UNIVERSITY OF SOUTHAMPTON

Ophthalmic Complications of Spina Bifida and Hydrocephalus

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Table of Contents

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ABSTRACT

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OPHTHALMIC COMPLICATIONS OF SPINA BIFIDA AND HYDROCEPHALUS

by Hannah Gaston

This thesis represents an attempt to further our knowledge of the ophthalmic complications of spina bifida and hydrocephalus by means of literature review and a long term clinical study, and to determine whether regular ophthalmic supervision can assist in the general management of affected children.

The ophthalmic complications of spina bifida have often been reported in the literature and thought to merit regular supervision of affected children, yet few centres currently offer this service.

In this study 322 children attending one regional centre were examined repeatedly over a six year period by one ophthalmologist. Ophthalmic complications were found to be very common. They frequently provided evidence of raised intracranial pressure due to shunt dysfunction even when other objective evidence was lacking.

Every spina bifida and hydrocephalus clinic should have an ophthalmologist in its medical team. Preservation of visual function and early diagnosis of raised intracranial pressure in these children should result from this arrangement.

Preface

The object of this study was to determine if regular, long term examination of children with spina bifida or hydrocephalus by an ophthalmologist working in the spina bifida clinic would assist in their general management, particularly in the diagnosis of shunt obstruction as it is often difficult to determine this by other means.

Summary

The majority of children with spina bifida and hydrocephalus require cerebrospinal fluid shunts at some time in their lives. These shunts commonly become obstructed. Shunt obstruction is followed by the development of an acute or chronic rise in intracranial pressure which may be difficult to diagnose early, as the symptoms and signs are often non specific. The main objects of this study were two-fold. Firstly, to determine whether papilloedema, as assessed by a trained ophthalmologist, is a reliable diagnostic sign of raised intracranial pressure and shunt malfunction in hydrocephalic children. Secondly, to determine the incidence of other ocular complications in children with hydrocephalus and whether recognition and treatment of these disorders significantly benefit their development, education and employment prospects.

A background review of the literature outlines the pathology and treatment of spina bifida, hydrocephalus and associated lesions, and evaluates previous observations of the ophthalmic complications of these conditions. In this study 322 children who were suffering from either spina bifida or hydrocephalus were examined over a six year period by one ophthalmologist working in the clinic alongside paediatric staff. Regular examinations were made of visual acuity and field, ocular motility and fundi, starting at the time of birth or diagnosis and repeated whenever possible, at each clinic visit or hospital admission. A particular effort was made to exam children about to have exploratory surgery for suspected shunt dysfunction or raised intracranial pressure to determine the frequency of positive eye findings in such cases.

The results of the study illustrate that, in 70% of proven episodes of shunt dysfunction in hydrocephalic children, ophthalmic complications of raised intracranial pressure, e.g. papilloedema, Parinaud's syndrome or convergent squint are valuable early diagnostic signs. Over all, 42% of the children had a manifest squint, 29% had developed a lateral rectus paresis, 14% had papilloedema and 17% had optic atrophy. Only 27% of children had normal visual function. Eighteen case

histories are used to illustrate and emphasise some of the clinical findings in this study. Evaluation of the results of treatment of ophthalmic complications of hydrocephalic children suggest that ophthalmic care could significantly improve educational and employment prospects.

It is concluded that every spina bifida and hydrocephalus clinic should have an ophthalmologist in its medical team. He should undertake regular examination of visual acuity and field, ocular motility and fundi starting at the time of birth or diagnosis, and tailor ophthalmic care to the special needs of these children. This should ensure that these children achieve and maintain the best possible standard of vision and that the earliest ophthalmic signs of raised intracranial pressure can be detected in patients where the diagnosis is in doubt. Immediate liaison between the Paediatrician and Opthalmologist is vital.

Introduction

At least one quarter of the children with spina bifida or hydrocephalus, living in Wessex are seen regularly at a clinic held weekly at Southampton General Hospital. The clinic is run by Paediatric Surgeons who also perform all the necessary operations for closure of neural tube defects and control of hydrocephalus at this hospital.

The Eye Hospital in Southampton is some two and a half miles distant from the General Hospital. In the past spina bifida children with eye problems had to attend ophthalmic outpatients as well as the general clinic, thus imposing an additional burden on their parents. Many children failed to attend for treatment, and the liaison between Paediatric Surgeon and Ophthalmologist was often poor because it depended on correspondence rather than immediate discussion. (Case histories 1,5 and 9). Several Ophthalmologists and their junior staff were involved in the childrens' care so that there was little familiarity of any one surgeon with the problems caused by spina bifida and hydrocephalus and little continuity of care. The detection of new ophthalmic problems was the responsibility of paediatric staff. Understandably, the plethora of other problems to be dealt with made ophthalmic examination receive low priority. Mental retardation, illness and nystagmus compounded the difficulties of examination in these children. Lack of equipment and reluctance to dilate pupils were additional factors.

Thus, it had been felt for some years that regular examination of these children by one ophthalmologist during their routine clinic visits might greatly improve the quality of care available to them. In addition it was felt that such examination might assist in the diagnosis of uncontrolled hydrocephalus for it is frequently difficult to make the diagnosis from other clinical evidence.

This thesis is based on observations made by one ophthalmologist working in the spina bifida and hydrocephalus clinic over a six year period.

Classification of disorders discussed.

1. Spina bifida occulta.
2. Spina bifida cystica alone (a) meningocele (b) myelomeningocele.
3. Spina bifida with hydrocephalus.
4. Occipital encephalocele.
5. Occipital encephalocele with hydrocephalus.
6. Anencephaly.
7. Primary hydrocephalus.
8. Secondary hydrocephalus.
9. Hydranencephaly.
10. Porencephaly.

Background and Survey of Previous Work

Spina Bifida

Spina bifida is a defect of the vertebral arches, most often dorsal. In spina bifida occulta this bony lesion is hidden from view. A defect limited to a single vertebra can be regarded as a normal variant, but if the overlying skin in abnormal, bearing a tuft of hair or a lipoma, there may be an associated intraspinal lesion referred to as occult spinal dysraphism.

In spina bifida cystica (aperta) there is a cystic swelling herniating through the vertebral defect. In meningocele the cystic swelling consists of meninges and fluid in continuity with the cerebrospinal fluid (CSF); it is usually covered by intact skin. In myelomeningocele the swelling also contains spinal cord or nerve roots. Some authors reserve the term myelomeningocoele for closed lesions which have a covering of skin and use the term myelocoele for the commoner open lesions. In many cases of myelomeningocele associated abnormalities of the spinal cord are to be found both cranial and caudal to the obvious lesion.

The incidence of spina bifida cystica (including still births) in the United Kingdom is 2.5/1000 births[1]. There is considerable regional variation so that in South Wales the incidence is 3.92/1000 births[2] and in Southampton 3.2/1000 total births[3]. There is a 5% risk of a neural tube defect arising in a sibling after a case of spina bifida or anencephaly[4]. It seems likely from available evidence that some environmental factor determines the development of spina bifida in genetically predisposed embryos[1].

The clinical effects of spina bifida vary enormously depending upon the size and site of the lesion and whether it is open or closed. Unfortunately there is no clear distinction between open and closed lesions as a closed lesion may have been open during early pregnancy[5]. Meningocele,accounting for up to 16% of spinal neural tube defects[6] usually carries a good prognosis although hydrocephalus may rarely supervene. However, in myelomeningocele there is usually a marked loss of motor, sensory function and autonomic function below the level of the lesion. 82% of myelomeningoceles are lumbar or sacral in site[7] so the common problems are paralysis and anaesthesia of the lower limbs, trophic ulcers of the feet, incontinence of urine and faeces, and back pressure on the urinary tract leading to progressive renal deterioration. Many children also have associated primary anomalies of the urinary tract.

Myelomeningocele produces a complex neurological picturee including upper and lower motor neurone lesions. Nor is it a static disability; a large number of patients show neurological change and deterioration with time and problems of balance, spatial orientation, vision and coordination cause further reduction of locomotor function[1].

Treatment of spina bifida. Early operation to close the back greatly increases the chances of survival by diminishing the risk of infection. In most centres the operation is carried out within twenty four to thirty six hours of birth, under general anaesthesia, in a special surgical centre. The neural tissue in the sac is dissected free and preserved, redundant membrane is excised and the dura closed over the neural plaque. Skin closure is then obtained. Meningitis occurs in 10-25% of patients and is often fatal. A CSF leak from the wound increased the risk of infection and may require insertion of a shunt.

Hydrocephalus in spina bifida.

Because of an associated Arnold-Chiari malformation i.e. herniation of brain stem and the tonsils and vermis of the cerebellum, the majority of open spina bifida patients also have clinical evidence of hydrocephalus. Some babies also have an associated aqueduct stenosis causing internal hydrocephalus. Hydrocephalus denotes enlargement of the ventricles of the brain and implies the presence of an increased quantity of CSF under increased pressure, either intermittently or persistently, currently or at some time in the past. Examination of the ventricles at birth reveals that they are enlarged in nearly all cases of spina bifida[8]. One quarter of all babies with spina bifida have significant hydrocephalus at birth, but one third have only a mild hydrocephalus that arrests spontaneously at an early stage[1].Following surgical closure of a meningomyelocoele there is often an increase in the severity of associated hydrocephalus: this then requires a CSF shunt. Infants who require a shunt rarely become independent of it later, though a little transependymal absorption of CSF may develop.

The diagnosis of infantile hydrocephalus is made on the basis of rapidly increasing head circumference, sunsetting, a bulging fontanelle, prominence of the scalp veins and separation of the cranial sutures. Apnoeic episodes may occur. Originally the degree of hydrocephalus was assessed by air studies; these have now been replaced by computed tomography (CT scan) and ultrasonography.

If hydrocephalus is allowed to progress the ventricles dilate, brain tissue atrophies and the skull becomes thin. Involvement of the pyramidal tracts causes wasting and spasticity of limbs, cerebellar ataxia, convulsions, mental deficiency, cranial nerve palsies, precocious puberty and finally coma may ensue. Severe untreated hydrocephalus is usually fatal.

Even when hydrocephalus appears to have arrested on the basis of ventriculography, normal CSF pressure, stable head size, non-progressive neurological symptoms and absent or clinically non-functioning shunt, the state may be, in reality, one of "normal pressure" hydrocephalus. Continuous intraventricular pressure measurement for fifteen to twenty hours in these patients may show elevations of CSF pressure on a normal baseline. Hence one determination of intraventricular pressure is not enough, especially in those children who demonstrate a discrepancy between performance and verbal intelligence quotient (IQ) studies. Such children may be improved by shunting [9,10]. True arrest of hydrocephalus can be proven only by continuous ventricular pressure monitoring; removal of the shunt without this precaution is unwise [11] and indeed may be fatal [12].

Treatment of Hydrocephalus.

The development of cerebrospinal fluid shunts has radically altered the prognosis of spina bifida and hydrocephalus. In the Spitz Holter system a proximal catheter is inserted into the lateral ventricle of the brain via a parietal burr hole and connected via a one-way valve to a distal catheter which is inserted via the common facial vein or the internal jugular vein into the right atrium of the heart. A Rickham cap interposed

between the proximal catheter and the valve overlying the burr hole allows occasional sterile taps to be performed for sampling of CSF and measurement of its pressure. Alternatively, the distal catheter may be passed into the peritoneal cavity or temporarily exteriorized.

Shunt complications are frequent and serious. Catheters may become disconnected and even float into the circulation. Proximal catheter blockage by choroid plexus or blood clot can occur. It sometimes does so repeatedly soon after insertion of a shunt. Distal catheter blockage tends to occur later and more slowly as the child grows and the catheter tip is drawn out of the atrium into the superior vena cava. Flow is then laminar and the shunt may be occluded by thrombus. Simpson[11] found the rate of blockage was 1 per 5-6 years of life with a shunt. Distal catheter blockage may be prevented by prophylactic shunt revision (vide infra).

Shunt disconnection and blockage cause raised intracranial pressure and the child may present with poor feeding, vomiting, headaches, drowsiness and bulging fontanelle. Although the diagnosis sounds easy such symptoms are non specific and may be due to urinary tract infections or other childhood ailments. The symptom complex varies with the child, but may be identical in each episode of obstruction in the same child. Pumping the valve may be diagnostic since in a distal catheter blockage the valve fails to empty on compression (the valve feels stiff) and in a proximal catheter blockage the valve empties but does not refill when the pressure is released. Skull and chest x-rays will help to exclude disconnection of the catheter. A CT scan may help in diagnosing a persistent partial

blockage of the system but the scan is often normal when the block is acute. A cap tap to measure intracranial pressure can be done but is an invasive procedure. Continuous measurement of the ventricular pressure via the Rickman cap is feasible and may be useful[1310] but not all children have a cap as part of their shunt system and the procedure is invasive.

When the distal catheter requires replacement and all the neck veins have been used before, the use of every conceiveable site has been described - for example, the pleural cavity, stomach, ureter or right atrium via direct puncture, successful control of the hydrocephalus becomes increasingly difficult and the outlook correspondingly grave.

Infection of the shunt may present with progressive anaemia, splenomegaly and pyrexia. Shunt nephritis also occurs. Frank septicaemia and ventriculitis are uncommon. Treatment is by removal of the entire shunt system and delayed or immediate replacement when the infection has been cleared by antibiotics.

Other shunt complications include perforation of the heart or abdominal viscera, necrosis of skin over the valve, subdural haematoma, craniosynostosis. and pulmonary emboli:- the latter may cause pulmonary hypertension and cor pulmonale. 74% of patients with a shunt require its revision at some time, and 55% of patients with shunts in spina bifida require revision within one year of their primary operation[14]. However, in many centres shunts are replaced prophylactically at intervals in all children (see Methods section). A total of 20 shunt revisions by the age of 12 is not all that uncommon[15].

The history of the treatment of spina bifida shows that the crucial factor is the management of associated hydrocephalus. Yet there is still no reliable method which allows intracranial pressure to be monitored by non-specialised doctors [16].

The Selective Policy

Early closure of the back combined with ventriculo-atrial or ventriculo-peritoneal shunting was enthusiastically practiced from the 1960's and the survival rates of infants with spina bifida and hydrocephalus greatly increased. However in 1971 it was shown that in spite of the progressively increasing survival rate the problems created were greater than those solved. Only 50% of those treated had survived [17]. Infection, hydrocephalus and renal failure were the common causes of death. There was a 2% annual mortality rate and 80% of the survivors had severe multi-system handicaps including incontinence, paraplegia, skeletal deformities, fractures, dislocations, trophic ulcers, blindness, fits and mental handicap. Few children would ever achieve competitive employment [18].

Since then infants with large high lesions, severe paraplegia, kyphosis, gross hydrocephalus or other malformations present at birth have not been treated surgically, the object of selection being to avoid treating those who would survive with very severe handicaps who, untreated, will almost certainly die during the first weeks of life [14,19]. The occasional child who survives a trial period of non-treatment usually develops hydrocephalus which then requires treatment [20,21].

Since the advent of the selective policy, genetic advice, amniocentesis and therapeutic termination of pregnancy, the number of children heavily handicapped by spina bifida has fallen, although some of the decline appears to be unrelated to these factors[22]; the national notification rate for spina bifida fell from 1.88 to 1.04 per thousand life births and still births between 1972 and 1981. Nevertheless, a cohort of those treated during the earlier years of active optimistic intervention remain to stretch the resources of the Health and Social Services.[23] Unemployment and social isolation are major problems in those reaching adulthood.

Occipital encephalocele. This is a herniation of meninges, or meninges and brain, through a defect in the occipital region of the skull. The size of the encephalocele varies and with the larger lesions the intracranial part of the brain is often malformed. The greater the volume of brain included in the lesion the worse is the prognosis. Early operation, as for spina bifida, is usually advised. Treatment of the associated hydrocephalus is required.

Occipital (myelo) meningocoele consists of a herniation of meninges (and neuroglial tissue) only.

Anencephaly. Anencephaly is due to a failure of fusion of the anterior neuropore and results in gross malformation. The infant is stillborn or dies within twenty four hours.

Primary hydrocephalus. A group of children often managed alongside those with spina bifida and hydrocephalus are those with primary hydrocephalus. This useful term may be applied to patients with congenital hydrocephalus which is not obviously secondary to neoplasm, meningitis or the Arnold Chiari malformation. Some centres (including Wessex) even use the term to include children with post meningitis hydrocephalus, but not tumoral hydrocephalus. Doubtless many children with "primary" hydrocephalus who are examined at post mortem could be shown to have a definite cause for their hydrocephalus e.g. aqueduct stenosis, the Dandy Walker syndrome, intrauterine infection, venous sinus thrombosis, choroid plexus papilloma and many more may have a communicating hydrocephalus secondary to birth trauma with intracranial haemorrhage[24]. Primary hydrocephalus is much less common than hydrocephalus associated with neural tube defects.

Hydranencephaly.- This is the extreme degree of hydrocephalus in which there are virtually no cerebral hemispheres, only basal ganglia and remnants of the mesencephalon above the tentorium. The long term prognosis is poor whether or not cerebrospinal fluid shunting is instituted. The present view is that hydranencephaly reflects profound cerebral hemisphere infarction in utero.

Porencephaly. This is a condition in which there is a focal loss of cortical tissue from the brain. The space left as a result is usually occupied by fluid which communicates with the ventricular system.

Ophthalmic complications of spina bifida, spina bifida with hydrocephalus and primary hydrocephalus.

This section reviews the existing literature on visual function and ophthalmic complications in these disorders. The findings in encephalocele, anencephaly and hydranencephaly are discussed separately.

Orbital anomalies. In untreated congenital hydrocephalus the dome of the skull may become so large that the eyes are displaced downwards and proptosis results[25]. The orbits become widely separated and depressed while the eyes are deep set with overhanging brows. Hydrocephalus is now usually controlled by shunting at an early age so that such orbital changes are rarely seen. Rabinowicz[26] found no gross alteration in orbital contour and commented that any downward displacement of the eye appeared to be due to paresis of upward gaze and disappeared when intracranial pressure was relieved.

Exposure keratitis. Corneal scarring is one cause of visual impairment in children with spina bifida[27]. In a survey of 100 school children with spina bifida four had corneal scarring[28]. Goddard[27] found two cases of corneal ulceration requiring tarsorraphy in her series of 251 children (0.8%). She commented that this complication is liable to occur at any age when the child's general condition is sufficiently poor. Such ulceration is rapidly progressive and may cause scarring with considerable visual impairment and a predisposition to recurrent corneal erosions.

Ocular motility disorders.

Setting sun phenomenon. True sunsetting is a forced downward deviation of the eyes so that the lower lid overlaps the cornea and a rim of sclera shows above [29]. It is a classic sign of hydrocephalus and kernicterus but may also occur in premature babies and transiently in normal full term babies. It may appear spontaneously or when one changes the infant's position. A child in whom sunsetting persists for more than three months will usually be found to have serious cerebral damage [30,31]

The setting sun phenomenon was originally thought to be mechanical in origin due to downward displacement of the eyes by the altered direction of the orbital plates, but even in gross hydrocephalus it may be corrected by surgical relief of the hydrocephalus [32] so it is the result of a pressure effect on the brain stem in the region of the anterior end of the third ventricle [33]. The presence or absence of the setting sun phenomenon may be a useful index of shunt patency [32].

Rabinowicz [26] elicited a history of sunsetting in 10% of his series of 100 hydrocephalic children but he suggests that this is an under-estimate of its incidence. 50% of those with a history of the setting sun phenomenon had unilateral or bilateral optic atrophy. It is thus a prognostic sign to be taken seriously and may be a reflection of both the frequency and degree of raised intracranial pressure in infancy. Rabinowicz did not observe the phenomenon after eighteen months of age. His findings also confirm the neurological explanation since he observed no gross alteration in orbital contour in his patients and found that

recovery to the normal position of the eyes occurred when raised intracranial pressure was relieved, the phenomenon recurring if the intracranial pressure rose again.

Goddard[29] found the setting sun phenomenon in 12% of hydrocephalic children under six months but in only one child older than this. She was able to observe gradual resolution of sunsetting in some patients and thought that this was due to effective treatment of the hydrocephalus or perhaps to death of those affected by it.

Cobbs and co workers[34] have also found forced downgaze in adults with decompensated hydrocephalus. Swash[32] described a common syndrome of disordered ocular movement in hydrocephalus which he called 'divergence with downward deviation'; in addition to resting downward deviation of the eyes there was absence of oculocephalic and optokinetic upward gaze and sometimes large, sluggishly reacting pupils and upper lid retraction. He postulated that this was due to dysfunction of decussating fibres in the posterior commisure caused by hydrocephalic distortion or compression from caudal herniation of an enlarged suprapineal recess. The sylvian aqueduct syndrome seems to be a partial manifestation of the disorder.

Sylvian aqueduct syndrome (Parinaud's syndrome)

This second syndrome of disturbed ocular motility occurs in older patients with hydrocephalus and seems to represent a partial manifestation of the disorder causing the setting sun phenomenon. Parinaud's syndrome consists of paralysis of voluntary conjugate vertical eye movements with or without pupillary anomalies and is due to a hypothetical supranuclear vertical

gaze centre lesion in the tectal or pretectal region of the mesencephalon[35]. It was once considered pathognomonic of a mass lesion in the region of the pineal gland or posterior third ventricle, but may also occur in decompensated hydrocephalus[33].

The full sylvian aqueduct syndrome, in addition to impaired conjugate upgaze and pupillary anomalies, consists of convergence nystagmus on attempted upgaze, retractory nystagmus, vertical nystagmus on downgaze and palsies of extraocular muscles[32]. This is due to distortion of periaqueductal structures by assimilation and enlargement of the rostral aqueduct with additional compression of this region either by herniation of an enlarged suprapineal recess or by secondary tentorial block from caudal herniation of the third ventricle and brain stem, occurring when hydrocephalus decompensates.

Shallat[36] confirmed that Parinaud's syndrome may be an early reliable sign of shunt malfunction and attributable to compression of the quadrigeminal plate by a dilated suprapineal recess with compression of the aqueduct. A cerebrospinal fluid shunt breaks the cycle and allows the aqueduct to open. Shallat[36] described four patients with hydrocephalus who developed this syndrome; all had upgaze palsies and one also had pupillary anomalies. Three of the patients had several episodes, all of which resolved when their hydrocephalus was controlled.

Cobbs, Schatz and Savino[34] described four adults with upgaze palsy, retractory nystagmus, forced downgaze and pupillary light-near dissociation. They attributed these signs to dilatation of the posterior third ventricle with assimilation of the aqueduct and downward pressure of

the ballooned supra-pineal recess. They suggested that prior episodes of third ventricular enlargement demonstrated in two of these cases, predisposed them to emergence of eye signs before apparent venticular enlargement.

Hemmer and Bohm[12] recorded one case of Parinaud's syndrome in obstructive hydrocephalus which decompensated. Copighnon and co workers[37] reviewed twenty eight published cases of Parinaud's syndrome due to hydrocephalus,without underlying tumour, some cases proven by autopsy. Most had aqueduct stenosis. They presented observations on two cases who developed Parinaud's syndrome cured by shunt revision. They emphasised that relief of shunt obstruction had to be prompt if irreversible changes in the brain stem were to be avoided.

Squint. Squint is a very frequent feature of congenital hydrocephalus. The incidence quoted differs somewhat. Goddard[27] found an incidence of 30%, Harcourt[38] 30%,Hunt[28] 59%, Rabinowicz[26] 54% Clements and Kaushal[24] 77 of 105 (i.e. 73%) Tew and Laurence[39] 18 of 55 (i.e. 33%). The variation is not surprising as the groups studied are not fully comparable in respect of age, diagnostic breakdown and mode of selection for study, (Table 1).

Clements and Kaushal[24] divided their cases of spina bifida into those with and those without hydrocephalus. Although they found that squint was commoner in those with hydrocephalus than in those without, nevertheless 52% of their cases with spina bifida without hydrocephalus had a squint. They concluded that many of these cases may have had subclinical hydrocephalus. They found no difference in the type of squint in the two groups.

The incidence of squint remains noteworthy when one considers that only two to three per cent of the general population squints[40]. The incidence of squint in spina bifida and hydrocephalus is similar to the incidence in children with cerebral palsy.

Convergent Squint. This was the commonest type of squint in the series. Rabinowicz[26] found an incidence of 44%, and Clements and Kaushal[24] 67%. The squint usually started early in life with a unilateral or bilateral lateral rectus palsy[26,29,41]. This was most often due to an acute rise in intracranial pressure and sometimes disappeared with relief of hydrocephalus[41]. An increased angle of squint was repeatedly seen as a sign of raised intracranial pressure[26] and was sometimes the first unequivocal sign of raised pressure[38, 38a]. After a prolonged episode of raised intracranial pressure the squint did not always revert to its previous angle[26, 38, 38a]. Harcourt also considered that lateral rectus palsy may have been the result of an abnormal antenatal development or meningitis. Mental retardation and defective vision were other possible underlying factors.

Though most squints are paretic in origin, concomitance develops early[26] and the squint is usually alternating in character. Fully accommodative squints are uncommon (13%[29, 24]) as are abnormal retinal correspondence and eccentric fixation[26]. Amblyopia is rare (3%[26], 1.5%[28, 24]).

A vertical element to the squint is a common finding (30% of over 2 year olds [29]). This may be secondary to large horizontal deviations or to involvment of third and fourth cranial nerves [24]. The A syndrome is extremely common. Rabinowicz found a 50% incidence [26]. It may be due to primary underaction of the lateral rectus muscle [26, 41, 42] or associated with downdrift in adduction due to a weakness of the inferior obliques [41]. Abduction defects and gaze paretic nystagmus are common as would be expected [26].

Squints are common in children of lower intelligence [28]. There is a significant association between ocular defect (mainly squint) and poor school performance [39]. Squint is more than a cosmetic disadvantage; it can affect the rate and quality of learning and may be an important determinant in eventually gaining employment [39, 27].

Treatment of convergent squint. Treatment of squint and amblyopia are often neglected when other severe handicaps are present [28,39]. Nevertheless all authors are agreed that treatment of squint should receive as much attention as in normal children. Goddard [29] alone suggested that surgical intervention should be early, and not delayed until hydrocephalus has arrested, on the grounds that intracranial conditions may be changing. This is a surprising view as a convergent squint may disappear with relief of hydrocephalus [38, 41, 26]. Wybar [41] more rationally suggested (as with any paretic squint) waiting until the squint has become concomitant and stable and until there is no prospect of alternation developing. He then prefers to recess the medial recti because lateral rectus resections may be wasted if hydrocephalus recurs. Clements and Kaushal [24] also proposed bimedial rectus recession as the

more logical procedure. If an A phenomenon is present surgical intervention should be planned to treat this also. If there is downdrift in adduction Wybar[33] suggested that the superior obliques should be weakened by partial posterior tenotomies, but if there is no downdrift (so that the phenomenon must be due to lateral rectus weakness) the insertions of medial recti should be raised at the time of recession.

Clements and Kaushal[24] reported five functional results in 22 operations and Goddard[29] one in three operations.

Divergent Squint. Many of these cases have diminished visual acuity often due to optic atrophy[24,26]. There is frequently a history of multiple procedures including shunt revisions and repeated urinary infections[26]. A few follow surgery for convergent squint (2 out of 10[24], 2 out of 10[24]). Divergence may follow convergence spontaneously[24].

Nystagmus. Nystagmus is generally agreed to be common in congenital hydrocephalus. Goddard[29] found horizontal and rotary nystagmus especially in patients with optic atrophy. Rabinowicz[26] found nystagmus in 30%(28% musculo paretic, 7% micronystagmus, 4% pendular). Those with pendular nystagmus usually had very poor vision, often less than 6/60, and those with micronystagmus usually had some visual impairment. The musculo-paretic nystagmus he noted was a manifestation of lateral rectus weakness and was often associated with a convergent squint. Clements and Kaushal[24] found nystagmus in 13 out of 130 cases (10%) mostly on lateral gaze; three had rotary nystagmus and one, latent nystagmus.

Retractory nystagmus has been mentioned already as part of the Sylvian aqueduct syndrome.

Refractive Errors. Refractive errors are not particularly common considering the high incidence of squint and the fact that nearly 50% of children with cerebral palsy (who have a similar incidence of squint) have refractive errors [40]. Hunt [26] found that 22 of 100 school children wore spectacles. Twenty five per cent of Tew's [39] series of 55 children (who were nearing their tenth birthday) had a refractive error requiring spectacles. Clements and Kaushal [24] found 7 out of 43 cases with convergent squint (16%) had significant refractive errors (anisometropia, hypermetropia greater than three dioptres or astigmatism). Fully accommodative squints are rare and correction of refractive errors does not appear to be an important part of squint management. Nevertheless refraction should always be done because of the help it may give in an individual case.

Papilloedema. Most authors have found papilloedema to be a rare event in congenital hydrocephalus. The great variety of reasons given for this is not surprising since the precise pathogenesis of papilloedema is still a controversial subject [43]. Hayreh [44] stated that papilloedema occurs only when the subarachnoid cerebrospinal fluid pressure is raised and not in obstructive hydrocephalus with distended ventricles. Goddard [29] attributed the rarity of papilloedema in congenital hydrocephalus to the ease with which the infant skull expands; this would indicate that the generalised intracranial pressure in congenital hydrocephalus is seldom raised. Nevertheless the intracranial pressure must by definition be raised at sometime in hydrocephalus, whether it is communicating or

obstructive, and indeed continuous intraventricular pressure monitoring may be used to diagnose uncontrolled hydrocephalus[11,12]. Perhaps, however, the intracranial pressure is never enough to cause papilloedema while the skull remains expansile.

Early insertion of the shunt is now often followed by premature skull fusion, so that the skull soon becomes non-expansile. Then shunt obstruction may be followed by an abrupt rise in intracranial pressure. Goddard[29] found one patient (out of a series of 251) with bilateral papilloedema which resolved with surgical relief of the obstruction. Rabinowicz[26] found 2% of his 100 cases had papilloedema and in one patient a solitary haemorrhage near the disc was seen prior to the diagnosis of shunt obstruction. Papilloedema developed in the presence of optic atrophy in one of his patients. Harcourt[38] described two cases (out of 70 = 3%) with papilloedema due to mechanical obstruction of their shunts; it resolved in both cases after shunt revision.

Pierre-Kahn et al[45] found that either an abnormal increase in head circumference or papilloedema, or both,were present in all children whose resting intracranial pressure exceeded 25cm of water. Plotkin and Smith[46] described four adult cases with normal pressure hydrocephalus (NPH), two of whom had papilloedema. Shallat et al[36] described four patients with Parinaud's syndrome in hydrocephalus and two of these also had papilloedema. Wybar[33] asserted that, althogh papilloedema is rare, it must be taken very seriously when it does occur. Papilloedema may be due to raised pressure in the infratentorial compartment of the skull even when the lateral ventricles are successfully drained by a shunt; thus a CT scan showing small ventricles may be misleading. Papilloedema may occur

on top of optic atrophy if arrested hydrocephalus decompensates . The atrophic nerve head is probably more resistent to oedematous changes so that papilloedema supervening on an atrophic disc usually reflects markedly raised intracranial pressure. More recently Ghose[47] found a 14.5% incidence of papilloedema in 200 consecutive cases of congenital hydrocephalus. He included suspected and incipient papilloedema in his 14.5%, as well as cases of developed papilloedema who still had pulsations of the central retinal vein. He may therefore have included cases which other authors would have excluded.

Papilloedema can be, but need not be, the prelude to serious visual decline[26]. One of Harcourt's[38] two cases went on to develop optic atrophy; the other child's papilloedema resolved without impairment of vision.

Optic Atrophy Optic atrophy is generally agreed to be common. Rabinowicz[26] reported an incidence of 31% although the optic atrophy was sometimes unilateral. An assessment of the true incidence is difficult because the optic disc is always pale in infants[29]. The vision may vary from 6/9 or better (28%) to perception of light. Sixty per cent of children with atrophic discs have a vision of 6/12 or worse[26]. Though optic atrophy may co-exist with good vision, these children are particularly vulnerable as a number of those with severe visual deterioration had shown marked optic atrophy with good vision at an earlier stage.

Optic atrophy is most often attributed to stretching of the optic nerves or their vascular supply[29,38,33]. This occurs when the brain stem shifts in position. A sudden drop in intracranial pressure, following a

shunt operation, may cause just such a change in position. Optic atrophy may also be due to an associated developmental anomaly. It may be secondary to raised intracranial pressure or meningitis[33]. Goddard[29] saw six patients with raised intracranial pressure developing after closure of the anterior fontanelle; they all later developed optic atrophy. Two of Harcourt's[38] four cases developed optic atrophy after shunt obstruction. One of these had had papilloedema; the other had not.

Optic atrophy is one of the main causes of blindness in hydrocephalus[27], although cortical blindness may co-exist[48, 49]. Such children frequently have specific learning difficulties (apart from those due to poor vision) because of associated brain damage[27].

Cortical Blindness. This may be another cause of visual failure in congenital hydrocephalus[38,26]. It is probably the result of oedema and swelling of the brain, secondary to hypoxia, septicaemia, meningitis, multiple operations, trauma and convulsions. Both posterior cerebral arteries are occluded on the tentorial edge[48, 33]. It may develop very suddenly. It may co-exist with optic atrophy. The diagnosis is only established with certainty by an absent visual evoked response.

Blindness. Lorber[49] described 13 cases of blindness in congenital hydrocephalus. This was presumably cortical blindness since only one had pale discs, according to the practitioner,who was not an ophthalmologist, who examined them. Blindness was precipitated by raised intracranial pressure in five (due to shunt obstruction or untreated hydrocephalus) and to shunt infection in eight. This blindness sometimes recovered months later.

Keen[50] found 17 children with myelomeningocoele and shunt-treated hydrocephalus who became blind over a period of one week to two years. Sudden blindness commonly followed shunt revision for acutely raised intracranial pressure. The blindness was sometimes transient so that in this situation urgent treatment of the raised intracranial pressure is mandatory. Some children had suffered gradual visual deterioration which highlights the need for regular ophthalmic examination including testing of the visual field. Keen[50] did not differentiate between blindness due to optic atrophy and cortical blindness. Rabinowicz[26] and Welch[51] also noted that blindness may occur acutely or insidiously, due to shunt malfunction or to operations on the shunt.

Visual evoked potentials. The absence of a visual evoked response (VER) is the only certain proof of cortical blindness. Lawton Smith et al[48] found absent visual evoked responses in two of their five cases with presumed cortical blindness in hydrocephalus. Sklar et al[52] found abnormal VERs to repetitive flash stimuli in ten patients with hydrocephalus; abnormalities included latency delays, fatiguability and asymmetries. Latencies and wave form disturbances improved after shunting. Clinical progression of the hydrocephalus occurred in several patients and this correlated well with worsening VERs. Ehle and Sklar[53] studying VER to flash stimuli in 15 patients with hydrocephalus showed increased latencies for P2 in all, compared with age and sex matched controls. P2 latency decreased after shunting.

Rapid eye movement (REM) sleep. In 30 children with more or less arrested hydrocephalus, intracranial pressure rose during REM sleep [45]. The intracranial pressure rise was a consequence of altered breathing during REM sleep.

Chorioretinitis. The association of hydrocephalus with congenital toxoplasmosis, chorioretinitis and intracranial calcification is well known. Nevertheless chorioretinitis is relatively uncommon in children with hydrocephalus. Goddard [29] found one patient in her series of 251, Rabinowicz [26] one of 100. Only 20 - 30% of the patients in these two series had hydrocephalus unassociated with neural tube defects.

Giles and Lewis [54] found five out of 31 (16%) institutionalised patients with advanced idiopathic hydrocephalus had extensive chorioretinitis, yet none of these could be proved by uveitis survey or pathological or bacteriological examination (in two cases) to have toxoplasmosis.

Other retinal lesions in hydrocephalus. Gerhard et al [55] described a family of four children, three of whom had hydrocephalus and died early. In two of these a histological examination of the eye showed retinal dysplasia, persistent hyaloid artery and defects of the primary and secondary vitreous. Warburg [56] described a patient with hydrocephalus who also had congenital retinal non-attachment and falciform folds causing near blindness. He was mentally retarded. His parents were first cousins which suggests an autosomal recessive inheritance.

Other ophthalmic anomalies in spina bifida and hydrocephalus.

Anophthalmos, microphthalmos, colobomas, absence of the optic nerves and ganglion cells of the retina, absence of the macula, optic atrophy and Horner's syndrome may occur in spina bifida and hydrocephalus. Powell[57] described a case of anterior lenticonus occurring in an infant with extensive thoraco-lumbar meningomyelocoele who survived for three days. Nasolacrimal duct obstruction occurred in about 8% of cases[29] which is similar to the incidence in normal infants.

Visual Acuity. Rabinowicz[26] found that 37% of his 100 patients had an acuity of 6/6 or better, 85% had 6/12 or better in one eye, 24% saw 6/18 or less in one eye and 7% had an acuity of 6/60 or worse. Thus only one third had "normal" vision and 7% were eligible for blind registration. He found blindness to be associated with marked dilatation of the third ventricle, infection (CSF or urinary), chiasmal traction, consecutive optic atrophy or cortical blindness. Blindness occurred suddenly or insidiously. He suggested that patients who already have optic atrophy are particularly vulnerable to raised intracranial pressure and may become blind quite rapidly from effects on their optic nerves or visual cortex. The 24% with an acuity of 6/18 or worse in one eye were a vulnerable group who almost always had optic atrophy.

Hammock et al[9] noted that visual acuity often improved after shunting of patients with normal pressure hydrocephalus associated with spina bifida.

Hunt[28] studied 100 school children with spina bifida. Of these 64 had visual defects and two were educationally blind.

Visual Field examination. Visual field examination is generally agreed to be important in these children. It is, nevertheless, often difficult if not impossible to perform and figures are often incomplete. Rabinowicz[26] found field defects in 9%; they included homonymous hemianopias and homonymous inferior quadrantinopias, suggesting posterior involvement of the optic radiation. Four per cent had extinction defects, where one side of the field is preferred and the other suppressed. He used composite pictures to detect these defects.

Pressure of the distended third ventricle on the chiasm may lead to bitemporal hemianopia[43]. Homonymous hemianopia due to infarction of the calcarine cortex when the posterior cerebral artery is stretched over the tentorium cerebelli during an episode of coning is rarely seen, presumably because the patient is usually in coma when this occurs[38].

Visual perception. At least 10% of children with hydrocephalus have perceptual problems[26]. They find it difficult to reject irrelevant information from a complex display (figure-ground discrimination). They copy shapes and complete figures poorly and tend to reverse letters or shapes (reverse orientation)[58]. They have problems with recognising left and right, top and bottom[27]. Those with ocular defects (mainly squint) in particular, have a specific weakness in visuo-spatial and visuomotor functioning revealed by a Wechsler performance IQ, although those without ocular defects still have a lower IQ score than normal controls[39].

Difficulty with shape recognition is a prime factor in learning disability. These children have impaired reading skills as well as

problems in dressing and sports . Visual perception scores in spina bifida and hydrocephalus can be improved by following the Frostig programme for the development of visual perception.

Perceptual problems probably represent damage to the projection areas of the visual cortex. Some of the patients also have constructional apraxia,dyscalculia and homonymous hemianopia which suggests involvement of the optic radiations and the parietal and parieto occipital cortex [26].

The Effect of Ophthalmic Problems on the Quality of Life. Children with ocular defects perform badly on intelligence, reading and mathematical tests and this seriously reduces their chances of future employment. 40% of a group of ten year olds with neural tube defects or hydrocephalus come into this category [39].

Visual problems add to difficulties at school. One of 100 school children attended a school for the blind and one was in a special class for the blind in a physically handicapped school. Twenty two needed spectacles - yet another item of equipment to be remembered by those already encumbered with calipers, sticks, wheelchairs, urinary appliances etc. Teachers may need to give extra help because of difficulties in spatial perception and eye-hand coordination. Those with squints (59%) appeared to have lower intelligence [28].

Blindness may bring unbearable problems and anxieties to the multiply handicapped child and his family [27]. The blind child's world contracts and anaesthetic parts of the body "disappear" since they can be neither seen nor felt. Learning is characteristically retarded. Braille is

especially difficult for those with perceptual problems since they find it hard to learn the comparative position of the six dots in the braille cell. Perception through touch sometimes takes a long time to develop. The child will take much longer to become independent, and being less mobile is especially likely to become obese.

The Role of the Ophthalmologist in hydrocephalus

Many authors agree that ophthalmic complications of spina bifida and hydrocephalus have been overlooked in the past and have emphasised the importance of frequent eye examinations [50,24,33,39,11] with special attention being paid to the range and extent of ocular movements, the presence of squint, the appearance of the optic discs and the field of vision [38]. The VER also may be a useful non-invasive technique for following patients with CSF shunts and those with presumed arrested hydrocephalus [52].

Ocular signs may be an early and reliable indicator of increased intracranial pressure due to shunt malfunction [36] and may be more reliable than CT scanning [34].

Now that the life and IQ of spina bifida and hydrocephalic children can be preserved, retention of normal vision becomes of paramount importance and the ophthalmologist may assist the paediatric surgeon to this end by preventing corneal scarring from exposure keratitis, by confirming papilloedema and ensuring early surgical intervention to prevent its progression to optic atrophy, and by treating ocular deviations and amblyopia [29]. The ophthalmologist should be part of the team serving the child and his family [1].

29

Goddard has also highlighted the need for a long term study correlating the general condition with the presence and development of ocular lesions, to establish a pattern of eye diseases and provide a basis for further investigation.

Despite these beliefs, few centres are organised to monitor the effects of deteriorating eyesight[15] and none of the studies available have been long term.

Ophthalmic features of Occipital Meningocoele and Encephalocoele.

Lorber[60] studied 45 cases with occipital encephalocoele and ten with occipital meningocoele. The presence or absence of neural tissue in the sac was the single most important prognostic factor. Hydrocephalus was a serious associated malformation. Occipital encephalocoele had a high mortality rate. Of twenty survivors, five were blind, four were partially sighted and one had a coloboma (site not specified). None of the ten occipital meningocoeles were blind or partially sighted. Septo-optic dysplasia may occur in occipital encephalocoele[61].

Ophthalmic features of Anencephaly. Hypoplasia or atrophy of optic ganglion cells, the nerve fibre layer and the optic nerves are typical pathological findings[61,62]. The retinal pigment epithelium and inner nuclear layer is normal. Atypical findings are uveal coloboma, retinal dysplasia, corneal dermoids and malformation of the anterior chamber angle.

Ophthalmic features of Hydranencephaly. Three cases studied clinically by Hill[63] showed absent visual responses apart from a dazzle reflex, sluggishly reacting pupils, white discs, avascular retinas and full ocular movements. He concluded that the blindness must have been cortical and the dazzle reflex a subcortical phenomenon. One of these cases was subjected to pathological study; retinal gliosis and absence of retinal vessels were found.

Jones[64] examined one case at four and twenty one months of age. Electroretinography (ERG) was normal and a VER was present. The visual performance improved subjectively and electrophysiologically between the two examinations. The discs were pale but the macula and the retinal blood vessels were normal. Septo optic dysplasia may occur in hydranencephaly[61].

Patients, Materials and Methods.

Patients. The study was based on 322 children seen over the six year period 1977 to 1983. Their diagnoses are shown in Tables 2 and 3.

Patients with spina bifida and arrested hydrocephalus, who had never been shunted were classified separately from those with spina bifida and hydrocephalus treated with a shunt.

The patients were all children aged 0 to 18 from the Wessex area who were attending Southampton General Hospital as either inpatients or outpatients. Every effort was made to see all children with these diagnoses whether or not they had, or were likely to have, ophthalmic abnormalities. Whenever possible they were seen regularly, at each clinic visit, starting either at the time of birth or when the diagnosis was made. In addition, they were seen on an emergency basis in the clinic or on the ward, in the hope that ophthalmic examination might assist in the diagnosis of raised intracranial pressure. However, children from remote areas of Wessex have their outpatient care undertaken at peripheral hospitals and only come to Southampton General Hospital for inpatient care and operations. Hence they could only be seen on the ward and examination of their eyes was necessarily sometimes incomplete. Anencephalics and children who did not survive the first few hours or days of life were not included in the study.

Care of children with spina bifida in Southampton follows conventional lines. The selective policy was begun in 1971 but was not fully implemented until the end of 1973. Only about 25% of children now come to early operation. About 65% develop hydrocephalus and, after computed tomography, a ventriculo-atrial shunt (Spitz-Holter type) is inserted, usually between the second and fourth week of life. Shunts are revised prophylactically at 18 months, 4-6 years and 8-12 years depending upon the clinical findings. Thereafter revision is only undertaken when necessary. Ventriculo-peritoneal and ventriculo-pleural shunts are sometimes used at revision operations when ventriculo-atrial shunting is impossible. The urinary tract is examined regularly by midstream urine examination, intravenous pyelography and micturating cystography. Urinary incontinence is managed by an appliance for boys, intermittent or continuous catheterisation for girls and occasionally urinary divertion. All children are seen by an Orthopaedic Surgeon. At the age of 18 all are passed to the care of Neurosurgeons and Urologists.

Methods. Where possible, children were examined at the start of the outpatient session before other tests and examinations, so that they were fresh and co-operative. At the first visit a history was taken with particular reference to the presence of headaches, vomiting and behaviour disturbance, visual disturbance, past ophthalmic history, family history of squint and amblyopia, the occurrence of sunsetting and parental assessment of visual function. Hospital notes were perused for past observation of sunsetting and papilloedema. Visual acuity was determined with the Snellen chart at six metres in older children or the Sheridan

Gardner test in children aged two and a half to four years. Vision in younger children was assessed by observing the response to Catford drum symbols, the ability to locate and pick up "hundreds and thousands" sweets and by eliciting following, menace, dazzle and preferential looking reflexes.

If the visual acuity was less than 6/9 in either eye, retinoscopy and subjective refraction were performed and spectacles prescribed if clinically indicated. Retinoscopy was also performed whenever the pupils were dilated for fundoscopy.

Confrontation field testing to hand movements or graded white hat pins was performed in children old enough to co-operate. In younger children binocular field testing to hand movements or observation of the response to toys moved in the peripheral field was performed.

The presence of a squint was determined by observation and cover testing. Particular regard was paid to the presence of an A syndrome. Eye movements and the presence of nystagmus were assessed as the patient followed a torch light and convergence from distant to near targets was observed and measured. Stereopsis was investigated with the Wirt stereo test.

Pupil responses to a torch light and accommodation from distant to near objects were assessed in the usual way. Pupil size during distant fixation in an illuminated room was assessed with a graticule.

The optic discs and peripapillary area were examined with a direct ophthalmoscope. If this was impossible, drops of cyclopentolate 1% (or 0.5% in infants) were instilled and the fundi examined with the direct or indirect ophthalmoscope. Complete fundus examination through dilated pupils with the indirect ophthalmoscope was also attempted whenever the cause of hydrocephalus was unknown. Otherwise mydriatics were used as little as possible as the stinging they induce adversely affects the doctor/patient relationship.

At subsequent visits a further brief history was taken and examination repeated with respect to visual acuity, squint, eye movements, pupils and optic discs. Where full examination had been incomplete at the first visit because of age or non co-operation, further investigation was usually possible at subsequent visits. Stereopsis and visual fields were investigated periodically. All observations were performed by the author.

Several children were already under the care of other Ophthalmologists and Orthoptists. In these cases detailed measurements of squint angle and stereopsis were requested.

Selected patients who were old enough to co-operate were referred for fundus photography and fluorescein angiography. The dose of fluorescein given was 9.5mgm/kg body weight and a West German Zeiss camera used with Kodak Ektachrome GPR 133-36 film. Selected children who were old enough to co-operate were refered for study of their visual evoked potentials and brain stem auditory evoked potentials. They were recorded on the Nicolet Path Finder II Evoked Potential Analyser.

Raised Intracranial Pressure Study.

The aim of this study was to determine the percentage of children with uncontrolled hydrocephalus who have ophthalmic evidence of raised intracranial pressure and to correlate the ophthalmic and general findings with the nature and duration of the obstruction and the height of the CSF pressure.

Cases were analysed if the following criteria were fulfilled;

1) An eye examination had been performed during a period of suspected raised intracranial pressure due to uncontrolled hydrocephalus.
2) At operation they were found to have a shunt obstruction requiring its revision or to have raised intracranial pressure requiring the insertion of a shunt for the first time.

Cases were selected for the study merely on the basis of their availablity for study of their eyes before operation. Many patients with acute symptoms came to surgery before such examination could be performed and could not be included in the study. Findings were classified as (a) positive - there was ophthalmic evidence of raised intracranial pressure or (b) negative - no such ophthalmic evidence.

Survey of Other Centres.

In September 1981 a letter (Appendix II) was sent to leading centres for the management of spina bifida and hydrocephalus to discover whether they employed routine ophthalmological screening of their patients, and if so, the nature and extent of such screening. The Paediatrician or Paediatric Surgeon concerned was asked (a) what proportion of children were seen by an ophthalmologist (b) how many ophthalmologists were involved (c) whether children were seen by an ophthalmologist and paediatrician during the same session.

The following definitions were made in order to classify the findings of the study.

Sunsetting. Forced downward deviation of the eyes, intermittently or continuously present.

Parinaud's syndrome. Some or all of the following - upgaze nystagmus, upgaze palsy, dilated pupils, downgaze nystagmus, downgaze palsy. (Table 5).

Convergent squint. A convergent squint manifest at least some of the time or in some position of gaze, at sometime during the study and not subsequently becoming or remaining divergent.

Divergent squint. A divergent squint manifest at least some of the time, or in some position of gaze, and never known to be proceeded by convergence.

Consecutive divergence. A manifest convergent squint becoming and remaining a manifest divergent squint either spontaneously or following squint surgery.

Amblyopia. A difference in Snellen visual acuity between the two eyes of one line or more, not corrected by glasses and without a structural cause.

Anisometropic amblyopia. Amblyopia due to unequal refraction of the two eyes.

Internuclear Ophthalmoplegia. Restricted adduction with nystagmus on abduction.

Lateral rectus palsy. Absent abduction of the eye.

Lateral rectus paresis. Reduced abduction of the eye

Musculo-paretic nystagmus due to lateral rectus paresis. A jerk nystagmus present on abduction of the eye.

Significant refractive error.

Myopia. greater than or equal to -1D

Hypermetropia greater than or equal to +2D

Astigmatism greater than or equal to 1D

Papilloedema. Unilateral or bilateral optic disc swelling with or without haemorrhages.

Optic atrophy. Unilateral or bilateral optic disc pallor.

Cortical blindness. Blindness in the presence of normal pupils and optic discs or in excess of that to be expected from pupillary and disc findings.

Roth's spot. a retinal haemorrhage with a pale centre.

Visual handicap.

Normal. Snellen visual acuity of 6/6 or better in each eye + full fields and stereopsis.

Minor. Snellen vision less than 6/6 in one or both eyes, but better than 6/18 or manifest squint, or absence of binocular vision or minor field defect.

Moderate. Snellen visual acuity 6/18 to 6/36 or major field loss in one eye.

Major. Partially sighted or blind, i.e. vision less than 6/36 in both eyes or major field loss in both eyes.

Unclassified. Snellen visual acuity not known because of age, mental handicap or incomplete eye examination.

General Handicap.

None No handicap.

Mild Orthopaedic problems e.g. use of stick or caliper or perceptual or emotional problems only.

Intermediate Occasional use of wheelchair. Well functioning diversion or appliance or hemiplegia, or mild mental retardation.

Severe Mainly confined to wheelchair and other problems or mental retardation, fits or incontinence.

Very severe Chair bound with other major problems, e.g. blindness, fits, renal failure, or severe mental retardation.

Not classified (NC) General handicap not assessed because of age or insufficient information.

The classification of general handicap for spina bifida children was taken from that of Castree[23]. Additional criteria were added to extend the application to children with hydrocephalus alone.

For visual and general handicap the most recent assessment available at the conclusion of the study was used.

Selection Pre - child born in 1970 or before.

Mid - child born between 1971 and 1973.

Post - child born in 1974 and onwards.

Results.

Orbital and facial anomalies. One child with congenital toxoplasmosis had the appearance of harlequin orbits on skull x-ray. One patient with spina bifida and hydrocephalus had a pronounced anti-mongoloid slant. Otherwise orbital and facial abnormalities were not a notable feature of the studied cases.

Exposure keratitis and corneal scarring. None of the 322 patients had this complication.

Sunsetting phenomenon. 53 patients (16%) had a history of this phenomenon. Some of the episodes were observed before the six year period. Five patients had repeated episodes. The age, diagnosis, visual and general handicaps of the patients are shown in Table 4. Eleven of these patients (21%) had developed optic atrophy by the end of the study but 41 (77%) had not. In one other patient the optic discs were never seen because of ring synechiae (preventing dilatation of the pupil) and lens or retrolental opacities secondary to retinopathy of prematurity.

Parinaud's syndrome. 22 patients (6.5%) showed this syndrome at some time during the six year period. The features of the syndrome and the diagnoses, visual and general handicaps of the patients are shown in Table 5. The age at observation ranged from three weeks to 16 years. The syndrome resolved completely in eleven patients (52%), often after shunt surgery, although in one of these it recurred on two subsequent occasions. In eight patients (38%) the syndrome persisted in all features. In one patient the syndrome resolved partially (upgaze nystagmus and dilated pupils persisted, downgaze nystagmus resolved). In the other patient there was insufficient follow up.

Pupil changes. Seven patients had dilated pupils, probably due to coning of the brain stem at a time when they were very ill. Three of these patients died shortly after this observation. Two of them had conjugate gaze palsies at this time and one other had papilloedema.

Four patients had dilated pupils along with other features of Parinaud's syndrome.

Three patients had permanently dilated pupils because of bilateral optic atrophy. Two children had unilateral afferent pupillary defects; in one this occurred shortly before revision of his shunt. One child had a history (from his parents) of constricted pupils during attacks of raised intracranial pressure. One child had posterior synechiae and iris bombe secondary to retinopathy of prematurity.

Manifest squint. 136 patients (42%) had a manifest squint at some time. It was convergent in 89, divergent in 27 and consecutively divergent in 20. In 45 (33%) the squint had been previously undiagnosed. 41 (30%) had had squint operations; 40 patients had operation for convergent squint and one for a divergent squint. The eyes of 14 of the patients with convergent squints who came to operation subsequently diverged, whereas the eyes of only 6 of the patients with convergent squint who never came to operation subsequently diverged.

Manifest Convergent Squint. 89 patients (28%) had a manifest convergent squint at some time. The diagnoses of the patients and the features of their squint are shown in Table 6.

Manifest Divergent Squint. 27 patients (8%) had a manifest divergent squint at some time. These patients had no history of a preceding convergent squint. Their diagnoses and the features of their squint are shown in Table 7.

Consecutive Divergence. 20 patients (6%) had a consecutively divergent squint. The eyes of 14 of these children (70%) became divergent after surgery for convergent squint. The eyes of 6 (30%) became spontaneously divergent. The diagnoses of the patients and the features of their squint are shown in Table 8.

Vertical Squint. 38 patients (11%) had a vertical component to their squint. 14 (40%) of them had had squint surgery but the vertical squint was known to be present prior to surgery in at least two cases.

Head Posture. A chin-up posture was common in these patients because a convergent squint with A syndrome was present. Thus the eyes were straight in depression but convergent in the primary position and elevation. Chin-down posture was also seen in patients with an A syndrome and divergent squint because the eyes were only straight in elevation.

A head tilt due to a vertical squint was seen in one case. A head turn in lateral rectus palsy was seen in seven cases but field defects, nystagmus and discomfort in the neck due to shunt procedures may have contributed to the abnormal head posture. Compensatory head postures were an additional handicap to children who were often already kyphotic and wheel-chair bound.

Nystagmus and disorders of ocular motility.

10 patients (3%) had horizontal pendular nystagmus. Six of these (60%) were blind or partially sighted (major visual handicap). Vision could not be definitely classified in the other four (40%) but two were thought to be blind. Table 9.

One patient (0.3%) with hydrocephalus had vertical pendular nystagmus for a short period from birth until his first shunt operation. He had many other ocular motility disorders. c.h.18.

Three patients (1%) had the signs of internuclear ophthalmoplegia c.h.l. They all had spina bifida with hydrocephalus. Visual handicap was minor in two and major in one. General handicap was intermediate in one, severe in one and very severe in one who later died. c.h.l.

One child with hydrocephalus had rotary nystagmus. Visual handicap was unclassified and general handicap intermediate.

Eleven patients (3%) had a conjugate lateral gaze palsy for a short time. Table 10. They were aged five days to six years and were severely ill with infected or blocked shunts at the time. Three patients died soon after this observation was made. Those who survived and were followed up usually showed gradual recovery of conjugate gaze.

Horizontal jerk nystagmus of second or third degree was seen in seven cases (2%). Table 11. (First degree nystagmus could not be confidently differentiated from musculo paretic nystagmus due to lateral rectus paresis in these children and is considered below.) The observation was made in children aged 18 months to 16 years who were severely ill with raised intracranial pressure shortly before surgery or death.

Lateral Rectus Palsy/Paresis/Musculoparetic Nystagmus. This was diagnosed in 93 cases (29%) at some time. Table 12. Only three cases of lateral rectus palsy with associated convergent squint completely resolved without residual paretic nystagmus (nystagmus on abduction) or residual squint. They were aged three, four and ten years and had all had evidence of at least low grade binocular single vision (i.e. Wirt Fly positive)

before this episode. The resolution took place after shunt surgery and without squint surgery. In other cases full abduction usually returned as time went on and the squint became concomitant, but nystagmus on abduction persisted. Only one patient had a permanent complete palsy, i.e. inability to abduct one eye beyond the midline. c.h.18.

Refractive Errors. 71 children (22%) had significant refractive errors. Table 13. One child had anisometropic amblyopia which responded to spectacles and occlusion.

Papilloedema. 44 children (14%) had papilloedema at some time. Two children had papilloedema on two separate occasions. Three of the episodes had been observed by others before the start of the six year period. Table 14. The age at diagnosis was six weeks to sixteen years. At least one child still had the anterior fontanelle open at the time that papilloedema was seen (the child being 19 months old). Seven children (16%) died later - some shortly after the papilloedema was seen. One case whose parents refused shunt surgery is still alive seven years after papilloedema was seen - the parents have also refused ophthalmic examination since then.

Papilloedema usually took several weeks to resolve in these children. In four it was still persistent after three months; in one of these it was still present to some extent after ten months. Three of the four cases were entirely symptom free and had no further shunt surgery during the period when papilloedema persisted. One of the four was admitted three months after shunt surgery (for papilloedema) with suspected valve dysfunction but had no further operations.

The optic discs often looked rather pale during and shortly after the period of resolution of papilloedema even when they later regained a normal colour. However six cases progressed to true optic atrophy and three cases clearly had atrophy when papilloedema developed. Eighteen cases regained normal discs and sixteen were lost to follow up.

Fluorescein Angiography. This was performed on two children with suspicious discs. Clinically one was thought to have pseudopapilloedema (due to hypermetropia) and the other was thought to have early disc oedema (due to raised intracranial pressure); the latter was found to have a blocked distal catheter at subsequent surgery. Both angiograms were negative, i.e. no abnormal leakage of fluorescein from the optic disc in late films.

Optic Atrophy. Fifty five children (17%) had optic atrophy at the time the study ended. Table 15. They often had a history of prematurity, multiple shunt operations, delayed surgical intervention at some stage, meningitis and septicaemia. Papilloedema was a causative factor in at least six cases.

Cortical Blindness. Nine patients (3%) had cortical blindness at some time. Eight (89%) also had a degree of optic atrophy Table 16.

Sudden Visual Loss. Three patients (1%) had a sudden decline in visual acuity at a time when shunt blockage was suspected for other reasons. One had papilloedema. In each case the visual loss was taken as an indication for urgent shunt surgery and the vision rapidly returned afterwards, at any rate for a while. The patient with papilloedema died a few months later.

Visual Evoked Potentials. Table 17. 19 patients had their visual evoked potentials and brain stem auditory evoked potentials recorded on one occasion towards the end of the study. They were aged 5 to 17 years. One had spina bifida with arrested hydrocephalus. 16 had spina bifida and hydrocephalus and two had primary hydrocephalus. Only six children had a VER which was normal in both wave form and latency at both occiputs. Four had spina bifida and two had primary hydrocephalus. Only two children had a BAEP normal in both wave form and latency; they both had primary hydrocephalus; they had normal visual function and the mildest of general handicaps.

Retinal Lesions. One child had disseminated choroiditis. He also had hydrocephalus with intracranial calcification due to congenital Toxoplasmosis. One other child with primary hydrocephalus had an isolated choroidal scar, possibly due to toxoplasmosis.

Three children had retinal haemorrhages seen shortly after birth and assumed to be due to delivery.

Four children (3 with SBH, 1 with H) had Roth's spots. One had an infected shunt system removed two weeks before the observation was made. The other three had recently had shunt obstructions without definite evidence of infection.

Two children with hydrocephalus had retinopathy of prematurity - one was completely blind with detached retinae and secondary anterior segment changes (grade V). The other was partially sighted with Grade II retinopathy.

One child had a unilateral macular scar possibly due to Toxocariasis.

Other Anomalies.

Ptosis. Two children had congenital ptosis; one had had surgery at the age of two years.

Horner's syndrome. Two children developed Horner's syndrome, one during a period of shunt obstruction and one after shunt surgery. In both cases the syndrome resolved gradually.

Facial Palsy. 8 children had a facial palsy before or after shunt surgery. Apart from one child with Pyle's disease (craniometaphyseal dysplasia) the palsies were mild and gradualy resolved and no child had exposure keratitis.

Optic Disc Changes. Two children had tilted discs. Three had glial tissue on one disc. Two others had myelinated nerve fibres adjacent to their discs.

Heterochromia Iridum. One child had this condition in one eye.

Nasolacrimal Duct Obstruction. One child had congenital epiphora.

Visual Function

86 children (27%) had normal visual function at the time the study ended.Table 18. However, 11 (13%) of these 86 children had had transient eye problems, e.g. squints, lateral rectus palsy, papilloedema, not affecting their final normal function.

74 children (23% of the total series) had a minor visual handicap at the time the study ended.Table 19. Squint, amblyopia, and minor degrees of optic atrophy with their effects on visual acuity, binocular single vision and visual field were the main reasons for impaired visual function.

18 children (5.5%) had a moderate visual handicap.Table 20. In 13 of the 18 the visual handicap was the result of amblyopia due to squint. One case had amblyopia associated with congenital ptosis. Three cases had significant optic atrophy as well as squint. One had a macular scar probably due to Toxocariasis.

11 children(3%) had a major visual handicap, i.e. they were eligible for registration as blind or partially sighted persons. Table 21. They all had severe or very severe general handicaps and two died during the study. Two children had spina bifida and hydrocephalus; they had bilateral optic atrophy which was seen to follow papilloedema in one. Delayed surgical intervention must have played a large part in the genesis of optic atrophy in these cases. c.h. 1 and 9. The other nine children had hydrocephalus alone. Five were born prematurely, two having retinopathy of prematurity and one other possibly so. Three had had infected shunts and in one the hydrocephalus was secondary to neonatal meningitis. One had severe hydrocephalus at birth due to sex-linked aqueduct stenosis (his sibling having died at birth of the same condition). One other had Pyle's disease.

133 children (41%) were too young or retarded for accurate assessment or were incompletely examined. Table 22.

Visual Field Defects. 15 children (5%) had field defects demonstrable on confrontation testing. Table 23. Five had generalised restriction, c.h.15, (very gross in one - see c.h.1.); they all had spina bifida and hydrocephalus. Two had inferior altitudinal defects in both eyes (one had spina bifida, c.h.11. the other had hydrocephalus). Two had quadrantanopias (one with SBAH, one with SBH), six had homonymous hemianopias.

Visual Perception. The testing of visual perception was not undertaken during this study but many children gave a history of visual perceptual problems noted at school by teachers or educational psychologists.

Correlation of General and Visual Handicap. The visual and general handicaps of the 322 children are summarised in tables 24 and 25 and they are correlated in tables 26 and 27.

The Effect of the Selective Policy on Visual Handicap. This is summarised in tables 28 and 29.

Deaths. There were 15 deaths during the six year study (5%). Table 30. The child with spina bifida occulta died of an unrelated muscle disorder. The seven children with spina bifida and hydrocephalus had all had multiple shunt problems. Of the 5 with hydrocephalus alone, four were born very prematurely and had had a stormy neonatal period. The other had gross hydrocephalus due to aqueduct stenosis at birth. Of the two with encephalocele and hydrocephalus one died of meningitis and the other of an infected shunt system.

Raised Intracranial Pressure Study. 53 episodes of raised intracranial pressure fulfilled the criteria for this part of the study. The clinical, operative and ophthalmic findings are shown in Table 31. 37 episodes (70%) had positive ophthalmological evidence of raised intracranial pressure. Papilloedema was the commonest positive finding. (27 episodes i.e. 51%). Other positive findings were the appearance of Parinaud's syndrome, or additional features of the syndrome in those who already had some features; the appearance of a convergent squint, or increased angle of squint; the appearance of a lateral rectus palsy (paresis) or increased degree of paresis; sunsetting and blurred vision.

The correlation between operative findings and the presence of positive ophthalmological evidence is shown in Table 32.

Survey of Other Centres.

Seven Paediatricians or Paediatric Surgeons working at nine hospitals replied to the letter. Only at two hospitals did spina bifida and hydrocephalic children receive screening by an Ophthalmologist as a matter of policy, although the replies did not state how often or how many times children were seen for this screening. At five hospitals most children were seen by an ophthalmologist at some time because of visual problems or squint. At one of these every child had full ophthalmoscopic assessment at every general clinic (presumably by a Paediatrician). At the two remaining hospitals children were only referred to an ophthalmologist as necessary, but at one of these all children were subjected to careful neurological follow up including inspection of the fundi.

At two of the nine hospitals the children could be seen by an ophthalmologist at the same session. Four centres mentioned one ophthalmologist and one centre mentioned two ophthalmologists who took a special interest in this group of children.

Discussion.

Unlike previous studies this reported work was long term. It covered a six year period during which many of the patients were examined repeatedly. 322 patients were studied making this a larger series than those of other authors. Table 2. The series consisted essentially of a random selection of patients studied because they were attending the paediatric outpatient department or were inpatients on the paediatric ward, irrespective of whether they were known to have, or suspected of having, eye problems. The main aim of the study was to determine the contribution an Ophthalmologist could make to the general management of these children. Observation of the incidence and severity of ophthalmic problems encountered was an essential part of the study. The active involement of the ophthalmologist during the six year period may have influenced the management of the children studied along side other changing factors including the evolution of the selective policy.

The children studied were a heterogeneous group including some with conditions which would have excluded them from some previous studies, e.g. spina bifida occulta, occipital encephalocele. However the details given under "Results" gives a diagnostic breakdown for each abnormality observed so that the effect of these inclusions on the overall incidence is apparent. The patients with spina bifida occulta acted, in effect, as a control group being virtually free of ophthalmic problems. Although patients with spina bifida cystica, their parents, and their paediatricians tended to resist the invitation to have regular ophthalmic examinations, they were included in the research because nearly all cases

of spina bifida have enlarged ventricles at birth, even if they never require treatment of clinically manifest hydrocephalus. Moreover, if the notes of patients who carry a diagnosis of spina bifida cystica alone, are studied in detail, it transpires that a few have obviously had clinical hydrocephalus which arrested at an early stage. Children with arrested hydrocephalus appear to be particularly vulnerable to decompensation at a later stage and fully justify continued observation. c.h. 1.

Orbital and facial anomalies. Like other studies[26] this research showed a very low incidence of orbital and facial anomalies and this demonstrates that control of hydrocephalus by shunting at an early stage should eliminate such deformities.

Exposure keratitis and corneal scarring. No cases were seen in this study. This contrasts with the o.8% and 4% increase in earlier reports[29,28]. Superior general management reducing episodes of coma and facial palsy is likely to be responsible for this improvement.

The Setting Sun phenomenon. 16% of this series had a history of sunsetting.c.h. 11,14,16,18. This is a somewhat higher incidence than previous series[26] (10% [29] 12%) even though the latter two series, unlike the present one, did not include children with spina bifida occulta and spina bifida cystica, who had a low incidence of sunsetting. More patients with a history of sunsetting had an underlying diagnosis of primary hydrocephalus than spina bifida with hydrocephalus, despite the fact that patients with spina bifida with hydrocephalus formed the largest diagnostic group in the series. This is probably because the development

of hydrocephalus in a case of spina bifida is anticipated, confirmed by serial measurements of head circumference, computed tomography or ultrasonography and treated at an early stage before some of the clinical manifestations such as sunsetting supervene. The diagnosis of primary hydrocephalus, on the other hand, may be delayed until a later stage when other clinical features such as sunsetting are obvious.

No case of persistent sunsetting was observed, but five patients (9%) had repeated episodes.c.h.16. Sunsetting was commonest in children under six months (83% of episodes). This is generally in agreement with the series of Rabinowicz[26] (who found no cases after eighteen months of age), and Goddard (only one case after six months); but 17% of episodes occurred after six months of age and one patient was 7 years old,c.h.11, when the phenomenon was observed. This accords with the cases of forced down gaze in adults with decompensated hydrocephalus described by Cobbs et al[34].

21% of cases with a history of sunsetting had developed optic atrophy by the end of the study. This is a much lower incidence than the 50% quoted by Rabinowicz[26] and is not much higher than the general incidence of optic atrophy in the series (17%). However an analysis of the visual handicap of those with a history of sunsetting and the series as a whole, shows a trend towards a worse visual prognosis in the former group. Table 33.

Parinaud's syndrome. 6.5% of patients showed this syndrome at some time during the six year period.c.h. 9,13,14,15,16. There are no figures available for comparison in the literature, although many authors have

described the syndrome in non-tumoral hydrocephalus. Partial manifestations of the syndrome were most often seen. In agreement with other reports, it was found that the syndrome could be cured by shunt revision if this was done promptly; this was the outcome in eleven patients (52%) c.h. 9,13,14, but in one case the syndrome persisted in some features, c.h.15, and in eight (38%) it persisted in all features that had developed c.h. 16. Persistence of the syndrome was presumably due to irreversible changes in the brain stem. None of the children who developed this syndrome had normal visual function at the end of the study. All the children had some general handicap. Table 5.

The appearance of Parinaud's syndrome in a child with hydrocephalus is, therefore, a useful indication of uncontrolled hydrocephalus. Urgent shunt surgery is required, but the syndrome may persist despite it, and the child is likely to have at least some visual and general handicap thereafter.

Pupils. Dilated pupils were seen as part of Parinaud's syndrome in four children c.h.13, due to optic atrophy in three cases, c.h. 5. and due to coning of the brain stem in nine children, c.h.10. Pupil changes, therefore, tend to occur late, only in association with other ophthalmic abnormalities or severe illness. Nevertheless, examination of the pupils should not be neglected and measurement of pupil size under standardised conditions assists in the detection of early changes.

Manifest squint. 136 patients (42%) had a manifest squint at some time during the study period. This accords with figures in the literature

(30-73%) considering that the series are not strictly comparable. The present series included some cases of spina bifida occulta and spina bifida cystica who had a low incidence of squint; such cases were excluded from other series Tables 1 and 2. Squint is thus extremely common in a group of children already burdened with multiple handicaps.

45 children (33%) were not known to have a squint until they were seen as part of this study, and a few had not received treatment for a recognised squint, c.h.12. Therefore, regular ophthalmic screening of this group seems justified in view of the high incidence of squint and the frequency with which the diagnosis is missed by those not regularly examining children with malaligned eyes.

Manifest convergent squint. This was the commonest type of squint noted. It occurred in 28% of the whole series. This is a lower figures than that of Rabinowicz[26] (44%) and Clements and Kaushal[24] (67%); the inclusion of patients with spina bifida occulta and older patients (who were more likely to have consecutive divergence) in the trial may account for this. Tables 1 and 2.

Not surprisingly convergent squint occurred much more frequently in patients with shunt dependent hydrocephalus than in those with arrested hydrocephalus or spina bifida cystica alone. There was evidence of lateral rectus palsy or paresis in 57% c.h. 4,14. showing that this is the commonest underlying mechanism for convergent squint. An A syndrome was present in 41%. This is a much higher incidence than is usual in squints (Stanworth[65] estimates that the A syndrome occurs in 1/10th to 1/15th

of all squints) and adds support to the theory that lateral rectus palsy is the usual cause of convergent squint in these cases. 14% had a positive family history of convergent squint, although the true figures may be higher as many spina bifida children are fostered or adopted and know nothing of their family. Significant hypermetropia was present in only 19% and fully accommodative squints were uncommon in this series as in others. Although major degrees of amblyopia were found to be uncommon (1%) c.h.12. as in other series, minor degrees were not at all unusual (36%). Treatment of amblyopia by conventional occlusion was successfully undertaken in some cases, c.h. 6,18, but failed in others.c.h.12. perhaps because such treatment is an insupportable burden for children and their mothers who already have multiple appliances and treatment regimens to be remembered. 26 children (29% of convergent squints) had had squint surgery with beneficial results. This must be balanced against the 14 children with divergent squint following convergent squint surgery. c.h.6. In addition two children with the A syndrome were divergent in downgaze after surgery for a convergent squint despite being straight or only minimally convergent in the primary position.

Manifest divergent squint. 27 patients (8%) had a manifest divergent squint without a history of preceding convergent squint. c.h.17. However, nine of these (33%) had signs of lateral rectus palsy or paresis c.h.3 and five (18.5%) had an A syndrome, suggesting that some of these squints were in fact cases of consecutive divergence. Eight (30%) had a major visual handicap which accords with the findings of diminished visual acuity in a high proportion of divergent squints in other series [24,26]. Significant myopia was present in only two patients (7%).

More patients with divergent squint had primary hydrocephalus or encephalocele with hydrocephalus than would have been expected from their incidence in the series as a whole. This may be because divergent squint was frequently associated with major visual handicap which was commoner in primary hydrocephalus and encephalocele with hydrocephalus than in spina bifida with hydrocephalus.

Consecutive divergence. 20 patients (6% of the total series) had a consecutively divergent squint. This followed convergent squint surgery in 14 (70%) c.h. 6. but occurred spontaneously in 6 (30%) c.h.14. Thus 14 (30%) of the 47 patients in this series with divergent or consecutively divergent squints had had squint surgery compared with the 20% incidence in Rabinowicz[26] and Clements and Kaushal's,[24] series. Lateral rectus palsy (75%) and the A syndrome (35%) were at least as common in this group as in those with convergent squint (57% and 41%). It is interesting to note that weakness of abduction does not protect against the development of consecutive divergence and it is important to realise that a high proportion of convergent squint operations in these children will be followed by this complication. There must therefore be a considerable predisposition in these children for the eyes to diverge. Recognition of convergence weakness might help to forewarn surgeons of the likelihood of consecutive divergence.

Spontaneous divergence was most often seen at about the age of ten years. Frequently there was a period of time during which the squint was only divergent in depression c.h. 14 and straight or convergent in the

primary position and elevation, before the squint became divergent in the primary position or all positions of gaze.

Vertical squint. 38 patients (11%) had a vertical component to their squint (Goddard[29] gives an incidence of 30%).

Head posture. A marked chin up position due to the A syndrome with convergent squnt was found in four children. A noticeable chin down posture due to a divergent squint with A syndrome was found in one child. The disabling effect of a compensatory head posture in children already handicapped by poor mobility and spinal deformities should highlight the need for this syndrome to be treated at the same time as convergent or divergent squint surgery.

Nystagmus and disorders of ocular motility. The following types of nystagmus were observed - pendular, jerk, micronystagmus and musculo paretic nystagmus. Ten children (3%) had horizontal pendular nystagmus c.h. 5,18. - a similar figure to that of Rabinowicz[26] (4%); all children so affected had poor vision when they were old enough for it to be tested. The underlying diagnosis was primary hydrocephalus in 90% and spina bifida with hydrocephalus in 10%. (In the series as a whole primary hydrocephalus formed 33.5% and spina bifida with hydrocephalus 44%). This was due, presumably, to the more frequent occurrence of major visual handicap in primary hydrocephalus than in spina bifida with hydrocephalus.

Some other ocular motility disorders were observed which were not reported in other series. These included internuclear ophthalmoplegia c.h.l.

conjugate lateral gaze palsy, c.h.10 and horizontal jerk nystagmus of second and third degree. The latter two disorders were seen only in severely ill patients, several of whom subsequently died. Conjugate lateral gaze palsy was seen mainly in children with primary hydrocephalus and in those with a major visual handicap.

Lateral rectus palsy and paresis and musculo paretic nystagmus. 93 cases (29%) had this complication. There are no figures for comparison in the available literature, although it is generally agreed that such a palsy is very common and is the underlying cause of the majority of squints and of nystagmus (28% [24]). Lateral rectus palsy was found in one case classified as having spina bifida cystica alone, suggesting that this child had had significant hydrocephalus at some time. Complete resolution of lateral rectus palsy or paresis was rare (3 cases 3%) despite surgical intervention to relieve raised intracranial pressure, c.h. 4,2,6,9,11,13,14,16,18. Most cases had an associated squint so that delay in treatment of uncontrolled hydrocephalus until a lateral rectus palsy supervenes will usually result in regrettable permanent sequelae such as squint and nystagmus.

This study has shown that almost any type of nystagmus and ocular motility disorder may occur in congenital hydrocephalus, especially in the severely and terminally ill child. The appearance of nystagmus or another ocular motility disorder in a previously affected child usually denotes uncontrolled hydrocephalus and shunt surgery is indicated, but this may not restore normal ocular motility. The appearance of nystagmus, in particular, may be noted by the child's parents and provide useful

objective evidence of the need to seek medical advice.

Refractive errors. These were found in 71 children (22%). This finding compares closely with the 16-25% incidence in other series,[24,39,28]. Most squints in these children appear to be based on lateral rectus palsy rather than on hypermetropia. However, a refraction should be carried out whenever there is either poor visual acuity or a squint because of the help it may give in individual cases. A poor school performance by these children may be falsely ascribed to mental deficiency when an easily remediable condition such as myopia exists.c.h.8. However, spectacles should not be prescribed unless the children are clearly likely to benefit for they are already burdened with so many other appliances.

In a few of these children, myopia may have been the only sign of the retinopathy of prematurity. Many of the primary hydrocephalics had been born prematurely and a few had other signs of retinopathy (vide infra).

Papilloedema. This was observed in 44 children (14% of total series) at some time c.h. 1,2,3,7,10,11,12,14,16,17. It was recurrent in two cases, c.h.12, so 46 episodes were observed, 43 of them during the six year period. This findings contrasts with the very low incidence quoted by Goddard,[29] (less than 1%), Rabinowicz[26] (2%), Harcourt[38] (3%) although Ghose[47] more recently found an incidence of 14.5%. It must be emphasised that the 14% incidence in this study is based on examinations performed (sometimes repeatedly) over a long period and that a figure devised from single examinations within a short space of time would necessarily be much lower.

At least one child in the seies still had an open anterior fontanelle at the time papilloedema was noted, c.h.7., so it cannot be assumed that the expansibility of the infant skull provides complete protection against the development of papilloedema.

16% of children with a history of papilloedema died within the six year period which emphasises the seriousness of this condition, c.h. 1,10,12. (5% of the total series died). In addition at least 14% subsequently developed optic atrophy, c.h.13,17. However, a few children recovered with no visual or general handicap, thus the prognosis need not be gloomy if treatment is prompt.

Although uncommon, papilloedema was of the greatest possible help in the pre-operative diagnosis of raised intracranial pressure which otherwise may have to be made on the basis of symptoms and non-specific signs. Indeed in one case the patient was without either symptoms or other signs c.h.2. papilloedema being discovered on routine examination. Papilloedema was sometimes present when the valve was pumping normally (vide infra) c.h.3. In this study papilloedema was taken as an absolute indication for shunt surgery.

The significance of papilloedema was not always appreciated by junior paediatricians even when the fundi had been inspected. Indeed at the start of the study it was rare to find any mention of fundus examination in the case notes even when the child had been admitted as an emergency with symptoms of raised intracranial pressure. However by the end of the

study ophthalmic opinion was much more often sought and sometimes used as the criterion for surgery or discharge from hospital.

Papilloedema usually took several weeks to resolve after an episode of raised intracranial pressure even when shunt surgery was judged to be successful on other evidence c.h.2,3. The diagnosis of raised intracranial pressure was thus more difficult during this period, but repeated careful examination of the discs was still helpful so that the clearing of haemorrhages and the lessening of oedema could be sought. Papilloedema was recurrent on two occasions, c.h.12 therefore a history of papilloedema may be helpful in denoting children who are particularly likely to suffer this diagnostic complication during future episodes of raised intracranial pressure. However, the absence of papilloedema provided no reassurance about intracranial pressure. c.h.13.

As expected, papilloedema was sometimes followed by the development of optic atrophy. Nevertheless, it was found that the optic discs often looked pale during and after the period of resolution of papilloedema so that atrophy should not be diagnosed ill advisedly. As in other studies there were a few (3) cases in which papilloedema was superimposed on optic atrophy, c.h.1,10; the presence of pale discs provides no excuse for failure to examine the fundi in future. Notably, these three cases with papilloedema on atrophy all died before the end of the study. Therefore, the finding is one of the utmost gravity.

Fluorescein angiography. At the start of the study it had been thought that fluorescein angiography would be a useful tool in the diagnosis of raised intracranial pressure. Nevertheless its use had to be restricted to children old enough to co-operate with fundus photography and well enough to travel to the Eye Hospital for this purpose. In practice few children satisfied these criteria and only two came to angiography. In both of these the angiograms were negative although one later had surgery for a blocked shunt. In view of these unhelpful findings and the slight inherent risks of the procedure as well as its impracticability in most of these cases, reliance was increasingly placed on clinical findings alone. Thus, those with developed papilloedema with haemorrhages were admitted for urgent shunt surgery. Those with slightly swollen discs (suspicious discs or suspected papilloedema) who were clinically unwell and who had other evidence of shunt malfunction were admitted for general assessment, cap tap and exploratory surgery if necessary. Those with suspicious discs who were clinically well and who had no other signs of shunt dysfunction were refracted to exclude pseudo papilloedema due to hypermetropia. The remainder were observed more frequently in the Outpatient Department and only admitted if new symptoms or signs developed.

Optic atrophy. Optic atrophy was quite common (17%) in this series. Rabinowicz[26] found a higher incidence of 31%. The incidence of atrophy in this study would have been higher if cases of transient pallor after an episode of papilloedema had been included.

No child with optic atrophy had normal visual function although 31% had

only a minor defect (Rabinowicz found 28% had a vision of 6/9 or better). 16% definitely had a major visual handicap (i.e. were eligible for blind or partially sighted registration), and some of the unclassified group will probably fall into this category eventually.

There was a history of papillodema in six of the 55 cases but it may have occurred unrecognised in many more. Gross cupping of the atrophic optic disc c.h.5, was found in ten cases (18%); this suggests a preceeding pathological lesion occurring at the optic disc itself, e.g. papilloedema rather than in the retrobulbar optic nerve, optic tract or lateral geniculate body.

A history of prematurity, meningitis, septicaemia, multiple shunt operations or delayed surgical intervention was also common in cases of optic atrophy. c.h. 1,5,6,9,10,17. Primary optic atrophy seems a dangerous diagnosis to make in congenital hydrocephalus.c.h.9.

Optic atrophy was seen more often in primary hydrocephalus than in cases of spina bifida with hydrocephalus, although the former formed a smaller proportion of the series as a whole. This may reflect the fact that primary hydrocephalus may be diagnosed at a later stage than hydrocephalus in spina bifida (vide supra) and that primary hydrocephalics were more likely to have been born prematurely and have had a stormy neonatal period c.h.10.

Optic atrophy was also found more often in those with several general handicap and the death rate of affected children was 18% compared with 5%

in the series as a whole.

Children with optic atrophy are therefore a visually and generally handicapped group, predisposed to further visual and general deterioration and death.

Cortical blindness. Nine patients (3%) of this series had cortical blindness. Eight of nine patients also had a degree of optic atrophy. They all had primary hydrocephalus or encephalocoele with hydrocephalus. Multiple severe medical problems in the neonatal period and delay in diagnosis and treatment may account for the occurrence of cortical blindness in this group but not in spina bifida with hydrocephalus. Two children with cortical blindness had died by the time the study ended and none of the survivors was without some general handicap.

Visual loss and blindness. Keen[50], Lorber[49], Rabinowicz[26] and Welch[51] all describe acute or insidious loss of vision (sometimes reversible) in hydrocephalus, usually secondary to shunt obstruction or infection. In this study only three cases of declining visual acuity were actually observed, c.h.15, and the vision returned rapidly after shunt surgery. However, other cases had a history of transient visual obscurations without measurable decline in visual acuity, c.h. 8,16,14. In only one case was the visual decline dramatic, (VA 6/9, 6/6 to counting fingers both eyes) c.h.15.

No case of measureable permanent visual decline in a child under regular observation occurred during the study and this must surely reflect the

increased attention paid to ophthalmic signs and symptoms and the greater willingness to operate for shunt dysfunction on the basis of disordered visual function which gradually developed as the research progressed. Nevertheless, the literature and the history of events occurring before the six year study c.h.5. demonstrate the vulnerability of the optic nerves to raised intracranial pressure. Neglect of a blocked shunt may result, not in a dead child, but in a permanently blind one

Visual evoked potentials. Only 2 of the 19 children tested had both a normal VER and BAEP. They were both primary hydrocephalics with normal visual function and the mildest of general handicaps. All the children with spina bifida and hydrocephalus had abnormal BAEPs and most had abnormal VERs as well. The abnormalities in BAEP latency and waveform were suggestive of abnormalities at the pontomesencephalic level or abnormal conduction throughout the brainstem.

Retinal lesions. As in other series [29, 26], congenital toxoplasmosis with disseminated chorioretinitis was a rare finding. Other retinal lesions were also uncommon. Retinal haemorrhages, assumed to be due to delivery, were seen in three cases and no permanent sequelae were detected in these children. Four children had haemorrhages with pale centres (Roth's spots); they all had shunt dysfunction at the time, but only one had a proven shunt infection.c.h.17. Nevertheless detection of such haemorrhages should certainly be sufficient grounds for blood and cerebrospinal fluid cultures and measurement of cerebrospinal fluid pressure to be undertaken.

Two children with primary hydrocephalus had the retinopathy of prematurity with an associated major visual handicap. Therefore, in addition to examination of the optic disc and peripapillary area in all hydrocephalic children at every clinic visit, children with primary hydrocephalus should have their fundi fully examined after pupillary dilatation at least once so that chorio-retinitis and the retinopathy of prematurity can be detected.

Other anomalies. Facial palsies were seen in eight cases and Horner's syndrome in two cases shortly before or after shunt surgery. Gradual resolution occurred and no child developed exposure keratitis as a result of facial palsy.

Other anomalies included congenital ptosis (2 cases) and naso lacrimal duct obstruction (1 case); there is no evidence that these conditions were causally related to the underlying diagnosis.

Visual function. Only 86 (27%) children had normal visual function. This compares fairly well with Rabinowicz'[26] finding of 37% with a vision of 6/6 or better, bearing in mind that he did not have an unclassified group and that all his cases had spina bifida with hydrocephalus or primary hydrocephalus. Similarly 34% of Hunt's[28] group of school children were without visual defects. However 13% of the "normal" group had had transient eye problems (squint, lateral rectus palsy, papilloedema) not permanently reducing visual function so that few children escaped without some effects on their eyes. Also, many of this "normal" group had spina bifida occulta or spina bifida cystica - conditions which should not be associated with eye complications.

23% of the children had a "minor" visual handicap. Squint, amblyopia, and minor degrees of optic atrophy, with their adverse affect on visual acuity, binocular vision and field were the underlying mechanisms.

5.5% of children had a moderate visual handicap. Amblyopia due to squint was the commonest cause of unilateral visual defect, c.h.12; optic atrophy c.h.9, macular scarring and amblyopia due to ptosis were the remaining mechanisms. In contrast, Rabinowicz,[26] attributed most of his "6/18 or worse in one eye" group to underlying optic atrophy and found amblyopia to be rare.

Eleven children (i.e. 3%) in this study had a major visual handicap. Rabinowicz[26] found 7% eligible for blind registration, (he did not have an unclassified group) and Hunt[28] found 2% of her group of school age children to be educationally blind. Major visual handicap was invariably associated with severe or very severe general handicap and 18% of the group died during the study period.

An analysis of the case histories of these eleven children suggests that some of the visual handicap could have been avoided in at least nine cases. Delayed surgical intervention was undoubtedly responsible for the development of optic atrophy in the 2 children with spina bifida and hydrocephalus c.h.1,5. Retinopathy of prematurity was responsible in two cases so that better control of arterial oxygen in the Special Care Nursery might have helped these cases. A third child with optic atrophy and nystagmus had been born prematurely. Again better treatment of infected shunts or neonatal meningitis might have salvaged four other cases. The

management of the children with severe hydrocephalus at birth due to sex-linked aqueduct stenosis, and of the child with Pyle's disease could probably not have been improved in the present state of medical knowledge.

Many children in this study (41%) fell into the unclassified group because their youth prevented testing of Snellen visual acuity or because they were lost to follow up before ophthalmic examination could be completed.

Thus, only a small proportion of children with hydrocephalus, who are old enough to be tested, escape without some visual disturbance and routine ophthalmic assessment is therefore justified.

Visual field defects. 5% of this series had field loss. Rabinowicz[26] found 9% to have field defects and the types of defects in both series were similar. A much higher incidence of defects may have been found if all the patients had been old enough for detailed testing.

Visual perception. Although not tested in this study, many children gave a history of visual-perceptual problems noted at school by teachers or educational psychologists, as would be expected from the literature. Since some of these were children with "normal" visual function as assessed on the basis of acuity, stereopsis and field, the number of children with hydrocephalus who escape without any damage to their vision or visual perception must be small indeed.

Correlation of visual and general handicap. Only 12% of the children studied had no general handicap. In 15.5% the handicap was mild, 23% intermediate, 21% severe, 15% very severe and 14% were too young to be classified. Thus more children prove to have general handicaps than visual handicaps (although more children could be classified generally than visually). Those with the severest general handicaps tended to have the severest visual handicaps too, although two children (1%) with very severe general handicaps had normal visual function and 13 children (4%) only a minor visual defect. A major visual handicap only occurred in those already crippled with severe or very severe general handicaps.

Effect of the Selective Policy. Despite the introduction of selection during 1971 to 1973, children with minor, moderate and major visual handicaps continue to be found. Further comparison of pre and post selection figures would probably be invalid since at present a very large proportion (34%) of those in the post-selective era are unclassifiable, whereas only 4% of those in the pre-selective era cannot be tested. Moreover it must be remembered that spina bifida and hydrocephalus are progressive diseases so that children in the pre-selective group (being older) are more likely to have acquired visual handicaps with time than the younger post-selective group.

Deaths. There were 15 deaths during the study period (5% total series) c.h.1,10,12. Apart from one child with spina bifida occulta who had an unassociated muscle disorder, they all had hydrocephalus (with spina bifida, primary or with encephalocoele). They were in the groups with more severe general and visual handicaps. No child with normal visual

function died. More of the children who died came from the post selective group presumably because death in these children is commonest in their early years, and all children in the pre selective era were at least seven years old when the study commenced.

Raised Intracranial Pressure Study. 70% of documented episodes had positive ophthalmological evidence of raised intracranial pressure and only 30% had negative eye findings. Since these episodes tended to be the less acute ones in which there was time for more investigations before surgery (and sometimes doubt about the necessity for surgery) this is a finding of considerable practical importance. In 17% of cases the shunt appeared tobe pumping normally although at surgery it was obviously blocked, and in 6% there were no signs and symptoms apart from those relating to the eyes.

In 51% the positive eye finding was that of papilloedema,c.h. 1, 2, 3, 7, 11,12,14,17. In the remainder sunsetting, blurred vision, c.h.15, or a new/increased Parinaud's syndrome c.h. 9,13,14, convergent squint, c.h.4,6 or lateral rectus palsy/paresis c.h.4,6, were the criteria for diagnosing raised intracranial pressure. In the most dramatic case a divergent squint became convergent, c.h.6. Whereas papilloedema and sunsetting should normally be diagnosed by Paediatric staff, detection of ocular motility problems probably requires an ophthalmologist already familiar with the patient.

Eye signs were present more often in those with distal catheter blockage, Table 32, perhaps because this tends to cause a slower rise in pressure

than proximal catheter blockage, giving time for slowly progressive lesions such as papilloedema to develop. However, all types of shunt dysfunction were associated with eye signs.

It was not apparent from the available data why papilloedema occurred in some cases of raised intracranial pressure and not in others. In one child, raised intracranial pressure had been suspected for six months on the basis of a new lateral rectus paresis, and for two months on the basis of the appearance of Parinaud's syndrome before papilloedema finally developed and shunt surgery was expedited c.h.16. Thus, papilloedema may develop only when raised intracranial pressure has attained a high level. (see also c.h.11). Because of the risk of consecutive optic atrophy, raised intracranial pressure should, if possible, be diagnosed before papilloedema develops.

Abnormal eye signs were found in 70% of documented episodes of raised intracranial pressure, including those in which there was no other evidence of shunt dysfunction. Nearly 50% of cases did not develop papilloedema, and so a completely normal eye examination does not exclude the presence of raised intracranial pressure.

Survey of Other Centres. The amount of ophthalmic attention given to children with spina bifida and hydrocephalus varied considerably from centre to centre. Only two out of nine centres attempted routine ophthalmic assessment. At the other seven centres, as in Wessex before the start of this study, children were referred as necessary, although in five of these it appeared that most children did see an ophthalmologist at

some time. At one of the centres where patients were referred only as necessary, fundus examination was routinely performed as part of the neurological assessment.

This study has shown that some 33% of squints had not been previously diagnosed; thus a considerable number of ophthalmic anomalies will be missed if reliance is placed upon detection by paediatric staff and subsequent referral to an ophthalmologist. Only 27% of children definitely had normal visual function and 13% of these had had transient, but potentially serious eye problems (e.g. papilloedema, squint). Since ophthalmic complications are not only a handicap in themselves, but may assist in the diagnosis of raised intracranial pressure, anything less than regular routine ophthalmic screening is surely unacceptable.

Only two centres stated that they offered paediatric and ophthalmic assessment during the same hospital visit. This seems strange when attendance at orthopaedic and paediatric clinics during the same session is the norm. The imposition of multiple clinic visits on these handicapped children and their parents is also unacceptable.

Four centres referred children to one interested ophthalmologist and one centre to two. The specialised nature of ophthalmic problems in these children, the need for familiarity with their baseline ophthalmic performance if new and significant defects are to be detected and the necessity for close liaison c.h. 1,5,9, with the paediatrician surely justify this degree of superspecialisation.

The Role of the Ophthalmologist in Spina Bifida and Hydrocephalus.

As in previous studies [50, 24, 33, 39, 11] the work reported here has confirmed the importance of frequent eye examinations in children with spina bifida and hydrocephalus. In particular it has highlighted the value of having one interested ophthalmologist involved in the care of these patients, and available in the paediatric clinic for immediate discussion. Many important conditions, e.g. squint and optic atrophy, remain undetected or their significance not appreciated, c.h. 1,5,9, unless children are seen routinely and frequently by the same ophthalmologist. If such screening is carried out, small changes in ocular motility, visual acuity and optic disc appearance, which would otherwise go undetected, may be noted and lead to the early diagnosis of raised intracranial pressure. Thus the ophthalmologist may not only help to preserve vision but also to maintain the general well-being of the child.

This study has shown that after an initial period during which eye examination was sometimes considered unnecessary by medical staff and parents, an increasing value was placed on the presence of an ophthalmologist in the clinic. Parents remembered other children who had gone blind and were anxious to protect their own offspring from a similar fate. Medical staff became more aware of the ophthalmic complications of hydrocephalus; they more frequently performed eye examinations themselves and more often requested examination by the ophthalmologist. The ophthalmologist's report was increasingly used as a criterion for shunt surgery (in the event of positive eye findings) and for discharge of the patient from hospital (if the findings were negative). Once the study was

established, no child under regular observation suffered a measurable, irreversible loss of vision.

CONCLUSIONS.

There is a very high incidence of ophthalmic complications in children with spina bifida and hydrocephalus. Hitherto eye problems have been neglected, yet good vision is vital if these children are to become independent and gain employment. Few children escape without some damage to vision or visual perception. A wide variety of complications are seen. The children with the most severe general handicaps tend to have the most severe visual handicaps. Children with primary hydrocephalus tend to have a higher incidence of ophthalmic complications, perhaps because their hydrocephalus is diagnosed at a later stage, or because the problems of prematurity, neonatal infection etc. associated with the development of hydrocephalus are also responsible for adverse effects on the eye. Children with spina bifida cystica alone may also develop ophthalmic complications presumably because of latent or arrested hydrocephalus. The introduction of the selective policy has not prevented the development of visual handicaps in surviving children.

Orbital, facial and corneal problems are rare nowadays. Sunsetting is a common late complication of hydrocephalus in infants and tends to be associated with a worse than average visual outcome. Parinaud's syndrome may appear in older children due to raised intracranial pressures, but does not always resolve completely after apparently successful surgery. Visual function is always impaired following its appearance.

Convergent squint is extremely common. It is most often due to lateral rectus palsy; there is frequently an associated A syndrome. The first

appearance of convergent squint is usually due to raised intracranial pressure but persistence is the rule despite successful shunt surgery except in older children who have established binocular single vision. Minor degrees of amblyopia are common; major degrees are rare. Treatment of squint in hydrocephalus should be tailored to the special needs of this group. Control of underlying hydrocephalus is the first priority; ophthalmic and orthoptic treatment take second place. Fundus examination should be performed at every visit to an Eye Department or Eye Hospital and never delayed (along with refraction under atropine) until the next attendance, c.h.l. Hospital visits should be minimised and occlusion therapy employed for the minimum time. Convergent squint surgery should rarely be performed because of the risks of consecutive divergence. The latter is common after surgery but may even occur spontaneously. If squint surgery is undertaken it should include correlation of the A syndrome.

Divergent squint is also quite common, particularly in primary hydrocephalus. It is associated with major visual handicap or with preceeding convergent squint.

Almost any type of nystagmus and ocular motility disorder may be found in hydrocephalus, especially in primary hydrocephalus and in those who are severely and terminally ill. The first appearance of such a disorder is usually due to raised intracranial pressure.

Papilloedema occurs in a significant number of cases when they are followed over a long period. It may be the only objective evidence of

raised intracranial pressure and is an absolute indication for shunt surgery. It is probably a late manifestation of raised intracranial pressure and its absence should not be interpreted as evidence that hydrocephalus is adequately controlled. Papilloedema resolves slowly after successful shunt surgery. The optic discs may appear pale for a while, but the majority of children do not develop optic atrophy. However, there is a higher than average mortality rate after an episode of papilloedema, and an early death is likely if papilloedema develops in an already atrophic disc. There is rarely any need for fluorescein fundus angiography in these children.

Optic atrophy is quite common, especially in primary hydrocephalics and those with severe general handicaps. It is usually associated with a history of prematurity, infections, multiple shunt operations, delayed surgical intervention and preceeding papilloedema. Children with optic atrophy are predisposed to further visual and general deterioration and early death. Optic atrophy without papilloedema may be associated with chronically raised intracranial pressure c.h. 1,5,9. Shunt function should always be investigated if the history of its development is unknown.

Cortical blindness appears in a few severely ill, severely handicapped children with primary hydrocephalus or encephalocoele and hydrocephalus.

Visual evoked potentials are abnormal in all cases of spina bifida and hydrocephalus. Further investigation of these responses is indicated.

The majority of episodes of raised intracranial pressure lead to some ophthalmic signs. Papilloedema is the most frequent positive finding but blurred vision, sunsetting, Parinaud's syndrome or the development of a squint may also be useful evidence. However, negative eye findings do not mean that the intracranial pressure is normal. Neglect of raised intracranial pressure due to a blocked shunt may lead not to death, but to the tragic addition of blindness to an already multiply handicapped child, c.h.5.

Few centres currently employ the type and extent of ophthalmic screening established in this study. However, the work reported here has confirmed the value of an ophthalmologist working in the spina bifida and hydrocephalus clinic. Regular long term eye examinations of these children assist in their general management particularly in the diagnosis of shunt obstruction as well as in the preservation of their visual function. Personal experience of working in such a clinic has confirmed that both paediatricians and parents are grateful when the skills of such an ophthalmologist are readily available.

Recommendations.

Every spina bifida and hydrocephalus clinic should have an ophthalmologist as a member of its medical team. The ophthalmologist should make regular examinations of visual acuity and field, ocular motility and fundi, starting at the time of diagnosis in all children with spina bifida cystica whether or not they have clinical hydrocephalus, and in children with primary hydrocephalus and encephalocoele. Examination of the optic

discs alone is insufficient and is sometimes impractical for the non-specialist working without the advantage of the indirect ophthalmoscope. Many abnormalities of ocular motility go undetected by the non-specialist and the significance of such problems cannot be appreciated unless baseline data have been established.

Ideally eye examination should be performed in the same hospital and during the same session as examination by the paediatrician so that immediate discussion of the findings and proposed management can take place. The ophthalmologist should also examine children on the ward if they have suspected shunt dysfunction.

These policies should ensure that these children achieve and maintain the best possible standard of vision and that the earliest ophthalmic signs of raised intracranial pressure are detected in patients where the clinical diagnosis is in doubt.

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Appendix I

Case Histories.

1. D.B. This child had a thoraco-lumbar meningomyolocoele closed at birth in June 1970. Hydrocephalus developed but arrested and he did not have a shunt inserted. He was referred to the Eye Hospital in November 1972 where a left convergent squint was diagnosed, but an atropine refraction was impracticable. Six months later he was noted to have a right to alternating convergent squint, a head turn, right amblyopia and nystagmus. In September 1973 a squint operation was performed on the right eye. He had very severe general handicaps, suffering epileptic fits, confined to a wheel chair and requiring a penile appliance. There is no mention of fundus appearance in his records until his Ophthalmic Surgeon noted pale optic discs in August 1975.

When first seen in this study in January 1977 he had optic atrophy, gross visual field loss, and internuclear ophthalmoplegia. A CT scan showed hydrocephalus and the question of shunt surgery was discussed, but his Paediatric surgeon preferred to wait for evidence of clinical deterioration.

In July 1977 he was admitted to hospital complaining of headaches and vomiting; his cranial sutures had separated and his anterior fontanelle had reformed. There was papilloedema superimposed on optic atrophy.His cerebrospinal fluid pressure measured more than 400 mm of water and a shunt was inserted. After this operation his general behaviour improved

and papilloedema subsided but his gross field loss persisted and impaired his ability to manoeuvre his wheelchair. His epileptic fits increased in severity and he died in 1982 after a severe convulsion.

2. H.T. This girl had a lumbar meningomyelocoele closed at birth in June 1971 and a Spitz Holter valve inserted one week later. She had multiple orthopaedic operations and one distal cather revision (1971). In August 1977 her distal catheter was noted to be short on chest x-ray but she remained asymptomatic. During routine examination as part of this study in July 1978, bilateral papilloedema as well as bilateral nystagmus on abduction was noted, although she was well and had no headaches. Her distal catheter was revised shortly afterwards. Her optic discs were still swollen two weeks later but eventually regained a normal appearance. Visual acuity remained at 6/6 6/6 Snellen but nystagmus on abduction persisted.

3. J.T. This girl had a thoraco-lumbar myelocoele closed at birth in July 1969. A Spitz Holter valve was inserted six days later and revised in October 1969 and July 1970. She was admitted to a Medical ward in September 1977 complaining of headaches. Her valve was still pumping normally. After two weeks of observation and investigations she was seen as part of the study and noted to have bilateral papilloedema. She also had reduced abduction in each eye despite the presence of a divergent squint. At operation she had a very high cerebrospinal fluid pressure and her distal catheter was revised. By November 1977 her papilloedema had not resolved and there was a new finding of nystagmus in elevation. In February 1978 she again developed headaches and a further distal catheter

revision was performed. Papilloedema gradually resolved although the disc margins were still blurred in November 1978. Her final visual acuity was 6/6 6/6.

4. M.W. This girl had primary hydrocephalus diagnosed at three months of age. A Spitz Holter valve was inserted in August 1973 and revised in June 1975. She was first seen as part of the study in February 1977 when she had no squint and binocular single vision was present (Wirt Fly positive). In March 1977 she was admitted with vomiting and drowsiness and a convergent squint. She had a therapeutic proximal catheter revision. In April 1977 she still had a small manifest right convergent squint and a right lateral rectus paresis. She was given spectacles to correct a small degree of hypermetropia but by July 1977 her eyes were straight and the lateral rectus paresis had completely resolved.

5. G.V. This boy had a lumbar myelocoele closed at birth in October 1969 and a Spitz Holter valve inserted two weeks later. The shunt was revised in October 1969, November 1969 and January 1970. In March 1974 he was seen at the Eye Hospital for a right convergent squint (noted by his mother from birth). Vision was 6/6 (Sheridan Gardner test) in each eye and his optic discs normal.

In August 1974 he was admitted to the General Hospital with chest and back pains. These were thought to be due to nerve root irritation and he was discharged four days later. In September 1974 he was seen again at the Eye Hospital and found to be completely blind with fixed dilated pupils and pale optic discs. He was admitted to the General Hospital three days

later with a history of fits and emotional disturbance in addition to the one week history of blindness. He was discharged from hospital three days later. Finally, fifteen days later (in October 1974) after neurological observations had been performed for five days he had surgery for a distal catheter blockage; his cerebrospinal fluid pressure was greater than 300mm of water.

After the operation he remained blind (bare perception of light in one eye only) with roving eye movements and pale cupped discs, though he was able to commence learning braille. His general handicaps included paraplegia, double incontinence, a weak left arm, recurrent urinary tract infections and hydronephrosis.

6. R.G. This child had a lumbo-sacral myelocoele closed at birth in August 1975 and a Spitz Holter valve inseted in September 1975 and revised in July 1976. He was seen at the Eye Hospital in July 1976 for a right convergent squint which had been noted a few months before. In May 1977 right convergent squint surgery was performed but he developed a right divergent squint soon after the operation. His distal catheter was revised prophylactically in August 1977. In December 1977 it became infected and was removed and replaced with a lumbar theco-peritoneal shunt.

In July 1978 he commenced a course of left occlusion because of right amblyopia. However, in October 1978 his mother reported a recent change in the squint and it was noted that he now had a left convergent squint with bilateral lateral rectus pareses and marked left amblyopia. A course

of right occlusion was commenced and the left vision improved but his squint remained convergent. In February 1979 an upgaze palsy developed. Shortly afterwards a ventriculo-atrial shunt was performed. CSF pressure was normal but there was an improvement in his general condition and school performance afterwards.

Left convergent squint surgery was performed in September 1979 with cosmetic improvement. At the close of the study he had visual acuity of R 6/18 L6/9 Snellen with a right convergent squint, A syndrome, bilateral lateral rectus pareses, nystagmus on upgaze and slightly pale optic discs.

7. R.S. This girl had a lumbo-sacral meningomyelocoele closed at birth (February 1978) and a Spitz Holter valve inserted eight days later. She had no prophylactic distal catheter revisions but in September 1979 her optic discs (which had previously been normal) were found to be swollen. Her anterior fontanelle was still open. Her distal catheter was lengthened shortly afterwards and the disc swelling resolved. At the close of the study vision was 6/5 in each eye and there was no squint or other ophthalmic problem.

8. T.S. At birth in January 1962 this boy was noted to have a solid lesion on his back. The lesion discharged at times. He walked unaided at two and a half years, but had an awkward gait and later required orthopaedic operations. He achieved daytime continence at four and a half, but later became incontinent and required a penile appliance and intermittent catheterisation.

In June 1970 the lesion (a myelomeningocoele) was explored and closed. In September 1976 a CT scan showed gross hydrocephalus. At times his back wound became swollen (presumably with cerebrospinal fluid). Learning problems at school had been attributed to his general condition. However, his mother (herself a myope) had taken him to an ophthalmic medical practitioner who prescribed spectacles for myopia and his school performance improved thereafter. His ophthalmic medical practitioner also noted that at times his optic discs became pink and ill defined (unlike that of most myopes).

In September 1979 he complained of headache and transient visual obscuration; his optic discs were slightly elevated and blurred. He was admitted for CSF manometry. However, the pressure was only 150mm of water and so no shunt was inserted. His headache and visual problems subsided spontaneously and his clinical condition remained unchanged to the end of the study.

9. L.V. This girl had a thoraco-lumbar myelocoele closed at birth in April 1964 and a Wade valve inserted three and a half months later. Her family moved home several times during her early years and the shunt was not revised until 1971. She first saw an ophthalmologist in 1973 when primary optic atrophy was diagnosed. She was first seen as part of this study in August 1980 when she was an inpatient complaining of headaches and had a short distal catheter on chest x-ray. Visual acuity was 6/36, 6/18 not improved by any lenses. She had a right convergent squint, bilateral lateral rectus pareses and nystagmus on upgaze. The optic discs were pale, the left being slightly blurred. Her distal catheter was

revised a few days later and the upgaze nystagmus disappeared but her vision did not improve, bilateral lateral rectus palsies persisted and the optic discs remained pale.

10. M.H. This boy was born in July 1974 at 29 weeks gestation. His early life was complicated by birth asphyxia, hyaline membrane disease, apnoeic attacks, pneumonia on two occasions, jaundice, anaemia, a bleeding tendency and septicaemia. He developed communicating hydrocephalus and a Spitz Holter valve was inserted in 1974 and revised in July 1975. He was severely mentally retarded and had convulsions.

When first seen in this study in March 1977 he had pale optic discs but no sign of retinopathy. A further shunt revision was performed in August 1978 but his condition deteriorated. He became unconscious with fixed dilated pupils, a left conjugate gaze palsy and papilloedema superimposed on optic atrophy. He died shortly afterwards.

11. D.R. This boy was born in January 1967 and had an operation for closure of a spina bifida lesion shortly afterwards. A shunt was inserted six weeks later. In August 1974 he developed sunsetting and a therapeutic revision of his distal catheter was carried out. A further distal catheter revision was performed in 1975. He had many orthopaedic operations and was confined to a wheelchair. He suffered from kyphosis, trophic ulceration of the feet, a neurogenic bladder and recurrent urinary tract infections.

He was first seen in this study in March 1977 when he had a left to alternating convergent squint with bilateral nystagmus on abduction and nystagmus on upgaze. By October 1977 he had lateral rectus pareses but remained generally well. In April 1978 he was admitted with suspected shunt dysfunction but was discharged without operation. By the end of July 1978 he had developed bilateral early disc swelling; at operation a few days later a discontinuity in the distal catheter was corrected. Cerebrospinal fluid culture also grew Staphylococcus albus. Post operatively he remained unwell with pyrexia and his papilloedema did not regress. He was discharged home but soon readmitted with headaches and drowsiness and had a respiratory arrest. The disc swelling had increased by this time. At operation CSF pressure was raised and an entirely new shunt system was inserted. Afterwards his condition improved, papilloedema and upgaze nystagmus disappeared, but his squint and nystagmus on abduction persisted. He also had bilateral inferior altitudinal field defects.

12. V.E. This girl had a lumbo sacral myelocoele closed shortly after birth in April 1973 and a Spitz Holter valve was inserted two weeks later. Over the next seven years she had fifteen therapeutic shunt operations including ventriculo-pleural and ventriculo-peritoneal shunts, exteriorisations (1975 and 1980) wound closure over the shunt and an intragastric shunt (1980). She had septicaemia due to Staph. aureus and E. coli in Septembner 1977 and Strep. viridans infection of the valve in December 1977. Nevertheless she was learning to walk and was relatively bright.

She was first seen in this study in January 1977 when a left convergent squint with left lateral rectus paresis was noted. This had been present for some time but no treatment arranged because of her many hospitalisations. She was found to have left amblyopia (RVA 6/6 LVA 6/36). Glasses to correct hypermetropia and right occlusion were prescribed but there was no improvement in the left visual acuity.

In June 1978 she presented with a complaint of headaches for twenty four hours and was found to have bilateral papillodema. At operation the same day CSF pressure was 250 mm of water and her distal catheter was revised (her 13th shunt operation). Afterwards papilloedema resolved and she remained well until May 1980 when she became severely ill and papilloedema recurred. Despite two further shunt operations she died.

13. T.G. This boy had a thoraco lumbar myelocoele closed at birth in October 1963 and a Spitz Holter valve inserted shortly afterwards. His distal catheter was revised in 1968 and the whole system changed in January 1974.

He was first seen in this study in June 1977, a severely handicapped child. He had already had a convergent squint operation, but remained convergent with bilateral nystagmus on abduction. In September 1977 he had a prophylactic distal catheter lengthening, but post-operatively developed headache and vomiting. His squint and ocular motility were unchanged and there was no papilloedema but the right pupil was dilated. However, he was discharged from the ward shortly afterwards and his pupil sizes equalised.

He was seen again in December 1977 with a history of a dilated right pupil for one month. At this time he also had complete bilateral lateral rectus palsy, nystagmus in upgaze and downgaze and pallor of the right optic disc. Next day the CSF pressure was 280mm of water; his distal catheter was found to be looped and so was shortened. He remained unwell, both pupils became dilated and only reacted to light sluggishly and left papiloedema with haemorrhages developed. A further therapeutic distal catheter revision was performed after which he recovered well. The Parinaud's syndrome resolved completely, but nystagmus on abduction persisted and both discs remained pale.

14. S.W. This boy had a meningomyelocoele closed two weeks after birth in July 1963, and a Spitz Holter valve inserted two months later. (Sunsetting had been present shortly before valve insertion.) He had a therapeutic distal catheter revision in 1970.

He was first seen in this study in September 1977. He had severe general handicaps. There was no squint but he had bilateral nystagmus on abduction and nystagmus on upgaze.

In June 1979 a spinal osteotomy and insertion of Harrington's rods for kyphoscoliosis was performed. Soon afterwards he developed headaches and a stiff neck and in August 1979 he suffered transient loss of vision. At this time he also developed a left convergent squint, left lateral rectus paresis, nystagmus on downgaze and bilateral papilloedema with haemorrhages. At emergency operation a disconnected distal catheter was

found. Afterwards he had impairment of speech for a while but gradually recovered. Upgaze and downgaze nystagmus and papilloedema resolved but nystagmus on abduction persisted and he had a divergent squint in downgaze - an example of spontaneous consecutive divergence.

15. M.W. This boy had a meningomyelocoele closed at birth in April 1966 and a Wade valve inserted three weeks later. He had Staph. albus septicaemia in May 1966 and therapeutic shunt revisions in 1969, 1970, 1972, June and November 1973. He was very severely handicapped.

He had first seen an ophthalmologist in 1973; convergent squint, nystagmus on abduction and upgaze were noted. When first seen in this study in May 1977 he also had visual field restriction and pale optic discs.

In June 1983 he complained of severe headaches and his speech was slurred. He had developed upbeating nystagmus in the primary position and sluggishly reacting pupils. He was admitted and treted for a urinary tract infection. He began to feel a little better. At this time it was observed that his vision was too poor to permit self-catheterisation, although his visual acuity was right 6/9 left 6/6. He was discharged home but readmitted in December 1983 with recurrence of headaches. His vision had fallen to counting fingers in each eye. A therapeutic proximal catheter revision was performed. Four days later his visual acuity was 6/60, 6/36 and one week later 6/9 6/6. His pupils had returned to normal but upbeating nystagmus persisted.

16. L.R. This girl had a spina bifida lesion closed at birth in December 1978 and a Spitz Holter valve was inserted three weeks later. (Sunsetting was present prior to valve insertion.) Sunsetting recurred in April 1979 with bilateral lateral rectus palsies and a therapeutic distal catheter revision was performed. She subsequently recovered full eye movement and had no squint and a visual acuity of 6/4, 6/4 (Sheridan Gardner).

In March 1983 she developed bilateral musculoparetic nystagmus on lateral gaze and in July 1983 nystagmus on upgaze, although she remained generally well. A "prophylactic" shunt revision was being considered when in September 1983 she developed headaches and complained of blurred vision. The right visual acuity was 6/9 Snellen, left visual acuity unobtainable and she had bilateral papilloedema. A therapeutic distal catheter revision was performed and papilloedema subsequently resolved, but nystagmus on abduction and upgaze persisted.

17. A.J. This boy had a lumbar myelocoele closed at birth in May 1973 and a Spitz Holter valve inserted one week later. Over the next five years he had a further 13 shunt operations.

When first seen in this study in January 1977 he had a right divergent squint, a pale and swollen right optic disc, and a Roth's spot near the left disc. His entire shunt system had been removed and replaced two weeks before (for staph albus infection). Over the next few months his general condition improved and the disc swelling and Roth's spot disappeared. In November 1977 he was admitted with headaches and vomiting and papilloedema. The CSF pressure was over 250 mm of water and his

proximal catheter was revised. Two further shunt operations were carried out over the next few months. He then recovered quite well but had very severe general handicaps. At the close of the study he had bilateral optic atrophy but could see 6/9 in each eye with glasses for myopia and astigmatism.

18. N.H. This child presented with poor feeding, enlarged head and sunsetting at three weeks of age in February 1982. Ultrasonography showed hydrocephalus and a large cyst in the posterior cranial fossa. A cerebrospinal fluid shunt with catheters in the right lateral ventricle and posterior fossa were inserted but revised one day later.

He was first seen in this study in March 1982; he had spontaneous pendular vertical nystagmus. No horizontal eye movements could be elicited. By May 1982 the vertical nystagmus had disappeared and full horizontal eye movement could be elicited. In August 1982 he had an intermittent left convergent squint with a left lateral rectus palsy. A therapeutic proximal catheter revision was performed in October 1982, but the shunt became infected with Staph. albus in November 1982. By February 1983 the left convergent squint had become constant; he had developed horizontal pendular nystagmus of the right eye and his vision seemed poor. The ventricular and cyst catheters were revised shortly afterwards and for a while he seemed better; the nystagmus resolved, his squint alternated and there was left lateral rectus paresis.

By March 1983 he again had nystagmus of the right eye and sunsetting at times. A further proximal and cyst catheter revision was performed, but there was only a transient improvement. After a further shunt exploration

one week later he became extremely ill with gastrointestinal bleeding. In April 1983 a craniotomy was performed and the arachnoid cyst fistularized into the third ventricle. After the operation he slowly improved but had a very marked left convergent squint (greater than 50 degrees) with complete left lateral rectus palsy and left amblyopia. As soon as he was well enough, part time right occlusion was commenced with recovery of left vision. At the close of the study the angle of squint had not lessened and he was on the waiting list for left squint surgery.

Appendix II

Letter to Paediatrician or Paediatric Surgeon.

Dear Dr.

For some years I have been screening all the children who attend the Spina Bifida and Hydrocephalus Clinic at Southampton General Hospital, for visual defects, particularly those (e.g. papilloedema) which might assist in the diagnosis of raised intracranial pressure.

I am under the impression that there are very few centres in the country where such children receive routine ophthalmological screening, and wonder if you would be kind enough to tell me whether this is the case at the ..... Hospital.

Yours sincerely,

Appendix III - Tables

Key		
	SBO	Spina bifida occulta
	SBC	Spina bifida cystica
	SBAH	Spina bifida with arrested hydrocephalus
	SBH	Spina bifida with hydrocephalus (shunt-treated)
	H	"Primary" hydrocephalus
	E	Occipital encephalocoele
	EH	Occipital encephalocoele with hydrocephalus

Table 1. Incidence of Squint in Spina Bifida and Hydrocephalus.

Author(s)	Number of Patients	Age of Patient	Source of Patient	Diagnosis of Patients	%	Incidence of Manifest Squint
(1) Clements & Kaushal[24]	130	0-12 years	Random (Geographically remote patients excluded)	SBC	27	52%
				SBAH	14	71%
				SBH	64	83%
				H	24	87%
						Total 73%
(2) Goddard[29]	251	0-11 years	Random + specific referrals to eye clinics	H & SBH		30%
(3) Harcourt[38]	70	?	?	SBH		30%
(4) Hunt[28]	100	5-14 years	Random (Survivors of consecutive series)	SBC & SBH		59%
(5) Rabinowicz[26]	100	3 months - 15 years	?	SBH	80	54%
				H	20	
(6) Tew & Laurence[39]	55	10 years	Random (Survivors of consecutive series)	SBC/SBH	94	33%
				E	4	
				H	2	
(7) Present series (Gaston)	322	0-18 years	Random	SBO	4	0
				SBC	10.5	1
				SBAH	6	3
				SBH	44	64
				H	33.5	31
				E	1	0
				EH	1.5	0
						Total 42%

Table 2. Diagnosis of 322 patients.

Diagnosis	No	%
Spina bifida occulta (SBO)	12	4
Spina bifida cystica alone (SBC)	34	10.5
Spina bifida with arrested hydrocephalus (SBAH)	19	6
Spina bifida with hydrocephalus (treated by a shunt) (SBH)	141	44
Hydrocephalus alone (H)	108	33.5
Occipital encephalocoele (E)	3	1
Occipital encephalocoele and hydrocephalus (EH)	5	1.5

Table 3. Aetiology of Hydrocephalus without spina bifida or encephalocoele.
108 Patients.

Diagnosis	No	%
Unknown	39	36
Intracranial haemorrhage/trauma	33	30.5
Post meningitic	12	11
Sex linked aqueduct stenosis	6	5.5
Porencephaly	5	5
Arachnoid cyst	4	4
Hydranencephaly	3	3
Toxoplasmosis	2	2
Irradiation	1	1
Tuberose sclerosis	1	1
Leucodystrophy	1	1
Pyle's disease	1	1

Table 4. Sunsetting Phenomenon.

	Total patients	53	16%
Number of patients with repeated episodes		5	9%
	Total episodes	58	
Age at observation -	less than 6/12	48	83%
	more than 6/12	10	17%
Diagnosis	SBC	1	2%
	SBAH	1	2%
	SBH	15	28%
	H	34	64%
	EH	2	4%
Visual handicap	Normal	9	17%
	Minor	9	17%
	Moderate	1	2%
	Major	4	7.5%
	Unclassified	30	56%
General handicap	None	4	7.5%
	Mild	10	19%
	Intermediate	11	21%
	Severe	8	15%
	Very severe	9	17%
	NC	11	21%
Deaths		4	7.5%
Subsequent optic atrophy	Yes	11	21%
	No	41	77%

Table 5. Parinaud's syndrome.

			Total patients 21	6.5%
Features	Upgaze nystagmus		11	52%
	Upgaze palsy		4	19%
	Upgaze nystagmus/palsy + dilated sluggish pupils		2	9.5%
	Upgaze nystagmus/palsy + downgaze nystagmus		2	9.5%
	Upgaze nystagmus/palsy + downgaze nystagmus + dilated pupils		2	9.5%
Diagnosis	SBH		17	81%
	H		4	19%
Visual handicap		Minor	11	52%
		Moderate	4	19%
		Unclassified	6	28.5%
General handicap		Intermediate	7	33%
		Severe	7	33%
		Very severe	6	28.5%
		NC	1	5%
Persistence	Yes	- totally	8	38%
		- partially	1	5%
	No		11	52%
	Insufficient follow up		1	5%

Table 6. Manifest Convergent Squint.

Total		89	28%
Diagnosis	SBC	1	1%
	SBAH	3	3
	SBH	57	64%
	H	28	31%
Unilateral or bilateral hypermetropia 2D or more		17	19%
Amblyopia of 1 line or more		32	36%
Amblyopia 5 lines or more		1	1%
Positive family history		13	14%
Lateral rectus palsy/paresis ever		51	57%
A syndrome present		37	41%
Previously undiagnosed		29	32.5%
Squint operation with improvement		26	29%
No operation		63	71%
Transient squint only		5	6%

Table 7. Manifest Divergent Squint.

Total		27	8%
Diagnosis	SBC	1	4%
	SBH	10	37%
	H	14	51%
	EH	2	7%
Myopia 1D or more		2	7%
Major visual handicap		8	30%
Positive family history		5	18.5%
Lateral rectus palsy		9	33%
A syndrome		5	18.5%
Previously undiagnosed		14	52%
Operation		1	4%

Table 8. Consecutive Divergence.

Total		20	6%
Diagnoses	SBC	1	5%
	SBH	12	60%
	H	7	35%
Hypermetropia 2D or more		2	10%
Amblyopia 1+ line		7	35%
Amblyopia 5+ lines		1	5%
Positive family history		3	15%
Lateral rectus palsy		15	75%
A syndrome		7	35%
Major visual loss		2	10%
Previously undiagnosed		2	10%
Previous operation for convergent squint		14	70%
No operation		6	30%

Table 9. Horizontal Pendular Nystagmus.

Total		10	3%
Diagnoses	SBH	1	10%
	H	9	90%
Visual handicap	Major	6	60%
	Unclass.	4	40%
General handicap	Severe	3	30%
	Very severe	6	60%
	NC	1	10%

Table 10. Conjugate Lateral Gaze Palsy.

Total		11	3%
Diagnoses	SBH	1	9%
	H	10	91%
Visual handicap	Major	2	18%
	Unclass.	9	82%
General handicap	Intermediate	3	27%
	Severe	1	9%
	Very severe	7	64%
Deaths		3	27%

Table 11. Hortizontal Jerk Nystagmus.

Total		7	2%
Diagnoses	SBH	3	43%
	H	3	43%
	EH	1	14%
Visual handicap	Minor	3	43%
	Major	1	14%
	Unclass.	3	43%
General handicap	Intermediate	1	14%
	Severe	2	28.5%
	Very severe	3	43%
	NC	1	14%
Deaths		2	28.5%

Table 12. Lateral Rectus Palsy/Paresis/Paretic Nystagmus.

Total		93	29%
Diagnoses	SBC	1	1%
	SBAH	1	1%
	SBH	67	72%
	H	22	23%
	EH	2	2%
Palsy ever		54	58%
bilateral		37	39%
unilateral		17	18%
Nystagmus only		39	42%
bilateral		36	39%
unilateral		3	3%
Squint. Manifest convergent		56	60%
Consecutive divergent		14	15%
Divergent		7	7.5%
Latent convergent		2	2%
Latent divergent		2	2%
None		12	13%
Transient palsy with transient convergent squint		3	3%
Normal visual function		5	5%

Table 13. Refractive Errors.

Total	71	22%
Myopia	15	5%
Hypermetropia	21	6.5%
Astigmatism	35	11%
Anisometropic amblyopia	1	

Table 14. Papilloedema.

Total patients		44	14%
Recurrent papilloedema		2	4.5%
Unilateral		4	9%
Bilateral		40	91%
Age		6/52	- 16 years
Diagnoses	SBH	34	77%
	H	9	20%
	EH	1	2%
Visual function	Normal	9	20%
	Minor	20	45%
	Moderate	2	4.5%
	Major	2	4.5%
	Unclassified	11	25%
General handicap	None	2	4.5%
	Mild	6	14%
	Intermediate	13	29.5%
	Severe	12	27%
	Very severe	9	20%
	Unclassified	2	4.5%
Deaths		7	16%
Development of optic atrophy	Yes	6	14%
	No	18	41%
	Already present	3	7%
	Died early	1	2%
	Insufficient follow up	16	36%

Table 15. Optic Atrophy.

Total		55	17%
Bilateral		39	71%
Unilateral		16	29%
Disc cupping		10	18%
Diagnoses	SBH	25	45%
	H	29	53%
	EH	1	2%
Visual handicap	Minor	17	31%
	Moderate	7	13%
	Major	9	16%
	Unclassified	22	40%
General handicap	None	1	2%
	Mild	3	5%
	Intermediate	7	13%
	Severe	16	29%
	Very severe	23	42%
	Not classified	5	9%
Deaths		10	18%

Table 16. Cortical Blindness.

Total		9	3%
Co-existing optic atrophy		8	89%
Diagnoses	H	8	89%
	EH	1	11%
Visual handicap at close of study			
	Major	3	33%
	Unclassified	6	67%
General handicap	Intermediate	2	22%
	Severe	1	11%
	Very severe	4	44%
	Not classified	2	22%
Deaths		2	22%

Table 17. Visual Evoked Potentials of 19 Patients.

VER	Normal	6	31.5%
	Abnormal	13	68.5%
BAEP	Normal	2	10.5%
	Abnormal	17	89.5%

Table 18. Normal Visual Function.

Total		86	27%
Diagnoses	SBO	7	8%
	SBC	18	21%
	SBAH	13	15%
	SBH	34	39.5%
	H	14	16%
Transient eye problems		11	13%
General handicap	None	15	17%
	Mild	28	32%
	Intermediate	26	30%
	Severe	13	15%
	Very severe	2	2%
	NC	2	2%
Deaths		None	

Table 19. Minor Visual Handicap.

Total			74	23%
Diagnoses		SBC	2	3%
		SBAH	3	4%
		SBH	58	78%
		H	11	15%
General handicap		None	10	13%
		Mild	6	8%
		Intermediate	22	30%
		Severe	22	30%
		Very severe	13	17.5%
		NC	1	1%
Deaths			2	3%

Table 20. Moderate Visual Handicap.

Total			18	5.5%
Diagnoses		SBC	1	5.5%
		SBAH	1	5.5%
		SBH	13	72%
		H	3	17%
General handicap		Mild	3	17%
		Intermediate	4	22%
		Severe	6	33%
		Very severe	3	17%
		NC	2	11%
Deaths			2	11%

Table 21. Major Visual Handicap.

Total		11	3%
Diagnoses	SBH	2	18%
	H	9	82%
General handicap	Severe	2	18%
	Very severe	9	82%
Deaths		2	18%

Table 22. Unclassified Visual Handicap.

Total		133	41%
Diagnoses	SBO	6	4.5%
	SBC	13	10%
	SBAH	1	1%
	SBH	36	27%
	H	69	52%
	E	2	1.5%
	EH	6	4.5%
General handicap	None	14	10.5%
	Mild	13	10%
	Intermediate	22	16.5%
	Severe	24	18%
	Very severe	21	16%
	NC	39	29%
Deaths		9	7%

Table 23. Field Defects.

Total	15	5%
Generalised restriction	5	33%
Inferior altitudinal defects	2	13%
Quadrantanopias	2	13%
Homonymous hemianopia	6	40%

Table 24. Summary of Visual Function.

Total	322	100%
Normal	86	27%
Minor	74	23%
Moderate	18	5.5%
Major	11	3%
Unclassified	13	41%

Table 25. Summary of General Handicap.

Total	322	100%
None	39	12%
Mild	50	15.5%
Intermediate	74	23%
Severe	67	21%
Very severe	48	15%
NC	44	14%

Table 26. Correlation of Visual and General Handicaps. Numbers.

General/Visual	Normal	Minor	Moderate	Major	Unclass.	Total
None	15	10	0	0	4	39
Mild	28	6	3	0	13	50
Intermediate	26	22	4	0	22	74
Severe	13	22	6	2	24	67
Very severe	2	13	3	9	21	48
NC	2	1	2	0	39	44
	86	74	18	11	134	322

Table 27. Correlation of Visual and General Handicaps. Percentages.

General/Visual	Normal	Minor	Moderate	Major	Unclass.	Total
None	5	3	0	0	4	12
Mild	9	2	1	0	4	15.5
Intermediate	8	7	1	0	7	23
Severe	4	7	2	1	7	21
Very severe	1	4	1	3	6.5	15
NC	1	0	1	0	12	14
	27	23	5.5	3	41	100%

Table 28. Effect of Selection on Visual Handicap. Numbers.

	Normal	Minor	Moderate	Major	Unclass.	Total
Pre	41	44	10	6	12	113
Mid	25	25	4	1	11	66
Post	20	5	4	4	110	143
	86	74	18	11	133	322

Table 29. Effect of Selection on Visual Handicap. Percentage.

	Normal	Minor	Moderate	Major	Unclass.	Total
Pre	13	14	3	2	4	35
Mid	8	8	1	0	3	20
Post	6	1.5	1	1	34	44
	27	23	5	3	41	100

Table 30. Deaths.

Total		15	5%
Diagnoses	SBO	1	7%
	SBH	7	46%
	H	5	33%
	EH	2	13%
General handicap	Intermediate	1	7%
	Severe	3	20%
	Very severe	8	53%
	NC	3	20%
Visual handicap	Minor	2	13%
	Moderate	2	13%
	Major	2	13%
	Unclassified	9	60%
Selective category	Pre	6	40%
	Post	9	60%

Table 31. Raised Intracranial Pressure Study.

Total patients	53	
Preoperative findings.		
Episode without general signs or symptoms	3	6%
Valve pumping normally	9	17%
Ophthalmic findings.		
Papilloedema	27	51%
Sunsetting	2	4%
New or increased Parinaud's	7	13%
New or increased squint	6	11%
New or increased LR palsy/paresis	9	17%
Blurred vision	2	4%
Total positive eye signs	37	70%
Total negative eye signs	16	30%
Operative findings.		
Raised ICP/no valve	2	4%
Distal block	28	53%
Proximal block	14	26%
Entire shunt abnormal (infected)	6	11%
Disconnection	3	6%

Table 32. Correlation Between Operative Findings and Positive Eye Signs.

Raised intracranial pressure - no valve	2 positive (100%)
Distal block	24 positive (86%)
Proximal block	7 positive (50%)
Entire shunt abnormal (infected)	2 positive (33%)
Disconnection	2 positive (67%)

Table 33.

Visual Handicap After Sunsetting.	%	Visual Handicap in Series as a Whole.	%
Normal	17		27
Minor	17		23
Moderate	2		5.5
Major	7.5		3
Unclassified	56		41