reported that spina bifida patients and trauma patients were shunted earlier than patients from the congenital and meningitis groups. Date of shunting failed to show a significant correlation with intelligence, even when the statistical effects of nature and etiology of hydrocephalus were controlled. However, they did not indicate how this control was exerted, nor was the correlation quantified. The authors concluded that "no crucial period after which shunting might result in relatively low intelligence" could be identified.

Upadhyaya et al. (1982; see 2.4.3.) reported that children operated upon before the age of three months appeared to have a better mental performance than children operated later, but that these inter-group differences were not significant. They did not comment, however, on the fact that in their post-meningitic group children operated between the ages of 6 and 12 months appeared to have the best outcome and they did not analyze their data in a way that allowed consideration of the variables pathology and age at
shunting independently and in interaction with each other (e.g., MANOVA).

Dennis et al. (1981) compared four types of shunts to see whether this variable had any significance with regard to intellectual development: children with only lumbo-peritoneal shunts, and children with shunts in the left, right, or left and right ventricles. The shunts in the latter three groups ended in either the peritoneum or atrium, but no further analysis was provided in terms of ventriculo-peritoneal versus ventriculo-atrial shunts. Dennis et al. reported that no significant effect of these four types of shunts was found for any IQ measure.

Keucher and Mealey (1979) have investigated 228 patients who were treated with either a ventriculo-peritoneal (VP) or a ventriculo-atrial (VA) shunt. They did not report on any intelligence assessments, but their findings are relevant in this context due to the role of other medical history variables. For example, children with VP shunts required fewer revisions than children with VA shunts and the latter group also had a greater number of complications and more serious complications. Although the statistical analyses in this study were unclear, these findings again indicate that one medical history variable can not be validly analyzed independently of other variables.

The degree of hydrocephalus at the time of shunt insertion has been assessed in a variety of ways and many authors have investigated the importance of these measures. The most common
Indices include a measurement of the distance between the
ventricles and the skull (ventriculo-skull distance, VSD), which
can also be used as an estimate of the thickness of the cerebral
mantle. These are absolute measures. There are also relative
measures, such as the Evans index (described by Tromp et al.,
1979) as the quotient of the maximum width of the lateral
ventricles and the maximum width of the inner table of the skull).
Because cerebral mantle is a linear measurement that does not
reflect brain mass accurately in the presence of head enlargement,
Shurtleff et al. (1973) have developed an index of volumetric
brain size that corrects for this. Their widely used but simple
formula utilizes only two biological measurements: frontal VSD and
maximum occipital-frontal circumference (OFC) of the skull. For a
discussion of a variety of other indices, see Tromp et al. (1979).

Shurtleff et al. (1973; see 2.2.) have analyzed the
predictive power of VSD, OFC, and their above-mentioned brain mass
ratio with regard to later intelligence. When the progression of
hydrocephalus was expressed by consideration of both brain
thickness (VSD) and head size (OFC), the children with brain mass
of 60 percent or less of normal mean for age were all retarded
(i.e., had an IQ < 80) and none with more than 60 percent of
normal brain mass was retarded. This relationship, however, only
held for their subgroup of 19 cases with congenital hydrocephalus
without myelomeningocele and without complications. In the other
subgroups, children were found that had a normal brain mass and
were still retarded. Shurtleff et al. concluded that an estimate
of brain mass that considers both OFC and VSD can be used to
generate a prediction of future intellectual performance in children with uncomplicated hydrocephalus. They also concluded that neither OFC nor VSD alone would yield as accurate a prediction, but they did not provide detailed information regarding the exact numbers of retarded versus non-retarded children correctly classified by each measure.

Young, Nulsen, Weiss, and Thomas (1973) have reported that IQ on the Stanford-Binet or the WISC can be related to the ultimate frontal cerebral mantle in hydrocephalic children. This is of course not a predictive variable, since in this study the cortical thickness was measured at the time of psychological assessment. These authors have suggested that achievement of a cerebral mantle of 2.8 cm at the time of treatment can be seen as the cutting point for attainment of an IQ > 80, but they did not analyze the mantle at the time of treatment and did not discuss how much mantle restoration can occur following treatment.

Raimondi and Soare (1974; see 2.4.1. for details) subjected the diagnostic radiological studies of 77 of their patients with internal hydrocephalus and hydrocephalus with myelomeningocele to an analysis for degree of hydrocephalus at the time of the first shunt. The ventricular dilation was graded on a scale from 1 to 4 (increasing in severity), based on visual inspection of the angiograms, pneumoencephalograms and ventriculograms done prior to initial surgery. The decision was based on the apparent size of the ventricles, thickness of the cerebral mantle, vascular deformity and displacement, and vascular integrity, but no quantitative formula was used. Chi-square analysis revealed that
the patients with myelomeningocele were significantly more often in the less severe category prior to surgery than the patients with internal hydrocephalus. A correlation between the severity of hydrocephalus and IQ was .01 and was not significant. The authors interpreted this as indicating that there was no relationship between the severity of hydrocephalus prior to initial surgery and later intelligence.

Hunt and Holmes (1975, 1976; see 2.4.2. for details) assessed the degree of hydrocephalus before treatment in a variety of ways. First, they measured OFC at birth, using norms that related OFC to birth weight. These data were available for 76 children, 67 of whom were shunted. Second, they measured the enlargement of the head in the first four weeks of life. This was dichotomized as an increase of more or less than 5 cm in four weeks (corresponding to an increase from the 10th to the 90th percentile). These data were available on the same 76 children. Third, in 41 children operated on in the first four weeks of life, the thickness of the pallium was measured just below the right parietal eminence at the time of operation. The authors reported that these three measures were not strongly related: OFC at birth did not relate significantly to the thickness of the pallium at operation, nor did enlargement of the head (statistical information not quantified). There was reportedly no close correlation between OFC and later intelligence, but this correlation was not quantified. The authors also reported that a rapid increase in OFC in the first month of life was not associated with poorer intellectual outcome, but did again not provide specific information concerning statistical
analysis. Pallium thickness was dichotomized as more or less than 2.5 cm at operation. This dichotomy was apparently subjected to Chi-square analysis (although the authors were not clear on this) and a significant relationship was found with subsequent intelligence. More children with a thin pallium were intellectually subnormal (defined as having an IQ < 80). This relationship did not persist in children operated on after four weeks of age.

Lonton (1979; see 2.3. for details) used a ventriculo-to-brain ratio (V/B %) as an index of the degree of hydrocephalus. This ratio was apparently based on the CT scan level that showed the largest area of the ventricles, but was not further specified in terms of quantitative computations. The degree of hydrocephalus was rated on a scale of 1 to 6, with the first five groups being classified in terms of successive 10 percent increases of V/B % and the sixth group having a V/B % of 51-100. Verbal and performance IQs were obtained for each group. Although the author reported a significant effect of V/B % on PIQ, but not on VIQ, the way he analyzed this was totally unclear. It appeared that the children in the sixth group were in general performing less well, but this publication did not report on any kind of correlational analysis over all groups combined. Lonton also reported that the WISC scales most affected by increasing ventricular size were Picture Arrangement, Block Design, Coding and Object assembly, but again, the way this was analyzed was not clear.
In a later publication, Lonton (1982) reported on numerous independent Pearson correlation coefficients between a variety of medical history variables and IQ on the WISC in a sample of 966 children with some form of spina bifida with or without hydrocephalus. Neonatal pallium, measured by air ventriculography, and V/B ratio on CT scan (measured by the method described in Lonton's previous study from 1979) were among the variables that yielded significant correlations. Although the author mentioned that "appropriate multifactorial analyses" were needed to further investigate the predictability of intelligence, he did not indicate why he had made no attempt at using such analyses himself.

Tromp et al. (1979; see 2.4.1. for details) used several indicators of severity of hydrocephalus before treatment. They used both absolute measures such as cortical thickness and width of the third ventricle, and relative measures, including Evans index and residual brain mass/skull volume ratios. For the latter ratios, they used both the method described by Shurtleff et al. (1973) and another ratio, developed by the authors themselves that was supposed to reflect ventricular dilatation more adequately. This new estimate was well described in the article. According to the authors, however, none of the preoperative severity measures correlated significantly with later IQ of the children (but no quantitative data were provided). The authors could specifically not corroborate a finding from their previous study (Tromp & Van den Burg, 1978) in which they reportedly found evidence that third ventricle width might be relevant in
predicting later intelligence. In that study, though, the significant effect of third ventricle width was only found when the analyses were restricted to a group of patients shunted within 110 days of birth. When the complete group of subjects was included in the analyses, none of the measurements of brain mass or ventricular dilatation showed significant correlations with IQ.

Dennis et al. (1981; see 2.3. for details) rated thinning of the cerebral mantle as mild, moderate, or severe in degree. A figure was provided to illustrate what "typical" thinning classifications would mean in terms of normal and dilated ventricular systems, but no quantitative data were mentioned. Assessments of the degree of thinning were made separately for the whole cortex, right and left frontal lobe, and right and left vertex-occipital lobe. Separate ANOVAs for each of these assessments did not reveal any statistically significant effect on any of the four IQ measures that were employed. In our opinion, this should have kept the authors from performing post-hoc analyses, but they decided to look at regional differences in cortical thinning.

Children with proportionally thinner vertex-occipital than frontal cortex were reported to have lower PIQ than children whose frontal-lobe thinning was greater than or equal to vertex-occipital lobe thinning (analysis by means of t-test). The authors concluded that a selectively thin cortex at the vertex and occipital lobe is directly related to the pattern of intellectual development in children with IVOH. We do not feel this conclusion is justified. First of all, there was a methodological problem
involved in analyzing pre-selected dichotomous groups after failing to find main effects on the variables used to dichotomise. Second, the effect found by these authors in their vertex-occipital versus frontal analysis was far from statistically significant (a \( p < .11 \) was reported). We therefore think that the Dennis et al. study did not demonstrate a significant effect of mantle thickness at the time of first shunt operation to later intellectual development.

McCullough and Balzer-Martin (1982; see 2.4.3. for details) used measurements of head circumference at birth (HC), VSD, and the index suggested by Shurtleff et al. (1973) as indices of the preoperative degree of hydrocephalus. Intellectual performance was classified as normal (IQ > 80), borderline (65 < IQ < 80), or retarded (IQ < 65). Chi-square analyses revealed that the mean VSD of children in the 'normal' group (2 cm) was significantly higher than that in the 'retarded' group (1 cm). Although the authors reported that the brain mass ratio of Shurtleff et al. did not achieve statistical significance, they also found that none of the patients with a brain mass of less than 60 percent than the normal mean for age (suggested by Shurtleff et al. as a critical cut-off point for normal development) had an IQ above 60. The authors concluded that poor IQ outcome in overt neonatal hydrocephalus can be predicted on the basis of mantle thickness and brain mass before therapy. Their results appear to support the first part of this conclusion (although the analysis was not optimal and the sample small), but are equivocal with regard to brain mass.
Thompson, Eisenberg, and Levin (1982) reported on 26 hydrocephalic infants between the ages of 4.2 and 29.1 months who were assessed with the Bayley Mental Scale and with computed tomography. Etiology was diverse, but brain tumor cases were excluded. On the basis of the CT scan cuts that best represented the lateral ventricles, a ventriculo-brain ratio (VBR) was computed for each patient by means of dividing the area of lateral ventricles by the area of total intracranial contents and multiply the result by 100. Of the 26 cases, 12 had uncomplicated hydrocephalus and 14 suffered from complications such as infection or hemorrhage. The median VBRs of these two groups were not statistically different, but there was a significant inverse relationship between the Bayley MDI (mental development index) and VBR according to the Spearman rank order correlation coefficient. The authors emphasized that this relationship appeared to be of equal strength in both complicated and uncomplicated patients. Although the correlations for the separate groups were indeed similar, they did not subject this to a statistical analysis of differences between correlations (e.g., Hotelling's t-test).

Thompson et al. indicated that their results differed from those of other studies (including Shurtleff et al., 1973, and Young et al., 1973) in which the relationship between mantle thickness and cognitive functioning was specific to children with uncomplicated hydrocephalus. The authors discussed several differences in patient selection between their study and the Shurtleff et al. and Young et al. studies that might have contributed to this difference. In addition, we think that the
fact that Thompson et al. administered the same test to every patient and had a rather restricted and young age range is also an important factor that distinguishes this study from other reports in the literature and may well have been partly responsible for the difference in results.

Jansen, Gloerfelt-Tarp, Pedersen, and Zilstorff (1982) assessed 26 patients with infantile hydrocephalus at ages ranging from 21-32 years with the WAIS. Only 11 of these patients had had operations for their hydrocephalus and ventriculo-peritoneal and ventriculo-atrial shunts were even more uncommon. The authors used a V/B ratio expressed as the relation in percentage between the cross-sectional area of the lateral ventricles and the brain just above the third ventricle. Reportedly, no correlation was found between V/B ratio on CT scan and IQ, but specific quantitative information was not provided.

Upadhyaya et al. (1982; see 2.4.3. for details) used the index suggested by Lonton (1979) to measure the degree of hydrocephalus. 62 children were assigned to one of five groups, based on their V/B %, which was stratified in five successive 20 percent increase ranges. The authors used multiple independent t-tests to analyze differences between these groups (which is not an adequate way of doing this) and found significant differences between the groups with relatively high and those with lower ratios, suggesting that mental performance was inversely proportional to V/B %. The authors also pointed out, however, that there may be important exceptions to this rule: three children with a V/B % between 41 and 60 and one child with a V/B between 61
and 80 had a mental performance above 90. Similar exceptions were also reported by Lonton (1979), who made mention of a patient with a V/B % rating of 95 and had a WISC VIQ of 143, PIQ of 99, and FSIQ of 126. The existence of cases like these resemble the false positives that Lorber (discussed in Lewin, 1980) has found with regard to his own selection criteria for treatment and suggest caution against a black-white interpretation of data like these.

Amacher and Wellington (1984) followed a group of children shunted for hydrocephalus for more than five years. The description of sample characteristics and analyses was extremely poor in this study and the techniques to measure IQ and cerebral mantle were not specified. Intellectual outcome was classified as either 'excellent' or 'poor' but it was not clear what this meant. The only information that was provided that 'excellent' outcome implied "normal intellect" (not further specified) and "no significant neurological deficit" (not further specified). The authors reported that there was no significant correlation between intellectual outcome and initial mantle thickness in their series, but did not specify the statistical analysis and its quantitative results. Although this study also commented on other medical history variables (e.g., shunt infections), we will not discuss these aspects in subsequent sections because of the gross inadequacies that were just mentioned.

The variables discussed so far (age at first shunt, type of shunt, and degree of hydrocephalus and cortical thinning at the time of the first insertion) can all be assessed at the time of first treatment. Many children with shunted hydrocephalus suffer
from complications following treatment and need periodic revision of the shunt, etc. These variables are the subject of our next discussion. Again, we would like to emphasize that many of these and other variables are related to each other and that their effect on later intelligence may be interactive or additive.

Several studies have investigated whether or not the number of shunt revisions has any impact on the intellectual achievement of hydrocephalic children. Raimondi and Soare (1974; see 2.4.1. for details) compared their groups of internal hydrocephalus, external hydrocephalus, and hydrocephalus with myelomeningocele on the numbers of revisions. The latter were categorized as "no revisions", "1 to 3 revisions", and "over 3 revisions". The results of an ANOVA (with number of revisions and diagnostic groups as independent variables) reportedly showed no significant main effect of number of revisions and no significant interaction effect (the effect of diagnostic group on intelligence was discussed in section 2.4.1.). The authors interpreted this as evidence that there was no relationship between the number of revisions and IQ. Raimondi and Soare did not comment on the fact that the mean number of revisions appeared to be different among the diagnostic groups (ranging from 1.9 in the 'external' group to 4.6 in the 'internal' group).

Hunt and Holmes (1975, 1976; see 2.4.2. for details) reported that the intelligence of 23 children who did not require a shunt revision did not differ markedly from those who required one or more revisions. The way this was analyzed was not clear, but the authors appeared to have subjected the groups of children with and
without revisions with and without an IQ < 80 to Chi-square analysis and found that the proportion of intellectually subnormal children did not differ with the need for shunt revisions. The authors also suggested that the need for repeated revisions had no impact on intelligence in younger children, but might have have consequences in older children, but this suggestion was not substantiated by sufficient quantitative data.

Tew and Laurence (1975; see 2.3. for details) also examined the role of shunt revisions. Of their 31 children who were treated with shunts only 9 did not need revisions. The authors reported that the mean IQ of those without revisions (74.4) was not significantly different from those with revisions (74.4) but did not describe their statistical analysis.

Halliwell, Carr, and Pearson (1980) assessed the WISC IQs in 138 11-year-old children who were born with neural tube defects. 39 of these children were without shunts and of this latter group 11 had shown signs of developing but spontaneously arresting hydrocephalus. The statistical analyses in this study were totally unclear, but the authors reported an increasingly deleterious effect on intelligence as the number of shunt revisions increased. Interestingly, the authors commented that this may be related to the interaction with other variables, such as age at revision and reason for revision, and they suggested that multivariate statistics would be required. They did not indicate why they did not use such statistics themselves.

Dennis et al. (1981; see 2.3. for details) computed linear correlations between the number of shunt revisions and each of
their intelligence indices. This is an important difference with other studies discussed so far, that only contrast children with and without revisions. Dennis et al. reported that none of the obtained coefficients was found to be statistically significant (although the actual coefficients were not provided). They interpreted this as evidence that neither the level nor the pattern of intelligence varies with the number of shunt revisions. No information was provided on whether the number of revisions varied with the etiology or site of initial obstruction of hydrocephalus.

McCullough and Balzer-Martin (1982; see 2.4.3. for details) found that the number of revisions was higher in the group of patients with an IQ > 80 than in the group with an IQ < 80. The authors did not report whether or not this difference was statistically significant (which it probably was not, given the fact that the difference was so small: 1.8 versus 1.4) but did indicate that the group with more revisions was also on the average 1.3 years older. The latter is a very important observation, since in many of the studies discussed so far the age range of the subject sample was very large. This may have important implications for the number of revisions that the subjects may have had.

Shunt revisions may occur for a number of reasons, both prophylactically and because of malfunction of the shunt. Shunt infection may be a complication of shunt revision but may also be a reason why a shunt needs to be revised. The studies on revisions that we discussed often did not comment on how well the shunt was
functioning at the time of assessment (with the exception of Hunt and Holmes, 1975, 1976, and Raimondi and Soare, 1974) and often did not comment on shunt infections. These are variables that will be discussed next.

The study by Brown and McLone (1981) that suggested that infections may lower IQ in shunted children was discussed in section 2.2. In section 2.3, we discussed the study by Shurtleff et al. (1973) in which they indicated that infections may have an effect on intelligence in hydrocephalic children, but this study did not discriminate between infections as a cause and as a complication of hydrocephalus. Mapstone et al. (1984; see 2.2.) included ventriculitis as one of their sources of complications in the analysis that revealed that complicated hydrocephalus was associated with lower IQ than uncomplicated hydrocephalus, but they did not specifically investigate the role of ventriculitis or other infections. Other studies have deliberately limited themselves to cases without complications (e.g., Prigatano et al., 1981) or concentrated the bulk of the analysis on uninfected cases (e.g., Hunt & Holmes, 1975, 1976).

Thompson et al. (1982), whose study was mainly concerned with the importance of V/B ratios, also commented on the influence of complications. Under this heading, they included infection, hemorrhage, and other variables. They did not discriminate whether these 'complications' had occurred before or after the insertion of the shunt. A Mann-Whitney U-test revealed a trend for the Bayley MDI scores of the uncomplicated patients to be higher than those of the complicated patients (significant at $p = .06$ level).
Raimondi and Soare (1974) used three categories to assess the maintenance of shunt. Children with a "functioning" shunt had all necessary revisions carried out immediately after the diagnosis of shunt obstructions (sleepiness, vomiting, irritability, etc.). Patients with a "questionable" shunt had incomplete or unavailable medical histories. The authors called their concept of "non-functioning" shunt self-explanatory, but they did not specify whether the children in this third group presented with any neurological symptoms at the time of intellectual assessment. They also did not indicate whether the number of shunt revisions was different for these three groups. ANOVA revealed a significant effect of shunt function. Newman-Keuls post-hoc comparisons showed that children in the "functioning" group were significantly brighter than those in the "non-functioning" or in the "questionable" groups. Raimondi and Soare further indicated that none of the diagnostic subgroups in the "non-functioning" category had a mean IQ within the normal range.

Hunt and Holmes (1975, 1976) reported that in 35 of their 67 cases with shunts and without infection had a shunt that was either not working or only working sluggishly (this was not further specified). The authors reported that the intellectual outcome did not appear to relate to the function of the shunt but provided no quantitative or statistical information.

In summary, we again have to conclude that the literature is full of discrepancies with regard to an important category of medical history variables and their relation to intelligence in hydrocephalic children. We cannot overemphasize the importance of
two facts: the methodological differences between studies are significant and the different variables are often analyzed independently, although they are clearly related. For example, children with hydrocephalus that is accompanied by myelomeningocele typically get operated earlier than other diagnostic categories (which has implications for the possible role of age at first operation) and appear to have more shunt revisions. Not all studies, however analyze myelomeningoceles as a separate group and very few provide medical history variables for specific groups or the relationship between variables.

Most studies do not find an effect of the age of the child at the time of the first shunt insertion and no effect of the number of shunt revisions. Infections and other complications may have a negative impact on intelligence, but in some studies it is not clear whether these variables were present before or after the insertion of a shunt. The biggest area of controversy concerns the measurement of the degree of hydrocephalus before treatment. Especially with regard to this variable, studies differ considerably in their methods of analysis and classification. The fact that the results from studies that use the same indices seem to contradict each other (e.g., Shurtleff et al., 1973 versus Tromp et al., 1979) makes this matter even more confusing.

Furthermore, even if significant effects of a given variable are found, important exceptions to the rule are provided by some authors. In our opinion this conflicting and complex state of affairs underlines the necessity of considering the role of medical history variables in a comprehensive way that appreciates
the additive and interactive effects that they may have on the intelligence of hydrocephalic children. This dissertation attempts to do that.


3.1. Methodological aspects.

Many of the comments that were made in section 2.1. with regard to studies that focus on the intellectual development of children with infantile hydrocephalus have relevance to the studies to be discussed in this section. The lack of adequate data analyses and the inclusion of varying causes and possible sample bias are important problems in this context. Some of the studies that will be discussed in this section were already commented on in sections on intellectual development and are therefore subject to the criticisms that were raised earlier, which will not be repeated here.

Most of the studies that have commented on cognitive or neuropsychological aspects of the development of hydrocephalic children focus on rather isolated functions or variables. For example, some studies deal exclusively with motor coordination (Anderson & Plewis, 1977; Grimm, 1976; Sand, Taylor, Hill, Kosky, & Rawlings, 1974), while other studies focus on memory functions (Cull & Wyke, 1984; Parsons, 1969; Richardson, 1978) or language functioning (Dennis, Hendrick, Hoffman, & Humphreys, 1986; Tew, 1979). There are relatively few studies that have administered comprehensive neuropsychological assessments to reasonably sized
samples of hydorcephalic children. Parts of the Halstead-Reitan battery were administered by Hammock, Milhorat, and Baron (1976) and by Hurley, Laatsch, and Dorman (1983), while other studies (e.g., Prigatano, Zeiner, Pollay, & Kaplan, 1983) administered more ideographic batteries.

To our knowledge, no study has yet been published that has explored the possibility of subtypes of neuropsychological patterns in children with shunted hydrocephalus. Multivariate classification on the basis of performance on a comprehensive battery of neuropsychological tests is one of the objectives of this dissertation, and will be outlined in a later section. The current section will focus on studies that report on performance on tests of psychological abilities. Studies that deal with psychiatric sequelae and social adjustment will be discussed in another section.

3.2. Language functions

In the section 2.3, we have mentioned that hydrocephalus appears to affect performance intelligence, as assessed by the Wechsler scales, relatively more than verbal intelligence. At the same time, the language of hydrocephalic children has been reported to have some characteristic features as well.

Reviews by Dennis et al. (1986) and by Tew (1979) reveal a substantial number of reports that in some way or another describe the language abilities of children with hydrocephalus as aberrant. Hadenius, Hagberg, Hyttnes-Bensch, and Sjogren (1962; discussed in Tew, 1979) are credited for the invention of the term 'Cocktail
Party Syndrome' (CPS) as a general denominator of the peculiarities of the speech of hydrocephalic children. Most reports in the literature have not surpassed the level of clinical or phenomenological descriptions of this syndrome, that is usually characterized in terms of hyperfluent chattering with little content.

Spain (1974; see 2.3. for details) described CPS as a "hyperverbal" syndrome with echolalia and continuous non-constructive chattering. She did not formulate objective criteria for the syndrome, but reported that 30 out of 87 (≈ 34 percent) children with spina bifida and shunted hydrocephalus could be described as hyperverbal. The proportion of hyperverbal children was particularly high among those with shunts and with performance scores on the Griffiths Mental Development Test below 80. Most importantly, these hyperverbal children did not do very well on some subtests of the Reynell Developmental Language Scale. Although Spain had expected that hyperverbal children would do better on the Language Expression Scale than on the Language Comprehension Scale, this prediction was not borne out. Grasp of syntax, as assessed by the Structure subtest, appeared to be normal for age, but scores on the Comprehension and Content subtests were poor. The latter, however, was true for all children with performance scores below 80 and was not specific to those rated as hyperverbal. We can therefore not subscribe to Spain's conclusion that below average scores on the Content and Comprehension subtests, combined with average scores for Syntax on
the Reynell reflects a "characteristic pattern" of hyperverbal hydrocephalic children.

Spain's study involved only three-year old subjects. Tew has suggested that the CPS may disappear with age (Tew, 1979; Tew & Laurence, 1979). He studied an unselected sample of 49 cases of spina bifida. Initial assessments were made when the children were on the average 5.6 years and follow-ups were done at ages seven and ten years. Of these children, 30 had a shunt inserted for hydrocephalus. A control group of children who were matched for sex, place in family, social class, and area of residence was also assessed. Children were classified as having CPS if they had four of the following criteria: (1) a perseveration of response, either echoing the examiner, or repitition of an earlier statement made by the child, (2) an excessive use of social phrases in conversation, (3) an over-familiarity in manner, not normally expected in a five-year-old child, (4) a habit of introducing personal experience into the conversation in irrelevant and inappropriate contexts, and (5) fluent and normally well articulated speech. It is obvious that these criteria are vague and hard to quantify.

Tew classified 20 (40.8 percent) of the 49 children in his series as showing the CPS. The remaining 29 were described in the analysis as spina bifida (SB). Tew used Chi-square analyses as the statistical technique to compare these two groups. 16 of the 20 CPS cases had shunts and one child with CPS was reported to have arrested hydrocephalus without a shunt. CPS was relatively more common in children with higher than lower spinal lesions and more in girls than in boys, although the latter difference was not
statistically significant. Children with CPS had significantly lower VIQ, PIQ and FSIQ on the WPPSI than children in the SB group. The highest subtest score in the CPS group was Vocabulary, but the mean (5.0) was still well below age-average. Furthermore, the scores of the CPS group on the Expressive Language Scale of the Reynell were approximately seven months below their scores on the Language Comprehension Scale of this test, and some two and a half years below their chronological age. The absence of good scores on the Reynell Expressive Scale in CPS children is consistent with the performance of Spain's (1974) hyperverbal children. Although the SB children were also slightly below age expectation on the Reynell scales, there were significant differences between this group and the CPS group on both the Expressive and Comprehension Scales.

At the same time it could be argued this difference may be a reflection of the generally poor intelligence scores of the children with CPS, whose mean IQ measures were all under 65. Thus, although Tew indicated that CPS is more common in hydrocephalic children with lower intelligence, he did not make clear whether it is just an expression of low intelligence or a qualitatively different and functionally independent phenomenon.

Tew also re-assessed the children at the ages of seven and ten years. The re-assessment at the age of seven dealt with academic achievement and will be discussed in section 3.3. At the age of ten, 17 of the original 20 children with CPS were still alive. Using the criteria that were described above, only nine of these children were still considered to show evidence of the
syndrome. Although the children who had "grown out" of the syndrome were of relatively higher intelligence, the exact causes for the disappearance of the syndrome remained unclear.

Dennis et al. (1987) studied the development of language in two samples of children and adolescents, 75 with hydrocephalus in the first year of life, and 50 normal controls. Their study had a three-fold purpose. The first concerned the effects of hydrocephalus and of age at testing on five different domains of language function (word finding, fluency and automaticity, immediate memory for sentences, understanding of grammar, and metalinguistic awareness). The second purpose was to assess the extent to which differences in language performance can be attributed to differences in intelligence. Exploration of the relationship between a set of medical variables associated with hydrocephalus and language functioning was the third purpose.

The age at testing ranged from 5.96 to 21.52 years. The specific tests used to assess each of the above-mentioned language functions were well described, but many of the tests were experimental measures, developed by the senior author in previous studies. Intelligence was assessed with the Wechsler scales appropriate for age. Formative pathologies were well documented. Medical history variables were well described.

Three separate analyses were performed for each of the five language domains. To assess the effects on test scores of hydrocephalus, of test age, and of the interaction between the two, an analysis of covariance (ANCOVA) was performed with presence or absence of hydrocephalus as a dichotomous variable and
test age as covariate. Multiple linear regression analyses were used to assess the effect of VIQ and PIQ on language test scores. Finally, the importance of medical history variables with regard to language functions was assessed by means of multiple linear regression analyses.

With regard to the effect of age at testing and of hydrocephalus on language function, the first analysis revealed that age was generally a more potent predictor of test performance than presence or absence of hydrocephalus. Age at testing was a significant factor for all five language functions, while clinical status was significant for only one domain. Test scores improved with age in both normal and hydrocephalic children. The authors repeatedly emphasized that this improvement with age was more rapid in the normal than in the hydrocephalic group and suggested several educational implications, but they apparently forgot that a significant interaction between test age and clinical status was found for only one of their measures, (i.e., for understanding of grammar). Therefore, while these authors appear to emphasize that the hydrocephalic children improved significantly slower in some areas than the controls, we were more impressed with the similarity in rate of improvement between the two groups. Furthermore, it has been demonstrated that it is neither appropriate nor effective to use ANCOVA to control for test age (Adams, Brown, & Grant, 1985).

Both VIQ and PIQ contributed significantly to language test scores in four of the five domains, with VIQ explaining relatively more variance than PIQ. The magnitude of the contribution of
either IQ score varied considerably among the individual test measures, but the amount of variance accounted for never exceeded 32 percent. This means, as Dennis et al. indicated, that IQs did contribute to observed language test results, but were not fully explanatory. Furthermore, it appeared that fluency and automaticity (described as "lower order language functions") were less influenced by IQ than other (so-called "higher-order") functions, but this difference was not subjected to any statistical analysis.

The third set of analyses revealed that some medical history variables were predictive of language functioning while others were not, which was rightfully interpreted by Dennis et al. as an indication that hydrocephalus is not a homogenous condition. The combination of variables that formed statistically significant predictive sets differed with each language function, but some variables appeared in many models. Age at testing was a significant variable in almost every model, always being associated with higher test scores. The type of hydrocephalus also appeared repeatedly in the predictive models and IVOH appeared to have detrimental effects on more language functions than EVOH. The presence of perinatal anomalies was generally associated with better language scores. The authors suggested that this did not implicate that such anomalies have a positive effect on intelligence, but that children with such a history will do better than those whose hydrocephalus stems from major, formative, intrauterine perturbations of the ventricular system. We do not feel that this conclusion is warranted, however, since the number of
children in their sample with antenatal problems was quite small and the presence of such problems did not enter significantly in the regression analyses.

In summary, it appears that some children with hydrocephalus have impairments in specific linguistic domains, but hydrocephalus in general does not appear to be a condition that leads to universally poor language skills. Descriptive terms such as "hyperveral" and "CPS" are in general poorly defined and, as Dennis et al. have concluded, do not indicate how well and how rapidly hydrocephalic children develop language skills. At the same time, those children described as being hyperveral or having CPS do not appear to have good language skills. The intellectual capacity of hydrocephalic children has an important but not all-explaining impact on their language development and language test scores appear to improve with age. Medical history variables that appear to affect a variety of language functions in hydrocephalic children include demographic variables (age at testing), and birth and early development variables (perinatal problems and type of hydrocephalus).

3.3. Academic achievement.

Tew and Laurence (1975; see 2.3. for details) have reported on the performance of 59 children with spina bifida and a control group on the Vernon reading test, the Schonell spelling test, and the Mathematics Attainment Test. 49 Percent of the children that were alive at age seven attended special schools. Quantitative data about the performance of the children on each of the tests
that were administered were not provided, but the authors did report the results of t-test comparisons between the children with and without shunts, and the controls. Shunt-treated children were significantly poorer readers than the controls. Their spelling scores and their arithmetic abilities were not only significantly poorer than those of controls, but also significantly poorer than those of children with spina bifida without hydrocephalus. The authors commented that the poor achievement scores of the hydrocephalic children paralleled the distribution of intelligence in their sample. Indeed, the mean IQ scores found by these authors were quite low for hydrocephalic children, which distinguishes this study from some other reports in the literature (see section 2.3.).

In a later study, Tew and Laurence (1978; see 2.4.2. for details) administered the Neale Reading Test to a sample of 10-year-old spina bifida children. Although the statistical analyses were generally unclear in this study, the authors did provide a clear outline of how they analyzed the scores on the reading test. Usually the reading achievement is indicated by the concordance between the children's reading age and their chronological age but, as the authors indicated, this method does not take into account variations from the normal range of intelligence. For this reason, the authors used regression equations to calculate a reading age that would be predicted on the basis of intelligence test scores and the result was compared to the actual score to obtain an impression how far the child was performing below expectation. When the correction for lower
measured intelligence was made (which of course resulted in lower expected reading ages) the spina bifida cases with ocular defects still had actual reading ages that were 15 months below their expected age. For spina bifida cases without ocular defects this difference was only two months. This was interpreted by the authors as an indication that ocular defects in spina bifida are associated with lower academic achievement.

In the study by Tew (1979) that was discussed in section 3.2., 18 of the initial 20 cases with CPS were alive at the age of seven years and available for investigation. Only three CPS cases were able to read even a single word on the Vernon Reading Test, compared to 19 of the SB group. Similar and statistically significant (Chi-square) differences were found for spelling ability on Schonell’s Test and arithmetic ability as assessed by the Mathematics Attainment Test, although the differences were less dramatic on those tests. The achievement scores of the CPS children appeared to be directly related to their low intelligence.

Halliwell et al. (1980; see 2.4.4. for details) administered the Neale Analysis of Reading Ability (a test that reportedly measures mechanical ability and comprehension) and the Basic Number Screening Test (which reportedly measures competence in four arithmetical operations) to their sample of 138 11-year-old subjects. Both reading and arithmetic attainments were reported to be significantly lower in children with spina bifida with and without shunts as compared to 55 control subjects, matched for
age, sex, social class, and ethnic origin. The statistical analyses were, however, unclear.

Prigatano et al. (1983) administered a series of tests to 18 children with shunted uncomplicated hydrocephalus and 18 age- and sex-matched controls. The age at testing ranged from 4 to 9 years. The Vocabulary and Block Design subtests of either the WPPSI or the WISC-R were given to estimate verbal and nonverbal intelligence. Our objections to this procedure were outlined in section 2.1.2. Although the hydrocephalic children had significantly lower Vocabulary scaled scores than the controls, their total reading centile scores on the Woodcock Reading Test were not significantly different (analysis by means of t-tests). This was interpreted as an indication that hydrocephalus without complications is compatible with average academic achievement.

Tew and Laurence (1984) have reported on the relationship between intelligence and academic achievements in 51 16-year-old survivors of spina bifida. The number of hydrocephalics in this study was not mentioned but from the tables presented it appeared to be 31. The sample and the statistical analysis were poorly described. The authors found a relationship between IQ scores on the WAIS and performance on some (unspecified) tests of reading and mathematical skills. Furthermore, three interrelated factors were reported to be responsible for poor academic achievement: special school attendance, shunt insertion, and severe physical handicap. Adequate statistical analyses to give support to this report were lacking.
In summary, the academic performance of hydrocephalic children appears problematic, but it is not clear to what extent this is due to decreased intelligence, school absenteeism due to illnesses and shunt revisions, or other factors associated with hydrocephalus. Academic abilities are in general an underreported aspect of psychological functioning in survivors of infantile hydrocephalus.

3.4. Perceptual, motor, and visuo-motor functions.

Several studies have commented on ocular, motor, and related deficits in hydrocephalic children. Some of the pathological mechanisms that may underly these deficits were discussed in section 1.5. Deficits in ocular and/or motor abilities have been proposed both as an expression (Tew & Laurence, 1975) and as a cause (Spain, 1974; Dennis et al., 1981) of the relatively poor performance intelligence in children with hydrocephalus. The latter interpretation may have more face validity, since the nature of the tasks that assess performance intelligence on the Wechsler scales allows for poor performance due to pure perceptual or motor factors. We will discuss some studies that have looked in more detail at these functions.

Miller and Sethi (1971a) administered the Bender Gestalt Test and the Frostig Developmental Test of Visual Perception to 14 hydrocephalic children of school age and with an IQ in the range of 70-105. Children with sensory handicaps were excluded. Control subjects were normal school children matched for age. The statistical analyses were not specified, but hydrocephalics were
reported to be significantly inferior on both tests relative to the controls. Two further experiments, involving motor-free delayed perceptual matching, also resulted in significantly lower scores by the hydrocephalic subjects. The authors concluded that children with hydrocephalus have a deficit in the perception of visuospatial relationships.

In another study, Miller and Sethi (1971b) investigated whether perceptual deficits were also present in the tactile domain, and, more specifically, whether there would be evidence for partial callosal dysfunction in hydrocephalic children. 16 Patients with hydrocephalus in the age range of 7 to 15 years and 16 controls matched for age and sex were presented with a task that required matching shapes felt by one hand with similar shapes presented to the same (uncrossed condition) or the different (crossed condition) hand. Analyses by means of t-tests revealed that in terms of the total number of correct matches over both conditions combined, hydrocephalics were significantly poorer than controls. Specifically in the hydrocephalic group, however, the number of correct crossed matches was significantly less than the number of uncrossed matches. The authors concluded that the spatial information processing deficit, previously demonstrated for the visual modality, was also present in the tactile modality. Furthermore, there might be a partial split brain effect in hydrocephalus due to stretching of the corpus callosum.

Sand, Taylor, Rawlings, and Chitnis (1973) administered the Frostig Developmental Test of Visual Perception (Frostig test) to a sample of 37 spina bifida subjects in the age-range of 4.0 to
16.7 years. For subjects older than 10 years of age an estimated scale score was calculated. Statistical analyses included t-tests. 59 Percent of the total sample fell below the cut-off point for deviant performance. Children with hydrocephalus had significantly lower perceptual quotients than non-hydrocephalic children, but there remained considerable overlap in the performances of these two groups. Subtest 1 of the Frostig test (Eye-hand coordination) was most likely to elicit deficient performance.

Wallace (1973) made an inventory of motor dysfunctions in a sample of 225 cases of myelomeningocele. Specific attention was paid to the correlation between upper-limb abnormalities (pyramidal tract dysfunctions, ataxia, chorea, dyspraxia) and adequacy of mobility in several groups divided on the basis of level of occipito-frontal circumference, presence or absence of a shunt, sex, and age at the time of examination. Chi-square analyses were used for statistical purposes, but in a rather unusual way: for each variable, expected values were calculated from those observed and, subsequently, the observed and expected values for all groups were summated. The authors did not make clear why they did not just do chi-square analyses on observed values. In any event, hydrocephalus appeared to be associated with a significant increase in abnormal findings in the upper limbs and with a significant reduction in mobility. In general, upper limb abnormalities were associated with reduced mobility.

Gressang (1974) assessed 29 children with myelomeningocele, 20 of whom had also shunted hydrocephalus. Ages ranged from 4.9 to 9.3 years. The children were reportedly selected on the basis of
diagnosis and IQ score > 70, but the instrument with which intelligence was assessed was not specified. The following tests were given to assess perceptual processes: Frostig Developmental Test of Visual perception (Frostig), Beery-Buktenica Developmental Test of Visual-Motor Integration (Beery VMI), Ayres Space Test (Ayres), and the Southern California Figure-Ground Test (SCFG).

The author used t-tests to determine significance of differences between the non-hydrocephalic and the hydrocephalic children. A statistically significant difference was found only on the Frostig test, with the hydrocephalic children scoring higher. The children with three shunt revisions scored higher than the ones with fewer revisions. The author indicated that the results of her study were opposite to expectation. She attributed this to the small sample size, heterogenous group, and lack of pre- and post-shunt testing.

Sand, Taylor, Hill, Krosky, and Rawlings (1974) administered the Developmental Hand Function Test (DEHFT) to a group of 25 myelomeningocele patients. The age range was large and varied from six to 19 years. Hydrocephalus was reportedly present in 15 of these patients but this was assessed in an unusual and questionable way: if current (i.e., at the time of testing) measurement of head circumference did not meet the criterion of exceeding two standard deviations of the mean for the child's chronological age, the child was classified as hydrocephalus-absent. Independent t-tests were used to contrast the hydrocephalus-present and the hydrocephalus-absent groups on all 14 measures of the DEHFT. Scores on the DEHFT were expressed as a deviation between the time needed by the child to complete a
task and the mean time taken by the normative sample of this test. The deviations tended to be much higher in the hydrocephalus-present group, which was interpreted as an indication of impaired hand function. This was more evident in the children with IQs < 80.

Sand et al. discussed several possible causal factors that might be responsible for the observed high presence of impairment of hand function in their sample. Among the suggested factors was that hand function impairment might be a direct or indirect effect of hydrocephalus. At the same time, eight of the ten children with myelomeningocele without hydrocephalus (at least as assessed by these authors) also had significantly impaired hand function. Although there appeared to be more hand function impairment in the group with higher (T10-T12) lesions than in the other groups, this was contaminated by the fact that all subjects in that group were hydrocephalic. The precise effect of hydrocephalus on hand function was therefore not clear in this study, partly because of a questionable definition of hydrocephalus.

Tew and Laurence (1975; see 2.3. for details) administered the Frostig test to their 59 patients. The pattern of scores on this test reportedly closely resembled that of the IQ scores. A correlation coefficient of .82 was found, indicating that impairment of perceptual functions was strongly associated with low intelligence. Tew and Laurence interpreted this as evidence that impairment of visual perception is "in all probability" an expression of low intelligence but they did not discuss the
alternative interpretation that low performance on the WPPSI might be caused by visual impairments.

Grimm (1976) assessed 17 children with myelomeningocele and hydrocephalus, aged six to eight years, with the DEHFT and the Tactile Perception Battery (TPB) from the Ayres Southern California Sensory Integration Test. The children were categorized in terms of motor lesion level (L2 and above versus L3 and below) and also according to the status of hydrocephalus (not-shunted, shunted, and shunted with complications). One-way t-tests were used to analyze differences between groups. The results showed statistically significant age- and sex-discrepant hand function, as assessed by the DEHFT in children with hydrocephalus and high lesions. Interestingly, the performance on a test for hand graphesthesia were relatively poor in children with hydrocephalus and high lesion levels, whereas performance on a task requiring localization of tactile stimuli was especially poor in children without shunts. Of the 48 product-moment correlations computed between the hand function and tactile perception measures used, only one was statistically significant. The author interpreted this as evidence that hand function and tactile perception in children with myelomeningocele are unrelated factors. This may, however, have been influenced by the type of tests used, the small size of the sample, and the fact that these factors were not analyzed per subgroup. On the other hand, the fact that only one of 48 correlations was statistically significant can also be seen as impressive, since with so many correlations there is a
reasonable probability that some will reach statistical significance on the basis of chance.

Lonton (1976) analyzed the distribution of hand preference in an unselected sample of 203 children with myelomeningocele and hydrocephalus. The children were simply asked to draw a picture of a man and to write their name, and the preferred hand was noted. Children who used both hands were termed 'mixed handers'. Ages ranged from four to nine years. A control group of 200 normal children in the same age range and with a similar social-class distribution was also studied. The author reported that the patient group had significantly more mixed and fewer right handers than the control group, but that the incidence of left-handedness did not reach statistical significance (analysis by means of binomial test). The latter finding was probably related to the fact that the incidence of left-handedness in the control group was surprisingly high (19 percent) as compared to other studies. Lonton further reported that the mixed handers were more likely to have the upper limit of their myelomeningocele in the thoracic vertebrae. He suggested that the latter finding suggested that handedness is not only determined by cortical factors but may also be influenced by asymmetrical motor or sensory dysfunction due to lesion of the thoracic cord.

Anderson and Plewis (1977) administered a task that involved dotting between two circles, for which both speed and accuracy were measured, to two samples: 20 7-10 year-old children with spina bifida cystica and hydrocephalus, and 20 normals matched for age, sex and IQ on the Columbia Scale of Mental Maturity. ANOVA
revealed significant impairment in dotting speed in the spina bifida group, although both groups improved with practice. If visual monitoring of this task was subsequently restricted, spina bifida children were initially more affected than the controls, but were able to recover. The latter was interpreted as evidence that kinesthetic ability is not markedly impaired in spina bifida children as a group. The authors did not discuss in detail to what degree ocular abnormalities might have contributed to the poor performance of the spina bifida children in the condition with visual feedback.

Minns, Sobkowiak, Skardoutsou, Dick, Elton, Brown, and Forfar (1977) administered a variety of non-standardized clinical "neurological" (two-point discrimination, finger-nose test, etc.) and "functional" (using a knife and fork, threading a needle, etc.) tests to 31 children with spina bifida with a mean age of 7.2 years, 23 of whom had shunts for hydrocephalus. A control group of 31 children referred for simple otolaryngological procedures was administered the same tests. Statistical analyses were not specified, but it was reported that on 11 of the 89 "neurological" items and on 16 of the 36 "functional" items there were significant differences in results between the two groups. Two factors that appeared to be especially prominent in children with poor neurological and functional performances were the presence of hydrocephalus and presence of a shunt. Type of lesion and level of lesion had reportedly no relationship to practical upper limb difficulties.
Soare and Raimondi (1977; see 2.2. for details) administered the Beery VMI to 59 of 133 children with myelomeningocele and hydrocephalus, to 38 of their siblings, to 29 of 40 children with only myelomeningocele, and to 12 of their siblings. They compared the perceptual-motor age, as assessed by the Beery VMI, to chronological age by means of $t$-test for related means. VMI ages were significantly lower than chronological ages for the group with hydrocephalus and for the group without hydrocephalus. At the same time, a similar discrepancy was found for the siblings.

To investigate whether there really were differences between patients and siblings, and between the two patient groups a selection of subjects was made on the basis of those who could be matched both according to chronological age (within two years of each other) and IQ (within 15 points of each other). This procedure provided 14 pairs of patients and siblings and 26 pairs of patients with, and patients without hydrocephalus. Again, $t$-tests for related means were used for the statistical analysis and the variable on which the comparison was made was the difference score (chronological age minus VMI age). Patients in general had significantly bigger difference scores than siblings and patients with hydrocephalus had significantly bigger difference scores than patients without hydrocephalus.

Soare and Raimondi also studied the relationship between perceptual-motor deficit, as assessed by the discrepancy score, and level of the myelomeningocele sac. The Fisher Exact Probability Test showed that there was a significant association between higher lesions and greater perceptual-motor difficulties.
The authors also commented on the relationship between Beery VMI scores and IQ. A correlation of .51 between VMI test age and IQ was found. Soare and Raimondi cautioned, however, against an interpretation that perceptual-motor deficits are only found in children with low IQs. The reason for this was a suggested "ceiling effect" for the siblings who also performed below age expectation on the Beery VMI. In our opinion, this is a weak argument that is not substantiated by further evidence. Furthermore, we don't see how a ceiling effect can lead to artificial discrepancies if the test used encompasses test ages up to 15 years, and the age range in the subject sample (< 12.4 years for the group with hydrocephalus) is included in that range. We therefore think that there is an important relationship between IQ and VMI age, and the two may very well be expressions of the same deficit. We agree with Soare and Raimondi that the fact children without hydrocephalus also have significant perceptual-motor deficits on the Beery VMI, and the fact that greater problems were associated with higher spinal lesion levels argue against an interpretation of these problems as just signs of cerebral damage. This is consistent with Lonton's (1976) arguments against a purely cerebral interpretation of motor dysfunctions in hydrocephalic children. The fact that the children with hydrocephalus were relatively more impaired than those without may be related to the fact that higher myelomeningocele levels were more often associated with hydrocephalus.

Tew and Laurence (1978; see 2.4.2. for details) investigated visuomotor skills with the Bender Gestalt test and Stott's Test of
Motor Impairment. The latter test includes placing a stylus into nine holes following a defined sequence. Spina bifida cases with ocular defects were inferior on both tasks as compared to cases without such defects.

Brunt (1981) administered thirteen test items from the Southern California Sensory Integration Tests to a group of 28 children with myelomeningocele in the ages between 4 and 9.10 years. These tests involved visual perception, tactile perception, graphic duplication, and movement responses by imitation. A multiple regression analysis was used to investigate how a number of selected variables, associated with myelomeningocele, related to the subjects' obtained perceptual-motor score. The variables used were IQ (test used not specified), hospitalization, visual problems, hydrocephalus (measured by the number of shunt revisions), CNS infection, and ambulation. The total amount of variance explained by these combined variables was 51 percent. IQ contributed 32 percent of this variance and the other variables did not make statistically significant incremental contributions.

Brunt suggested that children with myelomeningocele and a sub-average IQ should be repeatedly assessed for perceptual and motor skills and eventual therapy. The strong correlation he found between IQ and perceptual-motor scores concurred with the findings of other authors (e.g., Sand et al., 1974; Soare & Raimondi, 1977). Although Brunt emphasized that hydrocephalus had little direct bearing on the perceptual motor scores of his population, this result may have been caused by the fact that (a) hydrocephalus was operationally defined in terms of the number of shunt revisions
and (b) was entered at a rather late step in the regression analysis (which may artificially have reduced the amount of variance it could still explain) despite its high correlation with the dependent variable.

Connell and McConnel (1981; see 2.3. for details) also administered the Beery VMI to part of their sample of hydrocephalic children. They did not provide quantitative data on VMI ages or difference scores and did not use statistical analyses. Of the 35 children who had been administered the test, 28 were reported to score at a level below their mental age equivalent. Although not subjected to adequate analysis, this is an interesting way of looking at the data, especially because of the above-mentioned relationship between IQ and VMI test age. A total of 20 children was considered to have evidence of constructional apraxia. The authors also reported that on the Benton Visual Retention Test, only 10 out of 22 patients had scores that were "appropriate for age range" and only 14 out of 30 patients performed adequately on the Wepman Test of Auditory Discrimination.

In the study by Dennis et al. (1981; see 2.3. for details) only IQ test results were discussed and no data on specific perceptual-motor tests were presented. Nevertheless, these authors discussed several important symptom variables that might be related to the relatively low performance IQ in hydrocephalic children. Dennis et al. suggested that both visual dysfunctions (deficits of gaze and movement and/or refraction and accomodation) and motor problems (varying from mild coordination problems to
severe paraplegia) might contribute to this. It was suggested that hydrocephalus may impair the motor skills necessary for nonverbal intelligence by affecting gross motor function (by deforming the cerebellum), fine motor control (by disturbing the kinesthetic–proprioceptive basis of hand control), and bimanual motor function (by causing stretching of the corpus callosum).

Zeiner and Prigatano (1981) have looked more specifically at the possible role of partial callosal dysfunction in hydrocephalic children. They administered visual, tactile, and auditory information processing tasks to 21 children with uncomplicated hydrocephalus in the age range of 4–10 years, and to two control groups consisting of children with excessive letter reversals in their reading, and normal elementary school children matched for age and sex. Vocabulary and Block Design subtests of the WPPSI or WISC-R were used as an estimate of intelligence (see 2.1.2. for objections to this). To investigate whether the performance of hydrocephalic children would specifically be influenced by possible partial callosal dysfunction, the authors analyzed both intra- and inter-hemispheric errors made on the three tasks.

The visual task involved delayed matching of bilaterally asymmetrical letters (b, d, q, p) presented to either the temporal right or temporal left visual hemifield. A 'crossed' or 'inter-hemispheric' error was defined as an error occurring when stimuli presented to opposite visual hemifields had to be matched, whereas 'uncrossed' or 'intra-hemispheric' errors were supposed to occur when stimuli presented in the same hemifields had to be matched. In the tactile task the subjects had to match tactile
designs made with nailheads on wooden blocks. In the 'uncrossed' condition the match was done with the same hand that had felt the first design; in the 'crossed' condition the opposite hand had to make the match. The auditory task consisted of a task with monaural presentation of consonant-vowel syllables and a dichotic listening paradigm using the same stimuli. More errors in a given ear under the dichotic than under the monaural condition were interpreted as evidence of inter-hemispheric problems.

ANOVAs and paired t-tests were used for statistical analyses. There were no significant differences between the numbers of crossed versus uncrossed errors for the hydrocephalic group in the visual or in the tactile condition. Moreover, hydrocephalics made more 'uncrossed' errors than controls on the visual task, while the number of 'crossed' errors on that task was not significantly different. This is exactly opposite to a callosal deficit prediction. Only in the auditory condition was this prediction confirmed, with the hydrocephalics demonstrating more dichotic errors. The authors concluded that in cases of uncomplicated hydrocephalus there are no consistent indications of callosal dysfunction. They suggested that there are information processing deficits in all sensory modalities in these children, but that they are equally intra- as well as inter-hemispheric in nature. This is, in our opinion, an important finding because it contradicts what some other authors (e.g., Dennis et al., 1981; Miller and Sethi, 1971b) have suggested as a possible cause for the other deficits in hydrocephalic children.
Jansen et al. (1982; see 2.4.4. for details) administered the Finger Tapping Test and the Grooved Pegboard Test to their adult sample. None of their patients obtained scores that were within the normal range on both tests. The authors did not provide statistical analyses but concluded that poor dexterity and clumsiness were common problems in these long-term survivors of infantile hydrocephalus.

In summary, a variety of studies provides evidence for perceptual and motor deficits in children with hydrocephalus. The motor deficits are probably not solely due to cerebral factors, but may very well be influenced by high levels of myelomeningocele. Specific evidence for callosal dysfunction in hydrocephalic children is equivocal.

3.5. Memory.

Some of the studies discussed in the previous section (e.g., Connell & McConnel, 1981) and the section on neuropsychological tests (3.6.) have included memory tests. In this section, we will discuss studies that have focused primarily on memory functions in hydrocephalic children.

Parsons (1969) was one of the first authors who specifically investigated memory functions in children with hydrocephalus. He administered a list of fifteen words in nine different sequences to 40 children with hydrocephalus with or without spina bifida and 31 non-hydrocephalic control children. The first sequence was a simple sentence and in the following sequences the normal word
order was progressively broken down. The median number of words recalled was calculated for each group and a Chi-square analysis was performed on the numbers of subjects scoring above and below the median in each group. The author found no significant differences between groups. His findings may, however, have been influenced by the fact that he used a rather experimental, unvalidated technique. The test may have also been too difficult even for average subjects, and may thus not have discriminated between mildly impaired and average subjects. Furthermore, his statistical analysis was inadequate, since the medians might very well have been significantly different for both groups.

Richardson (1978) assessed two adults with spontaneously arrested congenital hydrocephalus, aged 18 and 22 years and compared them with 10 age-matched normal controls. He administered the Verbal part of the WAIS and two Performance subtests (Digit Symbol and Block Design). No reasons were given why the whole WAIS was not administered. A memory test was also administered. It involved ten lists of common nouns presented for immediate free recall. The mean number of correctly recalled words per list was calculated. The statistical analysis involved ANOVA, which is questionable in view of the small number of subjects in the hydrocephalic group. No significant difference in VIQ was found. The values, reported by the author were, however, quite high (119 for both groups) which raises some concerns about the representativeness of this sample. Nevertheless, the two hydrocephalic patients were significantly impaired, as compared to
the controls, on the free recall task. This relative impairment in the context of superior IQ is an important finding.

Tromp and Van den Burg (1982) administered a five-trial, 15-word free recall task to a sample of 80 children between the ages of 6 and 12 years who had been shunted for hydrocephalus in their first year of life, and to 214 controls matched for school grade and sex. The total number of correctly recalled words after five successive trials was defined as the immediate recall score (IR). After 20 minutes an additional, unwarned recall trial was given and the score on that trial was defined as the delayed recall trial (DR). Estimates of verbal intelligence were made by using four subtests from the WISC-R.

The first statistical analysis involved a hierarchical multiple regression analysis with age and hydrocephalus as independent variables. Separate analyses were done with IR and IQ as dependent variables. Age did not emerge as a statistically significant main effect in either analysis whereas hydrocephalus did in both. Only for IR was there a small but statistically significant interaction. The second statistical analysis involved analysis of covariance (ANCOVA) to see whether the impairment in IR in cases of hydrocephalus could be accounted for by IQ. This ANCOVA included the same independent variables, and IQ was used as a covariate. In addition, a correction for a possible unreliability of IQ was used, with the reliability estimated at .80. When IQ was thus covaried out, hydrocephalus and the interaction with age still remained statistically significant.
Therefore, the impairment in IR could not be explained completely by the impairment in IQ.

Hierarchical multiple regression analysis with age and hydrocephalus as independent variables and DR as the dependent variable again revealed statistically significant main effect of hydrocephalus and interaction with age. ANCOVA with both IQ and IR as covariates was carried out with .80 as an estimate of unreliability for both IQ and IR. The result was still a statistically significant effect for hydrocephalus, but no interaction with age. Tromp and Van den Burg concluded that the impairment in delayed recall could not be explained by the joint impairment in immediate recall and IQ. This conclusion was further supported by the finding that there was a statistically significant difference between partial correlations of IQ, IR, and DR with the variable hydrocephalus. The authors also ran the same analyses with only those patients that attended normal schools (considered to be more directly comparable to the controls) and the results were reportedly similar, except that there were no interactions with age.

This study by Tromp and Van den Burg is an example of adequate application of multivariate statistical techniques that illustrates the importance of considering inter-relations between variables. The study shows that a memory deficit can be demonstrated in hydrocephalic children (especially in the area of delayed recall) that can not be explained by their lower intelligence.
Cull and Wyke (1984) investigated memory functioning in three groups of 10 subjects in the age range of 7.6 to 9.0 years. IQs for all groups were obtained by means of four subtests of the WISC-R (Similarities, Vocabulary, Picture Completion and Block Design). Group 1 consisted of children with spina bifida and hydrocephalus who had been shunted soon after birth and whose IQ ranged from 59 to 111. Group 2 consisted of children with no physical handicaps but with IQ ranging from 59 to 108. Group 3 consisted of children with no physical disability and whose IQ ranged from 92 to 108. The authors administered two verbal and two pictorial experimental memory tasks to these groups. The tasks were well described. For each task, four measures were obtained: learning ability (acquisition over three successive trials), immediate recall (trial 4), delayed recall (after 24 hours) and reacquisition (after delayed recall). IQs were not significantly different (as analyzed by means of t-test) between groups 1 and 2. The memory data were analyzed by means of Mann-Whitney U-test. Group 1 was found to be significantly poorer than group 2 on only one of the verbal tests. This test involved free recall of 10 unrelated nouns. Again, this is a significant finding in the context of comparable IQs. Furthermore, the authors interpreted the coincidence of this finding with the absence of significant differences between groups 1 and 2 on the other verbal memory task (which involved immediate recall of a story) as a possible indication that hydrocephalic children may have a rather selective deficit in their ability to use appropriate semantic strategies at the level of encoding. Although this interesting hypothesis needs
further independent validation, this study indicates the usefulness of obtaining different estimates of verbal and pictorial memory.

In summary, although the number of studies that specifically have investigated memory functions in hydrocephalic children is rather small, there are indications that memory impairments in these children can exist even in the context of average IQs. Furthermore, there is evidence that memory impairment cannot completely be explained by intellectual impairment in hydrocephalic children.

3.6. Performance on specific neuropsychological tests.

Although some of the tests in some of the studies discussed in previous sections may be classified as neuropsychological tests, we considered it to be appropriate to discuss the following studies in a separate section because they used a more comprehensive battery of internationally used neuropsychological tests.

Hammock, Milhorat, and Baron (1976) performed pre- and post-operative neuropsychological assessments in eight patients with myelomeningocele and normal-pressure hydrocephalus, ranging in age from 2.6 to 13.5 years. Children under the age of five years received the Stanford-Binet, Peabody Picture Vocabulary Test (PPVT) and Beery VMI. Children over the age of five years were given the WISC, Bender Gestalt, Wheeler-Reitan Aphasia Screening Test, Reitan-Klove Sensory Perceptual Examination, Tactual
Performance Test (TPT), Trail Making Test (TMT), and Finger Tapping Test. Although the authors reported important improvements in performance post-operatively, specific quantitative data with regard to the neuropsychological tests were not provided. Only one patient that improved on TPT was described but this rather selective illustration is not sufficient to warrant these authors' conclusion that psychomotor performance can be expected to improve within three months following successful ventricular shunting operations.

Tew, Laurence, and Richards (1980) administered the Matching Familiar Figures Test (MFPT), an auditory Continuous Performance Test (CPT) and a Visual Scanning Test (VST) to 48 cases of spina bifida, 78 percent of whom had accompanying hydrocephalus, and to a sample of controls matched for sex, age, and IQ (test not specified). The subjects ranged in age from 6.8 to 15.8 years. Statistical analysis were not specified. On the MFPT, the spina bifida subjects had more incorrect choices than their matched controls. Likewise, the spina bifida cases had lower vigilance scores on the CPT than the controls. On the VST, however, the accuracy scores for spina bifida and control cases were similar, although the spina bifida subjects took more time to complete the task. The authors interpreted this latter finding as evidence that spina bifida children are not necessarily less vigilant than normal children if they can work at their own pace.

Hurley et al. (1983; see 2.3. for details) administered a variety of neuropsychological tests to their subjects, including the Halstead Category Test (HCT), TMT, TPT, Finger Tapping Test,
and Finger Agnosia and Dysgraphesthesia. Compared to the controls, the hydrocephalic patients were significantly slower with their left hand on Finger Tapping, but significantly better on total time on the TPT (analysis by ANOVA on each measure). No statistically significant differences between groups were found on the other measures. The authors interpreted the TPT findings as evidence that the impairment of hydrocephalic children on perceptual motor tasks is due more to visual perceptual and visual organization problems than motor difficulties per se. Although we agree that the better TPT scores of the hydrocephalic patients are an unusual finding, we cannot subscribe to their interpretation of this. As was indicated in section 2.3., their control group was very diverse. Furthermore, these authors did not assess "visual perceptual and visual organization" skills directly with specific tests in this study.

Prigatano et al. (1983; see 3.3. for details) used the following neuropsychological tests: Fuld Object-Memory Test (FOMT), Benton Visual Retention Test (BVRT), Lafayette Grooved Pegboard (GP), and TMT. The authors used t-tests to compare hydrocephalic and control children on each of the measures. The hydrocephalic children were significantly poorer than the controls on consistency-of-recall measure of the FOMT, the immediate recall of the BVRT, left and right hand of the GP, and part B of the TMT. The authors concluded that despite successful neurosurgical management of hydrocephalus and normal-range estimates of intelligence, hydrocephalic children still present with significant neuropsychological impairments. Although the IQ
estimates of these authors can be criticized (see 3.3.), their finding of a diversity of neuropsychological deficits in children with uncomplicated hydrocephalus is significant, since complications may lead to even more impairments.

Laatsch, Dorman, and Hurley (1984) assessed 28 patients with myelomeningocele and two with meningocele. 21 of these had had a shunt at birth and 6 were shunted later. The ages ranged from seven to 16 and up. The subjects were assessed at least three times per year over a two-year period with a variety of neuropsychological tests. These included Finger Tapping, Grip Strength, BVRT, Finger Agnosia, Frostig Test, and the Coding and Mazes subtests of the WISC-R. Statistical analyses involved a "modified t-test" based on the average score over several assessments between the first and the second year. The authors reported that Finger Tapping, Grip Strength, and Coding were the most reproducible. This meant that the scores on these tests remained stable between the two years with supposedly no neurological changes. The authors suggested that such "stable" tests should be used to evaluate functioning in children who have stabilized neurological functioning. We do not totally agree with this, because what they have actually done is a test-retest reliability study for a very specific and small sample.

Furthermore, some cognitive (i.e., not involving motor manipulation) sequelae of hydrocephalus may keep changing after there are no indications of further neurological change and, moreover, some deficits in higher and more complex cognitive functions may only appear later in childhood.
In summary, tests that have been specifically developed to reflect impaired brain functioning often appear to reflect deficits in hydrocephalic children even though their IQs may be in the average range. This suggests that an evaluation of the development of hydrocephalic children should not only include an assessment of intelligence but also of specific neuropsychological functions. This dissertation attempts to do that.

4. Social and emotional aspects.

4.1. Psychiatric sequelae in the child.

Studies that have dealt with the social and emotional aspects of hydrocephalus and/or myelomeningocele can be divided into two main groups: studies that focus primarily on the various forms of psychiatric problems in the surviving children and studies that concentrate more on the impact on the family of having a hydrocephalic child. This is, of course, a global distinction based on relative (not absolute) differences and some studies pay attention to both aspects of the problem. Because physical disabilities due to spinal lesions have been extensively studied as a possible factor with regard to independence and adjustment, we will review some studies that provide little information about hydrocephalus and relatively more about myelomeningocele. In this section we will discuss the studies that focus primarily on the patients themselves.

Kolin, Scherzer, New, and Garfield (1971) conducted "semi-structured" interviews with 13 children with
myelomeningocele and with their parents. The ages of the patients ranged from 7 to 11 years, but there were large differences in the degree of physical impairment (ranging from "mild" to "severe") and the degree of mental impairment (WISC FSIQs ranging from 69 to 117). No information was provided about hydrocephalus or other aspects of the medical history. The interviews appeared to be rather analytically oriented. Based on subjective ratings of adaptation of the children and of the parents, a good to fair adaptation was found in seven of the children and 5 of the parents (criteria poorly defined). The authors found that the severity of physical impairment was a less important determinant of adaptation than psychological factors in both the child (such as anxiety and depression) and the parents (such as overprotection, rejection, etc.). Kollin et al. also reported that they considered quality of communication between the physician and the family a crucial factor in the adjustment process.

Hunt (1973) provided a rather anecdotal description of the implications of myelomeningocele for both the myelomeningocele child and the family. He described most families as making great effort and sacrifice for their child. However they reportedly often obtained disappointing results: the combination and severity of handicaps typically made independence of the children hard to achieve.

Seidel, Chadwick, and Rutter (1975) investigated whether the psychiatric disorders that may be found in children with cerebral lesions are the result of organic brain dysfunction per se, or to the presence of associated physical handicaps. Selection criteria
for the subjects were (a) ages between 5 and 15 years, (b) having a chronic, visibly crippling disorder of any type, and (c) an IQ (variety of tests used) of 70 or more. On the basis of these criteria 83, subjects were selected of whom 33 had a disorder due to cerebral involvement (cerebral palsy or hydrocephalus) and 43 had a disorder due to some other cause (e.g., congenital dislocation of the hip, muscular dystrophy, etc.). Interviews with the parents and with the child and a questionnaire administered to the teachers were used to assess psychiatric disorder. The groups with cerebral and with peripheral disorders were matched with regard to age, sex, social class, overcrowding in the home, and broken home. Statistical techniques included Chi-square and t-tests. When an overall rating of psychiatric disorder based on all instruments used was made, the 'cerebral' group showed a prevalence of psychiatric disorder of 24 percent while the 'peripheral' group had a rate of 12 percent. This difference was not statistically significant, but the latter was rapidly disregarded by the authors because they found that the rates were comparable to other studies. They concluded that brain damage was responsible to the children's increased vulnerability to emotional problems. Although this study suggested such a trend, the absence of a statistically significant effect makes this conclusion somewhat premature. In addition, it must be noted that three quarters of the children with cerebral injuries did not have psychiatric problems.

Of interest in the Seidel et al. study was that psychiatric disorder tended to occur less often in those children with severe
physical handicaps. According to the authors this finding could not be explained in terms of age, sex, type of lesion (cerebral or peripheral), IQ, reading attainment, or social class but it was not clear how they analyzed this. Reportedly, the only relevant item associated with this finding was that the mothers of severely disabled children tended to have lower scores on the Rutter Malaise Inventory (Rutter, Tizard, & Whitmore, 1970). In general, psychiatric disorder was more common in children from broken homes, overcrowded homes, families with marital discord, or with a mother with psychiatric problems.

McAndrew (1979) conducted "semi-structured" interviews with 37 survivors of spina bifida, aged 14 to 25 years old. Statistical analyses were not included in this mostly descriptive study. Mobility and continence problems were well documented. Walking and urinary incontinence were the dominant problems most commonly acknowledged. Those with ileal conduits frequently reported both social embarrassment and management difficulties. For employment, intellectual ability appeared to be more important than physical disability. The author suggested that the ability of a patient with spina bifida to cope with his impairments is not so much determined by the severity of the disability, but by available environmental support systems.

In his study of children with the Cocktail Party Syndrome (CPS), that was described in section 3.2., Tew (1979) also commented on the social adjustment of these children. At the age of five and a half years, children with CPS showed statistically significant retardation in social skills, as assessed by the
Vineland Social Maturity Scale, as compared to other children with spina bifida (analysis by means of t-test). The author suggested that reasons for this poor social maturity of the CPS children might be the combined effects of low intelligence and severe physical handicap on the one hand and over-solicitous maternal care on the other. The latter was, however, not formally assessed or quantified. At the age of seven years the teachers of these children rated those with spina bifida with or without CPS as showing a significant excess of behavior problems as compared to normal controls. The children with CPS did not have more problems than those with spina bifida and no CPS.

Connell and McConnel (1981; see 2.3. for details) used the questionnaires and individual, semi-structured interviews with both children and parents that were initially described by Rutter and colleagues (Graham & Rutter, 1968; Rutter & Graham, 1968). These procedures were used to assess the prevalence and extent of psychiatric disorder and also allowed affected children to be placed in one of three categories: those in whom symptoms of anxiety predominated (neurotic disorder), those characterised by anti-social behavior (conduct disorder), and a group with features common to both (mixed disorder). Subjective assessment of parental attitudes was supplemented by the Rutter Malaise Inventory for parents. The questionnaires and interviews indicated a 44 percent rate of psychiatric disorder in the hydrocephalic children, which is about four times as much as in the normative peer population. Conduct disorders were significantly more common than neurotic disorders (analysis by means of Chi-square). Boys had more, and
more severe disorders than girls, which corresponds to a similar pattern in the general population. There were no significant differences between the disturbed and the non-disturbed groups in intellectual level or physical handicap, but there were important differences in home background, parental attitudes and management. The latter aspects were described in terms of over-protection, rejecting attitudes, parental psychiatric disorder, and parental interpersonal difficulties in the disturbed group.

Hunt (1981) provided a qualitative analysis of the implications of having spina bifida for a series of 100 children. Information about variables associated with hydrocephalus was very limited, but 86 children were reported to have shunts which appeared to be clinically malfunctioning (not quantified) in half of the children. Hunt mentioned wheelchair dependency and incontinence as important factors that precluded the attendance of ordinary secondary schools for many of the children. Many children reportedly had learning difficulties, but these were poorly described. Complications that appeared to be of importance at school included visual defects (e.g., strabismus), epilepsy, and liability to injury, the latter being related to distribution of sensory loss.

Lenton (1981) investigated what proportion of spina bifida children were integrated into ordinary schools and what the main obstacles to such integration were. For this purpose, the school placement of 1235 patients with spina bifida (with or without hydrocephalus) aged between 2 and 29 years was studied. The WISC was used as a measure of intelligence and "suitable alternative
tests" were used for children outside the age range of the WISC. ANOVAs and correlation coefficients were used as statistical analyses. It appeared that IQ was the best predictor of school placement out of all the various physical, intellectual, and social parameters examined (i.e., it had the largest correlation with school placement). Of the physical factors, wheelchair dependence (alone, or in combination with incontinence and/or valve dependency) was the most important obstacle to integration into ordinary schools.

Johnson (1984) assessed the psycho-social development of "latency-aged" children with spina bifida. Interviews and the Children's Apperception Test were used. The characteristics of the methodology were very poorly described (e.g., no mention was made how many cases were seen, no information on the level of physical and mental disability was provided, etc.). The author reported that by the sixth month in first grade, all of the children had "adapted satisfactorily to school" but did not specify what this meant. Although the author concluded that spina bifida did not put a ceiling on the child's psycho-social development, there is no way in which the reader can verify this on the basis of this poorly described study.

Tew and Laurence (1985) administered the Rogers Personal Adjustment Inventory (RPAI) to 44 ten-year-old spina bifida children, 19 of whom had shunt-controlled hydrocephalus, and to 52 controls matched for sex, social class, and area of residence. Teachers assessed the school behaviour of these children on the Bristol Social Adjustment Guide (BSAG). Statistical analyses
included t-tests and Pearson product-moment correlation. The authors reported that spina bifida children and controls had similar scores on the BSAG (indicating similar school adjustment) but appeared to have significantly more emotional problems on the RPAI. Although the authors further analyzed the RPAI scores by scale and in relation to such factors as sex, severity of handicap, etc., these results appear irrelevant to us for two reasons. First, the spina bifida and control children differed significantly in FSIQ on the Wechsler scales (means 79.8 versus 107.5), and second, the authors themselves reported that "...the Rogers Inventory lacks basic psychometric properties, notably reliability, validity, and comprehensive norms...".

In summary, psychiatric sequelae of hydrocephalus and/or spina bifida appear not to be uncommon, although the majority of these patients do not present with significant emotional or behavioral disturbances. There is no evidence to suggest that the incidence of disorder is either greater or less than in comparably disruptive other neurodevelopmental conditions. The psychiatric symptoms that appear are most common include adjustment to disability and conduct disorders. The problem with many of the studies in this area is that the results are typically obtained from "semi-structured" interviews and questionnaires of dubious reliability and validity. To our knowledge, no study has yet attempted to assess psychiatric sequelae of hydrocephalus by means of well-standardized comprehensive personality inventories. The latter is one of the objectives of this dissertation.
4.2. **Impact on the family.**

Walker, Thomas, and Russell (1971) described the results of a study that used questionnaires and interviews to document the initial responses of 107 families to the birth of an infant with spina bifida. This study was mostly descriptive. Many mothers indicated that they immediately appreciated that something was wrong with their child, although the malformation tended to be minimized by attending professionals. The majority of the mothers were dissatisfied with the way they were initially informed about the nature and extent of the child's abnormality, with complaints including professional jargon and lack of information about functional implications. The majority of the mothers were also dissatisfied with the help received from general practitioners. The authors stressed the need for genetic counseling and family planning, and for continued social case work.

Hare, Payne, Laurence, and Rawnsley (1972) administered an abbreviated form of the Maudsley Personality Inventory (MPI) to 146 mothers of spina bifida children and 78 control mothers of normal children. The control mothers formed a series matched for age, parity, social class, and place of residence. The MPI supposedly measures two aspects of personality: extraversion (E) and Neuroticism (N) (Eysenck, 1958; cited in Hare et al., 1972). These authors used an abbreviated form of this 24-item questionnaire, selecting six questions concerning E and six N. The questionnaire was administered shortly after birth, after one month, after one year, and after two years. Data were analyzed by means of ANOVA with repeated measurements. One factor in this
analysis was severity of handicap, rated in four categories from absent to severe (criteria not specified) and the other factor occasion of interview. The controls were included as a fifth group in the severity-of-handicap factor. Separate analyses were done for E and N scores. There was no significant effect for either severity of handicap or occasion for either E or N scores. This means that in general, there were no differences between patient and control mothers in E or N scores. For the analysis of N scores a significant interaction effect was found, indicating that these scores tended to increase in mothers of children suffering from moderate or severe handicap.

Hare et al. also looked at the relationship between N scores and the number of "nervous symptoms" experienced by the mother and between N scores and the severity of the child's handicap. "Nervous symptoms" were assessed by asking whether the mother had suffered from any of the following six symptoms: depression, anxiety, fatigue, poor sleep, headaches, or poor appetite. This is a strange procedure, not only because of its simplicity but also because questions regarding these specific symptoms are also present in the MPI (e.g., "Do you often feel miserable or depressed?", "Do you often have bad headaches?", etc.). The finding that those with few (< 2) symptoms had lower N scores than those with many (> 4) is therefore rather tautological. Furthermore, this finding was not specific for the patient group. Chi-square analyses revealed that there was a significant association between number of symptoms and severity of handicap at one year, but this relationship did not hold up at two years. The
authors did not discuss how this would relate to their reported significant interaction effect in the ANOVA. Although we appreciate that the severity of a handicap a child may influence the state of mental well-being of the parents, these authors appeared to overlook that they did not find significant differences on their measures between spina bifida cases and controls.

Richards and McIntosh (1973) interviewed the parents of 86 spina bifida patients, aged between two and six years. 46 of these children had shunted hydrocephalus. Other complications (e.g., urinary tract infections, strabismus, etc.) and severity of physical handicap were well documented. Many of the results were comparable to that of the Walker et al. (1971) study: the majority of the parents indicated they had received insufficient and inadequate information about the nature and consequences of spina bifida. With regard to professional support, the major complaints in this study appeared to be lack of genetic counseling and family planning advice, as was also found in the Walker et al. study.

Richards and McIntosh also reported in more detail on the attitudes and problems of the parents. Feelings of guilt and episodic wishes that the child would not have survived proved to be common, although with only two exceptions, the parents came gradually to accept the situation and to want the child to survive. At the time of assessment most of the parents expressed worries about the child's future, but also expectations that the handicap could be "cured" or at least greatly reduced. Common problems experienced were difficulties with lifting the children,
incontinence, financial problems, and inadequate housing facilities.

Tew and Laurence (1973) assessed 59 children with spina bifida, their 44 siblings, and 59 age- and SES-matched control children and their 63 siblings. The children's ages ranged from two to 15 years. The siblings' behavioral adjustment in school was assessed with the Bristol Social Adjustment Guide (BSAG; Stott, 1963; cited in Tew & Laurence, 1973) which is based on teachers' ratings of appropriate behaviors. A score of 9 or less on this scale is interpreted as "normality", scores between 10 and 19 are supposed to reflect "unsettled behavior", and scores of 20 or more are supposed to indicate "maladjustment". Chi-square analyses revealed that the siblings of spina bifida children were significantly more likely to show evidence of maladjustment than the siblings of control children. There were no significant differences in BSAG scores of siblings according to whether the spina bifida child was at a residential school or not and these scores did not vary with birth order. The authors reported an unexpected effect of severity of physical handicap: the siblings of children with very slight handicaps were most likely to be maladjusted, followed by the severely handicapped group (not subjected to statistical analysis).

Tew and Laurence assessed mental health of the mothers with the Rutter Malaise Inventory. The authors compared the scores of the mothers of spina bifida children to those of parents of children with psychiatric problems, brain disorder, and physical handicaps that were described by Rutter et al. (1970). They
reported that mothers of spina bifida children had higher stress levels. They should have also administered this inventory to the mothers of the control children. The authors also reported a significant association between the mothers' Malaise score and the siblings ESAG score but did not indicate how this was analyzed, and provided no quantitative information. We were also struck with the very large standard deviations in Malaise scores.

Tew, Payne, and Laurence (1974) examined the quality of marital relationships of parents with a child with a major neural tube malformation. Control families in which the child did not have such a malformation, matched for domicile, social class, sex of the child, and family size were also assessed. Marital harmony was assessed by a social worker on the basis of home interviews and rated on a five-point scale, ranging from satisfactory to broken marriage. Chi-square analyses revealed that significantly more parents in the patient group had marital difficulties than in the control group. The divorce rate of the former families was twice that of the controls and that of the national average. The authors also reported that severity of the child's handicap was not found to influence the quality of the marital relationship, but did not provide statistical analyses.

Dorner (1975) investigated the relationship of severity of physical handicap to family life by means of interviewing the parents of 63 adolescents (age-range 13 to 19 years) with spina bifida living at home. Children with only meningocele were excluded from this study. Little information was presented with regard to parameters regarding hydrocephalus. In addition to the
Interview, the mothers were asked to fill out the Rutter Malaise Inventory. The adolescent patients themselves were also interviewed, but these findings were not reported in this study.

On the basis of the interviews, the following findings were reported. 50 percent of the patients were judged to be severely socially isolated. Interestingly, Chi-square analyses revealed that children going to ordinary schools were significantly less likely to be isolated than those attending special schools, but they were also much less handicapped as a group than those at a special school. There also appeared to be a relationship of social isolation to sex in mobile patients with urinary incontinence: Fisher Exact Probability Tests revealed that boys were much more likely to be socially isolated than girls in that specific group. 66 percent of the patients were considered to have had periods of depression, which was more likely to occur in girls and also appeared to be related to problems of mobility and social isolation. 42.5 percent of the parents reported experiencing restrictions in their social life and a statistically significant relationship with the patients' physical mobility was found: parents of children with severe mobility problems reported more social isolation than those whose children had only mild or no mobility problems (analysis by means of Chi-square).

Depression, as assessed by the Rutter Malaise Inventory, was common but Dorner reported that maternal depression was not related to the patients' degree of mobility, nor to other aspects of the child's physical condition. No statistical analyses were reported to support this report. On the basis of interviews, it
appeared that parents who admitted to depression rarely attributed it directly to problems associated with their handicapped child and the author concluded that the most likely explanation would be that the presence of a handicapped child within the family increases the vulnerability of mothers to other stressful life-events. Dorner also reported that marital disharmony or discord did not occur more frequently than in the general population (no quantitative data provided), which is consistent with the findings of Richards and McIntosh (1973) but at odds with the findings of Tew et al. (1974) that were discussed earlier.

Tew and Laurence (1975) administered the Rutter Malaise Inventory to 51 mothers of spina bifida children, including meningoceles, myelomeningoceles, and encephaloceles. The ages of the patients ranged from 9.3 to 15.8 years. Statistical analyses were done by means of t-tests. Severity of physical handicap was rated as mild, moderate, or severe, on the basis of levels of mobility and bladder control problems. Significantly higher Malaise scores were associated with severe locomotor disability, incontinence, an IQ less than 80 (test was not specified), and attendance of a special rather than a normal school. In this context it appeared to be important that pupils in special schools tended to be more severely handicapped. Although girls tended to be more severely handicapped than boys, this did not influence the Malaise scores of the mothers significantly.

Joosten (1979) investigated the impact of having a spina bifida child on the family by means of semi-structured interviews and questionnaires. These were administered to 22 families of
spina bifida children between the ages of four and six years, and 22 control families having a normal child of the same age group and matched for age of the mother, social class, and housing area. No statistical techniques were mentioned in this study so the evidence is rather anecdotal. It was found that mothers of spina bifida children were engaged in child care twice as long per day as mothers of normal healthy child. They also depended more on external resources such as friends, parents, and neighbors. Mothers with spina bifida children appeared not to differ from mothers with normal healthy children in their sense of well-being, but did experience more stress and felt more tired.

Nielsen (1980; see 2.4.1. for details) conducted serial interviews with the parents of 30 myelomeningocele patients at the ages of 6 months, 18 months, 3 years, and 6 years. The interviews were reportedly analyzed "qualitatively" to elucidate responses to certain age-specific problems met by all families with a myelomeningocele child, but possibly reacted to quite differently. The observations were divided into three main categories: birth, infancy, and preschool period. On the basis of case examples, Nielsen described a "developmental trend" in the way the parents dealt psychologically with the stresses of having a myelomeningocele child. The first half year was described as a period of "shock" with confusion and apathy being common. The period covering the second half of the baby's first year and the second year was described as the one during which many families had the hardest time getting through because of physical constraints on both the child and the parents and intense
emotional fluctuations. After that, an improvement in well-being was described and the child typically developed "along psychosocially quite normal lines".

Carr, Pearson, and Halliwell (1983) conducted interviews with the parents of 125 spina bifida children (no information about hydrocephalus variables) of 11 years of age and living with the parents. Classifications of locomotor disability and incontinence were well described. Statistical analyses were unclear. Although 45 percent of the mothers indicated that they felt that their other children had suffered from the presence of the disabled child (mainly in terms of limitations on outings, attention, etc.) the siblings were not reported to have any more behavioral problems than control children (assessed in another study). Although it would have been preferable if the authors had included their own control group in this study, this finding nevertheless contrasts with some other reports that found more problems with siblings of spina bifida children (e.g., Tew & Laurence, 1973).

In general, parents of severely disabled children were more restricted in their social activities than those of slightly disabled children. 22 Percent of the mothers of spina bifida children indicated they felt run-down and depressed and this figure was only 3 percent in the control group (again from another study). Interestingly, however, the mother's scores on the Malaise Inventory did not show a statistically significant relationship with any of the measures of the child's disability level, dependence or amount of hospitalisation, or measures of social
restriction. This is consistent with the findings of Dorner (1975) but at odds with the findings of Tew and Laurence (1975).

In summary, a substantial number of parents of hydrocephalic or spina bifida children admit to some degree of personal distress, usually in the form of social isolation and general stress. Again, most studies obtain their information from unstandardized techniques. Furthermore, the results of the studies are contradictory with regard to whether or not specific physical and social variables (especially the level of disability of the child) have any impact on the level of distress of the parents. An investigation of the relationship of those variables to parental distress in families with hydrocephalic children is one of the objectives of this dissertation.

5. **Statement of the problem.**

Although the intellectual, neuropsychological, and socio-emotional sequelae of shunted infantile hydrocephalic children have been the subject of numerous studies, they remain to a large degree equivocal. There are significant discrepancies both within and between studies with regard to which variables have a significant impact on the development of hydrocephalic children. These discrepancies are at least partly caused by the various methodological inadequacies of the studies that were discussed. Since the critical review by Raimondi and Soare (1974), in which some of these inadequacies were discussed, the general methodology in the literature has improved somewhat, but several aspects
remain problematic in many studies. These include particularly the selection of independent and dependent measures and data analysis.

In the area of intellectual development, the literature is very inconclusive about which variables have statistically significant predictive or discriminative value with regard to later attainment. This situation appears to be due to (among other factors) the fact that different studies do not use the same measures of intelligence, do not administer the same measures to all of their subjects, and especially do not include the same independent predictor variables. The main problem in this area is that there is a scarcity of reports that have used multivariate statistical procedures. Adequate statistical analyses are lacking in many studies reported in the literature; analyses that allow for an investigation of the role of specific variables in concert with other variables are particularly rare. The few studies that have used correlational or regression analyses have typically not made full use of these techniques and/or have not documented the quantitative results in satisfactory ways. To our knowledge, no study has yet been published that has included detailed, acceptable reports on the quantitative results of multiple regression analyses or discriminant analyses. We used such analyses to investigate the role of a variety of medical history variables (including demographic, pathologic, symptom, and therapeutic variables) with regard to intellectual development in children with shunted infantile hydrocephalus.

With regard to neuropsychological sequelae, the literature also shows lack of agreement. This is partly a result of the same
methodological problems that are apparent in many of the studies on intellectual sequelae. The fact that different studies use widely varying techniques to assess specific functions makes it especially hard to compare the results of one study to those from another. Furthermore, many studies focus on rather isolated and specific functions, and very few studies have administered comprehensive assessment batteries to the subjects involved. Many of the measures that have been employed are experimental and unstandardized.

We were unable to find any study in the literature that has attempted to identify subtypes of neuropsychological development in children with shunted infantile hydrocephalus. We administered a comprehensive battery of neuropsychological tests (assessing such functions as expressive and receptive linguistic abilities, verbal and visual memory, problem-solving and concept-formation, visuospatial functions, and sensory and motor functions) to a substantial number of patients. We investigated the existence of possible subtypes by means of multivariate statistical procedures, i.e., cluster analyses on the basis of performance on the tests administered.

Methodological problems and contradictions among studies are also characteristic of the area of socio-emotional sequelae of shunted infantile hydrocephalus. The use of experimental and unstandardized techniques is a particularly important problem. To our knowledge, no study has yet included standardized, well-normed, comprehensive assessments of psychiatric functioning in both patients with treated hydrocephalus and their parents. We
assessed socio-emotional functioning in such patients and their parents by means of standardized personality inventories. We investigated whether the different neuropsychological subtypes showed different patterns of emotional development and whether distress in the patients' parents could be related to specific medical history variables (such as degree of mobility, incontinence, socio-economic status, etc.).

In summary, the present investigation of psychological sequelae of infantile hydrocephalus had three aims (specific hypotheses are outlined below): (1) to determine which medical history variables are statistically relevant with regard to intellectual outcome, (2) to identify neuropsychological subtypes among these children, and (3) to characterize their emotional functioning and that of their parents.

The first purpose was to investigate how intellectual outcome after shunted infantile hydrocephalus was related to a wide variety of medical history variables associated with this cerebral abnormality. For this purpose, stepwise discriminant analyses were used to discriminate selected levels of intellectual attainment on the basis of medical history variables. The medical history variables were selected on the basis of a review of the literature and an attempt was made to include only those variables that had been investigated by at least two studies.

Separate analyses were performed for VIQ, PIQ, and FSIQ to determine sets of optimal discriminators for each estimate of intelligence, a procedure suggested by Dennis (1985a) who used
regression analyses. It was hypothesized that the discriminant analyses would identify whether and to what extent medical history variables are or are not discriminative regarding later intelligence. Due to the contradictions in the literature and the fact that adequate reports on such analyses had not yet appeared in other studies, no further specifications were made as to which variables would and which would not emerge as statistically significant.

Although predictions concerning the predictive or discriminative value of specific medical history variables were difficult to make, it was possible to formulate a hypothesis regarding the pattern of IQ scores in hydrocephalic children. The literature is rather consistent with regard to the finding that PIQ tends to be less well developed in hydrocephalic children than is VIQ. Therefore, a t-test for paired observations was planned to compare VIQ and PIQ in the total sample. It was hypothesized that PIQ would be significantly less well developed than VIQ.

Because of the contradictions in the literature concerning the role of sensory and motor factors with regard to performance on most intelligence tests, we correlated performance on selected sensory and motor measures to each of the obtained IQ scores.

The second purpose of this dissertation was to classify patients with shunted infantile hydrocephalus into homogenous groups on the basis of their performance on a variety of neuropsychological tests. The purpose here was to identify subtypes of children such that the similarity between subtypes
would be minimal and the similarity between different children in specific subtypes would be maximal.

Cluster analysis and Q-type factor analysis could be used for such purposes. No cogent case has been made for why psychologists should employ either of these techniques instead of the other (Adams, 1985). Based on evaluations of reports on the relative merits and problems of both techniques (Adams, 1985; Everitt, 1980; Fletcher & Satz, 1985; Morris, Blashfield, & Satz, 1981), we decided to employ cluster analysis. This choice was mainly based on the fact that factor analysis allows for subjects to load on more than one factor and to have negative factor loadings. The latter appeared to violate our intention to generate subtypes that are readily interpretable and that have minimal similarity between them. At the same time, we realized that this choice was still subjective. Some of the objections to factor analysis (e.g., often, only a relatively small proportion of the subjects will be classified) could also be raised against cluster analysis (which classifies all subjects and may force subjects into clusters of questionable validity).

For the purposes of subtype analysis, then, we employed cluster analysis of the hierarchical agglomerative type, utilizing an average-linkage method for combining subjects into groups or clusters and a Euclidean distance measure as the similarity measure. Determination of the number of clusters was done on the basis of inspection of the hierarchical tree (dendogram) and the semipartial R² (an indication of the loss of accounted-for variance, resulting from merging two clusters).
The hierarchical agglomerative procedures are the most popular of the clustering methods (Morris et al., 1981). The review by Milligan (1981) indicates that the average-linkage method is one of the clustering techniques with the best overall performance in terms of its replicability. The procedure to determine the number of clusters is subjective because no adequate objective technique has been proposed for solving this problem (Blashfield, 1980).

The choice of a similarity measure is an important issue in cluster analysis. Euclidean distance and product-moment correlation are two ways to measure similarity between subjects. A product-moment correlation is relatively more sensitive to profile shape (and its scatter) than is Euclidean distance and it has been argued by some authors (e.g., Del Dotto & Rourke, 1985; Joschko & Rourke, 1985) that product-moment correlation should therefore be used when one is interested in the pattern of scores over different variables. This relates to the fact that the correlation will be high when there is a high degree of similarity in pattern, no matter how far apart the profiles are in absolute level. This can be considered as an argument in favor of correlation as a measure of similarity.

At the same time, two children may have the exact same pattern of performances (a correlation of 1.00), but if one performs at a superior level, while the other is at a deficit level, they may be very different from a clinical point of view (Morris et al., 1981). This can be considered as an argument against correlation as a measure of similarity. Everitt (1980) has
summarized other criticisms of the use of correlation coefficients in cluster analysis.

Everitt indicated that two profiles may even have a correlation of 1.00 if they are not parallel. All that is required for such a perfect correlation is that one set of scores be monotonically related to a second set. Another criticism of correlation coefficients, provided by Everitt, is that in its use as a similarity coefficient in cluster analysis it utilizes the computation of a 'mean' over disparate variables of one object (equal weighing of characters). In general, this would seem to have little meaning (Everitt, 1980). For these reasons, Euclidean distance was chosen as the measure of similarity for this dissertation.

Dependent measures were the scores on a variety of neuropsychological tests (converted to $T$ scores on the basis of norms for the general population). Variables were selected on the basis of three criteria: (a) psychometric properties of standardization, demonstrated reliability and validity, and availability of age norms; (b) theoretical relevance in the sense that the total battery would allow a comprehensive assessment of different neuropsychological functions and (c) heuristic purposes, i.e., limiting the number of variables so that subtypes would remain interpretable and differences between subtypes might be maximized, without violating criterion (b).

Although no study in this area had, to our knowledge, yet been published that included identification of subtypes through multivariate statistical procedures, our review of the literature
suggested the possibility of at least two subtypes: children with deficits in the visuo-perceptual and visuo-constructive (visuospatial) domain and children with linguistic deficits. These two subtypes are of course not mutually exclusive: a "visuospatial" subtype may still have some linguistic problems and vice versa. One can also argue that performance on many linguistic tasks has often more to do with overlearned strategies, whereas performance on many visual-spatial tasks requires novel strategies (e.g., Rourke, 1982). The expected subtypes might therefore also be classified as having difficulties with either overlearned skills, or novel problem-solving. In the following text, the subtypes will be referred to as "linguistic" and "visual-spatial" subtypes, with the understanding that an alternative classification in terms of selective deficiencies in overlearned or novel strategies might also be applicable. Regardless of the interpretation of the classification, it was hypothesized that subtypes, defined by relatively poor performance within these specific domains would be found, and possibly a mixed form with deficits in both domains.

The reliability of the identified clusters was assessed in several ways. According to Everitt (1980), a valid solution should reappear with different clustering methods (which would basically indicate concurrent validity). Both another hierarchical agglomerative procedures and a different method of clustering were used. Specifically, hierarchical agglomerative methods with respectively centroid linkage and Ward's minimum variance, as well as a non-hierarchical K-means iterative partitioning method were
used. The centroid linkage and Ward methods were chosen on the basis of the fact that several studies (e.g., Del Dotto & Rourke, 1985, Edelbrock, 1979) had found high correlations between the clusters produced by average linkage and centroid linkage methods. In addition, several studies (e.g., Edelbrock, 1979, Milligan, 1981) recommended Ward's method as a technique that is approximately as reliable as average linkage. K-means iterative partitioning was chosen because, by means of this method, it is possible to check clusters and relocate any misassigned subjects to a more appropriate cluster (Dennis, 1985b; Morris et al., 1981). In this way, an idea can be obtained about the stability of the clusters. The sample was considered to be too small to assess the reliability of the clusters by means of a split-sample design or a Monte Carlo design.

Any statistical analysis that involves comparisons of subtypes on specific measures has to take into account that multiple comparisons among means may become a necessity. If such multiple comparisons among means are conducted in an analysis of variance (ANOVA), one must make sure that all the contrasts are planned. This is because the chance of making at least one Type I error (i.e., rejecting a true null hypothesis) becomes much higher than the defined alpha. In other words, it becomes more likely to find a statistically significant effect on the basis of chance. The Bonferroni additive inequality is a procedure that corrects for this error. This procedure is available in SAS computer programs (BON option in GLM and ANOVA procedures) and was used when necessary.
The extent to which the identified subtypes differed in intelligence was also analyzed. Because subjects were classified to maximize similarity in neuropsychological profiles in which IQ was not incorporated, it was possible to test the validity of the classification in this way. If subtypes differed significantly from each other in FSIQ, the neuropsychological deficits would be difficult to interpret and might be secondary to differences in general over-all intelligence only. On the other hand, it was considered to be plausible that a subtype with predominant linguistic dysfunctions would have a relatively low VIQ, whereas a subtype with more prominent visuospatial dysfunctions would have a relatively low PIQ. To investigate these possible differences, a MANOVA was planned. It was hypothesized that (a) the "linguistic" and the "visuospatial" subtypes would not differ significantly in FSIQ, (b) the "linguistic" subtype would have a significantly lower VIQ than the "visuospatial" subtype, and (c) the "visuospatial" subtype would have a significantly lower PIQ than the "linguistic" subtype. These planned analyses are, of course, not truly independent since the IQ scores were related indices provided by one task.

Another method of validation included comparison of the subtypes on "linguistic" and "visuospatial" measures (or measures of overlearned versus novel strategies) that were not included in the subtype analysis. For this purpose, groups of tests were selected in such a way that, for each measure included in the cluster analysis there would be another (validation) measure that could be supposed to appeal to the same area of psychological
functions. The clusters could then be compared on those validation measures. In this way, it was possible to compare subtypes with dysfunctions in specific domains on comparable measures. MANOVAs were planned to compare subtypes on the validation measures. It was hypothesized that (a) the "linguistic" subtype would perform significantly poorer on validation measures of language or overlearned strategies than the "visuospatial" subtype, and (b) the "visuospatial" subtype would perform significantly poorer on the validation measures of visuospatial or novel problem-solving skills than the "linguistic" subtype.

A comparison of the subtypes on academic measures was also planned. Problems with academic achievement had been reported in several studies of hydrocephalic children (see section 3.3.). Therefore, measures of academic achievement were obtained in this study. At the same time, it appeared inappropriate to include those measures in the subtype analysis because the sample included a large proportion of five- and six-year-old children with no or little formal academic education. We therefore planned to do a separate analysis on those children that had received at least six months of formal education. This analysis would include comparison of those children on a variety of measures of academic achievement by means of MANOVA. This analysis would only be performed if more than half of the sample could be included in it.

The third purpose of this dissertation was to investigate the prevalence of psychiatric disturbance in the subject sample and whether the identified neuropsychological subtypes presented with different profiles of personality functions. We assessed
personality functioning and psychiatric disturbances in the subjects by means of the Personality Inventory for Children - Revised (PIC-R; Lachar, 1982; Wirt, Lachar, Klinedinst, & Seat, 1984), which is an empirically constructed instrument providing comprehensive personality descriptions. The main purpose here was to see whether the different neuropsychological subtypes would display different personality profiles on the PIC-R, and whether PIC-R subtypes could be found (to be identified with the same cluster analyses as outlined above). Such PIC-R subtypes could then be compared on medical history variables, to determine if there was anything in their physical or social status that discriminated these subtypes.

In addition to maladjustment in the patients, another area of interest was the level of distress in the parents of the patients. We assessed this by means of the Symptom Check List-90-Revised (SCL-90-R; Derogatis, 1977). This is a short and well-researched self-report inventory with sound psychometric properties which assesses psychogenic distress (Tennen, Affleck, & Herzberger, 1985). We related the level of distress, as measured by the SCL-90-R scores, to a variety of selected physical and social variables measured at the time of assessment (i.e., these physical and social variables were not the predictive variables used for the intellectual development aspect of the study). We used multiple regression and discriminant analyses to investigate the relation of these variables to the parents' distress. Due to the lack of agreement in the literature it was difficult to generate hypotheses concerning many of these variables (e.g., degree of
mobility), but it was hypothesized that at least the level of maladjustment in the children would correlate positively with the level of distress in the parents.

In summary, the following hypotheses were tested in this dissertation:

Null hypothesis 1:

"Medical history variables do not predict or discriminate in a statistically significant or systematic way the value of VIQ, PIQ, or FSIQ as assessed by the WISC-R or WPPSI".

If this null hypothesis were to be rejected, it was expected that a set of medical history variables could be identified that were statistically significant predictors or discriminators of intellectual achievement in children with shunted infantile hydrocephalus (this "best predictive" or "best discriminative" set would represent an important reduction in data redundancy in terms of the number of medical history variables included).

Null hypothesis 2:

"There will be no difference between VIQ and PIQ (as assessed by the WISC-R or WPPSI) in the sample".

If this null hypothesis were to be rejected it was expected that, in the total sample, the average PIQ would be significantly lower than the average VIQ, as assessed by the WISC-R or WPPSI.
Null hypothesis 3:

"No reliable subtypes of hydrocephalic children can be identified on the basis of their neuropsychological test-performance".

If this null hypothesis were to be rejected, it was expected that at least two subtypes of children with shunted infantile hydrocephalus could be identified based on their performance on a comprehensive battery of neuropsychological tests. One subtype was expected to demonstrate a preponderance of expressive and receptive linguistic dysfunctions, and difficulties with overlearned strategies. A second subtype was expected to present with relatively more prominent visuo-perceptual and visuo-constructive dysfunctions, and difficulties with novel problem-solving.

Null hypothesis 4:

"If subtypes can be identified, none of them will differ significantly from each other in FSIQ, as measured by the WISC-R or WPPSI".

If we were to not reject this null hypothesis, the interpretation of neuropsychological patterns of different subtypes would become problematic.
Null hypothesis 5:
"If subtypes can be identified, none of them will differ significantly from each other in VIQ or PIQ as measured by the WISC-R or WPPSI".

If this null hypothesis were to be rejected, it was expected that the subtype with predominant dysfunctions in the linguistic or overlearned domain would have a significantly lower mean-VIQ and a significantly higher mean PIQ, as assessed by the WISC-R or WPPSI, than the subtype with predominant dysfunctions in the areas of visuo-perceptual and visuo-constructive skills and novel problem-solving.

Null hypothesis 6:
"If subtypes can be identified, none of them will differ significantly from each other on any other measures that are not included in the subtype classification".

If this null hypothesis were to be rejected, it was expected that the subtype with predominant linguistic dysfunctions would have significantly lower scores on measures of linguistic abilities or overlearned strategies not included in the subtype classification than the subtype with predominant visuo-perceptual and visuo-constructive dysfunctions. In addition, it was expected that the subtype with predominant visuo-perceptual and visuo-constructive dysfunctions would have significantly lower scores on measures of visuospatial and novel problem-solving.
abilities not included in the subtype classification than the subtype with predominant linguistic dysfunctions.

Null hypothesis 7:
"Social and physical variables are not statistically significant predictors of level of distress in parents of hydrocephalic children, as assessed by the SCL-90-R".

If this null-hypothesis were to be rejected, it was expected that the level of maladjustment in the subjects would be among the significant predictors of the level of distress in the parents.
CHAPTER II

METHOD

Subjects.

119 potential subjects between the ages of 5.6 and 8.6 years were selected from a sample available at the Department of Neurosurgery, Children's Hospital of Michigan, Detroit, Michigan. This age range was chosen because all of the selected psychological tests have age norms that cover this age range. Furthermore, it appeared desirable to limit the age range to a distribution that could be considered somewhat homogenous on theoretical grounds and that was equivalent to divisions made by other authors (e.g., Rourke, Bakker, Fisk, & Strang, 1986).

From these 119 candidates, subjects were further selected according to the following criteria:

1.

Hydrocephalus documented by radiological studies and treated with surgical implantation of a ventricular shunt before the age of 12 months.

This time criterion was selected to exclude cases in which the hydrocephalus became apparent after a measurable development of specific neuropsychological functions. It was also a limitation that is common in the literature. One case of spontaneously arrested hydrocephalus was excluded for this reason. Nine other
Children who had been operated after the age of one year (including all tumor cases) were also excluded.

3. Absence of significant additional brain injuries, disease, or psychopathology since the time of shunting.
This criterion was selected because additional lesions might obscure the effect of hydrocephalus in the statistical analyses. Five children were excluded because of additional brain injuries after their first year (two with closed head injury, two with anoxic episodes after the first year, and one with Sturge-Weber syndrome). Two children had expired before the time of assessment. Two other children were physically ill when twice contacted. In two cases, there was documented evidence for major psychiatric problems in the family.

4. English-speaking language background.
This criterion was selected to ensure that all the dependent measures could be administered in a valid way and that the norms would be applicable. One case in which Arabic was the major language spoken at home was excluded for this reason.

5. Statistical analyses involving comparisons on neuropsychological test measures or pattern of IQ scores would include only children whose Full Scale IQ exceed 70.
This criterion was suggested by Dennis et al. (1981) for the purpose of ensuring that variations in IQ pattern are related to
the characteristics of hydrocephalus and not just to the properties of the IQ distribution. It also ensures that all subjects included in the analyses have IQs within two standard deviation from the normative mean, which reduces the chance of "outliers". However, cases with a FSIQ < 70 can and were included in analyses that involved discrimination between discrete groups (stepwise discriminant analyses), and in analyses of questionnaires filled out by the parents.

Consideration of criteria 1. through 4. led to the exclusion of a total of 22 cases. In addition, 48 other cases could not be evaluated, for various reasons. Nine of the parents contacted declined to participate in the study. For twelve patients, an appointment was scheduled at least twice, and on both occasions the appointment was not kept, without notice (new appointments were not made in those cases). Five patients were found to live outside the states of Michigan and Ohio, and were not contacted. Finally, we were unable to contact 22 additional cases because a correct current address or phone number was not available in the hospital records.

The final subject sample therefore consisted of 49 cases. The data from one of these children were omitted from the analyses because the patient became sick during the assessment and the mother was unwilling to reschedule. Fourteen of the evaluated children were retarded to the degree that psychometric tests could not be administered to them in a standard manner. However, PIC-R and SCL 90-R data were obtained in these cases.
In summary, full neuropsychological assessments were obtained from 34 children. PIC-R and SCL 90-R data were obtained from the parents of 48 children. The PIC-R data from five of these cases were excluded from the analyses because the profile was considered to be invalid (i.e., L-scale had a T-score that exceeded 70).

**Measures.**

**Medical history variables.**

The variables that were selected on the basis of a review of the literature are presented in Table 2. The distinction in demographic, symptom, pathology, and therapeutic variables was originally suggested by Dennis et al. (1981).

Most of the variables were coded in terms of presence or absence (P/A), reflected by a numerical value of 1 (presence) or 2 (absence). Age at testing was coded as a continuous variable, in months. Sex was coded as 1 (male) or 2 (female); handedness as 1 (right-handed) or 2 (left-handed). Race was classified in terms of nominal descriptive categories and was therefore coded as a 'dummy' (D) variable (Cohen, 1968).

Dummy coding involves giving the subject a rating of 1 (applicable) or 2 (not applicable) on each of the categories that are included to describe the sample. There is always one dummy level less than there are sample categories to be identified. That is because this one category can be identified by a scoring of 2 (not applicable) on each of the other levels.
Table 2.

**Medical history variables assessed in hydrocephalic sample.**

<table>
<thead>
<tr>
<th>VARIABLE NAME</th>
<th>DESCRIPTION</th>
<th>MEASUREMENT</th>
</tr>
</thead>
<tbody>
<tr>
<td>Demographic variables</td>
<td></td>
<td></td>
</tr>
<tr>
<td>1. Age at testing</td>
<td>Test date - birth date.</td>
<td>Months.</td>
</tr>
<tr>
<td>2. Sex</td>
<td>Male or Female.</td>
<td>1/2:</td>
</tr>
<tr>
<td>3. Race</td>
<td>Black, White, or other.</td>
<td>D.</td>
</tr>
<tr>
<td>4. Handedness.</td>
<td>Right- or left-handedness.</td>
<td>1/2.</td>
</tr>
<tr>
<td>5. S.E.S.</td>
<td>Socio-Economic Status.</td>
<td>D.</td>
</tr>
<tr>
<td>Symptom variables</td>
<td></td>
<td></td>
</tr>
<tr>
<td>6. Prenatal and</td>
<td>Toxemia, intra-uterine infections, antepartum</td>
<td>P/A</td>
</tr>
<tr>
<td>perinatal problems.</td>
<td>hemorrhages, prematurity, meconium-stained fluids, and/or low birth weight.</td>
<td></td>
</tr>
<tr>
<td>7. Neonatal and</td>
<td>Hypoxia, respiratory distress, Rhesus defects, head trauma, cerebral hemorrhages, seizures and/or CNS infections in first year of life.</td>
<td></td>
</tr>
<tr>
<td>infancy problems.</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
Table 2. (Continued)

**Medical history variables assessed in hydrocephalic sample.**

<table>
<thead>
<tr>
<th>VARIABLE NAME</th>
<th>DESCRIPTION</th>
<th>MEASUREMENT</th>
</tr>
</thead>
<tbody>
<tr>
<td>8. Ocular defects.</td>
<td>Disorders of gaze, movement, refraction and/or accommodation.</td>
<td>P/A.</td>
</tr>
<tr>
<td>9. Motor problems.</td>
<td>Fine motor incoordination, ataxia, gait problems, and/or paraplegia.</td>
<td>D.</td>
</tr>
<tr>
<td>10. Seizures.</td>
<td>Any seizures occurring after age 1 year.</td>
<td>P/A.</td>
</tr>
<tr>
<td>11. L.S.D.</td>
<td>Lacunar skull deformities.</td>
<td>P/A.</td>
</tr>
<tr>
<td>12. Site of sac.</td>
<td>Vertebral location of the myelomeningocele sac.</td>
<td>D.</td>
</tr>
<tr>
<td>13. Sensory level.</td>
<td>Spinal sensory level recorded at birth.</td>
<td>D.</td>
</tr>
</tbody>
</table>

**Pathology variables.**

<table>
<thead>
<tr>
<th>14. Type of hydrocephalus</th>
<th>Intra-ventricular or extra-ventricular obstructive hydrocephalus.</th>
</tr>
</thead>
<tbody>
<tr>
<td>15. Aqueductal lesion</td>
<td>Stenosis, atresia, and/or gliosis of the cerebral aqueduct.</td>
</tr>
</tbody>
</table>
Table 2. (Continued).

**Medical history variables assessed in hydrocephalic sample.**

<table>
<thead>
<tr>
<th>VARIABLE NAME</th>
<th>DESCRIPTION</th>
<th>MEASUREMENT</th>
</tr>
</thead>
<tbody>
<tr>
<td>malformation.</td>
<td></td>
<td></td>
</tr>
<tr>
<td>17. Dandy-Walker</td>
<td>Cystic distention of the fourth ventricle.</td>
<td>P/A.</td>
</tr>
<tr>
<td>syndrome.</td>
<td></td>
<td></td>
</tr>
<tr>
<td>18. Bleeds.</td>
<td>Subdural hematoma, subarachnoid hemorrhage, and/or intra-ventricular</td>
<td>P/A.</td>
</tr>
<tr>
<td></td>
<td>hemorrhage.</td>
<td></td>
</tr>
<tr>
<td>19. Cerebral</td>
<td>Meningitis, encephalitis, and/or arachnoiditis.</td>
<td>P/A.</td>
</tr>
<tr>
<td>infection.</td>
<td></td>
<td></td>
</tr>
<tr>
<td>20. Cyst.</td>
<td>Subdural hygroma, paraventricular cyst, or arachnoid cyst.</td>
<td>P/A.</td>
</tr>
</tbody>
</table>

**Therapeutic variables.**

<table>
<thead>
<tr>
<th></th>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>22. Left shunt.</td>
<td>Shunt in left hemisphere.</td>
<td>P/A.</td>
</tr>
</tbody>
</table>
Table 2. (Continued)

Medical history variables assessed in hydrocephalic sample.

<table>
<thead>
<tr>
<th>VARIABLE NAME</th>
<th>DESCRIPTION</th>
<th>MEASUREMENT</th>
</tr>
</thead>
<tbody>
<tr>
<td>23. Right shunt.</td>
<td>Shunt in right hemisphere.</td>
<td>P/A.</td>
</tr>
<tr>
<td>24. Head size</td>
<td>Occipital-frontal circumference at the time of first-shunt insertion.</td>
<td>Mm.</td>
</tr>
<tr>
<td>25. Shunt revision.</td>
<td>Number of shunt revisions.</td>
<td>N.</td>
</tr>
<tr>
<td>26. Shunt infection.</td>
<td>Number of shunt infections.</td>
<td>N.</td>
</tr>
</tbody>
</table>

Race was thus classified as 'white' or 'black', with all other races described with a 2 on each of these categories. Socio-economic status (SES) was classified according to Hollingshead's Index (Hollingshead & Redlich, 1958). The Hollingshead index utilizes two factors (occupation and education) to determine social position, and five social classes can be distinguished on the basis of ranges of index scores. Because of the nominal nature of this classification, SES was coded as a 'dummy' variable, using Hollingshead's five class levels.

Motor problems at the time of assessment were assessed in terms of six nominal descriptive categories (modified from Dennis et al., 1986) and was therefore coded as a 'dummy' variable: 'fine motor coordination problems' (including limb ataxia, dyspraxia, intention tremor and/or dysdiadochokinesia), 'truncal ataxia
and/or balance problems', 'gait problems' (due to ankle clonus, equinua, etc.), 'mild paraplegia' (involving splints or short leg braces), or 'severe paraplegia' (involving wheelchair or crutches). Absence of any motor problems was scored by means of a 2 (not applicable) on each of these categories.

Site of the myelomeningocele sac was classified according to the criteria suggested by Soare and Raimondi (1977) and was coded as a 'dummy' variable: 'lumbo-sacral and sacral', 'lumbar', 'thoraco-lumbar and thoraco-lumbo-sacral', or 'occipito-cervical and cervical'. Absence of a myelomeningocele was coded by means of a 2 (not applicable) on each of these categories. Sensory level at birth was also classified according to criteria suggested by Soare and Raimondi and was also coded as a 'dummy' variable: 'L5-S5', 'L1-L4', or 'T12 and up'. Absence of any sac or sensory loss was coded by means of a 2 (not applicable) on each of these categories.

Type of hydrocephalus was coded as 1 (intra-ventricular obstructive) or 2 (extra-ventricular obstructive). Due to the fact that all tumor cases were operated after the age of one year (and thus were excluded from the study), tumor was not included under the pathology variables.

Occipito-frontal circumference was selected as an indirect measure of the degree of hydrocephalus at the time of the first shunting procedure. Several other measures, including frontal ventriculo-skull distance and brain mass have been used in the literature (e.g., Shurtleff et al., 1973). However, results with such measures have been typically inconsistent (e.g., Tromp et
Moreover, ventriculo-skull distance was not measured routinely at Children's Hospital of Michigan, and CT scans were not available for all subjects. Since a measure of the occipito-frontal circumference at the time of first shunting was available for all subjects, this measure was included in the study. The numbers of shunt revisions and shunt infections were coded as discrete variables, representing the actual numbers (n) of occasions.

Psychological test variables.

The Wechsler Intelligence Scale for Children - Revised (WISC-R; Wechsler, 1974) and the Wechsler Primary and Preschool Scale of Intelligence (WPPSI; Wechsler, 1967) were selected as the measures of intelligence that would provide the dependent measures for the regression or discriminant analyses involving the medical history variables as independent variables. The inclusion of these two different intelligence tests was considered appropriate since they have very much the same format, and share about two thirds of common variance. The Wide Range Achievement Test - Revised (WRAT-R; Jastak & Wilkinson, 1984) was used as a measure of academic achievement.

In line with the criteria outlined in the section entitled "Statement of the problem", a variety of psychological tests were selected for purposes of subtype classification and validation. Additional considerations in the selection of these tests included: (1) to have equal numbers of tests aimed at assessing
Table 3.

Psychological and sensory-motor measures included in individual assessments.

Psychological tests

1. PEABODY = Peabody Picture Vocabulary Test - Revised (Dunn & Dunn, 1981).
2. VOCAB = WISC-R or WPSSI Vocabulary subtest.
4. FLUENCY = MSCA Verbal Fluency Test
5. SPATHEM = KABC Spatial Memory subtest.
6. STORY = MSCA Verbal Memory II subtest.
7. TARGET = Target Test (Reitan & Davison, 1974).
8. REMIND = Selective Reminding Test (Morgan, 1982).
9. MATRIX = KABC Matrix Analogies subtest.
10. RIDDLES = KABC Riddles subtest.
11. PROGFIG = Progressive Figures Test (Reitan & Davison, 1974).
13. CLOSURE = KABC Gestalt Closure subtest.
14. BLOCKDES = WISC-R or WPSSI Block Design subtest.
15. MATCHFIG = Matching Figures Test (Reitan & Davison, 1974).
Psychological and sensory-motor measures included in individual assessments.

Tests of tactile and motor functions.

17. FSYMBOL = Finger Tip Symbol Writing Test (Reitan & Davison, 1974).
18. FEGS = Grooved Pegboard Test (Rourke et al., 1983).
19. FAGNOS = Finger Agnosia Test (Reitan & Davison, 1974).
20. TAPPING = Finger Tapping Test (Reitan & Davison, 1974).

Abbreviations: KABC = Kaufman Assessment Battery for Children (Kaufman & Kaufman, 1983); MSCA = McCarthy Scales of Children's Abilities (McCarthy, 1972).

Linguistic abilities on one hand, and tests aimed at assessing visuospatial abilities on the other hand; (2) to include tests for both receptive/perceptive and for expressive abilities in each of these categories; (3) to include both 'verbal' and 'spatial' tests (i.e., involving either verbal or visual stimuli and requiring primarily either verbal or spatial processing) in domains such as memory and problem-solving; and (4) to include measures of tactile and motor functions. The selected tests are listed in table 3.
It is important to realize that there are many subjective ways in which the psychological tests in table 3 can be classified. For example, RIDDLES might be considered a linguistic test, but also a problem-solving task. Similarly, BLOCKDES could be labeled a visuospatial task, but also a test of novel problem-solving. Factor analysis was used in this study as one of the ways to classify tests into specific groups.

The Personality Inventory for Children - Revised (PIC-R; Lachar, 1982; Wirt, Lachar, Klinedinst, & Seat., 1984) was selected as a measure of personality functioning in the patient sample. The Symptom Check List 90-Revised (SCL 90-R; Derogatis, 1977) was selected as a measure of malaise or distress in the parents of the children.

The obtained performances on each of the psychological and sensory-motor test variables were converted to T-scores, based on the means and standard deviations of the normative sample of each test (i.e., the norms for the general population; not for specific patient populations, as is possible with some of these tests). This was done in such a way that better than average performances were indicated by T-scores over 50 and below average performances by T-scores below 50. Furthermore, these scores were based on the most conventional indices provided by each test (e.g., scaled scores for subtests of the WISC-R, standard scores for some of the other tests), with the exception of the scores in the tactile and motor domain.

Usually, performance on measures of sensory and motor abilities are reported in terms of performance on both sides of
the body separately. Because of the diffuse effects of hydrocephalus (which will make clearly lateralized sensory or motor findings unlikely) and because of the possible contamination by spinal and peripheral factors, we considered it to be undesirable to report performance on tactile and motor measures for the left and right hand separately. Instead, a mean of the left and right hand T-score performance was used. Combining the scores from the right and left hand on the same test was suggested by Francis (1985). He argued that a composite score, based on both lateral indices, will increase test reliability and does not reduce the information provided by the measures (unless one is specifically interested in the discriminative validity of lateral measures). The purpose of including sensory and motor measures in this study was to investigate the relationship between sensory-motor functioning and IQ test performance.

**Physical and social variables**

On the basis of our review of the literature, a variety of physical and social measures were selected that could be related to the level of distress of the parents by means of multiple regression or discriminant analyses. These measures are listed in Table 4.

The first four variables (age, sex, socio-economic status, and motor disability) were assessed and coded in the manner described in the section on medical history variables. Bladder incontinence, bowel incontinence, and seizure disorder were coded as 1 (present) or 2 (absent). The level of general
maladjustment in the patient was defined as the T-score of the patient on the four Factor scales of the PIC-R, coded as discrete variables. School attendance was coded as 1 (special school) or 2 (regular school). The number of siblings was coded as a discrete variable. Broken home was defined as a single-parent family (regardless of cause) and was coded as 1 (applicable) or 2 (not applicable).

Table 4.

**Current physical and social variables assessed in patient and parent sample.**

1. Age of patient.
2. Sex of patient.
5. Bladder incontinence in patient.
7. General maladjustment in patient.
10. Number of siblings.
11. Broken home.
Statistical analyses

The computer programs that were used for the statistical analyses are capitalized in this section and are, unless otherwise indicated, all documented in the SAS User's Guide (1985 Edition) or in the SYSTAT manual (Wilkinson, 1987).

We had initially planned to use only children with FSIQ scores that exceeded 70 in the analyses of the effect of the medical history variables. The plan was to use all coded medical history variables of these children as independent variables in separate stepwise multiple regression analyses with either VIQ, PIQ, or FSIQ as a dependent variable, and to find best regression models for each of these three IQ estimates. However, inspection of our subject sample revealed that this would lead to the exclusion of a large number of subjects (14 children who could not be assessed with an IQ test because of severe retardation, and four children with FSIQ below 71). The distribution of IQ scores for the sample is shown in tables 5, 6, and 7. Two children had a VIQ below 71 and four children a PIQ below 71, yielding four children with FSIQ below 71 (i.e., the children with PIQ < 71 were the same children that had FSIQ < 71, and two of these children also had a VIQ < 71).
Table 5.

*Distribution of Verbal IQ scores in total sample (n = 48)*

<table>
<thead>
<tr>
<th>Value</th>
<th>Frequency</th>
<th>Proportion</th>
</tr>
</thead>
<tbody>
<tr>
<td>NT</td>
<td>14</td>
<td>29.17</td>
</tr>
<tr>
<td>59</td>
<td>1</td>
<td>2.08</td>
</tr>
<tr>
<td>68</td>
<td>1</td>
<td>2.08</td>
</tr>
<tr>
<td>75</td>
<td>1</td>
<td>2.08</td>
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<td>77</td>
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<td>2.08</td>
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<tr>
<td>80</td>
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<td>2.08</td>
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<tr>
<td>84</td>
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<td>2.08</td>
</tr>
<tr>
<td>99</td>
<td>1</td>
<td>2.08</td>
</tr>
</tbody>
</table>
Table 5 (continued).

Distribution of Verbal IQ scores in total sample (n = 48)

<table>
<thead>
<tr>
<th>VALUE</th>
<th>FREQUENCY</th>
<th>PROPORTION</th>
</tr>
</thead>
<tbody>
<tr>
<td>100</td>
<td>3</td>
<td>6.25</td>
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<tr>
<td>101</td>
<td>2</td>
<td>4.17</td>
</tr>
<tr>
<td>102</td>
<td>3</td>
<td>6.25</td>
</tr>
<tr>
<td>103</td>
<td>2</td>
<td>4.17</td>
</tr>
<tr>
<td>107</td>
<td>1</td>
<td>2.08</td>
</tr>
<tr>
<td>109</td>
<td>1</td>
<td>2.08</td>
</tr>
<tr>
<td>111</td>
<td>1</td>
<td>2.08</td>
</tr>
<tr>
<td>114</td>
<td>2</td>
<td>4.17</td>
</tr>
<tr>
<td>119</td>
<td>1</td>
<td>2.08</td>
</tr>
<tr>
<td>124</td>
<td>2</td>
<td>4.17</td>
</tr>
<tr>
<td>145</td>
<td>1</td>
<td>2.08</td>
</tr>
</tbody>
</table>

Abbreviation: NT = Not Testable
Table 6.

Distribution of Performance IQ scores in total sample (n = 48)

<table>
<thead>
<tr>
<th>VALUE</th>
<th>FREQUENCY</th>
<th>PROPORTION</th>
</tr>
</thead>
<tbody>
<tr>
<td>NT</td>
<td>14</td>
<td>29.17</td>
</tr>
<tr>
<td>48</td>
<td>1</td>
<td>2.08</td>
</tr>
<tr>
<td>58</td>
<td>1</td>
<td>2.08</td>
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<td>63</td>
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<td>67</td>
<td>1</td>
<td>2.08</td>
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<tr>
<td>75</td>
<td>1</td>
<td>2.08</td>
</tr>
<tr>
<td>77</td>
<td>2</td>
<td>4.17</td>
</tr>
<tr>
<td>80</td>
<td>2</td>
<td>4.17</td>
</tr>
<tr>
<td>81</td>
<td>3</td>
<td>6.25</td>
</tr>
<tr>
<td>85</td>
<td>2</td>
<td>4.17</td>
</tr>
<tr>
<td>87</td>
<td>1</td>
<td>2.08</td>
</tr>
<tr>
<td>88</td>
<td>2</td>
<td>4.17</td>
</tr>
<tr>
<td>89</td>
<td>1</td>
<td>2.08</td>
</tr>
<tr>
<td>90</td>
<td>1</td>
<td>2.08</td>
</tr>
<tr>
<td>91</td>
<td>2</td>
<td>4.17</td>
</tr>
<tr>
<td>93</td>
<td>1</td>
<td>2.08</td>
</tr>
<tr>
<td>96</td>
<td>2</td>
<td>4.17</td>
</tr>
<tr>
<td>97</td>
<td>1</td>
<td>2.08</td>
</tr>
</tbody>
</table>
Table 6 (continued).

Distribution of Performance IQ scores in total sample ($n = 40$)

<table>
<thead>
<tr>
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<th>PROPORTION</th>
</tr>
</thead>
<tbody>
<tr>
<td>100</td>
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<td>2.08</td>
</tr>
<tr>
<td>101</td>
<td>1</td>
<td>2.08</td>
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<td>105</td>
<td>1</td>
<td>2.08</td>
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<tr>
<td>106</td>
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<td>2.08</td>
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<td>108</td>
<td>1</td>
<td>2.08</td>
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<td>115</td>
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<tr>
<td>119</td>
<td>1</td>
<td>2.08</td>
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<tr>
<td>123</td>
<td>1</td>
<td>2.08</td>
</tr>
</tbody>
</table>

Abbreviation: NT = Not Testable
Table 7.

Distribution of Full-Scale IQ scores in total sample (n = 40)

<table>
<thead>
<tr>
<th>VALUE</th>
<th>FREQUENCY</th>
<th>PROPORTION</th>
</tr>
</thead>
<tbody>
<tr>
<td>55</td>
<td>2</td>
<td>4.17</td>
</tr>
<tr>
<td>70</td>
<td>2</td>
<td>4.17</td>
</tr>
<tr>
<td>76</td>
<td>1</td>
<td>2.08</td>
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<tr>
<td>81</td>
<td>2</td>
<td>4.17</td>
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<tr>
<td>84</td>
<td>1</td>
<td>2.08</td>
</tr>
<tr>
<td>87</td>
<td>1</td>
<td>2.08</td>
</tr>
<tr>
<td>88</td>
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<td>92</td>
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<td>4.17</td>
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<td>93</td>
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</tr>
<tr>
<td>99</td>
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<td>2.08</td>
</tr>
</tbody>
</table>
Table 7 (continued).

Distribution of Full-Scale IQ scores in total sample (n = 48)

<table>
<thead>
<tr>
<th>VALUE</th>
<th>FREQUENCY</th>
<th>PROPORTION</th>
</tr>
</thead>
<tbody>
<tr>
<td>105</td>
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<td>4.17</td>
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<tr>
<td>106</td>
<td>2</td>
<td>4.17</td>
</tr>
<tr>
<td>111</td>
<td>2</td>
<td>4.17</td>
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<tr>
<td>112</td>
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<td>122</td>
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<td>2.08</td>
</tr>
<tr>
<td>129</td>
<td>1</td>
<td>2.08</td>
</tr>
</tbody>
</table>

Abbreviation: NT = Not Testable

The exclusion of 18 subjects from a total sample of 48 was considered to be unacceptable for several reasons. First, this would greatly compromise the subject/variable ratio in any multivariate statistical procedure. Second, this would make the external validity of the study questionable because it would remove more than a third of the subject of an already selective sample. Moreover, if one considers the question of parents or professionals with regard to developmental outcome over a
several-years span, it is not hard to imagine that knowledge of
the risk of significant mental retardation is an important factor.

For these reasons, it was decided to use stepwise multiple
discriminant analyses instead of regression analyses.
Discriminant analysis addresses the question how a priori defined
groups can be differentiated by other variables that were not
included in the a priori discrimination process. Because of the
specific distribution of IQ scores (see tables 5, 6, and 7) and
the size of the total sample, it was decided to divide the subject
sample in two groups for each IQ measure: one group that included
all children that were either not testable or had specific IQ
scores below 71 (N = 16 for VIQ; N = 18 for PIQ and FSIO), and one
group that included all children that had specific IQ scores above
70 (N = 32 for VIQ; N = 30 for PIQ and FSIO). These groups will
hereafter be referred to as groups LOW VIQ (< 71) and HIGH VIQ (> 70), and groups LOW PIQ (< 71) and HIGH PIQ (> 70) respectively.
Since the utilization of these 'cut-off' points resulted in the
same groups for PIQ and FSIO, no additional classification needed
to be made for FSIO.

It would be ideal if all coded medical history variables
could be included in one discriminant analysis for each IQ
measure. As can be seen in table 2, however, the number of
independent variables (26) was rather large in comparison to the
number of subjects (48) and this is problematic in discriminant
analysis. The number of variables was furthermore inflated due to
the use of dummy variables. Therefore, we elected to divide the
variables from table 2 into six categories, and to perform
separate stepwise multiple discriminant analysis for each of these groups of measures. This division was done on a subjective basis (i.e., not the result of factor analytic procedures). The six variable categories are presented in Table 8.

Table 8.

**Categories of independent variables, used in discriminant analyses for intellectual outcome.**

<table>
<thead>
<tr>
<th>CATEGORY</th>
<th>VARIABLES INCLUDED (total measures)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Demographic</td>
<td>Age at testing, sex, race, handedness, Socio-Economic status (9).</td>
</tr>
<tr>
<td>2. Motor symptoms</td>
<td>Motor coordination and ambulation problems (5).</td>
</tr>
<tr>
<td>3. Spinal cord symptoms</td>
<td>Site of the myelomeningocele, and sensory level at birth (7).</td>
</tr>
<tr>
<td>4. Other symptoms</td>
<td>Perinatal problems, infancy problems, lacunar skull deformities, and current ocular defects and seizures (5).</td>
</tr>
<tr>
<td>5. Pathology</td>
<td>Type of hydrocephalus, and etiology (7).</td>
</tr>
<tr>
<td>6. Treatment</td>
<td>Age at first shunt insertion, hemispheric location of shunt, occipito-frontal circumference at first shunt insertion, number of shunt revisions, and number of shunt infections (6).</td>
</tr>
</tbody>
</table>
Separate analyses were performed for VIQ and PIQ measures, for each of the six variable categories; the discrimination criterion being groups (N = 2; LOW versus HIGH) in each analysis. PSIQ was not analyzed separately, since its distribution in terms of LOW versus HIGH was identical to that of PIQ. These discriminant analyses resulted in six best discrimination models (one for each variable category) for each IQ estimate. Finally, those and only those variables that remained in those best discrimination models were all included in final stepwise multiple discriminant analyses to arrive at over-all best discrimination models for VIQ and PIQ. The SAS STEPDISC procedure with BACKWARD elimination was used for these discriminant analyses. Only variables that were statistically significant at the .05 level were retained in the discrimination models. All 48 subjects (divided in groups LOW and HIGH) were included in all these analyses.

The raw VIQ and PIQ scores of the 30 subjects in the sample who had IQ scores that exceeded 70 were analyzed by means of t-test for paired observations. The SYSTAT STATS procedure was used. This is of course a dependent t-test, since the variables are limited to two related observations per subject, based on the nature of the task.

The raw scores on all the WISC-R / WPPSI, WRAT-R, and neuropsychological tests were converted to T-scores (based on the means and standard deviations of the normative sample of each test) to allow for comparison between tests. The SCL 90-R and
PIC-R raw scores were converted to T-scores in the manner that is standard with these instruments. The T-scores for all measures were used as independent variables in all further statistical analyses. It is important to realize that for all intelligence, achievement, and neuropsychological measures higher T-scores reflect better performance, whereas for the PIC-R and SCL 90-R data, higher T-scores reflect more significant problems.

In order to assess the relationship between sensory-motor functioning and performance on intelligence tests, the SAS CORR procedure was used to compute Pearson product-moment correlations and Kendall Tau correlations between the various sensory and motor measures on one hand (FSYMBOL, PEGS, PAGNOS, and TAPPING) and IQ scores on the other (VIQ, PIQ, and FSIQ). The T-scores for each of these measures were used in this analysis. Only the 30 subjects with FSIQ > 70 were included in this analysis.

Subtyping (cluster) analyses were performed for two groups of data: One involving neuropsychological test measures, and another involving the PIC-R data. Only subjects with complete neuropsychological testings and for whom all IQ scores exceeded 70 (i.e., only the 30 children from the HIGH PIQ group) were included in the subtyping analyses with neuropsychological data. All subjects on whom valid PIC-R profiles were obtained from the parents (N = 43) were included in the subtyping involving PIC-R data. The same cluster analytic procedures were utilized for the neuropsychological data as for the PIC-R data. Specifically, the SAS and SYSTAT CLUSTER procedures were used, with the AVERAGE
option for initial subtyping and the CENTROID, WARD, and K-Means options for analysis of reliability. The AVERAGE and CENTROID options are available both in SAS and SYSTAT. Only SAS provides the WARD option, and only SYSTAT provides the K-means option.

The AVERAGE, CENTROID, and WARD options all perform a cluster analysis of the hierarchical agglomerative type. This means that the procedures start with all n individual cases, and at each successive (hierarchical) stage combines together the two subjects or clusters that are closest (i.e., most similar) until finally all cases are combined in one cluster. For each of these options, squared Euclidean distance was used as a metric of similarity.

The AVERAGE option uses average linkage as a procedure to assign subjects to clusters. The term 'linkage' pertains to the 'link', or the smallest distance between an entity and any other entity in the set. In average linkage cluster analysis, the distance between two clusters is defined as the average of the distance between all pairs of entities in the two clusters, and each member of a cluster has a smaller average dissimilarity with other members of the same cluster than with members of any other cluster (Lorr, 1983).

The CENTROID option uses centroid linkage as a procedure to assign subjects to clusters. This produces clusters in which the members have a higher average similarity to the centroid of their own cluster than to that of any other cluster (the centroid being a set of variable means across the members of the cluster; Lorr, 1983).
The WARD option is based on the method of 'minimum variance'. It forms clusters in such a way that the sum of squared within-cluster deviations around the cluster mean of each variable is minimized for all variables at the same time (Lorr, 1983).

The K-means option performs a non-hierarchical or single-level cluster search method that involves iterative partitioning of subjects. The value of K represents the number of groups. It can check how many subjects of initially defined clusters are relocated as the result of the partitioning process (Morris et al., 1981).

For the AVERAGE, CENTROID, and WARD methods, the level of the semipartial R2 (provided by the SAS CLUSTER procedure) was analyzed for the amount of accounted-for variance that was lost by joining clusters at each successive step. In addition, a post-hoc inspection of the dendogram for the AVERAGE solution (provided by the SYSTAT CLUSTER procedure) was performed for the location of 'peaks and valleys'. These two strategies were used to determine the optimal number of clusters. The inspection of the dendogram is subjective and was only used as a cross-validation of the analyses of the semi-partial R2. The latter index was utilized to estimate the optimal number of clusters in the following way: A loss of more than 10% of the accounted-for variance in two successive steps in the clustering process was considered to represent the combination of clusters into a heterogenous cluster. Such an increase was given consideration as a possible 'cut-off point' to stop merging clusters further.
The K-means iterative partitioning method was also used for analysis of reliability of clusters. The maximum number of clusters allowed in this latter analysis was defined as the number of clusters that was suggested by the other procedures. It was felt that the number of subjects was too small to also test the reliability of the cluster solutions with a split-sample design.

For the neuropsychological test data, a factor analytic procedure was first performed to reduce the number of test variables that could be used to generate subtypes. The latter was considered to be desirable because the number of subjects who had complete neuropsychological testings and whose IQ scores exceeded 70 was small (N = 30) in comparison to the number of neuropsychological test measures (N = 16). The factor analysis was performed to select two subsets of measures: One that could be used to generate subtypes with the above-mentioned cluster analyses, and one that could be used to validate the obtained clusters. The planned procedure for the latter involved comparing the identified clusters on the other ('validation') variables by means of MANOVA. The SAS GLM procedure (with the BON option to use Bonferroni alpha correction for multiple comparisons) was planned for this purpose.

The SAS FACTOR procedure was used to perform a principal component analysis (PRINCIPAL method, with VARIMAX orthogonal rotation) on the neuropsychological measures that were presented in table 3. Principal component analysis derives a small number of linear combinations (principal components) from a set of quantitative variables. In this way, it is distinguished from
common factor analysis, which attempts to explain correlations among variables by means of an unobservable, hypothetical variable that cannot be linearly computed on the basis of the original ones (Kshirsagar, 1972). Because we were primarily interested in reduction of variable redundancy, principal component analysis was used. Rotation can be performed with both common factor analysis and principal component analysis to produce a factor pattern in which all the coefficients are close to 0, 1 or -1. This makes the factors easier to interpret. Orthogonal and oblique rotations are possible, with the difference being that rotated factors are uncorrelated in the former, and correlated in the latter. There are no generally accepted statistical reasons for choosing either method (Harman, 1976). We chose an orthogonal rotation because in principal component analysis the eigenvectors are also orthogonal.

In our principal component analysis, we included all psychological test measures that were presented in table 3. The tactile and motor measures were not included for two reasons. First, it was considered desirable to keep the number of variables limited in comparison to the number of subjects. Second, the purpose of the procedure was to find best representatives of verbal and visuospatial functioning, or of rote versus overlearned abilities and it was anticipated that inclusion of sensory and motor measures would only confuse the picture.
Table 9.

Loadings of 16 selected measures on rotated factors, derived by principal component analysis.

<table>
<thead>
<tr>
<th>VARIABLE</th>
<th>FACTOR 1</th>
<th>FACTOR 2</th>
<th>FACTOR 3</th>
<th>FACTOR 4</th>
</tr>
</thead>
<tbody>
<tr>
<td>VOCAB</td>
<td>.8454</td>
<td>.2487</td>
<td>.0611</td>
<td>.0296</td>
</tr>
<tr>
<td>BLOCKDES</td>
<td>.2143</td>
<td>.7502</td>
<td>.1931</td>
<td>-.1326</td>
</tr>
<tr>
<td>PEA BODY</td>
<td>.9499</td>
<td>.0909</td>
<td>.0639</td>
<td>-.0714</td>
</tr>
<tr>
<td>TOKENS</td>
<td>.6537</td>
<td>.1865</td>
<td>.3552</td>
<td>.3581</td>
</tr>
<tr>
<td>FLUENCY</td>
<td>.2095</td>
<td>.0631</td>
<td>.1703</td>
<td>.8461</td>
</tr>
<tr>
<td>SPATHEM</td>
<td>.1837</td>
<td>.7255</td>
<td>.4239</td>
<td>.1213</td>
</tr>
<tr>
<td>STORY</td>
<td>.4919</td>
<td>.0451</td>
<td>.4560</td>
<td>.4187</td>
</tr>
<tr>
<td>TARGET</td>
<td>-.0830</td>
<td>.8456</td>
<td>-.1517</td>
<td>.3607</td>
</tr>
<tr>
<td>REMIND</td>
<td>-.0642</td>
<td>.0212</td>
<td>.8017</td>
<td>.1989</td>
</tr>
<tr>
<td>MATRIX</td>
<td>.4016</td>
<td>.2775</td>
<td>.6785</td>
<td>.0903</td>
</tr>
<tr>
<td>RIDDLES</td>
<td>.7308</td>
<td>.3133</td>
<td>.1804</td>
<td>.0976</td>
</tr>
<tr>
<td>PROFIG</td>
<td>.2105</td>
<td>.6602</td>
<td>.1123</td>
<td>.0961</td>
</tr>
<tr>
<td>WORDFIND</td>
<td>.7307</td>
<td>.0933</td>
<td>.2884</td>
<td>.4146</td>
</tr>
<tr>
<td>CLOSURE</td>
<td>.4329</td>
<td>.1683</td>
<td>.5494</td>
<td>-.3597</td>
</tr>
<tr>
<td>MATCHFIG</td>
<td>.4644</td>
<td>.4193</td>
<td>-.1382</td>
<td>.1723</td>
</tr>
<tr>
<td>DRAWDES</td>
<td>.2667</td>
<td>.7793</td>
<td>.0752</td>
<td>-.2357</td>
</tr>
</tbody>
</table>
The results from the principal component analysis on the neuropsychological test data are presented in table 9. Four factors with Eigenvalues greater than 1.00 were found, accounting for 71 percent of the variance. The factor scores in table 9 represent loadings on the rotated factors (i.e., after the VARIMAX orthogonal rotation). The factor pattern was inspected for tests that loaded highly on one factor, and considerably lower on all other factors. In addition, the pattern was inspected with the intention of reducing the 16 variables to two smaller sets that would both include a few 'verbal' and a few 'visual' measures, as well as tests requiring overlearned skills and tests of novel problem-solving and recall. With these considerations in mind, FACTOR 1 and FACTOR 2 were found to be most relevant in the context of this study.

VOCAB, PEABODY, TOKENS, RIDDLES, and WORDFIND all loaded highly on FACTOR 1, and much lower on all other factors. All of these tests require some form of processing of verbal material. VOCAB and PEABODY appear adequate representatives of overlearned verbal skills, whereas RIDDLES and WORDFIND both require some problem-solving on the basis of verbal material. BLOCKDES, SPATMEM, TARGET, PROGFIG, and DRAWDES all loaded highly on FACTOR 2, and much lower on all other factors. These tests pertain to visual perceptual, visual constructional, and visual organizational skills. Both SPATMEM and TARGET require recall of new information. Several of these tests (e.g., BLOCKDES, PROGFIG) can also be considered to require problem-solving.
Because of this particular rotated factor pattern, it was decided to select two sets of test measures, one for initial subtype classification, and the other for validation. This selection was performed on the basis of the loadings of each test on FACTOR 1 and FACTOR 2, and because of theoretical considerations. The latter included having in each set equal numbers of 'verbal' and 'visual' variables, and measures of overlearned skills as well as novel problem-solving and recall.

PEABODY, SPATMEM, RIDDLES, and DRAWDES were selected as variables on which to cluster subjects initially. VOCAB, TARGET, WORDFIND, and BLOCKDES were selected as the measures for validation. Thus, two linguistic and two visuospatial measures were included in each group. In addition, each group included a measure of recall of new information, as well as measures of verbal and nonverbal problem-solving. Indeed, the homologous measures in each group appeared very similar to each other from a theoretical point of view. PEABODY and VOCAB both pertain to single word knowledge, and reflect overlearned verbal abilities. SPATMEM and TARGET both involve immediate recall of new visual patterns. RIDDLES and WORDFIND both require identification of a concept on the basis of verbally presented cues, requiring verbal problem-solving. DRAWDES and BLOCKDES are both tasks where the child has to construct a complex pattern on the basis of a model, which requires some form of nonverbal problem-solving.

In summary, on the basis of the principal component analysis four tests were selected for initial subtyping (PEABODY, SPATMEM, RIDDLES, and DRAWDES), and four for validation (VOCAB, TARGET,
WORDPIND, and BLOCKDES). A MANOVA was planned to compare the subtypes on the latter four measures, supplemented with Bonferroni T-tests for individual inter-group comparisons on each of the dependent variables.

We also decided to perform an additional subjective selection of tests on the basis of a division between overlearned versus novel problem-solving skills. The reason for this was to evaluate if there would be any difference between clusters in their ability to complete tasks assessing rote abilities versus tasks requiring new, adaptive strategies. It is important to realize that this selection was based on face validity and clinical experience with the tests, and not on statistical grounds. Tests that were included in the initial subtyping were, of course, excluded from this selection.

VOCAB, FLUENCY, MATCHFIG, and TAPPING were thus selected as tests of overlearned abilities. WORDPIND, REMIND, MATRIX, and PEGS were selected as tests of problem-solving. This selection does not pretend to be exhaustive. Other tests might be added or substituted (e.g., BLOCKDES could also be considered a test of novel problem-solving task). We simply limited the number of tests to the same number as selected for subtype classification and validation. In addition, we attempted to select tests in such a way that for each test of 'overlearned' abilities, there would be another test of 'problem-solving' that could subjectively be considered to require processing of similar task materials but at a higher level of complexity.
VOCAB was considered to assess rote passive vocabulary, whereas WORDFIND supposedly requires verbal reasoning. FLUENCY requires the naming of words, belonging to specific categories, for which the subject may use overlearned semantic strategies. On the other hand, REMIND requires the learning of a list of unrelated words, requiring active encoding and recall of new items of information. MATCHFIG requires simply the matching of geometric line-drawings, whereas the child has to analyze the component parts of a pattern and reason on the basis of analogy on MATRIX. Finally, TAPPING is a relatively simple measure involving finger oscillation, whereas the child has to put 1-cm grooved pegs into a form board on PEGS. The latter requires not only motor dexterity, but also integration of different forms of sensory (e.g., tactile, proprioceptive, visual) information. A MANOVA was planned with the identified clusters as independent variables, and VOCAB, WORDFIND, FLUENCY, REMIND, MATCHFIG, MATRIX, TAPPING, and PEGS as dependent variables. Bonferroni T-tests were planned for comparisons of the clusters on each of the individual dependent variables.

For the PIC-R data, it was considered unnecessary to perform a factor analysis, because the PIC-R provides its own four factor scales: Undisciplined / Poor Self Conrol (Factor I), Social Incompetence (Factor II), Internalization / Somatic Complaints (Factor III), and Cognitive Development (Factor IV). It was decide to use these four factor scales in the cluster analyses, rather than the 12 clinical scales (which would have resulted in a much less satisfactory subject/variable ratio). Since no other
child personality measures were included in this study, validation with personality variables could not be performed for the PIC-R clusters. Validation on the basis of the twelve PIC-R clinical scales would be invalid because the factor scales are based on those scales (i.e., this would not be an independent comparison). However, several physical and social variables were considered to be potentially relevant with regard to the PIC-R profile.

Sick and physically handicapped children are known to have elevations on the PIC-R Psychosis scale, because of their lack of independence (Lachar, 1982; Wirt et al., 1984). Therefore, the following variables were included for validation: Severe Paraplegia, Mild Paraplegia, Bladder Incontinence, Bowel Incontinence, and Seizures. We also considered the fact whether children were cognitively impaired or were going to a special school important with regard to the Achievement and Intellectual Screening scales of the PIC. However, because not all 43 children in this sample were testable with our selected IQ tests, IQ scores were not used for validation. However, the variable School was included. Finally, the variable Broken Home was selected because of its presumed relevance to the Family Relations scale of the PIC.

It is important to realize that all these variables were selected on the basis of subjective reasons only, and not by any statistical criterion. Because of the nominal nature of these variables, nonparametric statistics were indicated for comparison of the clusters. The SYSTAT TABLES program was used to perform separate Chi-Square tests for each selected dependent variable.
(Severe Paraplegia, Mild Paraplegia, Bladder Incontinence, Bowel Incontinence, Seizures, School, and Broken Home). PIC-R clusters formed the grouping (independent) variable in each of these analyses. The Chi-Square test is a non-parametric analysis of the significance of differences in frequencies. For analyses with small frequencies, Guilford (1973) recommends a procedure known as Yates' correction for continuity (which has the effect of making Chi-Square, based on discrete frequencies, more conservative). We therefore used Chi-Square with Yates' correction.

In order to evaluate whether there would be a relationship between psychometric intelligence and PIC-R profile, it was decided to compute Pearson product-moment correlations between the four PIC-R Factor scores and the three IQ scores. Only subjects whose FSIQ exceeded 70 and whose PIC-R profiles were valid (n = 24) were included in this analysis. Since the ADJ scale of the PIC-R is often considered a general screening measure (Wirt et al., 1984), Pearson product-moment correlations between ADJ T-score and IQ scores were also computed.

Initially, we had planned to analyze the academic performance of the different neuropsychological subtypes by means of a MANOVA with subtypes as independent variables, and the Reading, Spelling, and Arithmetic performances on the WRAT-R (converted to T-scores) as dependent measures. However, only 14 of the 30 children included in the subtyping analyses had had more than six months of formal academic education. This number was considered to be too small to allow further analysis.
Table 10.

Categories of independent variables used in discriminant analyses for SCL 90-R data.

<table>
<thead>
<tr>
<th>CATEGORY</th>
<th>VARIABLES INCLUDED (total measures)</th>
</tr>
</thead>
<tbody>
<tr>
<td>2. Non-somatic Child</td>
<td>Age at testing, sex, type of school, general adjustment (7).</td>
</tr>
</tbody>
</table>

To investigate the relationship of specific physical and social variables to the level of distress in the parents, the SAS STEPDISC procedure with BACKWARD elimination was employed to perform stepwise multiple discriminant analyses, and the SAS STEPWISE procedure with BACKWARD elimination to perform stepwise multiple regression analyses.

The manual for the SCL 90-R provides strict criteria for considering a case 'positive' (i.e., indicative of clinically significant distress). These criteria include: a GSI (general stress index) T-score > 62, or at least two of the other clinical scale T-scores > 62. According to these criteria, the 48
available SCL 90-R cases were divided in two groups: Group
POSITIVE included all cases that met the above criteria for being
a 'positive' case (N = 21), and group NEGATIVE included all other
cases (N = 27). The SCL 90-R does not include validity scales;
therefore all cases were included in the analyses.

Three separate STEPDISC discriminant analyses with BACKWARD
elimination were performed, using three separate sets of
variables. The three sets of variables were selected from the
ones presented in table 4. The subdivision in these three sets
was subjective (i.e., not based on a factor analysis). The three
sets of variables are presented in Table 10. Only variables that
were statistically significant at the .05 level were retained in
the discrimination models. Groups (POSITIVE versus NEGATIVE) were
the discrimination criteria in each of these analyses.

In addition, three STEPWISE multiple regression analyses with
BACKWARD elimination were used, with the same three subsets of
variables as independent variables, and the T-score on the GSI
measure of the SCL 90-R as the dependent variable. Only variables
that were statistically significant at the .05 level were retained
in the best regression models. Only these variables were then
entered in a final STEPWISE regression analysis with BACKWARD
elimination, in order to find one final best regression model for
the GSI index of the SCL 90-R.

Finally, it is important with any planned statistical
analysis to make sure one has enough subjects to be able to reject
the null hypothesis. This relates to the 'power' of a statistical
test (which can be interpreted in much the same way as a
reliability coefficient). Because of the fact that all measures in this study were converted to T-scores (with a mean of 50 and a standard deviation of 10), it was decided to compute the power of a one-way test of the significance of a 10-point difference between two mean T-scores.

Glass and Hopkins (1984) provide a formula for determination of the parameter Phi (representing the non-centrality of a distribution), which by means of provided tables can be used to arrive at an estimate of the power of the F test. They suggested that the power should exceed .70 in order for the obtained significant difference between means to be considered reliable.

In the formula that was provided by Glass and Hopkins, Phi was described as the square root of the following computation:

\[ n((M_1 - M_2)^2 + (M_2 - M_3)^2) / k(sd)^2 \]

where \( n \) = the number of cases in each group
\( k \) = the number of groups
\( M_1 \) and \( M_2 \) = the means of the two groups
\( M_d = (M_1 + M_2) / 2 \)
\( sd \) = the standard deviation of the population.

We investigated the power of a one-tailed F-test with regard to analyzing a difference of 10 points in mean T-scores (e.g., 60 and 50) between two groups. Two values of \( n \) were chosen: 15 and 20, and power was computed for each \( n \), with all other parameters being equal. In other words, in both computations, \( k \) was equivalent to 2, \( M_d \) equaled 55, and \( sd \) was 10. The values of \( n = \)
15 and n = 20 were chosen because of the total sample sizes for some of the measures (e.g., a total of 30 children in the HIGH PIQ group, and 43 children with reliable PIC-R data).

Application of the formula and tables of Glass and Hopkins yielded a power of .73 when n was 15, and .86 when n was 20. These values appeared quite acceptable. We therefore considered between 15 and 20 subjects per group adequate to perform statistical inter-group comparisons of means.
CHAPTER III

RESULTS

Summary characteristics of the medical history variables that were documented for this sample are presented in Tables 11 and 12. Means and standard deviations of all medical history variables that were measured as continuous measures are summarized in Table 11. Sample frequencies and proportions of applicability of all medical history variables that were measured in terms of presence vs absence (including dummy variables) are presented in Table 12. Average T-scores for all test measures are presented in Table 13. All statistical analyses were done on the basis of T-scores. The only exception to this is the t-test on the significance of VIQ-PIQ discrepancy, for which the conventional raw IQ scores were used. The n values in Table 13 reflect the number of subjects for whom reliable and valid results were obtained on each measure.

Stepwise Discriminant Analyses: Discriminating between LOW versus HIGH VIQ and PIQ groups with medical history variables.

For details regarding the measurement and selection of the respective variable groups, the reader is referred to Tables 3 and 5. We will first report on the results of six sets of initial stepwise discriminant analyses (SDA), as described in Table 8.
As was outlined in the 'method' section, we first performed six separate SDAs with respectively demographics, motor symptoms, spinal cord symptoms, other symptoms, pathology variables, and treatment variables. Separate analyses were performed for VIQ and PIQ, with the criterion being the HIGH and LOW groups for each of these measures. The variables that remained in the best discrimination models with each of these six analyses were then

Table 11.

Means and standard deviations for selected medical history variables.

<table>
<thead>
<tr>
<th>VARIABLE</th>
<th>MEAN</th>
<th>SD</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age at first shunt surgery (days)</td>
<td>68.21</td>
<td>89.17</td>
</tr>
<tr>
<td>Occipito-frontal circumference (mm)</td>
<td>401.13</td>
<td>59.92</td>
</tr>
<tr>
<td>Shunt revisions</td>
<td>2.48</td>
<td>3.87</td>
</tr>
<tr>
<td>Shunt infections</td>
<td>.25</td>
<td>.57</td>
</tr>
<tr>
<td>Age at testing (months)</td>
<td>78.90</td>
<td>14.15</td>
</tr>
</tbody>
</table>
Table 12.

Frequencies (n) and sample proportions (%) of occurrence of medical history variables.

<table>
<thead>
<tr>
<th>VARIABLE</th>
<th>n</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>DEMOGRAPHIC VARIABLES</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Male sex</td>
<td>29</td>
<td>60.42</td>
</tr>
<tr>
<td>Female sex</td>
<td>19</td>
<td>39.58</td>
</tr>
<tr>
<td>White race</td>
<td>36</td>
<td>75.00</td>
</tr>
<tr>
<td>Black race</td>
<td>10</td>
<td>20.83</td>
</tr>
<tr>
<td>Other race</td>
<td>2</td>
<td>4.17</td>
</tr>
<tr>
<td>Right-handedness</td>
<td>33</td>
<td>68.75</td>
</tr>
<tr>
<td>Left-handedness</td>
<td>15</td>
<td>31.25</td>
</tr>
<tr>
<td>Socio-Economic Class I</td>
<td>3</td>
<td>6.25</td>
</tr>
<tr>
<td>Socio-Economic Class II</td>
<td>8</td>
<td>16.67</td>
</tr>
<tr>
<td>Socio-Economic Class III</td>
<td>11</td>
<td>22.92</td>
</tr>
<tr>
<td>Socio-Economic Class IV</td>
<td>19</td>
<td>39.58</td>
</tr>
<tr>
<td>Socio-Economic Class V</td>
<td>7</td>
<td>14.58</td>
</tr>
</tbody>
</table>
Table 12 (continued).

**Frequencies (n) and sample proportions (%) of occurrence of medical history variables.**

<table>
<thead>
<tr>
<th>VARIABLE</th>
<th>n</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>MOTOR SYMPTOM VARIABLES</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>No motor problems</td>
<td>13</td>
<td>27.08</td>
</tr>
<tr>
<td>Fine motor coordination problems</td>
<td>6</td>
<td>12.50</td>
</tr>
<tr>
<td>Truncal ataxia / balance problems</td>
<td>3</td>
<td>6.25</td>
</tr>
<tr>
<td>Gait problems</td>
<td>6</td>
<td>12.50</td>
</tr>
<tr>
<td>Mild paraplegia</td>
<td>3</td>
<td>6.25</td>
</tr>
<tr>
<td>Severe paraplegia</td>
<td>17</td>
<td>35.42</td>
</tr>
<tr>
<td><strong>SENSORY SYMPTOM VARIABLES</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>No myelomeningocele sac</td>
<td>34</td>
<td>70.83</td>
</tr>
<tr>
<td>Lumbo-sacral / sacral sac</td>
<td>6</td>
<td>12.50</td>
</tr>
<tr>
<td>Lumbar sac</td>
<td>5</td>
<td>10.42</td>
</tr>
<tr>
<td>Thoraco-lumbar sac</td>
<td>2</td>
<td>4.17</td>
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<tr>
<td>Occipito-cervical sac</td>
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<td>2.08</td>
</tr>
</tbody>
</table>
Table 12 (continued)

Frequencies (n) and sample proportions (%) of occurrence of medical history variables.

<table>
<thead>
<tr>
<th>VARIABLE</th>
<th>n</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

SENSORY SYMPTOM VARIABLES (continued)

| No sensory loss              | 35 | 72.92 |
| Sensory loss T12 and up      | 2  | 4.17  |
| Sensory loss L1 - L4         | 8  | 16.67 |
| Sensory loss L5 - S5         | 3  | 6.25  |

OTHER SYMPTOM VARIABLES

| Prenatal & perinatal problems | 25 | 31.25 |
| Neonatal & infancy problems   | 29 | 60.42 |
| Lacunar skull deformities     | 12 | 25.00 |
| Ocular defects                | 22 | 45.83 |
| Seizures                      | 13 | 27.08 |
Table 12 (continued)

Frequencies \((n)\) and sample proportions \((\%)\) of occurrence of medical history variables.

<table>
<thead>
<tr>
<th>VARIABLE</th>
<th>n</th>
<th>(%)</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>PATHOLOGY VARIABLES</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Intra-ventricular obstruction</td>
<td>34</td>
<td>70.83</td>
</tr>
<tr>
<td>Extra-ventricular obstruction</td>
<td>14</td>
<td>29.17</td>
</tr>
<tr>
<td>Aqueductal lesion</td>
<td>15</td>
<td>31.25</td>
</tr>
<tr>
<td>Arnold-Chiari malformation</td>
<td>9</td>
<td>18.75</td>
</tr>
<tr>
<td>Dandy-Walker syndrome</td>
<td>4</td>
<td>8.33</td>
</tr>
<tr>
<td>Bleeds</td>
<td>12</td>
<td>25.00</td>
</tr>
<tr>
<td>Cerebral Infection</td>
<td>2</td>
<td>4.17</td>
</tr>
<tr>
<td>Cyst</td>
<td>8</td>
<td>16.67</td>
</tr>
<tr>
<td><strong>TREATMENT VARIABLES</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Left hemisphere shunt</td>
<td>11</td>
<td>22.92</td>
</tr>
<tr>
<td>Right hemisphere shunt</td>
<td>48</td>
<td>100.00</td>
</tr>
</tbody>
</table>
Table 13.

**Performance (in T-scores) on psychological test measures, and nature of original score on which T-scores were based.**

<table>
<thead>
<tr>
<th>VARIABLE</th>
<th>NATURE OF SCORE</th>
<th>N</th>
<th>M</th>
<th>SD</th>
</tr>
</thead>
<tbody>
<tr>
<td>VIQ</td>
<td>WISC-R/WPPSI Verbal IQ</td>
<td>30</td>
<td>51.56</td>
<td>9.35</td>
</tr>
<tr>
<td>PIQ</td>
<td>WISC-R/WPPSI Performance IQ</td>
<td>30</td>
<td>45.56</td>
<td>8.82</td>
</tr>
<tr>
<td>FSIQ</td>
<td>WISC-R/WPPSI Full-Scale IQ</td>
<td>30</td>
<td>48.38</td>
<td>8.73</td>
</tr>
<tr>
<td>WRAT-R READ.</td>
<td>Standard Score</td>
<td>14</td>
<td>49.42</td>
<td>17.64</td>
</tr>
<tr>
<td>WRAT-R SPEL.</td>
<td>Standard Score</td>
<td>14</td>
<td>47.13</td>
<td>15.58</td>
</tr>
<tr>
<td>WRAT-R ARIT.</td>
<td>Standard Score</td>
<td>14</td>
<td>43.33</td>
<td>12.30</td>
</tr>
<tr>
<td>PEABODY</td>
<td>Standard Score</td>
<td>30</td>
<td>49.44</td>
<td>9.96</td>
</tr>
<tr>
<td>VOCAB</td>
<td>Scaled Score</td>
<td>30</td>
<td>51.78</td>
<td>9.29</td>
</tr>
<tr>
<td>TOKENS</td>
<td>Total items correct</td>
<td>30</td>
<td>45.74</td>
<td>12.77</td>
</tr>
<tr>
<td>FLUENCY</td>
<td>Total words in four trials</td>
<td>30</td>
<td>53.70</td>
<td>10.30</td>
</tr>
<tr>
<td>SPATHEM</td>
<td>Scaled Score</td>
<td>30</td>
<td>44.00</td>
<td>8.94</td>
</tr>
<tr>
<td>STORY</td>
<td>Total items correct</td>
<td>30</td>
<td>49.00</td>
<td>8.29</td>
</tr>
<tr>
<td>TARGET</td>
<td>Total items correct</td>
<td>30</td>
<td>36.40</td>
<td>10.87</td>
</tr>
<tr>
<td>REMIND</td>
<td>Consistent long-term recall</td>
<td>30</td>
<td>44.30</td>
<td>8.18</td>
</tr>
<tr>
<td>MATRIX</td>
<td>Scaled Score</td>
<td>30</td>
<td>51.22</td>
<td>6.81</td>
</tr>
<tr>
<td>RIDDLES</td>
<td>Standard Score</td>
<td>30</td>
<td>50.24</td>
<td>9.13</td>
</tr>
<tr>
<td>PROGFIG</td>
<td>Time for completion</td>
<td>30</td>
<td>47.90</td>
<td>11.19</td>
</tr>
<tr>
<td>WORDFIND</td>
<td>Total items correct</td>
<td>30</td>
<td>49.62</td>
<td>10.79</td>
</tr>
</tbody>
</table>
Table 13 (continued).

Performance (in T-scores) on psychological test measures, and nature of original score on which T-scores were based.

<table>
<thead>
<tr>
<th>VARIABLE</th>
<th>NATURE OF SCORE</th>
<th>n</th>
<th>M</th>
<th>SD</th>
</tr>
</thead>
<tbody>
<tr>
<td>CLOSURE</td>
<td>Scaled Score</td>
<td>30</td>
<td>45.33</td>
<td>9.08</td>
</tr>
<tr>
<td>BLOCKDES</td>
<td>Scaled Score</td>
<td>30</td>
<td>46.00</td>
<td>9.72</td>
</tr>
<tr>
<td>MATCHFIG</td>
<td>Time for completion</td>
<td>30</td>
<td>51.10</td>
<td>7.12</td>
</tr>
<tr>
<td>DRAWDES</td>
<td>Total score on nine items</td>
<td>30</td>
<td>42.26</td>
<td>11.24</td>
</tr>
<tr>
<td>FSsymbol</td>
<td>(Left + Right errors)/2</td>
<td>30</td>
<td>52.54</td>
<td>8.52</td>
</tr>
<tr>
<td>PEGS</td>
<td>(Left + Right time)/2</td>
<td>30</td>
<td>34.64</td>
<td>23.63</td>
</tr>
<tr>
<td>Fagnos</td>
<td>(Left + Right errors)/2</td>
<td>30</td>
<td>46.36</td>
<td>11.08</td>
</tr>
<tr>
<td>Tapping</td>
<td>(Left + Right total)/2</td>
<td>30</td>
<td>39.75</td>
<td>12.44</td>
</tr>
<tr>
<td>PIC-R I</td>
<td>Regular T-score</td>
<td>43</td>
<td>56.23</td>
<td>10.43</td>
</tr>
<tr>
<td>PIC-R II</td>
<td>Regular T-score</td>
<td>43</td>
<td>54.98</td>
<td>10.72</td>
</tr>
<tr>
<td>PIC-R III</td>
<td>Regular T-score</td>
<td>43</td>
<td>58.63</td>
<td>12.40</td>
</tr>
<tr>
<td>PIC-R IV</td>
<td>Regular T-score</td>
<td>43</td>
<td>79.86</td>
<td>28.92</td>
</tr>
<tr>
<td>PIC-R L</td>
<td>Regular T-score</td>
<td>43</td>
<td>52.26</td>
<td>9.10</td>
</tr>
<tr>
<td>PIC-R F</td>
<td>Regular T-score</td>
<td>43</td>
<td>71.14</td>
<td>19.67</td>
</tr>
<tr>
<td>PIC-R DEF</td>
<td>Regular T-score</td>
<td>43</td>
<td>46.33</td>
<td>10.59</td>
</tr>
<tr>
<td>PIC-R ADJ</td>
<td>Regular T-score</td>
<td>43</td>
<td>57.65</td>
<td>11.83</td>
</tr>
</tbody>
</table>
Table 13 (continued).

Performance (in T-scores) on psychological test measures, and nature of original score on which T-scores were based.

<table>
<thead>
<tr>
<th>VARIABLE</th>
<th>NATURE OF SCORE</th>
<th>n</th>
<th>M</th>
<th>SD</th>
</tr>
</thead>
<tbody>
<tr>
<td>PIC-R ACH</td>
<td>Regular T-score</td>
<td>43</td>
<td>62.47</td>
<td>17.43</td>
</tr>
<tr>
<td>PIC-R IS</td>
<td>Regular T-score</td>
<td>43</td>
<td>80.00</td>
<td>29.20</td>
</tr>
<tr>
<td>PIC-R DVL</td>
<td>Regular T-score</td>
<td>43</td>
<td>68.16</td>
<td>20.45</td>
</tr>
<tr>
<td>PIC-R SOM</td>
<td>Regular T-score</td>
<td>43</td>
<td>59.61</td>
<td>15.58</td>
</tr>
<tr>
<td>PIC-R D</td>
<td>Regular T-score</td>
<td>43</td>
<td>53.33</td>
<td>9.32</td>
</tr>
<tr>
<td>PIC-R FAM</td>
<td>Regular T-score</td>
<td>43</td>
<td>50.54</td>
<td>9.98</td>
</tr>
<tr>
<td>PIC-R DLQ</td>
<td>Regular T-score</td>
<td>43</td>
<td>51.33</td>
<td>12.08</td>
</tr>
<tr>
<td>PIC-R WDL</td>
<td>Regular T-score</td>
<td>43</td>
<td>54.81</td>
<td>11.63</td>
</tr>
<tr>
<td>PIC-R ANX</td>
<td>Regular T-score</td>
<td>43</td>
<td>52.54</td>
<td>10.01</td>
</tr>
<tr>
<td>PIC-R PSY</td>
<td>Regular T-score</td>
<td>43</td>
<td>74.63</td>
<td>23.77</td>
</tr>
<tr>
<td>PIC-R HPR</td>
<td>Regular T-score</td>
<td>43</td>
<td>51.28</td>
<td>12.09</td>
</tr>
<tr>
<td>PIC-R SSK</td>
<td>Regular T-score</td>
<td>43</td>
<td>51.51</td>
<td>10.45</td>
</tr>
<tr>
<td>SCL 90-R GSI</td>
<td>Regular T-score</td>
<td>48</td>
<td>55.25</td>
<td>10.27</td>
</tr>
<tr>
<td>SCL 90-R PST</td>
<td>Regular T-score</td>
<td>48</td>
<td>55.00</td>
<td>10.24</td>
</tr>
<tr>
<td>SCL 90-R PSDI</td>
<td>Regular T-score</td>
<td>48</td>
<td>51.75</td>
<td>8.78</td>
</tr>
<tr>
<td>SCL 90-R SOM</td>
<td>Regular T-score</td>
<td>48</td>
<td>52.92</td>
<td>9.95</td>
</tr>
<tr>
<td>SCL 90-R OBS</td>
<td>Regular T-score</td>
<td>48</td>
<td>54.88</td>
<td>8.85</td>
</tr>
<tr>
<td>SCL 90-R INT</td>
<td>Regular T-score</td>
<td>48</td>
<td>56.15</td>
<td>9.04</td>
</tr>
<tr>
<td>SCL 90-R DEP</td>
<td>Regular T-score</td>
<td>48</td>
<td>56.58</td>
<td>9.87</td>
</tr>
</tbody>
</table>
Table 13 (continued).

**Performance (in T-scores) on psychological test measures, and nature of original score on which T-scores were based.**

<table>
<thead>
<tr>
<th>VARIABLE</th>
<th>NATURE OF SCORE</th>
<th>n</th>
<th>M</th>
<th>SD</th>
</tr>
</thead>
<tbody>
<tr>
<td>SCL 90-R ANX</td>
<td>Regular T-score</td>
<td>48</td>
<td>51.98</td>
<td>9.44</td>
</tr>
<tr>
<td>SCL 90-R HOS</td>
<td>Regular T-score</td>
<td>48</td>
<td>56.17</td>
<td>9.25</td>
</tr>
<tr>
<td>SCL 90-R PHO</td>
<td>Regular T-score</td>
<td>48</td>
<td>49.04</td>
<td>7.96</td>
</tr>
<tr>
<td>SCL 90-R PAR</td>
<td>Regular T-score</td>
<td>48</td>
<td>51.71</td>
<td>9.32</td>
</tr>
<tr>
<td>SCL 90-R PSY</td>
<td>Regular T-score</td>
<td>48</td>
<td>54.00</td>
<td>9.34</td>
</tr>
</tbody>
</table>

entered into two final SDAs (one for VIQ, one for PIQ) to establish final, overall best sets of discrimination variables.

Nine demographic variables were entered in the first SDA: Age at testing, Sex, Race (two dummy levels: Black, White), Handedness, and Socio-Economic Status (four dummy levels: I, II, III, IV). None of these variables proved to be statistically significant for VIQ. However, Handedness (Partial $R^2 = .0982$, $F(1,47) = 5.000$, $p = .0301$) was found to be a statistically significant discrimination variable for PIQ.

Five motor symptom variables were entered in the second SDA, in dummy levels 'Fine motor coordination problems', 'Truncal
ataxia and/or balance Problems', 'Gait Problems', 'Mild Paraplegia', and 'Severe Paraplegia'. 'Severe Paraplegia' was the single significant variable in the models for VIQ (Partial $R^2 = .1603$, $F(1,47) = 8.784$, $p = .0048$), and PIQ (Partial $R^2 = .1732$, $F(1,47) = 9.635$, $p = .0003$).

Seven spinal cord symptom variables were entered in the third SDA: Vertebral Location of the Myelomeningocele Sac (four dummy levels: 'Lumbo-Sacral and Sacral', 'Lumbar', 'Thoraco-Lumbar and Thoraco-Lumbo-Sacral', and 'Occipito-Cervical and Cervical'), and Sensory Level at Birth (three dummy variables: 'L5-S5', 'L1-L4', and 'T12 and up'). None of these variables discriminated significantly between HIGH and LOW groups for either VIQ or PIQ.

Five other symptom variables were entered in the fourth SDA: Prenatal and Perinatal Problems, Neonatal and Infancy Problems, Lacunar Skull Deformities, Ocular Defects, and Seizures. A three-variable model emerged for VIQ, with Neonatal and Infancy Problems (Partial $R^2 = .1023$, $F(1,47) = 5.012$, $p = .0303$), Ocular Defects (Partial $R^2 = .1434$, $F(1,47) = 7.366$, $p = .0095$), and Lacunar Skull Deformities (Partial $R^2 = .0925$, $F(1,47) = 4.485$, $p = .0399$) as significant discrimination variables. A two-variable model was found for PIQ, including Neonatal and Infancy Problems (Partial $R^2 = .1595$, $F(1,47) = 8.540$, $p = .0054$) and Ocular Defects (Partial $R^2 = .2356$, $F(1,47) = 13.867$, $p = .0005$).

Seven pathology variables were entered in the fifth SDA: Type of Hydrocephalus, Aqueductal Lesion, Arnold-Chiari Malformation, Dandy-Walker Syndrome, Bleeds, Cerebral Infection, and Cyst. A five-variable model emerged for VIQ with discrimination variables
Aqueductal Lesion (Partial R2 = .1164, F (1,47) = 5.402, p = .0252), Arnold-Chiari Malformation (Partial R2 = .1252, F (1,47) = 5.866, p = .0199), Bleeds (Partial R2 = .1721, F (1,47) = 8.521, p = .0057), Cerebral Infection (Partial R2 = .1204, F (1,47) = 5.611, p = .0226), and Cyst (Partial R2 = .1369, F (1,47) = 6.505, p = .0146). No significant discrimination variables were found for PIQ.

Six treatment variables were entered in the sixth SDA: Age at Treatment, Left Shunt, Right Shunt, Head Size, Shunt Revisions, and Shunt Infections. No significant variables were found for either VIQ or FSIQ.

After having performed these separate initial SDAs for each of the categories of medical history variables, the results were used to analyze what would be the overall best discrimination models for VIQ and PIQ. To establish the best set of discrimination variables over all of the documented medical history variables, two final SDAs (one for VIQ, and one for PIQ) were performed, using only those variables that had remained as significant discriminators in the six initial SDAs. For VIQ, this included Severe Paraplegia, Neonatal and Infancy Problems, Ocular Defects, Lacunar Skull Deformities, Aqueductal Lesion, Arnold-Chiari Malformation, Bleeds, Cerebral Infection, and Cyst. For PIQ, this included Handedness, Severe paraplegia, Neonatal and Infancy Problems, and Ocular Defects. The resulting best discrimination models are presented in Table 14.

As can be seen in Table 14, a three-variable best discrimination model emerged for VIQ. Presence of each of these
three variables (Neonatal and Infancy Problems, Ocular Defects, and Lacunar Skull Deformities) in the child's medical history made it more likely that the child would be in the LOW VIQ group. This model correctly classified 40 of the 48 cases in the sample (83.33%). Two of the 16 cases in group LOW VIQ were misassigned to group HIGH VIQ, and 6 of the 32 cases in group HIGH VIQ were misclassified in group LOW VIQ.

Table 14.

<table>
<thead>
<tr>
<th>VARIABLES</th>
<th>R^2</th>
<th>F</th>
<th>df</th>
<th>P</th>
</tr>
</thead>
<tbody>
<tr>
<td>Neonatal and Infancy Problems</td>
<td>.1023</td>
<td>5.012</td>
<td>1,47</td>
<td>.0303</td>
</tr>
<tr>
<td>VIQ Ocular Defects</td>
<td>.1434</td>
<td>7.366</td>
<td>1,47</td>
<td>.0095</td>
</tr>
<tr>
<td>Lacunar Skull Deformities</td>
<td>.0925</td>
<td>4.485</td>
<td>1,47</td>
<td>.0399</td>
</tr>
<tr>
<td>PIQ Neonatal and Infancy Problems</td>
<td>.1595</td>
<td>8.540</td>
<td>1,47</td>
<td>.0054</td>
</tr>
<tr>
<td>Ocular Defects</td>
<td>.2356</td>
<td>14.867</td>
<td>1,47</td>
<td>.0005</td>
</tr>
</tbody>
</table>
A two-variable best discrimination model was found for PIQ. Presence of each of these two variables (Neonatal and Infancy Problems, and Ocular Defects) in the child's medical history made it more likely that the child would be in the LOW PIQ group. This model correctly classified 42 of the 48 cases in the sample (87.50%). Two of the 18 subjects from group LOW PIQ were misclassified, and four of the 26 subjects from group HIGH PIQ were misclassified.

Verbal - Performance IQ discrepancies.

The average raw IQ scores on the WISC-R (N = 15) or WPPSI (N = 15) of the subjects in the sample who attained VIQ, PIQ, and FSIQ levels above 70 (i.e., the HIGH PIQ group) are presented in Table 15. A paired-samples t-test revealed that the difference between the VIQ and PIQ measures for this total group was statistically significant (t (29) = 3.848, p < .001).

Correlations between sensory-motor performance and intelligence.

The correlations between sensory and motor measures (based on the average of the two T-scores for each hand), and the VIQ, PIQ, and FSIQ measures for all subjects with FSIQ > 70 are presented in Table 16. All correlations involving TAPPING and PEGS are Pearson product-correlations, those involving FSsymbol and FAGNOS (variables with a skewed distribution) represent Kendall Tau coefficients. The latter index does not rest on the assumption of
Table 15.

Means and standard deviations of IQ scores of subjects with FSIQ > 70.

<table>
<thead>
<tr>
<th>VERBAL IQ</th>
<th>PERFORMANCE IQ</th>
<th>FULL-SCALE IQ</th>
</tr>
</thead>
<tbody>
<tr>
<td>m</td>
<td>SD</td>
<td>m</td>
</tr>
<tr>
<td>102.33</td>
<td>14.02</td>
<td>93.33</td>
</tr>
</tbody>
</table>

a normal distribution and represents a ranking-method correlation coefficient. As can be seen in this Table, the only significant correlation was between the measures PIQ and PEGS, with PEGS accounting for about 25% of the variance in PIQ (better performance on PEGS being associated with higher PIQ scores).

For some of the psychological test measures it might also be argued that they have a strong sensory-motor component. For example, manipulation of a pencil, visual inspection of detail, and fine motor movements, are all to some extent involved in TARGET, PROGFIG, and DRAWDES. It was therefore decided to compute correlations between these measures and the above-described tactile and motor measures. The choice of these three
Table 16.

**Correlations between sensory and motor measures, and IQ scores.**

<table>
<thead>
<tr>
<th></th>
<th>VIQ</th>
<th>PIQ</th>
<th>FSIQ</th>
</tr>
</thead>
<tbody>
<tr>
<td>FSYMBOL</td>
<td>0.1664</td>
<td>-0.0221</td>
<td>0.0441</td>
</tr>
<tr>
<td>FAGNOS</td>
<td>0.0970</td>
<td>0.2264</td>
<td>0.1967</td>
</tr>
<tr>
<td>TAPPING</td>
<td>-0.1006</td>
<td>0.0642</td>
<td>-0.0202</td>
</tr>
<tr>
<td>PEGS</td>
<td>-0.0800</td>
<td>0.4952 (*)</td>
<td>0.2147</td>
</tr>
</tbody>
</table>

(*) $p < .006$

Table 17.

**Correlations between sensory and motor measures, and selected neuropsychological test scores.**

<table>
<thead>
<tr>
<th></th>
<th>TARGET</th>
<th>PROFIG</th>
<th>DRAWDES</th>
</tr>
</thead>
<tbody>
<tr>
<td>FSYMBOL</td>
<td>0.0001</td>
<td>0.0949</td>
<td>-0.0551</td>
</tr>
<tr>
<td>FAGNOS</td>
<td>0.2321</td>
<td>0.0037</td>
<td>0.0667</td>
</tr>
<tr>
<td>TAPPING</td>
<td>0.4122 (*)</td>
<td>-0.0273</td>
<td>0.1123</td>
</tr>
<tr>
<td>PEGS</td>
<td>0.4164 (*)</td>
<td>0.2724</td>
<td>0.4093 (*)</td>
</tr>
</tbody>
</table>

(*) $p < .05$
neuropsychological measures was, of course, subjective (i.e., sensory and motor functions may also influence performance on other tests). The selection was merely made as an example of the relation of tactile and motor variables to performance on complex visual-spatial tasks, without the pretention of being exhaustive. The results are presented in Table 17. Again, all correlations are of the Pearson product-moment type, with the exception of those involving FSYMBOL and FAGNOS (Kendall Tau).

As can be seen in Table 17, there was a significant correlation between TAPPING and TARGET, explaining about 17% of the variance (better faster performance on TAPPING being associated with fewer errors on TARGET). PEGS had a significant correlation with both TARGET and DRAWDES, accounting for approximately 17% of the variance in both cases. Faster performance on PEGS was associated with better scores on both TARGET and DRAWDES.

Cluster analysis on selected neuropsychological measures.

Details regarding the clustering process that was performed with the four selected neuropsychological variables are presented in Table 18. The hierarchical tree dendogram from the Average Linkage analysis is presented in Figure 1.
Table 18.

Cluster solutions of three different hierarchical agglomerative techniques, with clustering variables PEABODY, SPATHEM, RIDDLES, and DRAWDES, and squared Euclidean distance similarity measure.

<table>
<thead>
<tr>
<th>n</th>
<th>AVERAGE</th>
<th></th>
<th>CENTROID</th>
<th></th>
<th>WARD</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>FREQ</td>
<td>SPRSQ</td>
<td>FREQ</td>
<td>SPRSQ</td>
<td>FREQ</td>
<td>SPRSQ</td>
</tr>
<tr>
<td>10</td>
<td>4</td>
<td>.0209</td>
<td>2</td>
<td>.0119</td>
<td>4</td>
<td>.0209</td>
</tr>
<tr>
<td>9</td>
<td>3</td>
<td>.0167</td>
<td>4</td>
<td>.0209</td>
<td>5</td>
<td>.0212</td>
</tr>
<tr>
<td>8</td>
<td>16</td>
<td>.0586</td>
<td>3</td>
<td>.0167</td>
<td>10</td>
<td>.0268</td>
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<td>.0271</td>
<td>17</td>
<td>.0271</td>
<td>4</td>
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</tr>
<tr>
<td>6</td>
<td>2</td>
<td>.0199</td>
<td>6</td>
<td>.0414</td>
<td>14</td>
<td>.0385</td>
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<td>6</td>
<td>.0414</td>
<td>20</td>
<td>.0807</td>
<td>5</td>
<td>.0501</td>
</tr>
<tr>
<td>4</td>
<td>20</td>
<td>.0807</td>
<td>2</td>
<td>.0199</td>
<td>17</td>
<td>.0585</td>
</tr>
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<td>3</td>
<td>22</td>
<td>.0737</td>
<td>22</td>
<td>.0737</td>
<td>8</td>
<td>.0963</td>
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<td>8</td>
<td>.0895</td>
<td>8</td>
<td>.0895</td>
<td>22</td>
<td>.1043</td>
</tr>
<tr>
<td>1</td>
<td>30</td>
<td>.4215</td>
<td>30</td>
<td>.4215</td>
<td>30</td>
<td>.4215</td>
</tr>
</tbody>
</table>

Abbreviations: n = Number of clusters

AVERAGE = Average Linkage method

CENTROID = Centroid Linkage method

WARD = Ward's Minimum Variance method

FREQ = Number of subjects of new cluster

SPRSQ = Semi-partial R2 of new cluster
Figure 1.

Average Linkage cluster solution with PEABODY, SPANEM, RIDDLES, and DRAWDES as clustering variables, and squared Euclidean distance as similarity measure.
Inspection of Table 18 reveals that a loss of more than 10\% of the accounted-for variance did not occur until the last two clusters were merged. This pattern was evident for all three clustering methods. A two-cluster solution was therefore considered, with 22 subjects in one cluster, and eight subjects in the other. Inspection of the dendogram in Figure 1 suggested that this would be compatible with the distribution of the subjects in Euclidean space. Further inspection of the cluster membership of specific cases revealed that the AVERAGE, CENTROID, and WARD analyses were in 100\% agreement with regard to the assignment of individual subjects to two different clusters. Therefore, a non-hierarchical K-Means iterative partitioning analysis was performed, with K specified as 2. This resulted in the reassignment of only case (3.33\%).

On the basis of these findings, the two-cluster Average Linkage solution was considered to be reliable. The average T-scores of the two clusters on the initial subtyping and validation variables are presented in Tables 19 and 20. The average IQ scores (represented in the conventional manner) are presented in Table 21.

The large difference in cluster size, and especially the small n in the smaller cluster raised some concern about the feasibility of performing statistical analyses of the differences in performance between the clusters. As was mentioned above (see Method section), about 15 subjects were felt to be needed in each group to be able to reject the Null hypothesis. Any test of the
differences in performance between the two neuropsychological clusters found in this study should therefore be considered a most conservative estimate of the importance of those differences. We chose to perform MANOVA's to evaluate the presence of an main effect of cluster membership. Individual comparisons between means of the two clusters on each dependent measure were done by means of Bonferroni T-tests.

Table 19.

Results of Average Linkage solution with two neuropsychological clusters for selected neuropsychological measures included in classification, and minimum significant difference for Bonferroni (BON) T-test (significant comparisons indicated by *).

<table>
<thead>
<tr>
<th></th>
<th>CLUSTER 1 (n = 8)</th>
<th>CLUSTER 2 (n = 22)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>M</td>
<td>SD</td>
</tr>
<tr>
<td>PEABODY</td>
<td>61.17</td>
<td>9.53</td>
</tr>
<tr>
<td>SPATHRM</td>
<td>51.25</td>
<td>7.96</td>
</tr>
<tr>
<td>RIDDLES</td>
<td>60.33</td>
<td>8.20</td>
</tr>
<tr>
<td>DRAWDRES</td>
<td>54.25</td>
<td>9.47</td>
</tr>
</tbody>
</table>
As can be seen in Table 19, the performances of Cluster 2 on the clustering variables were, on the average, about one standard deviation or more below those of Cluster 1, with specifically impaired performance on one of the visuospatial measures (DRAWDES). A MANOVA was performed with Clusters (two) as the independent variable and Tests (PEABODY, SPATHMEM, Riddles, and DRAWDES) as dependent variables. A significant main effect of

Table 20.

T-scores of two neuropsychological clusters on selected neuropsychological measures not included in classification, and minimum significant difference for Bonferroni (B)N T-test (significant comparisons indicated by *).

<table>
<thead>
<tr>
<th></th>
<th>CLUSTER 1 (n = 8)</th>
<th>CLUSTER 2 (n = 22)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>M</td>
<td>SD</td>
</tr>
<tr>
<td>VOCAB</td>
<td>61.67</td>
<td>11.27</td>
</tr>
<tr>
<td>TARGET</td>
<td>40.26</td>
<td>6.50</td>
</tr>
<tr>
<td>WORDFIND</td>
<td>56.96</td>
<td>9.57</td>
</tr>
<tr>
<td>BLOCKDES</td>
<td>53.33</td>
<td>6.17</td>
</tr>
</tbody>
</table>
Clusters was found ($F(4,25) = 18.92$, $p < .0001$). Bonferroni comparisons revealed that the performance of Cluster 1 was significantly better than that of Cluster 2 on all dependent variables.

Inspection of Table 20 reveals that differences of about one standard deviation between the average level of performance of Cluster 1 and Cluster 2 also occurred on the variables that were selected for validation. The difference was less explicit for TARGET, which proved to be a relatively poor score in both clusters. A MANOVA was performed with Clusters (two) as the

Table 21.

**Average IQ scores of two neuropsychological clusters, and minimum significant difference for Bonferroni (BON) T-test (significant comparisons indicated by *)**.

<table>
<thead>
<tr>
<th></th>
<th>CLUSTER 1 ($n = 8$)</th>
<th>CLUSTER 2 ($n = 22$)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>$M$</td>
<td>$SD$</td>
</tr>
<tr>
<td>VERBAL IQ</td>
<td>118.00</td>
<td>13.50</td>
</tr>
<tr>
<td>PERFORMANCE IQ</td>
<td>107.63</td>
<td>8.07</td>
</tr>
<tr>
<td>FULL SCALE I.Q</td>
<td>113.63</td>
<td>9.87</td>
</tr>
</tbody>
</table>
Independent variable and Tests (VOCAB, TARGET, WORDFIND, and BLOCKDES) as dependent variables. A significant main effect of Clusters was found ($F(4,25) = 5.569, p < .002$). Bonferroni comparisons revealed that the performance of Cluster 1 was significantly better than that of Cluster 2 on VOCAB, WORDFIND, and BLOCKDES, but there was no significant difference on TARGET.

Inspection of Table 21 reveals that the average IQ scores of Cluster 1 were also all at least one standard deviation higher than those of Cluster 2. A MANOVA was performed with Clusters (two) as the independent variable and IQ scores (three) as dependent variables. The main effect of Clusters was significant ($F(3,26) = 12.470, p < .001$). Bonferroni comparisons revealed that Cluster 1 had significantly higher scores than Cluster 2 on all three IQ measures.

The average T-scores of the two clusters on the variables that were subjectively selected as measures of 'overlearned' skills on one hand and 'novel problem-solving' abilities on the other are presented in Table 22. A MANOVA was performed with Clusters (two) as the independent variable and Tests (VOCAB, FLUENCY, MATCHFIG, TAPPING, WORDFIND, REMIND, MATRIX, and PEGS) as dependent variables. The main effect of Clusters was significant ($F(8,21) = 4.215, p < .004$). Bonferroni comparisons revealed that Cluster 1 obtained significantly higher scores than Cluster 2 on two of four measures 'overlearned' skills (VOCAB and MATCHFIG), as well as on two of four measures of 'novel problem-solving' (WORDFIND and MATRIX). No significant differences were found on the other dependent measures.
Table 22.

**T-scores of two neuropsychological clusters on selected neuropsychological measures of overlearned skills and of novel problem-solving, and minimum significant difference for Bonferroni (BON) T-test (significant comparisons indicated by *).**

<table>
<thead>
<tr>
<th></th>
<th>Cluster 1 (n = 8)</th>
<th>Cluster 2 (n = 22)</th>
<th>BON</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>M</td>
<td>SD</td>
<td>M</td>
</tr>
<tr>
<td><strong>Overlearned</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>VOCAB</td>
<td>61.67</td>
<td>11.27</td>
<td>48.18</td>
</tr>
<tr>
<td>FLUENCY</td>
<td>51.58</td>
<td>7.68</td>
<td>54.47</td>
</tr>
<tr>
<td>MATCHFIG</td>
<td>55.49</td>
<td>6.67</td>
<td>49.51</td>
</tr>
<tr>
<td>TAPPING</td>
<td>36.00</td>
<td>11.35</td>
<td>41.11</td>
</tr>
<tr>
<td><strong>Novel</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>WORDFIND</td>
<td>56.96</td>
<td>9.57</td>
<td>46.96</td>
</tr>
<tr>
<td>REMIND</td>
<td>44.56</td>
<td>9.81</td>
<td>44.21</td>
</tr>
<tr>
<td>MATRIX</td>
<td>55.42</td>
<td>3.96</td>
<td>49.70</td>
</tr>
<tr>
<td>PEGS</td>
<td>42.34</td>
<td>13.22</td>
<td>31.84</td>
</tr>
</tbody>
</table>
Table 23.

**Means and standard deviations of T-scores of two neuropsychological clusters on the PIC-R.**

<table>
<thead>
<tr>
<th></th>
<th>CLUSTER 1 (n = 8)</th>
<th>CLUSTER 2 (n = 22)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>M</td>
<td>SD</td>
</tr>
<tr>
<td><strong>FACTOR SCALES</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>I</td>
<td>54.75</td>
<td>10.49</td>
</tr>
<tr>
<td>II</td>
<td>47.50</td>
<td>5.86</td>
</tr>
<tr>
<td>III</td>
<td>52.75</td>
<td>20.84</td>
</tr>
<tr>
<td>IV</td>
<td>50.38</td>
<td>16.51</td>
</tr>
<tr>
<td><strong>VALIDITY SCALES</strong></td>
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<td></td>
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<tr>
<td>L</td>
<td>50.88</td>
<td>11.73</td>
</tr>
<tr>
<td>F</td>
<td>57.50</td>
<td>15.20</td>
</tr>
<tr>
<td>DEP</td>
<td>44.63</td>
<td>8.80</td>
</tr>
<tr>
<td>ADJ</td>
<td>48.25</td>
<td>9.79</td>
</tr>
</tbody>
</table>
Table 23 (continued).

Means and standard deviations of T-scores of two neuropsychological clusters on the PIC-R.

<table>
<thead>
<tr>
<th></th>
<th>CLUSTER 1 (n = 8)</th>
<th>CLUSTER 2 (n = 22)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>M</td>
<td>SD</td>
</tr>
<tr>
<td>CLINICAL SCALES</td>
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<td></td>
</tr>
<tr>
<td>ACH</td>
<td>43.13</td>
<td>2.95</td>
</tr>
<tr>
<td>IS</td>
<td>54.13</td>
<td>14.47</td>
</tr>
<tr>
<td>DVL</td>
<td>48.88</td>
<td>7.40</td>
</tr>
<tr>
<td>SOM</td>
<td>47.75</td>
<td>10.40</td>
</tr>
<tr>
<td>D</td>
<td>48.38</td>
<td>13.39</td>
</tr>
<tr>
<td>FAM</td>
<td>48.13</td>
<td>12.97</td>
</tr>
<tr>
<td>DLQ</td>
<td>43.75</td>
<td>9.10</td>
</tr>
<tr>
<td>WDL</td>
<td>44.38</td>
<td>5.37</td>
</tr>
<tr>
<td>ANX</td>
<td>50.50</td>
<td>14.32</td>
</tr>
<tr>
<td>PSY</td>
<td>51.38</td>
<td>15.24</td>
</tr>
<tr>
<td>HPR</td>
<td>56.88</td>
<td>8.76</td>
</tr>
<tr>
<td>SSK</td>
<td>45.00</td>
<td>7.69</td>
</tr>
</tbody>
</table>
The personality characteristics of each cluster, as assessed by the PIC-R, are presented in Table 23. Note that, unlike neuropsychological measures (where higher T-scores indicate better performance), higher PIC-R T-scores reflect more problems. Subjective inspection of this Table reveals that, on the average, the PIC-R profile of Cluster 2 children was relatively more elevated, as compared to that of Cluster 1 children on one of the factor scales (IV, Cognitive Development) and two of the clinical scales' (IS, Intellectual Screening, and PSY, Psychoticism). However, the standard deviations were also very large in both groups on these scales. A MANOVA with Clusters (two) as the independent variable and PIC-R Factor Scales (four) as dependent variables did not reveal a statistically significant main effect of Clusters ($F(4,25) = 1.210, p > .33$). Bonferroni comparisons (critical value of $T = 2.05$, $df = 28$) revealed that there were no significant differences between the two clusters on the first three Factors, but that Cluster 2 scored significantly higher than Cluster 1 on Factor IV.

**Cluster analysis on PIC-R factor scores.**

Details regarding the clustering process that was performed with the four PIC-R factor scores are presented in Table 24. The hierarchical tree dendogram from the AVERAGE analysis is presented in Figure 2.

Inspection of Table 24 reveals that a loss of more than 10% of the accounted-for variance did not occur until the last two
Table 24.

Cluster solutions of three different hierarchical agglomerative
techniques, with clustering variables PIC-R FACTORS I, II, III,
and IV, and squared Euclidean distance similarity measure.

<table>
<thead>
<tr>
<th>n</th>
<th>AVERAGE</th>
<th></th>
<th>CENTROID</th>
<th></th>
<th></th>
<th>WARD</th>
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</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>FREQ</td>
<td>SPRSQ</td>
<td>FREQ</td>
<td>SPRSQ</td>
<td>FREQ</td>
<td>SPRSQ</td>
<td></td>
</tr>
<tr>
<td>10</td>
<td>13</td>
<td>0.0211</td>
<td>13</td>
<td>0.0211</td>
<td>12</td>
<td>0.0116</td>
<td></td>
</tr>
<tr>
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<td></td>
</tr>
<tr>
<td>8</td>
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<td>20</td>
<td>0.0405</td>
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<td>0.0190</td>
<td></td>
</tr>
<tr>
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<td>0.0309</td>
<td>2</td>
<td>0.0094</td>
<td>13</td>
<td>0.0211</td>
<td></td>
</tr>
<tr>
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<td>2</td>
<td>0.0127</td>
<td>17</td>
<td>0.0655</td>
<td>17</td>
<td>0.0367</td>
<td></td>
</tr>
<tr>
<td>5</td>
<td>25</td>
<td>0.0838</td>
<td>18</td>
<td>0.0221</td>
<td>15</td>
<td>0.0403</td>
<td></td>
</tr>
<tr>
<td>4</td>
<td>15</td>
<td>0.0403</td>
<td>20</td>
<td>0.0463</td>
<td>22</td>
<td>0.0448</td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>26</td>
<td>0.0305</td>
<td>21</td>
<td>0.0249</td>
<td>6</td>
<td>0.0546</td>
<td></td>
</tr>
<tr>
<td>2</td>
<td>17</td>
<td>0.0655</td>
<td>22</td>
<td>0.0753</td>
<td>21</td>
<td>0.0762</td>
<td></td>
</tr>
<tr>
<td>1</td>
<td>43</td>
<td>0.5816</td>
<td>43</td>
<td>0.5841</td>
<td>43</td>
<td>0.5952</td>
<td></td>
</tr>
</tbody>
</table>

Abbreviations: n = Number of clusters
AVERAGE = Average Linkage method
CENTROID = Centroid Linkage method
WARD = Ward's Minimum Variance method
FREQ = Number of subjects of new cluster
SPRSQ = Semi-partial R2 of new cluster
Figure 2.

Average Linkage cluster solution with PK-R Factors I, II, III, and IV as clustering variables, and squared Euclidean distance as similarity measure.
clusters were merged. This pattern was evident for all three clustering methods. Inspection of the dendogram in Figure 2 suggested that a two-cluster solution would be compatible with the distribution of subjects in Euclidean space. However, this did not resolve the issue that the number of cases assigned to two final clusters differed with various solutions: The AVERAGE method suggested a 26 - 17 distribution, whereas the CENTROID and WARD methods both suggested 22 - 21 distributions.

Further inspection of cluster membership with each solution revealed that the CENTROID method assigned to a different cluster 5 of the 26 cases that would be included in the larger of the two AVERAGE clusters (n = 17 + 5 = 22 for the larger CENTROID cluster), with all other assignments corresponding to those of the AVERAGE solution. With the WARD method, 4 of the same 5 cases were assigned to a different cluster (n = 17 + 4 = 21 for the smaller WARD cluster), with all other assignments corresponding to those of the AVERAGE solution. In other words: there was a 88.37% agreement between the AVERAGE and CENTROID solutions, and a 90.70% agreement between the AVERAGE and WARD solutions.

A K-Means iterative partitioning method, where K was set at 2, yielded a 24 - 19 distribution. This solution reassigned two subjects from the clusters of the AVERAGE analysis (95.35% agreement), three from the CENTROID analysis (93.02% agreement), and two from the WARD analysis (95.35% agreement).

The K-Means results were not felt to provide strong reasons for preferring either of the three hierarchical cluster solutions.
In addition, the differences between the solutions were felt to be small (the smallest rate of agreement being more than 88%). For these reasons, the two-cluster AVERAGE solution was considered to be sufficiently reliable to be used for further analysis.

The average T-scores of the two PIC-R clusters identified with the AVERAGE solution are presented in Table 25. Note that the clusters were found with Factor scales I, II, III, and IV as the clustering variables. The Validity and Clinical scales are added to Table 25 only for the reader’s convenience, to further illustrate the findings.

Inspection of Table 25 reveals that, among clustering variables, Cluster 1 was most distinguished from Cluster 2 on FACTOR 4 (Cognitive Development), with Cluster 1 scoring much higher. In the context of conventional interpretation of PIC-R elevations (where T-scores above 70 are considered clinically significant), this elevation on FACTOR 4 was accompanied by clinically significant elevations for Cluster 1 on the Clinical scales that form the 'cognitive triad': ACH (Achievement), IS (Intellectual Screening), and DVL (Development). There was also a high elevation for Cluster 1 on Clinical scale PSY (Psychoticism).

Because of the particular pattern of elevations of Cluster 1, it was decided to compute Pearson product-moment correlations between PIC-R Factor scores and the ADJ screening scale on one hand, and IQ scores on the other. Only subjects whose IQ scores exceeded 70 and whose PIC-R was considered valid (n = 24) were included in these analyses. The results are presented in Table 26.
Table 25.

_T-scores of two PIC-R clusters (C) on Factor, Validity, and Clinical scales of the PIC-R._

<table>
<thead>
<tr>
<th></th>
<th>CLUSTER 1 (n = 17)</th>
<th>CLUSTER 2 (n = 26)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>M</td>
<td>SD</td>
</tr>
<tr>
<td><strong>FACTOR SCALES</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>I</td>
<td>56.53</td>
<td>12.20</td>
</tr>
<tr>
<td>II</td>
<td>63.29</td>
<td>9.47</td>
</tr>
<tr>
<td>III</td>
<td>64.12</td>
<td>13.58</td>
</tr>
<tr>
<td>IV</td>
<td>110.77</td>
<td>11.33</td>
</tr>
<tr>
<td><strong>VALIDITY SCALES</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>L</td>
<td>54.82</td>
<td>9.38</td>
</tr>
<tr>
<td>F</td>
<td>89.29</td>
<td>15.03</td>
</tr>
<tr>
<td>DEF</td>
<td>46.82</td>
<td>11.19</td>
</tr>
<tr>
<td>ADJ</td>
<td>64.29</td>
<td>11.68</td>
</tr>
</tbody>
</table>
Table 25 (continued).

**T-scores of two PIC-R clusters (C) on Factor, Validity, and Clinical scales of the PIC-R.**

<table>
<thead>
<tr>
<th></th>
<th>CLUSTER 1 (n = 17)</th>
<th>CLUSTER 2 (n = 26)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>M</td>
<td>SD</td>
</tr>
<tr>
<td><strong>CLINICAL SCALES</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>ACH</td>
<td>77.59</td>
<td>14.17</td>
</tr>
<tr>
<td>IS</td>
<td>108.35</td>
<td>12.99</td>
</tr>
<tr>
<td>DVL</td>
<td>88.24</td>
<td>13.34</td>
</tr>
<tr>
<td>SOM</td>
<td>67.82</td>
<td>13.88</td>
</tr>
<tr>
<td>D</td>
<td>58.35</td>
<td>10.13</td>
</tr>
<tr>
<td>FAM</td>
<td>49.29</td>
<td>6.85</td>
</tr>
<tr>
<td>DLQ</td>
<td>55.77</td>
<td>11.74</td>
</tr>
<tr>
<td>WDL</td>
<td>62.94</td>
<td>10.72</td>
</tr>
<tr>
<td>ANX</td>
<td>53.35</td>
<td>12.05</td>
</tr>
<tr>
<td>PSY</td>
<td>97.12</td>
<td>14.87</td>
</tr>
<tr>
<td>HPR</td>
<td>48.59</td>
<td>12.60</td>
</tr>
<tr>
<td>SSK</td>
<td>58.29</td>
<td>10.25</td>
</tr>
</tbody>
</table>
There were significant correlations between Factor IV and all IQ scores. In each instance, higher IQ scores were associated with lower T-scores on this PIC-R scale. A similar relationship was found for the ADJ scale, although in this case the correlations were significant only for VIQ and FSIQ. None of the other correlations was significant.

The number of subjects in the PIC-R clusters was considered to be sufficient to perform formal statistical analyses on selected physical and social variables for purposes of validation.

<table>
<thead>
<tr>
<th></th>
<th>VIQ</th>
<th>PIQ</th>
<th>FSIQ</th>
</tr>
</thead>
<tbody>
<tr>
<td>FACTOR I</td>
<td>-.12</td>
<td>-.07</td>
<td>-.15</td>
</tr>
<tr>
<td>FACTOR II</td>
<td>-.17</td>
<td>-.05</td>
<td>-.11</td>
</tr>
<tr>
<td>FACTOR III</td>
<td>-.25</td>
<td>-.02</td>
<td>-.14</td>
</tr>
<tr>
<td>FACTOR IV</td>
<td>.47 *</td>
<td>.68 **</td>
<td>.61 **</td>
</tr>
<tr>
<td>ADJ</td>
<td>.62 **</td>
<td>.28</td>
<td>.52 **</td>
</tr>
</tbody>
</table>

* $P < .05$

** $P < .01$
Table 27.

**Frequencies (n) and proportions (%) of occurrence of selected medical history variables in two PIC-R clusters, and results of Chi Square tests (X²).**

<table>
<thead>
<tr>
<th>VARIABLE</th>
<th>CLUSTER 1</th>
<th>CLUSTER 2</th>
<th>X²</th>
<th>df</th>
<th>p</th>
</tr>
</thead>
<tbody>
<tr>
<td>Severe Paraplegia</td>
<td>10</td>
<td>5</td>
<td>50.02</td>
<td>5</td>
<td>19.23</td>
</tr>
<tr>
<td>Mild Paraplegia</td>
<td>1</td>
<td>1</td>
<td>5.88</td>
<td>1</td>
<td>3.85</td>
</tr>
<tr>
<td>Bladder Incontinence</td>
<td>12</td>
<td>11</td>
<td>70.59</td>
<td>11</td>
<td>42.31</td>
</tr>
<tr>
<td>Bowel Incontinence</td>
<td>12</td>
<td>8</td>
<td>70.59</td>
<td>8</td>
<td>30.77</td>
</tr>
<tr>
<td>Seizures</td>
<td>8</td>
<td>2</td>
<td>47.06</td>
<td>2</td>
<td>7.69</td>
</tr>
<tr>
<td>Broken Home</td>
<td>5</td>
<td>7</td>
<td>29.41</td>
<td>7</td>
<td>26.92</td>
</tr>
<tr>
<td>Special School</td>
<td>13</td>
<td>6</td>
<td>76.47</td>
<td>6</td>
<td>23.08</td>
</tr>
</tbody>
</table>

of the clusters. Details regarding the applicability of these variables for the two clusters, as well as the results from the Chi-Square analyses (with Yates' correction for continuity) that were performed for each of these variables are presented in Table 27.
Inspection of Table 27 reveals that children in Cluster 1 were significantly more likely than children in Cluster 2 to have severe paraplegia, bowel incontinence, and/or seizures. Children in the former group were also significantly more likely to be attending a special school than children in the latter group.

**Discrimination and prediction of SCL 90-R scores.**

The average SCL 90-R T-scores of parents in the POSITIVE and NEGATIVE groups are presented in Table 28. As was indicated in Table 13, the average T-score of the total sample of 48 on the GSI scale of the SCL 90-R was 55.25, with a standard deviation of 10.27.

It is important to realize that the POSITIVE and NEGATIVE groups were selected on the basis of criteria that involved the GSI scale and all clinical scales of the SCL 90-R (i.e., only the PST and PSDI scales were not included in the selection process). Statistical analyses of the differences between the two groups on each of the SCL 90-R scales were therefore not performed.

Three separate stepwise discriminant analyses (SDA) with BACKWARD elimination were performed, using the variable categories described in Table 10, to distinguish between POSITIVE and NEGATIVE cases. The same variable categories were used to perform three separate stepwise multiple regression analyses (SRA) with BACKWARD elimination, with the T-score on the GSI scale as the criterion.
Table 28.

T-scores of parents with 'positive' (n = 21) and 'negative' (n = 27) SCL 90-R profiles.

<table>
<thead>
<tr>
<th>SCL 90-R Scale</th>
<th>POSITIVE</th>
<th>NEGATIVE</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>M</td>
<td>SD</td>
</tr>
<tr>
<td>GSI</td>
<td>63.91</td>
<td>3.74</td>
</tr>
<tr>
<td>PST</td>
<td>62.10</td>
<td>5.68</td>
</tr>
<tr>
<td>PSDI</td>
<td>59.14</td>
<td>4.88</td>
</tr>
<tr>
<td>SOM</td>
<td>59.43</td>
<td>7.11</td>
</tr>
<tr>
<td>OBS</td>
<td>60.71</td>
<td>6.09</td>
</tr>
<tr>
<td>INT</td>
<td>62.00</td>
<td>7.97</td>
</tr>
<tr>
<td>DEP</td>
<td>64.24</td>
<td>6.91</td>
</tr>
<tr>
<td>ANX</td>
<td>58.81</td>
<td>5.58</td>
</tr>
<tr>
<td>HOS</td>
<td>62.48</td>
<td>5.33</td>
</tr>
<tr>
<td>PHO</td>
<td>52.81</td>
<td>9.27</td>
</tr>
<tr>
<td>PAR</td>
<td>58.62</td>
<td>7.26</td>
</tr>
<tr>
<td>PSY</td>
<td>60.81</td>
<td>7.77</td>
</tr>
</tbody>
</table>
Eight Somatic Child variables were entered in the first SDA and SRA: Motor coordination and ambulation problems (in five dummy levels: 'Severe Paraplegia', 'Mild Paraplegia', 'Gait Problems', 'Truncal Ataxia and/or Balance Problems', and 'Fine Motor Coordination Problems'), Bladder Incontinence, Bowel Incontinence, and Seizures. None of these variables discriminated significantly between the POSITIVE and NEGATIVE groups. However, a one-variable best regression model was found for GSI, which consisted of 'Fine Motor Coordination Problems' ($R^2 = .0832$, $F(1,47) = 4.17$, $p = .0469$).

Seven Non-Somatic Child variables were entered in the second SDA: Age at Testing, Sex, Type of School, and T-scores on the four PIC-R Factor scales. None of these variables discriminated significantly between the POSITIVE and NEGATIVE groups, nor did any of these variables emerge as a significant regressor with regard to GSI.

Six Non-Child variables were entered in the third SDA: Number of Siblings, Broken Home, and Socio-Economic Status (in four dummy levels: I, II, III, and IV). None of these variables discriminated significantly between the POSITIVE and NEGATIVE groups. However, a one-variable best regression model was found for GSI, which consisted of 'Number of Siblings' ($R^2 = .0958$, $F(1,47) = 4.87$, $p = .0323$).

Because no variables were found that discriminated significantly between parents with 'positive' SCL 90-R profiles and parents with 'negative' SCL 90-R profiles, no further SDA was performed. To find the overall best regression model for the GSI
T-score on the SCL 90-R, a final SRA was performed with only those variables that had entered significantly in the three initial analyses. This included 'Fine Motor Coordination' and 'Number of Siblings'. Only the latter variable remained in the best overall regression model for GSI (R² = .0958, F (1,47) = 4.87, p = .0323).
CHAPTER IV

DISCUSSION

Medical history variables and intelligence.

The first purpose of this study was to consider whether (and, if so, which) medical history variables can discriminate between hydrocephalic children with various levels of intellectual development. For this purpose, level of intellectual development was dichotomized as HIGH (less than two standard deviations below the normative mean) versus LOW (two standard deviations or more below the normative mean). Many medical history variables did not discriminate between these HIGH and LOW groups. 'Neonatal and Infancy Problems' and current 'Ocular Defects' proved to discriminate between HIGH and LOW groups for both VIQ and PIQ measures. For VIQ, the presence of 'Lacunar Skull Deformities' in the first few months of life was also a significant discriminator. Presence of any of these variables in the child's medical history was associated with a greater likelihood of poor intellectual outcome.

The fact that best discrimination models were found for VIQ and PIQ allows rejection of null hypothesis 1. For some variables, our findings confirmed those of other studies in this area. However, for many other variables, the results of this study differed from other reports in the literature.
This is not surprising, given the many contradictions in the literature. In addition, the fact that we used a multivariate statistical approach with inclusion of many variables contributes to the fact that our findings differ from many other published reports that used few variables and/or less sophisticated methodologies or statistical analyses. For example, many more variables proved to be significant in the six separate initial discriminant analyses than the two or three that remained in the final best discrimination models. Had we only considered a small group of a priori selected variables (as many studies have done), our findings would probably have been quite different. We will briefly discuss some of the communalities and discrepancies between the present findings and those from other studies.

**Significant medical history variables**

The finding that 'Ocular Defects', such as gaze and movement defects or refraction and accommodation deficits, are often associated with poor intellectual outcome is consistent with the findings of Dennis et al. (1981) and Tew and Laurence (1978). However, unlike Dennis et al. (who only found an effect on PIQ), we found that 'Ocular Defects' were associated with lower absolute levels of both VIQ and PIQ. This suggests that it is not just the idiosyncracy of the task (e.g., requiring detailed and accurate visual inspection on such tasks as the WISC-R or WPPSI Picture Completion subtest) that causes many hydrocephalic children with ocular problems to have low IQ scores. Rather, ocular defects may represent an indication of generally greater structural cerebral
involvement, either at the level of the brainstem (e.g., superior colliculus), cortex (occipital pole), or white matter. Diffuse cerebral involvement (related to the pressure effects of the hydrocephalus) may be associated with a generally greater likelihood of cognitive difficulties.

We also have to disagree with Dennis et al. regarding the importance of 'Neonatal and Infancy Problems'. Hypoxia or respiratory distress and seizures were the most common problems in this category in our sample, and their presence tended to be associated with poor intellectual outcome. Dennis et al. concluded that these factors did not seem to compromise intelligence. Several studies have concluded that anoxic episodes in and of themselves have few long-term effects on intellectual development per se (see the review by Kopp & Krakow, 1983). However, some studies have also suggested that it may be the combination of several postnatal problems that puts children at risk for problematic intellectual development (e.g., Kitchen et al., 1980). This makes sense in our sample: Hypoxia is a common etiology for neonatal seizures, and the presence of either hypoxia or seizures in addition to the fact that the child is hydrocephalic may place him or her at greater risk for mental retardation (additive risks). As with 'Ocular Defects', the presence of 'Neonatal and Infancy Problems' may simply suggest that the child's brain has been compromised to a greater extent.

The selective effect of 'Lacunar Skull Deformities' (LSD) on VIQ adds to the widespread disagreement in the literature regarding the importance of this symptom with regard to later
intelligence. Stein et al. (1974) suggested that the presence of LSD should be an important predictor in the selection for surgery for hydrocephalic children. On the other hand, Hunt and Holmes (1975, 1976) and Lonton et al. (1975) failed to find a significant impact on intelligence. None of these studies reported on VIQ or PIQ specifically, and they all had serious methodological problems. The meaning of the selective effect of LSD on VIQ in this study remains unclear.

We also found that many medical history variables did not discriminate between LOW and HIGH intellectual attainment. Again, there are similarities and differences with other reports in the literature. The most important differences will be discussed briefly.

**Neutral medical history variables**

None of the demographic variables was found to be significant. The fact that Socio-Economic Status (SES) was neutral with regard to intellectual outcome in this study may seem especially surprising. Correlations between social class and intelligence have been reported in many studies, and some have even suggested that SES is a more important determinant of development than many medical or physical variables (see the review by Sameroff and Chandler, 1975). Although the distribution of SES strata in our sample appeared fairly representative of the general population, for the Detroit region there was probably an under-representation of Black subjects (who are also more likely to have lower SES). This may reflect a form of sample bias.
Several other possible explanations for the neutrality of SES with regard to intellectual outcome might be proposed. A statistical artifact, related to the restriction of the distribution of SES strata (five levels) as compared to that of intelligence is unlikely, since the discriminant analysis involved only two broad categories of intellectual outcome (retarded versus not retarded). It is more likely that, in cases of severe brain injury, the nature and quality of the child's physical and social environment does not matter very much with regard to intelligence: Structural lesions may compromise intellectual potential regardless of the background or environment. The implication may be that, all other things being equal (especially the quality of medical care), additional physical difficulties such as respiratory distress or ocular defects are more important for a hydrocephalic child's intellectual potential than his or her SES.

Another demographic variable for which no significant discrimination effect was found with regard to intelligence was handedness. However, this was a somewhat remarkable variable in this study, since there was a high incidence of left-handedness in this sample. The proportion of 31% left-handers among our subjects is almost four times that of the general, normal population. Although handedness did not emerge as a significant variable in the final best discrimination models for VIQ and PIQ, the majority of the left-handed subjects (i.e., 10 out of 15) were found in the LOW IQ groups. This raises the possibility that some of these children may represent cases of 'pathological left-handedness' (Silva & Satz, 1979). However, that syndrome is
usually associated with early cerebral lesions that are lateralized to the left hemisphere. This is usually not the case in hydrocephalus. Moreover, shunts are routinely placed first in the right hemisphere. Only five left-handed subjects had left shunts in addition to their right shunts, and the left shunts were typically placed after the first year of life. Therefore, pathological left-handedness does not appear to be an adequate explanation for the high incidence of sinistrality in this sample. The possibility remains that higher proportions of left-handedness just represent a common symptom of some form of cerebral dysfunction. This interpretation would be consistent with the generally high proportions of left-handedness in clinical populations (see the review by Hardyck & Petrinovich, 1977).

We did not find any systematic effect of motor or sensory symptoms on intellectual outcome. Especially with regard to motor disability, our results deviate from reports in the literature that found lower IQ scores in children with motor impairment (e.g., Badell-Ribera et al., 1963; Dennis et al., 1981). However, the vast majority of our subjects did have some form of motor impairment. This is important, because it indicates that many children with significant motor impairment are not mentally retarded. Although motor coordination problems may still contribute to a child's (especially Performance) IQ score, there is nothing in these data to suggest that motor impairment is detrimental with regard to intellectual functioning.

The literature shows perhaps its greatest inconsistencies with regard to the importance of the pathology of hydrocephalus.
As was noted in section 2.4.3., many of the discrepancies are related to methodological inadequacies. From our multivariate analysis, no single etiology or type of hydrocephalus can be identified that is associated with especially poor (or, for that matter, good) intellectual outcome.

Finally, we did not find any significant effect of treatment variables. The fact that the age at the time of the first shunt insertion and the number of shunt revisions proved neutral with regard to intellectual outcome is consistent with many reports in the literature (e.g., Dennis et al., 1981; Holmes et al., 1975; Raimondi & Soare, 1974; Tromp et al., 1979). In our sample, shunt infections also proved to be neutral with regard to intellectual outcome, which deviates from the findings of some other studies (e.g., Brown & McLone, 1981; Shurtleff et al., 1973). This may be related to the fact that shunt infections were a relatively uncommon occurrence in this sample. However, several children who had sustained a shunt infection still had IQ scores in the average range, which argues against a general detrimental effect of such an occurrence.

Occipito-frontal circumference (OFC) at first shunt insertion was also not a significant discriminator in this study. Since many authors have contradicted either other studies (e.g., Shurtleff et al., 1979 versus Tromp et al., 1979) or have failed to replicate their own earlier findings (e.g., Tromp et al., 1979 versus Tromp & Van den Burg, 1978) regarding OFC or similar measures, this is not a surprising finding. In a multivariate context, the absolute degree of ventricular expansion (indicated
indirectly by head size) may be less important than is the structural damage that this expansion (e.g., causing 'Ocular Defects') and/or additional medical problems (e.g., 'Neonatal or Infancy Problems') may have caused.

Accuracy of discrimination

Apart from the similarities and differences with other reports in the literature, it is important to consider exactly how well our best discrimination models were able to distinguish very poor (LOW group) from relatively better (HIGH group) intellectual outcome. The correct classification rate was fairly high (i.e., exceeding 80%) for both VIQ and PIQ. In most cases where a subject was misclassified, the statistical analysis underestimated the child's intellectual outcome. This is an important finding, since it illustrates that statistical significance cannot be equated with clinical significance. For example, one cannot conclude from these findings that one should tell parents of hydrocephalic children that have had a hypoxic or convulsive episode in the first few days of life that the child will probably end up being mentally retarded.

In our opinion, one can conclude from the findings of the discriminant analyses that 'Neonatal and Infancy Problems', 'Ocular Defects' and, to a lesser extent, 'Lacunar Skull Deformities' place the child at a greater risk for impaired intellectual development. All children in the LOW groups had at least one of these symptoms, and most of them (i.e., 14 out of 16 for LOW VIQ, and 16 out of 18 for LOW PIQ) had two or more of
these symptoms. However, not all children that had one or more of these symptoms ended up being retarded. A good example was a girl with Dandy-Walker syndrome who was born cyanotic, underwent open heart surgery shortly after birth (followed by several episodes of respiratory distress), received right and left shunts which were revised a total of 24 times, had a paralysis of upward gaze and right exotropia at the time of testing, and obtained a VIQ of 95 and a PIQ of 88. The implication seems to be that the above-mentioned factors need to be regarded as risk factors, and, consequently, that children with those symptoms be followed somewhat more closely. All children with hydrocephalus (even those with multiple risk factors) deserve to be treated efficiently. The results from the present study may be useful as guidelines, not for selection for treatment, but for alerting caregivers to those children who are 'at risk'.

**Verbal-Performance Discrepancies.**

On the average, PIQ scores tended to be somewhat lower than VIQ scores. The fact that this 9-point difference was statistically significant allows rejection of null hypothesis 2. The magnitude of the VIQ-PIQ discrepancy was commensurate with that reported in several other studies (e.g., Badell-Ribera et al., 1966; Connell & McConnel, 1981; Dennis et al., 1981; Tew and Laurence, 1975). However, a 9-point difference between two IQ scores is usually not considered clinically significant in every-day practice. In addition, the average PIQ of the sample was well within the average range, unlike the scores reported in
most other reports in the literature. This difference in findings may be partly due to the fact that we only included in this analysis cases with IQ scores above 70 (unlike most other studies).

Our findings are consistent with the rest of the literature in the sense that the average PIQ of hydrocephalic children is less than their VIQ. However, in this relatively small (and selective) sample, the amount of the difference is modest. Only in the context of other findings (e.g., poor performance on other visual-spatial measures not included in the PIQ score) would we feel comfortable attributing clinical significance to this difference. A discrepancy of between the two IQ measures of more than 14 points (which is conventionally considered clinically significant) occurred in 10 of the 30 subjects in the HIGH PIQ group. In nine of these cases, the VIQ was a standard deviation or more higher than the PIQ. This adds credibility to the above-mentioned VIQ-PIQ discrepancy in the total sample but, again, other evidence for deficits in visual-spatial processing or novel problem-solving would be needed to support any interpretation of this discrepancy.

**Sensory-motor performance and intelligence**

The average time to complete two trials of the Grooved Peg-Board showed a significant correlation with PIQ, accounting for about 25% of the variance. All other correlations between sensory-motor measures and IQ scores were not significant. The Grooved Peg-Board requires rapid placement of 1-cm grooved pegs
into their corresponding spaces on a form board. It is a complex task that requires fine motor coordination, with integration of various sensory (e.g., visual, tactile) inputs. This makes it much more a true sensory-motor measure than many of the other tests that have been used in previous studies, such as the Frostig Test (Miller & Sethi, 1971a; Sand et al., 1973; Tew & Laurence, 1975) or the Beery VMI (Connell & McConnel, 1981; Soare & Raimondi, 1977) in which perceptual and constructional abilities are much more involved.

Although our findings agree with many of the above studies in the sense that there is a source of common variance between sensory-motor performance and PIQ, it is also clear that this relationship is imperfect. Many simple sensory and motor measures did not correlate with PIQ, and for the one in this study that did (PEGS), at least three quarters of the variance in PIQ involved factors that are apparently not related to 'pure' sensory-motor performance. A similarly modest relationship was found between sensory and motor measures on one hand and performance on some neuropsychological tests involving visual and motor activities (e.g., Target Test, Draw-a-Design) on the other.

It should be noted that the low correlations between sensory-motor performance on one hand and scores on IQ tests or neuropsychological tests on the other may also be related to the range and distribution of the former measures. The distributions of scores on PAGNOS and FSsymbol were quite skewed. In addition, the range was quite large for PEGS (i.e., a standard deviation of
more than 20 T-points). The low correlations may have been in part a statistical artifact of these phenomena.

On the other hand, the imperfect relationship between sensory-motor performance and intelligence is consistent with the fact the 'Ocular Defects' were not only lawfully related to PIQ, but also to VIQ: There is simply no evidence for a strong selective effect of 'pure' sensory or motor factors on the PIQ of hydrocephalic children. Therefore, our results would not support the contention of others, such as Spain (1974) and Dennis et al. (1981) who suggested that deficits in ocular and/or motor abilities might be the cause of the relatively poor PIQ in these children.

**Neuropsychological subtypes**

Another major purpose of this study was to investigate whether reliable subtypes of hydrocephalic children could be identified on the basis of their scores on selected neuropsychological tests. For this purpose, cluster analyses were performed with tests selected on the basis of a factor analysis of our comprehensive battery. Two reliable clusters could be identified, but they appeared to differ only in level of performance: the smaller cluster had scores on cluster variables, validation variables, and IQ scores that were almost invariably one standard deviation or more higher than the corresponding scores of the larger cluster. In addition, when the subtypes were compared on subjectively selected tests of 'overlearned' versus 'novel problem-solving' skills, statistically significant
differences were found equally frequent with the former as with the latter variables.

Null hypothesis 3 (and, consequently null hypotheses 4 through 6) could therefore not be rejected: We were unable to identify specific 'linguistic' and 'visuospatial' subtypes. There was also no evidence for selective deficits of any of the subtypes in either overlearned skills or novel problem-solving. The fact that we did not find subtypes characterized by different patterns (instead of level) of neuropsychological performance may have been due in part to the fact that the number of subjects included in this analysis (30) was fairly small. However, the subject/variable ratio (30/4) was considered acceptable. Consideration of the general distribution of IQ scores among the children further illustrated the importance of sample size.

**FSIQ distribution**

The distribution of FSIQ scores in our sample indicated that the range of intellectual performance in hydrocephalic children is very wide: from severely retarded to very superior. About 37.5% of our sample was mentally retarded (i.e., FSIQ < 71). About 6.25% of our sample performed in the superior range (i.e., FSIQ > 114). The bulk of the children (about 56.25% of our sample) performed at intermediate levels, with most of them (almost 48% of the sample) obtaining FSIQ scores within one standard deviation of the normative mean. With the exclusion of the mentally retarded group from the cluster analyses there may simply not have been enough subjects to identify consistent patterns of performance in
this population because of the wide range in general level of performance. The possibility of the existence of specific subtypes of neuropsychological skills and abilities in a hydrocephalic population (i.e., subtypes with distinctive patterns of performance on neuropsychological tests) can therefore not be determined on the basis of our findings.

**Target Test**

One aspect of the neuropsychological findings that was somewhat outstanding was the children's performance on the Target Test. This task was designed to assess concentration and immediate visual recall; the child is asked to reproduce, after a three-second delay, with a pencil on paper a sequence that was tapped by the examiner in a dot matrix. It was the only neuropsychological test that revealed a T-score (averaged over all subjects) in the impaired range. In addition, it was a relatively poor score even in the cluster with the generally high level of overall performance. The Target Test was also the strongest representative of the second factor in the performed principal component analysis (a factor on which several tests of visual-spatial skills loaded highly). These findings suggest that this test adds a rather unique contribution to the battery that was used in this study.

One interpretation of the poor Target Test performance in this sample may be that the test is just too hard for these children. However, that would not explain why it is so hard. In addition, such an interpretation would be difficult to reconcile
with the fact that other tests for which the norms were derived from the same sample as for the Target Test (e.g., Progressive Figures Test, Matching Figures Test) did not appear to be a general stumbling-block for our subjects.

Another interpretation might be that the children had generally defective concentration or immediate recall skills. However, this would not be consistent with the fact that children in this sample performed much better on some other tests of such functions, such as recalling a meaningful narrative (STORY) or a list of unrelated words (REMEMD).

Consideration of the general pattern of performance of hydrocephalic children across a variety of tests suggests a different interpretation of their low Target Test score. The larger of the two clusters not only performed quite poorly on the Target Test, but also on the Draw-a-Design subtest from the McCarthy Scales. In addition, for both subtypes, performance was consistently higher on 'verbal' measures (PEABODY, RIDDLES, VOCAB, and WORDFIND) than on any of the 'visual-spatial' measures (SPATHMEM, DRAWDES, TARGET, and BLOCKDES) that were selected from the factor solution. These findings, in combination with the fact that the PIQ was lower than VIQ in both subtypes may suggest relatively reduced efficiency of visual-spatial skills in these hydrocephalic children.

**Visual-spatial skills and novel versus overlearned strategies**

Our data do not provide consistent evidence that these hydrocephalic children had general difficulties with tasks
requiring new, adaptive strategies to deal with complex or unfamiliar material. Indeed, both subtypes had means and standard deviations well within the average range on several tests that required problem-solving on the basis of either verbal (e.g., RIDDLES) or visual (e.g., MATRIX) materials. Rather, the results from this study appear to suggest the possibility of selective mild difficulties with the processing of complex visual or spatial patterns.

Several cautions need to be made with regard to this interpretation. First of all, this is a small sample, and the findings with the neuropsychological tests need independent replication. Second, the importance of the word 'relatively' cannot be overstated. By no means do we wish to imply that all hydrocephalic children do poorly on every 'visual-spatial' task. We merely imply that, in general, they tend to perform somewhat less efficiently on tasks involving complex visual-spatial skills than on tests of verbal skills. Third, it is the combination and consistency of several findings that gives rise to this consideration. This relates to the above-mentioned qualification of the interpretation of the VIQ-PIQ discrepancy: Only in the context of other evidence that hydrocephalic children have difficulty with visual-spatial tasks does the 9-point VIQ-PIQ discrepancy become meaningful.

We do feel confident that the relatively poor performance of the hydrocephalic children on many visual-spatial measures is not just due to ocular or fine motor dysfunctions. The fact that 'Ocular Defects' were also related to VIQ and the fact that more
than three quarters of the variance in PIQ was not related to
sensory-motor factors argues against this. The stimulus materials
of some of the tasks at hand also illustrates that other factors
must be involved. For example, one has to be almost legally blind
not to be able to discriminate the simple 3x3 5-cm dot design on
the Target Test. In addition, the motor involvement on the Target
Test and the KABC Spatial Memory subtest is of such a low level of
complexity that even children with significant upper extremity
motor coordination problems can often perform the pointing or
drawing aspects of the task without much difficulty.

In summary, then, clear neuropsychological subtypes could not
be identified in this study. Most of the differences between
subjects in terms of neuropsychological test scores reflected
different levels, rather than patterns, of performance. In
general, the pattern of both IQ scores and factor-analytically
selected neuropsychological measures suggests that hydrocephalic
children may have relatively reduced efficiency in complex
visual-spatial processing, which is not attributable to 'pure'
sensory or motor factors.

We can only speculate about the meaning of the suggested
selective mild impairment of complex visual-spatial skills in
hydrocephalic children. There is a possibility that early damage
to the posterior quadrants of the right hemisphere (associated
with the insertion of a shunt) may compromise the development of
these cognitive skills. However, the data from this study did not
allow a direct evaluation of the relationship between lesion
localization and neuropsychological outcome. More important, in
our opinion, is what the implications of having mild visual-spatial difficulties at ages between five and eight years may be. Selective deficiencies in visual perceptual and organization abilities are one of the characteristics of older children with non-verbal learning disabilities (Rourke, 1987). In addition, some effects of early brain damage or early visible mild impairments may not become 'full-blown' until the child becomes older, i.e., when the level of complexity of (especially academic) tasks increases (Rourke et al., 1983). There is, therefore, a possibility that hydrocephalic children who do exhibit mildly impaired visual-spatial skills at an early age are at increased risk for subsequent development of further problems in nonverbal problem-solving, adapting to novel or complex situations, etc. This is a question that can only be answered in cross-sectional studies with additional age groups, or in longitudinal studies.

Performance on other neuropsychological measures

There are very few other studies that have examined the performance of hydrocephalic children on neuropsychological tests. With regard to language skills, Tew (1979) reported a fairly high incidence of the so-called Cocktail Party Syndrome (CPS) in his sample. This concept has been convincingly criticized by Dennis et al. (1987), and its diagnostic or descriptive usefulness appears limited. Even when we did apply Tew's criteria to our sample (which was about the same size as Tew's) we could identify only two children that were testable and that had a distinctive pattern of verbal perseveration, frequent provision of task
irrelevant information, and tangential answers to open-ended questions. However, these children were also considered to have a general Attention Deficit Disorder. In any event, this proportion would be nowhere near the 40% incidence reported by Tew. We therefore strongly doubt the existence of a specific 'hydrocephalic' pattern of speech or linguistic development.

We agree with Dennis et al. (1987) that language difficulties are not widespread among hydrocephalic children. On the average, our subjects performed well on such tests as the Peabody Picture Vocabulary Test - Revised, the Verbal Fluency subtest of the McCarthy Scales, and the Token Test for Children. The latter result differs somewhat from the findings of Dennis et al. who found a significant effect of hydrocephalus on understanding of grammar. This difference may be due to the fact that they used a different, possibly more complex, test to assess oral sentence comprehension.

Visual perceptual and constructional abilities have often been assessed in other studies with the Frostig Test or the Beery VMI, with several studies (Miller & Sethi, 1971a; Sand et al., 1973; Tew & Laurence, 1975; Connell & McConnel, 1981; Soare & Raimondi, 1977) reporting relatively poor performance by hydrocephalic children. The Draw-a-Design (DRAWDES) subtest from the McCarthy Scales that was used in our study is quite similar in requirements to the Beery VMI: making paper-and-pencil copies of geometric line drawings. Although the sample as a whole obtained an average T-score on DRAWDES in the lower end of the average range, it was a test on which the larger of the two
neuropsychological clusters obtained significantly impaired results. We therefore feel that this is a result that is consistent with those obtained in several other studies.

Memory functions have also been assessed in several studies. Tromp and Van den Burg (1982) and Cull and Wyke (1984) reported deficient performance of hydrocephalic children on memory tests. However, their measures were experimental in nature, without adequate age norms. In our sample, children performed quite well on some age-normed memory measures, such as STORY and REMIND. The latter task is somewhat similar to the tasks used in the other studies cited above, and our results argue against a general memory deficit in hydrocephalic children (although the norms for REMIND are far from perfect). The significance of the outstandingly poor performance of our sample on the Target Test was discussed above.

Finally, impaired performance of hydrocephalic children on sensory-motor tests such as Finger Tapping (TAPPING) or Lafayette Grooved Peg Board (PEGS) has been reported in several studies (e.g., Hurley et al., 1983; Prigatano et al., 1983) and our results are consistent with those of such studies: On the average, our subjects obtained impaired performances on those tests. Many children with hydrocephalus appear to have difficulty with fine motor tasks but, as was discussed above, this would not appear to be the sole reason for their PIQ tending to be relatively depressed.
Personality subtypes

In order to investigate whether reliable subtypes of behavioral adjustment could be identified in this sample, cluster analyses were performed with the four Factor scales of the PIC-R. Two clusters were found. Cluster 1 was comprised of 17 subjects who were identified by their parents as having major difficulties in their intellectual development, personal independence, and somatic health. Among these variables, impaired intelligence, reflected in a very high elevation on the Factor IV scale (Cognitive Development) was the most outstanding characteristic of cluster 1. Cluster 2 was comprised of 26 subjects, described by their parents as doing generally well in areas of cognitive, physical, and social development.

Analyses of the medical history of the two PIC-R clusters further illustrated the nature of the difficulties of cluster 1. These children often displayed one of the following symptoms: needing a wheelchair, being incontinent for bowel movements, suffering from a seizure disorder, and/or attending a Special Education program. Most of these children were classified in the LOW VIQ or LOW PIQ group. This is consistent with the fact that this cluster had significant elevations on the 'cognitive triad' of the PIC-R (ACH, IS, and DVL), and with the significant (albeit imperfect) correlation between scores on PIC-R Factor IV and IQ scores. The latter is important because the clustering variable on which cluster 1 differed most outstandingly from cluster 2 was Factor IV. None of the other PIC-R Factor scores had a significant correlation with any of the IQ measures. This
confirms the fact that these other Factors pertain to different aspects of children's development. In general, the selectivity of the significant negative correlations between Factor IV and IQ scores confirms that the factor pattern of the PIC-R.

It is important to realize that the elevation on the PSY scale in these children does not mean that they are psychotic. In this sample of children, this elevation typically reflects the lack of physical independence, delay in reaching developmental milestones, and social isolation (often due to physical handicaps).

Of course, the current findings do not reflect different 'personality' subtypes in the conventional sense of the word. The PIC-R in this sample was apparently much more frequently affected by the physical consequences of hydrocephalus and related problems (e.g., spina bifida) than by behavioral or emotional issues. One might argue that, because of its sensitivity to physical problems, the PIC-R might not be an appropriate instrument for hydrocephalic children, but we would disagree with that. First of all, it is one of the very few psychometrically 'sound' instruments in the area of child personality assessment. Second, in several individual cases, the PIC-R proved to be quite helpful in detecting other than physical issues. For example, in two cases, a sharp elevation on the PAM scale alerted us to disagreements between the parents about disciplining and socialization issues (which disagreements had not been spontaneously reported verbally). In another case; a sharp elevation on the DLQ scale appeared to reflect oppositional and acting-out behavior in
specific circumstances (also not reported on personal interview). The PIC-R also proved helpful in confirming clinical diagnoses of Attention Deficit Disorder in three cases, and of a general withdrawal/distress syndrome in three additional cases. In one of the latter cases, this appeared related to the recent death of a parent, and in another to teasing by peers about a mild physical handicap; the etiology was unclear in the third case. Therefore, although no clear general 'personality' subtypes were found in this study, the PIC-R was an important addition to our assessment in individual cases. The fact that specific emotional-behavioral 'syndromes' had a low frequency in this sample probably explains why they were not detected by the cluster analyses.

One might consider the above-mentioned seven cases of oppositional behavior, Attention Deficit Disorder, and withdrawal/distress syndrome as instances of 'true positives' with regard to behavioral maladjustment. This would represent a proportion of 16% in the sample for whom valid PIC-R profiles were available. This compares quite favorably with the rates of psychiatric disorder reported in some other studies (e.g., 44% in Connell & McConnel, 1981). However, comparisons with other studies are difficult to carry out because most of them relied on interviews or experimental rating scales. In addition, in some children who were severely motorically and mentally retarded, some behavioral problems (e.g., head banging) were obscured by the general elevation of the PIC-R Psychosis scale (common in motorically impaired children). There were also two other children which were clinically considered to meet the criteria for
Attention Deficit Disorder, but who were not motorically hyperactive and were thus not identified as such by the PIC-R. Therefore, it would appear that PIC-R profiles need to be considered in the context of a comprehensive parent interview and adequate behavioral observations during the assessment.

To our knowledge, this is the first study that has examined in a systematic way the profile of hydrocephalic children on a well-standardized and psychometrically sound inventory. On the basis of our findings, it is concluded that the majority of hydrocephalic children do not present with significant psychiatric problems. In those who do show behavioral adjustment difficulties, the issues do not always appear to be directly related to their hydrocephalus.

**Distress in the parents**

The nature and level of distress of the parents was assessed with the SCL 90-R. Formal, direct comparisons with other reports in the literature are again difficult to carry out, since most studies relied on interviews or experimental inventories. To our knowledge, this is the first study that has analyzed the pattern of responses of parents of hydrocephalic children on a standardized questionnaire of documented reliability and validity. However, some general considerations of similarities and differences with other studies can be made.

A 'positive' profile was obtained in 21 out of 48 cases (about 44%). This is consistent with many other reports in the literature that have reported a fairly high incidence of distress
In parents of children with hydrocephalus and/or myelomeningocele (e.g., Dorner, 1975; Richards & McIntosh, 1973).

None of the selected physical and social variables discriminated between 'positive' and 'negative' profiles. We did not find any relationship between level of distress of the parents and level of disability in the patient. This means that hypothesis 8 was not confirmed. In this respect, our findings agree with those of Carr et al. (1983), Dorner (1975), and Richards and McIntosh (1973), but disagree with those of Tew and Laurence (1975). The neutrality of the variable 'broken home' with regard to distress in the parents is consistent with the findings of Dorner (1975) and Richards and McIntosh (1973) but not with those of Tew et al. (1974). The only variable that had some meaningful relationship with the distress of the parents appeared to be the number of siblings in the family (with more siblings being associated with higher levels of distress). However, the amount of variance explained by this variable was quite small (i.e., less than 10%).

Consideration of the profile of average T-scores in the POSITIVE group revealed a mild level of general distress (as indicated by the GSI score), and relative elevations on the Depression, Hostility, and (to a lesser extent) the Interpersonal Sensitivity Scale. Such a pattern could be interpreted as reflecting feelings of inadequacy and hopelessness, with particular difficulties in the area of coping with daily interpersonal stressors, leading to a generally dysphoric mood.
In all cases where 'positive' profiles were obtained, the nature of distress was further discussed with the parents in clinical interviews. The documented dysphoria was subjectively reported to be related to a wide variety of issues, ranging from specific guilt about the child's physical condition to loss of a job. In the context of the absence of strong relationships with selected physical and social variables, these results may suggest that the high incidence of distress in parents of hydrocephalic children is not so much related to isolated physical or social variables per se. Rather, as Dorner (1975) has suggested, the presence of a child with medical problems in the family may increase the vulnerability of the parents to general daily stressors. Requirements such as having to make special transportation arrangements, accommodating frequent medical appointments and sometimes hospitalizations, etc. may put so much pressure on the parents that they have less resources to cope with other problems. As a result, their level of subjective distress may be increased.

**The psychological profile of hydrocephalic children: some general conclusions.**

This study set out to investigate how well hydrocephalic children and their parents are doing in a variety of areas when the children are about five to eight years after their initial surgery. In our opinion, the most striking finding was the wide variety in outcome: Some children are severely motorically and mentally retarded whereas others (with often similar medical
histories) appear unimpaired in all aspects of adaptive functioning.

This study has demonstrated the importance of consideration of the complete medical history of these children. Consideration of only isolated medical history variables may lead to many erroneous conclusions or expectations with regard to the intellectual prognosis in infantile hydrocephalus. This is, to our knowledge, the first attempt at using a comprehensive, multivariate, statistical approach to investigate intellectual sequelae of infantile hydrocephalus. Our results suggest that this is a useful approach which may help to clarify some of the confusions in the literature. In addition, the results from this study suggest caution in making long-term predictions: "Risk factors" can be identified, but they are not invariably associated with impaired intellectual development. Risk factors can and should be used as guidelines for frequent follow-up. In that way, symptoms of developmental problems may be detected more efficiently, with associated opportunities for early intervention.

One important prospect for further research is to investigate whether different risk factors can be identified for development at different age levels. Rourke et al. (1983) have suggested that some sequelae of early brain lesions may not become manifest until the child gets older, when environmental demands and complexity of academic tasks increase. It may be possible that some hydrocephalic children who do not exhibit significant cognitive or behavioral dysfunctions when they are five or six years of age may encounter adaptive and academic difficulties when they become nine
or ten years old. Early evidence for mild impairment of complex visual-spatial processing may be a predictor of increased risk for later, more significant cognitive or behavioral dysfunctions. This is an issue to be addressed in future research.

The results of this study have not been conclusive with regard to the possibility of neuropsychological subtypes. The pattern of the performances of hydrocephalic children across a variety of intelligence and neuropsychological measures suggests that they may have reduced efficiency with regard to complex visual-spatial processing. Larger sample sizes are needed to accurately investigate the possibility of specific differences in pattern of performance, in addition to the differences in level of performance found in the current study.

Finally, we considered emotional adjustment in the children and their parents. Clinically significant levels of behavioral maladjustment were found in some children although the majority of this sample did not present with general adjustment problems. Standardized parent questionnaires such as the PIC-R appear to provide very significant information about behavioral adjustment in many cases. However, the results always need to be considered in the context of their general clinical presentation (e.g., level of motor impairment, evidence for attention deficits without hyperactivity, etc.). The same is true for the use of the SCL 90-R as an index of distress in the parents. Almost half of the parents appeared to experience some significant form of distress, but this did not appear to be meaningfully related to specific aspects of the physical and social aspects of the patient and his
or her family. There is a possibility that having to take care of a child with many medical problems (sometimes involving frequent hospitalizations for shunt revisions, eye surgery, urinary tract infections, etc.) may render parents more vulnerable to general psychosocial stressors in their daily life. This needs to be addressed in future research. The inclusion of measurements of actual activities of daily living of hydrocephalic children and developmental demands which they face should also be considered in future research. For example, administration of the Vineland Adaptive Behavior Scales (Sparrow, Balla, & Cicchetti, 1984) to the primary caretaker would be potentially very informative.

It is concluded that the cognitive and behavioral possibilities for a newborn with hydrocephalus are often wide open. Efficient medical management of all patients, with frequent medical and psychological follow-up (including consideration of the adjustment of the parents), can be of great value with regard to fostering optimal neurobehavioral outcome.
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APPENDIX A

RAW DATA RECORD FORM

The raw data were documented for each subject with the record form, presented below. Details regarding the measurement of most of the variables are discussed in the 'Method' section, and are summarized in Table 2 (page 157), Table 3 (page 163), and Table 4 (page 167). We will briefly discuss those variables that were not included in those tables.

Variable 001 (SUBJECT) represents the subject number that was used for identification of the individual cases. This number represents a discrete variable, that indicates in a chronological manner the cases that were assessed and retained in the study.

Variable 032 (TUMOR) was included because all 119 potential subjects were rated with the demographic, symptom, pathology, and therapeutic variables that are listed on this form. However, no tumor cases were included in the final analyses (see 'Method' section).
Variables 039 (CEREBRAL MANTLE) and 040 (VENTRICULO-SKULL) were included because at the beginning of this study it was not yet clear whether or not these indices could be obtained on sufficient subjects.

Variable 049 (IQTEST) was included to indicate for purposes of convenience which intelligence test was used. Cases assessed with the WPPSI were indicated with a score of 1 on this variable, cases assessed with the WISC-R with a score of 2, and cases that could not be tested with a score of 3.

Variables 053 through 067 represent the scaled scores on the various subtests of the WISC-R and WPPSI.

Variables 085 through 092 represent the left and right hand scores on the various sensory and motor measures. However, only indices based on the average of both hands were used in the statistical analyses.

Missing data (e.g., IQ scores for children who could not be administered psychological tests) were indicated with a period. The order of variables on the record form represents the order in which they were entered in the computer. The specific raw data for each individual subject are presented in the same order in Appendix B. These data constituted the complete source for all statistical analyses in this study.
### RAW DATA RECORD FORM

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**PHYSICAL AND SOCIAL VARIABLES**

**INTELLIGENCE TEST VARIABLES**

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094 FACTOR II
095 FACTOR III
096 FACTOR IV
097 L
098 F
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101 ACH
102 IS
103 DVL
104 SOM
105  D
106  PAM
107  DLQ
108  WDL
109  ANX
110  PSY
111  HPR
112  SSK

SCL 90-R SCALES

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114  PST
115  PDSI
116  SOM
117  OBS
118  INT
119  DEP
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121  HOS
122  PHO
123  PAR
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APPENDIX B

RAW DATA PER SUBJECT

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| 50 | 53 | 57 | 49 | 46 | 49 | 52 | 54 | 44 | 41 | 44 |
VITA AUCTORIS

Jacobus Donders was born on August 26, 1959 in Tilburg, The Netherlands. In June, 1977 he graduated cum laude from Copenhagen High School, Tilburg, The Netherlands. In September, 1977, he enrolled at Tilburg University. He graduated cum laude with the Doctorandus of Psychology degree in June, 1984. Since September, 1985 he has been enrolled in the Doctoral programme in Clinical Neuropsychology at the University of Windsor.