Orthoptic Findings and Visual Fixation in children in general and in children with surgically treated hydrocephalus

Eva Aring

Department of ophthalmology
Institute of Neuroscience and Physiology
The Sahlgrenska Academy at Göteborg University
"When one is a bear with a very little brain and think about things, one discovers that an idea that seemed like a real idea in your brain, is different when it comes out in the open and other people witness it."

Pooh
Abstract

**Background:** The ocular motor system is complex and dependent on the cooperation of many different areas of the brain. Ocular motor functions may be disturbed by various factors resulting in strabismus, and abnormal fixational behaviour, which may interfere on a multitude of visual functions. To study binocular functions orthoptic methods may be used and when studying visual fixation eye-trackers may be used. However, previous orthoptic studies often lack precise definitions, and studies of visual fixation behaviour in children are rare. In a multidisciplinary study of children with surgically treated hydrocephalus (HC), a variety of functions, including ophthalmologic and orthoptic functions were to be studied.

**Aim:** The aim of this study was to obtain results of commonly used orthoptic tests and visual fixation behaviour in a sample of children, for use as comparison in studies of different patient groups, and to compare these results, with the same variables in a population-based group of children with surgically treated HC. We also wished to study the effects of the onset and the aetiology of HC, associated neurological impairments and the ventricular width on orthoptic variables and visual fixation behaviour.

**Materials and methods:** Papers I and III: The comparison group consisted of children aged 4−15 years from different schools and pre-schools in the area of Göteborg. Papers II and IV: The HC group consisted of a population-based group of children aged 7−12 years, with surgically treated HC. All children in both groups were evaluated with the same test battery of orthoptic test commonly used in clinical practice. An infrared eye-tracker (Orbit) was used to evaluate visual fixation behaviour.

**Results:** Paper I: In the comparison group, five children (3.5%) had heterotropia, and one child had abnormal ocular motility. Heterophoria was found in 37 children (26%) and was four times more common at near than at distant fixation. The near point of convergence was ≤6 cm in 97% of the children, and 97% had stereo acuity of 60” or better. The results of the AC/A calculations with the methods used were regarded as unreliable in this age group. No anomalous head postures or nystagmus were observed.

**Paper II:** Heterotropia (69%), abnormal head posture (41%), nystagmus (44%), and motility abnormalities (60%) were significantly more common in children with HC (p<0.001) than in the comparison group. Children with overt HC at birth had a significantly higher frequency of orthoptic abnormalities compared to children developing HC during the first year of life.

**Paper III:** In the comparison group, both fixation time (p=0.02) and fixation density around the centre of gravity of fixation (p<0.01) increased with increasing age, while number of intruding saccades decreased (p<0.01) with increasing age. Blinks and drifts could not be shown to be age dependent.

**Paper IV:** As a group children with HC had shorter fixation time (p<0.01) and higher frequency of drifts (p<0.01) than the comparison group. However, children with myelomeningocele (MMC), did not differ from the comparison group in these variables, while children with associated neurological impairments, were more affected than those without.

**Conclusion:** We have obtained results from commonly used orthoptic tests and described the fixation behaviour in children. Fixation stability improves with age in children in general. Children with HC with aetiologies other than MMC, a group with more associated neurological impairments, had more abnormal fixation behaviour than those with MMC. However, regarding orthoptic abnormalities children with MMC were equally affected indicating the vulnerability of the binocular system in children with HC.

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Original papers

This thesis is based upon the following original papers, which will be referred to in the text by their Roman numerals:


IV. Aring E, Andersson S, Hellström A, Ygge J. Abnormal visual fixation in children with hydrocephalus. *In manuscript.*

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### Abbreviations and acronyms

<table>
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<tr>
<th>Abbreviation</th>
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<tr>
<td>AC/A</td>
<td>ratio of accommodative convergence to accommodation ratio</td>
</tr>
<tr>
<td>aCT</td>
<td>alternate cover test</td>
</tr>
<tr>
<td>CNS</td>
<td>central nervous system</td>
</tr>
<tr>
<td>CP</td>
<td>cerebral palsy</td>
</tr>
<tr>
<td>CSF</td>
<td>cerebrospinal fluid</td>
</tr>
<tr>
<td>CT</td>
<td>(monolateral) cover test</td>
</tr>
<tr>
<td>D</td>
<td>diopeter</td>
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<tr>
<td>EP</td>
<td>epilepsy</td>
</tr>
<tr>
<td>Eph</td>
<td>esophoria</td>
</tr>
<tr>
<td>ET</td>
<td>esotropia</td>
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<tr>
<td>HC</td>
<td>hydrocephalus</td>
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<tr>
<td>ICP</td>
<td>intracranial pressure</td>
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<tr>
<td>IQ</td>
<td>intelligence quotient</td>
</tr>
<tr>
<td>IR</td>
<td>infrared</td>
</tr>
<tr>
<td>IXT</td>
<td>intermittent exotropia</td>
</tr>
<tr>
<td>LogMAR</td>
<td>logarithm of the minimum angle of resolution</td>
</tr>
<tr>
<td>MMC</td>
<td>meningo(myelo)cele or myelomeningocele</td>
</tr>
<tr>
<td>NPC</td>
<td>near point of convergence</td>
</tr>
<tr>
<td>n.s.</td>
<td>non-significant</td>
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<tr>
<td>pCT</td>
<td>prism and cover test</td>
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<tr>
<td>PD</td>
<td>prism dioptre</td>
</tr>
<tr>
<td>PHHC</td>
<td>post-haemorrhage hydrocephalus</td>
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<tr>
<td>RAF</td>
<td>Royal Air Force ruler</td>
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<tr>
<td>SD</td>
<td>standard deviation</td>
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<td>VA</td>
<td>visual acuity</td>
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<tr>
<td>Xph</td>
<td>exophoria</td>
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<tr>
<td>XT</td>
<td>exotropia</td>
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Glossary

**Accommodation (A):** Ability of the eye to change refractive power of the lens and automatically focus objects at various distances on the retina.

**Amblyopia:** Reduced visual acuity (VA), due to abnormal visual experience in childhood.

**Cover test (CT):** The main method of detecting strabismus. Each eye is covered, while the examiner looks for movement of non-covered eye in order to reveal any manifest strabismus.

**Diopter (D):** Unit to measure the power of a lens.

**Esophoria (Eph):** Latent convergent strabismus, kept in check by the fusion mechanism in binocular viewing.

**Esotropia (ET):** Convergent strabismus (i.e. one eye deviates towards the nose).

**Exophoria (Xph):** Latent divergent strabismus, kept in check by the fusion mechanism in binocular viewing.

**Exotropia (XT):** Divergent strabismus (i.e. one eye deviates away from the nose).

**Fusion:** *Sensory fusion:* Ability to appreciate two similar images, one from each eye, and interpret them as one. *Motor fusion:* Ability to maintain a single fused image during vergence movements.

**Heterophoria:** Latent ocular misalignment, kept in check by the fusion mechanism in binocular viewing.

**Heterotropia:** Manifest deviation not kept in check by fusion.

**Hydrocephalus (HC):** Progressive ventricular dilatation caused by disturbance in circulation of the cerebrospinal fluid.

**Intruding saccade:** An exaggregation of the normal microsaccadic movement associated with fixation.

**Nystagmus:** Involuntary, rhythmic, pendular or jerky conjugate oscillations of the eyes.

**Ocular motility:** Ability to use the extra-ocular muscles (EOMs) to move the eye in different directions.

**Prism diopter (PD):** Unit used for measurement of the size of ocular deviations. For example, 1 PD displaces the visual axes, referred to the “centre of rotation”, by 1 cm at a distance of 1 m, and is equal to 0.57°.

**Stereo acuity:** Quantitative measurement of the degree of disparity of two images fused into one and measured in seconds of arc (”).

**Strabismus:** Misalignment of the visual axes.
Introduction

Human vision is a complex system involving two eyes and many different cortical and subcortical areas of the brain. This system must move both eyes synchronously, keep them stably together in different gaze positions, and transfer a clear image to the areas of the brain concerned with perception of the world.

Orthoptics (Greek, orthos = straight, ops = vision) is an ophthalmological field concerned with the diagnosis and treatment of patients with strabismus and eye movement disorders. Strabismus may cause amblyopia and because the sensitive period for developing and treating amblyopia is during childhood, the majority of orthoptic patients are children.

Orthoptic treatment, to restore binocular single vision became popular in the 1930’s and, the theory was that the defect binocular function was the cause of strabismus and could be improved by training (Lyle and Wybar 1970). It was later realised that defect binocular function, could also be the result of a deviation and not the cause. Orthoptic exercises with training of binocular control are nowadays mainly used to increase control before or after strabismus surgery. Pleoptics is a treatment used to eradicate non-central or excentric fixation in the deviating eye. However, the use of this treatment has decreased or ceased in recent years. Pleoptics includes daily treatment sessions over a number of weeks, in which the patients are trained to encourage foveal fixation (Loudon 2007). Another task for the orthoptist was, and still is today to examine vision and if necessary treat amblyopia.

In Sweden, orthoptic education began in 1973, with influences from both the English and the German schools. Most of the original orthoptic examination methods are still widely used and clinically well established, probably because they are well functioning and easy to perform. However, it is difficult to evaluate previous orthoptic studies, since the methods used are poorly and rarely described, the tests may be used in more than one way, and thus reference values differ between study groups.

A cohort of children aged 4-15 years, created to represent a random sample of the general population, was investigated with a battery of clinically commonly used orthoptic tests (Rowe 2004). To complement the orthoptic evaluation, we included a quantitative measurement of fixation stability with an infrared ocular motor tracker (Bolzani et al. 1998; Ygge et al. 1999). In the second part of the thesis, a population-based cohort of children surgically treated for HC was evaluated with the same tests as the comparison group.

In Sweden, epidemiological studies of childhood HC have been performed for many decades (Hagberg et al. 1963; Fernell et al. 1986; Fernell et al. 1987; Hagberg et al. 1988), while ophthalmological/orthoptic investigations have been few
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(Lennerstrand et al. 1990; Caines and Dahl 1997). Children with HC have a high frequency of neurological, cognitive, and behavioural problems and, a majority of HC children have low average IQ or mental retardation (Heinsbergen 2002; Lindquist et al. 2005). Children with HC developing before one year of age, is a group of children with brain lesions occurring during development of binocular functions and fixation. This group is known to have a high incidence of strabismus, ocular motility defects and nystagmus due to the effect the disorder has on the central nervous system (Gaston 1985; Tamura and Hoyt 1987; Gaston 1991; Biglan 1995; Hoyt and Good 1995). Thus an orthoptic evaluation was an important part of this population-based multidisciplinary study, aiming to obtain comprehensive knowledge of the abilities/disabilities of the children in this group. The information gained may be used to compare outcome of regimes and treatments in children developing HC during the first year of life and to design habilitation.

In this study a comprehensive ophthalmologic investigation was performed in both the comparison group (Grönlund, Andersson et al. 2006) and among HC children (Andersson et al. 2006), and neurological, and neuropsychological investigations were also performed in the HC group, and those results have been presented elsewhere (Lindquist 2007; Persson 2007).

The aim of this study was twofold. Firstly, we wished to obtain comparison values for clinically commonly used orthoptic variables and fixation behaviour in children. Secondly, we wished to examine orthoptic functions and fixation behaviour in a multidisciplinary approach including paediatric, neurophysiologic and neuroradiologic evaluations, in a population-based group of children with HC surgically treated before one year of age. We also wanted to study the association between both the orthoptic variables and fixation behaviour and the time of onset and the aetiology of HC, associated neurological impairments, and the ventricular width.
Background

Ocular alignment and visual fixation are complex functions involving many different areas of the brain. These functions may be disturbed by a variety of causes, such as abnormal early visual development or underlying neurological or neuromuscular disease. The spectrum of disease in children differs from that in adults (Cassidy et al. 2000); in children, the disease affects an immature brain at different developmental stages, while in adults a mature and fully developed brain is affected. The infant brain has great plasticity and ability to reorganize and recover after injury, and so, naturally, the age at injury affects the adaptive mechanism (Johnston 2003). In infancy, there is a close relationship between visual impairment and ocular motor disorders; not only strabismus and nystagmus, but also ocular motility, fixation, saccadic, and pursuit movements may be affected (Jacobson et al. 1996; Jacobson and Dutton 2000; Jan et al. 2001; Salati et al. 2002). Thus, cerebral maldevelopments and lesions from a variety of causes, especially insults to the parietal-occipital cortex and underlying white matter (geniculostriate projections or optic radiations) may result in impairment of these functions (Hertle 2005). Periventricular and intraventricular haemorrhage in the neonatal period and retinopathy of prematurity increases the prevalence of infantile strabismus and abnormal fixation (Bremer et al. 1998; O’Keefe et al. 2001; Salati et al. 2002; VanderVeen et al. 2006). Low birth weight (≤2500 g), Down’s syndrome, and other mental handicaps are also associated with increased risk (Castane et al. 1995; Pott et al. 1999).

Associated systemic disease has been noted in about one-third of patients with heterotropia (Lorenz 2002). Heredity is an important risk factor for strabismus, and approximately 30% of children born to strabismic parents will develop strabismus themselves (Abrahamsson et al. 1999). Family history of strabismus has also been shown to be a risk factor for inferior oblique muscle overaction, dissociated vertical deviation, and latent nystagmus in children with infantile esotropia (Taira et al. 2003). A relationship between refractive errors and strabismus has been reported, where hyperopia has been shown to be more pronounced in children with esotropia than in children with exotropia at the time of detection of strabismus, and anisometropia frequently develops after onset of esotropia (Abrahamsson et al. 1992).

An extensive cortical and sub cortical network is involved in controlling the ocular motor system coding for accuracy and stability of visual fixation. Lesions in these brain areas may thus affect the stability of visual function. Such lesions could give rise to extraneous eye movements such as saccades and drifts. Blinks may also affect the stability of visual fixation. For the best visual acuity, it is important that the image is brought close to, and held steady at the centre of the fovea (Leigh and Zee 1999).

So far, few studies on fixation behaviour have been performed in children. It is
well known that children with CNS lesions such as, intraventricular haemorrhage, and periventricular leukomalacia may have unstable fixation (Jacobson et al. 1998; Salati et al. 2002). In addition children with neurodevelopmental disorders, such as autism, attention-deficit/hyperactivity disorder, and dyslexia, may have a defective ocular motor control (Fischer and Hartnegg 2000; Gould et al. 2001; Sweeney et al. 2004; Nowinski et al. 2005), which could present as an unstable fixation. A consequence of cerebellar pathology may be the occurrence of square wave jerks, i.e. intruding saccades, while impaired flocculus and paraflocculus functions may present with centripetal ocular drifts during gaze holding in lateral gaze (Zee et al. 1976; Dell’Osso et al. 1977; Zee et al. 1980). In addition, frequent intruding saccades when fixating has been described in children with albinism without nystagmus (Timms et al. 2006). In the adult patient, a high frequency of intruding saccades during fixation has been described in connection with disorders such as progressive supranuclear palsy, Friedreich’s ataxia and focal cerebral lesions (Sharpe et al. 1982; Abadi and Gowen 2004).

At birth the ability to fixate steadily is not yet developed, and stable fixation is usually acquired during the first six months of life (von Noorden and Campos 2002), paralleling the maturation of the fovea and the CNS. Ocular and visual defects per se, i.e. congenital cataract, strabismus, and amblyopia can also give rise to unstable fixation (Leigh et al.1989; Varma et al. 2006; Siepmann et al. 2006; Abadi et al. 2006). Three components are important for visual fixation; the first is suppression of unwanted saccades, the second is the ability to detect image drift on the retina and program corrective eye movements, and the third is the recalibration of eye movements to optimise gaze stability (Leigh and Zee 1999).

**Hydrocephalus (HC)**

Hydrocephalus is a clinical entity in which a disturbance of the cerebrospinal fluid (CSF) circulation causes the accumulation of intraventricular CSF, resulting in progressive ventricular dilatation (Mori et al. 1995). Hydrocephalus is characterised by increased intracranial pressure, increased CSF volume and dilatation of the CSF space. The size of the ventricles forms the basis of the diagnosis; however, enlarged ventricles may also be due to atrophy of the brain, which must be distinguished from HC. Distortion and compression of structures (the grey and white matter) as well as blood vessels may be the result of HC, and this affects the physiology, the biochemistry and, the ultra structure of the brain (Persson 2007). For the purpose of this thesis HC is defined as a head circumference of >2 standard deviation scores (SDSs) above the body length SDS, combined with enlarged ventricles. Figure 1 shows the ventricles and the CSF flow.

There are number of different aetiologies to HC, children with myelomeningocele (MMC) have known malformations located in the spinal cord, brain stem, and/or posterior fossa regions caused by a developmental defect of the neural tube. Approximately one-third of children with HC have MMC (Persson et al. 2005). Hy-
Background
drocephalus developing before one year of age with other aetiologies than MMC will be referred to as infantile hydrocephalus (IH). Children with IH often have parenchyma defects due to haemorrhage and/or infarctions in a variety of brain regions. The most common causes of IH are intraventricular haemorrhage due to premature birth (post-haemorrhage hydrocephalus (PHHC)), malformations such as aqueduct stenosis, arachnoidal cysts, Dandy Walker syndrome, lipoma, tumours and infections (Hoyt 2005). Since children with aetiologies other than MMC are known to have a poorer outcome than children with MMC (Heinsbergen 2002), we decided to divide the group of children in the present study into these two aetiological groups i.e. children with MMC and those with other aetiologies (IH).

Hydrocephalus may be categorised in a five-step classification system defined by age at development of HC, the “Perspective Classification of Congenital Hydrocephalus”. Stages IV to V are the main periods for developing infantile hydrocephalus (Oi et al. 1994).
I: Weeks 8-21. The main process is neuronal proliferation.
II: Weeks 22-31. The main processes are cell differentiation and migration.
III: Weeks 32-40. The main process is cell differentiation.
IV: Weeks 0-4 postnatally. The main process is dendritic maturation.
V: Weeks 5-50 postnatally. The main process is myelination.
Hydrocephalus can be communicating or non-communicating. In the communicating type, the blockage is outside the ventricular system, and fluid within the ventricular system communicates with the spinal subarachnoid space and basal cisterns. In non-communicating HC, the ventricular fluid does not communicate with CSF in the spinal subarachnoid spaces or in the basal cisterns.

**Treatment of Hydrocephalus**

The use of CSF shunts in children with HC, to lower the intracranial pressure (ICP) became common around 1960. Endoscopic third ventriculostomy is an alternative to shunt treatment, which has been used more frequently during the past decade (Persson et al. 2006, accepted for publication). In the present study, the majority of children have CSF shunts (Fig 2).

![Diagrammatic drawing of child with hydrocephalus and cerebrospinal fluid shunt.](image)

Figure 2. *Diagrammatic drawing of child with hydrocephalus and cerebrospinal fluid shunt.*

**Ventricular width in children with hydrocephalus**

When inserting a shunt, the pressure decreases, and the ventricular width will be reduced, and in some of the cases, the ventricles may collapse. A number of different neuroradiological examinations can be used for diagnosis and for
follow-up of HC. In small children with open fontanel, ultrasonography is the screening procedure, while in older children with closed fontanel, and in follow-up after shunting, computed tomography (CT) was the most common investigative method during the study period. However, in general magnetic resonance imaging (MRI) is preferred, as it is the most sensitive of the available methods for studying parenchyma abnormalities.

**Associated neurological impairments in children with hydrocephalus**

Cerebral palsy (CP) is a group of disorders of the development of movement and posture. The motor disorders are often accompanied by disturbance of sensation, cognition, perception, and/or behaviour, and/or by a seizure disorder (Bax et al. 2005). In previous Swedish studies in children with HC, CP has been reported in 26-47% of the children. Prematurely born children had a higher rate than those born at full term (Fernell et al. 1988a; Fernell et al. 1988b; Hagberg et al. 1988), reflecting a higher frequency of parenchyma lesions in the preterm born children.

Epilepsy (EP), is defined as two or more unprovoked epileptic seizures. There are different types of seizures; partial or general, and with or without loss of consciousness (Lindberg and Lagercrantz 1999). Most of the seizures in HC are of the partial type. The presence of neuroradiological abnormalities in the brain parenchyma, and the aetiology other than MMC, are risk factors for developing epilepsy (Persson 2007). Epilepsy has been reported in 22% of full-term HC children and 33% of premature HC children (Fernell et al. 1988a; Fernell et al. 1988b).

“Learning disability” is defined as an intelligence quotient (IQ) score of <70, according to both the International Statistical Classification of Diseases and Related Health Problems, 10th revision (ICD-10), and the American Psychiatric Association’s Diagnostical and Statistical Manual of Mental Disorders (DSM-IV). In children with HC, as a group the intellectual performance is affected, the overall IQ is low-average or below (IQ 70-84) (Lindquist et al. 2005) and the verbal intelligence is often better preserved than the non-verbal. Behavioural problems and autism spectrum disorders are also common (Heinsbergen 2002; Lindquist et al. 2006).

**Ophthalmological findings associated with hydrocephalus**

Obstructed CSF can create a multitude of neuro-ophthalmic problems. Problems such as defect ocular motility, visual field defects, optic atrophy, and loss of visual acuity (VA) have been reported (Harcourt 1968; Clements and Kaushal 1970; Rothstein et al. 1973; France 1975; Houtman et al. 1981; Holman and Merritt 1986; Mankinen-Heikkinen 1987; Lennerstrand et al. 1990; Gaston 1991; Biglan
1995; Altintas et al. 2005). Strabismus, mostly esotropia, A- and V-syndromes, oblique muscle dysfunction, and lateral rectus weakness are other common findings among children with HC (Gaston 1985; Tamura and Hoyt 1987; Biglan 1990; Lennerstrand et al. 1990; Gaston 1991). In addition, nystagmus, gaze palsies, dorsal mid-brain syndrome, and acutely occurring strabismus can coincide with HC (Corbett 1986; Hoyt and Good 1995). Esotropia and visual loss have been described in association with slit-ventricle syndrome (Nguyen et al. 2002). Convergent squints, lateral rectus palsy, divergence palsies, and other ocular motility disorders can also be useful indices of shunt dysfunction and raised ICP (Rabinowicz 1976; Gaston 1991; Altintas et al. 2005). Shunt treatment may improve several of these dysfunctions; however, these disorders are not always completely resolved, and revisions of the shunt may be a risk factor for both strabismus and amblyopia (Altintas et al. 2005).

Definitions and classifications

Heterotropia is a manifest ocular misalignment (manifest strabismus), which cannot be corrected by fusional vergence (Leigh and Zee 1999; Ansons and Davis 2001; von Noorden and Campos 2002; Rowe 2004), it results in a deviation of the non-fixating eye. Classification of heterotropia relies on to the direction of the deviation, the accommodative convergence/accommodation (AC/A), the time of onset, the combination with muscle paralysis, the pattern of the deviation, and whether it is a primary, secondary, or intermittent deviation (von Noorden and Campos 2002). The occurrence of manifest strabismus in the general population has been reported to be 3-8% (Turacli et al. 1995; Gupta et al. 2000; Kvarnström, et al. 2001; Ohlsson et al. 2001).

Heterophoria is a latent deviation (latent strabismus) where the eyes are kept in alignment by the fusional mechanism during binocular viewing (Leigh and Zee 1999; Ansons and Davis 2001; von Noorden and Campos 2002; Rowe 2004). Classification of heterophoria is by the direction of the deviation, the distance where the deviation occurs, and by the distance/near relationship (Ansons and Davis 2001; Rowe 2004).

Orthophoria/orthotropia is present when there is no apparent deviation of the visual axes, but the eyes are dissociated and fusion is suspended (von Noorden and Campos 2002; Rowe 2004).

Children may present with an abnormal head posture to compensate for ocular muscular palsy, mechanical restrictions of ocular movements, nystagmus, defective eye movements (saccades or pursuit movements) or strabismus or refractive errors (astigmatism). The cause of abnormal head posture can also be non-ocular, including contracture of the sterno-cleido-mastoid muscle in the neck. The components of head posture are described as head tilt, face turn and chin elevation or depression (Rowe 2004).
**Background**

_Stereoaucuity_ is the quantitative measurement of the degree of disparity of two images fused to one. It is measured in seconds of arc ("'). Measurement of stereoaucuity is mainly based on two principles; the random dot and linear polarisation (von Noorden and Campos 2002). The random dot test employs images formed by dots, which are displaced in relation to each other. Two disparate pictures are extracted from the dots, and one is present to each eye; stereoaucuity is deemed to occur if the two images are fused. Common tests are the TNO test and the Lang test (von Noorden and Campos 2002; Rowe 2004). In the linear polarisation test, pictures with two Polaroid surfaces are presented to the patient, who wears glasses with horizontal and vertical filters. Two disparate images are presented, one to each eye, and stereoaucuity is deemed to occur if the images are fused. Common tests are the Titmus/Wirt stereo test and Randot test (von Noorden and Campos 2002; Rowe 2004).

_Accommodative convergence to accommodation ratio_

The relationship between accommodation (A) of the lens in dioptres (D) and accommodation-linked convergence (AC) in prism dioptres (PD) can be expressed as a ratio, the AC/A ratio (Leigh and Zee 1999; Rowe 2004). Measurements of AC/A is recommended in patients with convergence excess esotropia and in patients with intermittent exotropia. The findings may influence both treatment and outcome (Ansons and Davis 2001). The normal range of AC/A is considered to be between 3/1 and 5/1 (Leigh and Zee 1999; Fawcett et al. 2000; von Noorden and Campos 2002; Fawcett and Birch 2003); however, different studies use different methods, and most studies include both children and adults. There are a number of different methods of calculating the ratio. Two common clinical methods are the gradient method, i.e. (near deviation in prism dioptres (PD) −near deviation in PD with addition of +3 D lens)/3 D, and the heterophoria method, i.e. interpupillary distance in cm + (near deviation in PD −distance deviation in PD)/3 D (Ansons and Davis 2001; von Noorden and Campos 2002; Rowe 2004). The size of the deviation is measured with the alternating prism and cover test, and with habitual glasses or best correction.

_Near point of convergence_

Near point of convergence (NPC) is the point closest to the nose where fusion can maintain fixation with both eyes and is measured in cm. In the present study the Royal Air Force (RAF) ruler is used (von Noorden and Campos 2002; Rowe 2004).

_Versions_ are conjugate movements of both eyes in the same direction by equal amounts (Leigh and Zee 1999; Ansons and Davis 2001; von Noorden and Campos 2002; Rowe 2004).

_Ductions_ are movements of one eye (Leigh and Zee 1999; von Noorden and Campos 2002; Rowe 2004).
**Nystagmus** may be manifest or latent (i.e. occurring only when one eye is occluded) and is defined as involuntary oscillatory movements initiated by slow phases. A description of nystagmus usually includes the waveform, the direction, the frequency and the amplitude (Dell’Osso and Daroff 2005; Jacobs and Dell’Osso 2004). The direction is described by the direction of the quick phases (Leigh and Zee 1999).

**Intrusive saccades** are occasional inappropriate saccadic movements that may intrude on steady fixation (Herishanu and Sharpe 1981; Abadi 2004). In the present study, intruding saccades were defined as rapid eye position changes of more than 3º amplitude in the recordings. For an example of saccades see figure 5.

**Drifts** are involuntary slow eye movements taking the fixation away from the fixation target. In the present study, drifts were defined as slow changes of eye position with amplitude of more than 3º. For an example of drifts see figure 5.

A **blink** is a brain stem reflex, which activates the orbicularis oculi and the levator palpebrae muscle. The normal spontaneous blink rate is two blinks per minute in early infancy, with an increase during childhood up to age 14 or 15 (Zametkin et al. 1979); adult frequency has been described as 20 blinks per minute (Leigh and Zee 1999). In the present study, blinks were defined as large rapid changes of eye position, back and fourth in all four channels (horizontal and vertical in both eyes) without intersaccadic interval. For an example of blinks see figure 5.
Aims

To investigate a sample of children aged 4-15 years, from four different schools and pre-schools, with a well-defined comprehensive test battery of clinically often used orthoptic tests, in order to obtain results for use as comparison in clinical studies of different patient groups (paper I).

To investigate a population-based group of children with surgically treated HC, with a well defined comprehensive test battery of clinically often used orthoptic tests, to determine the frequency of orthoptic abnormalities, and to relate the results to those of an age- and sex-matched comparison group (paper II).

To correlate the frequency of orthoptic abnormalities in children with HC with the time of onset and the aetiology of HC, with associated neurological impairments, and with the ventricular width at neuroimaging (paper II).

To determine visual fixation behaviour in a sample of children aged 4-15 years, from four different schools and pre-schools, in order to obtain results for use as comparison in studies of different patient groups (paper III).

To determine visual fixation behaviour, in a population-based group of children with surgically treated HC and to relate the results to those of an age- and sex-matched comparison group (paper IV).

To correlate visual fixation behaviour in children with HC with the time of onset, and the aetiology of HC, with associated neurological impairments, and with the ventricular width at neuroimaging (paper IV).
Subjects and Methods

Subjects
The papers are based on two groups of children, one group without major illnesses (the comparison group) and one population-based group with surgically treated HC (Fig 3). Both groups were investigated with the same battery of tests.

Paper I: The comparison group consisted of 143 children (67 girls) aged 4–15 years (median age 9.4 years) from four different schools and preschools in the area of Göteborg, Sweden. All children except five (3.6%) were born at a gestational age ≥37 weeks, and the birth weight was in mean 3 158g (range 1 000-5 470g). Information regarding socio-economic background was obtained from the parents; these data were compared with data from Västra Götaland, other communities in Sweden, and the total Swedish population ((SCB) - Statistiska Centralbyrån). Neither living conditions nor socio-economic group differed from either those in other communities in Sweden or the Swedish standard.

Paper II: The subjects in paper II comprised a population-based group of 75 children aged 7–12 years (median age 9.7 years) (34 girls), with overt HC at birth or developing during the first year of life, and requiring surgical treatment for HC. Subjects were born between April 1989 and April 1993 in a well-defined area in southwestern Sweden (i.e. the counties of Västra Götaland, Halland, and Värmland) and surgical treatment was performed at the Queen Silvia Children’s Hospital, Göteborg, Sweden. Nineteen children (25.3%) were born at < 37 weeks of gestation and the birth weight was in mean 3 585g (range 1 540-4 970g). The total eligible group contained 92 children; 15 of these declined to participate, and two children who had moved away from the region could not be contacted. The results were compared with those from an age-and sex-matched comparison group, selected from study I (n=140).

Paper III: The study group consisted of 135 of the 143 children from paper I. The children (63 girls) were aged 4–15 years (median age 9.8 years). Eight children were rejected from the material, three children because of insufficient cooperation, and five children for technical problems with the recording procedure.

Paper IV: Thirty-five of the 52 children with HC, who were investigated at the Queen Silvia Children’s Hospital, where the eye-tracker was located, were included in the study group. Ten children could not participate in the recording procedure (all with VA<0.2, European decimal; >0.7 logMar), and a further seven excluded due to problems with the eye-tracker. The results were compared with those from an age-and sex-matched comparison group, selected from study I (n=117).
Orthoptic examinations in paper I and II

All children in both groups were examined using an appropriate combination of the following tests, table 1.

Comments to the tests

Test 1: The Hirschberg reflex test was used to screen for heterotropia and to estimate the size of a deviation by noting any displacement of the corneal light reflection from a pen light (Ansons and Davis 2001).

Test 2a: Heterotropia was defined with the monolateral cover test (CT) (Ansons and Davis 2001) while fixating targets at 3 and/or 0.33 m distance. If a child who was unable to take the CT demonstrated a deviation of >10 deg in Hirschberg’s test, the child was considered to have heterotropia. The CT was performed with the head held straight and a black paddle occluder as a cover. Detailed fixation objects were used as targets. At distance, the CT was performed in four additional positions of gaze (i.e. the child was ordered to look in the different directions). For distance, fixation targets were at 1 m (18.5 deg) up, down, right, and left, respectively, of the straight-ahead target.

All children were examined without optical correction as well as with their habitual glasses or with best correction.
### Table 1: Type of tests used and outcomes.

<table>
<thead>
<tr>
<th>Test No.</th>
<th>Type of test</th>
<th>Outcome</th>
<th>Value/result</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Hirschberg reflex test</td>
<td>Orthotropia; heterotropia</td>
<td>Symmetrical or degree of decentration</td>
</tr>
<tr>
<td>2a</td>
<td>Cover test (monolateral)(CT)</td>
<td>Orthotropia; heterotropia</td>
<td>ET, XT, hypertropia or hypotropia</td>
</tr>
<tr>
<td>2b</td>
<td>Alternating cover test (aCT)</td>
<td>Orthophoria; heterophoria</td>
<td>Eph, Xph, hyperphoria or hypophoria</td>
</tr>
<tr>
<td>2c</td>
<td>Alternate prism and cover test (pCT)</td>
<td>Size of the angle of deviation</td>
<td>Prism dioptre (PD)</td>
</tr>
<tr>
<td>3</td>
<td>Inspection</td>
<td>Occurrence of nystagmus and/or abnormal head posture</td>
<td>Nystagmus/ no nystagmus Type of abnormal head posture</td>
</tr>
<tr>
<td>4</td>
<td>TNO, Titmus tests and Lang 1</td>
<td>Stereo acuity</td>
<td>Seconds of arc (&quot;)</td>
</tr>
<tr>
<td>5a</td>
<td>To fixate and follow a penlight in an upward, downward, right and left gaze</td>
<td>Ocular motility; versions</td>
<td>Descriptive</td>
</tr>
<tr>
<td>5b</td>
<td>To fixate and follow a penlight in the main directions of action of each muscle, with the non-tested eye occluded</td>
<td>Ocular motility; ductions</td>
<td>Nine-point scoring system, where 0 = normal, scores of −1 to −4 indicate underaction and +1 to +4 indicate overaction</td>
</tr>
<tr>
<td>6</td>
<td>RAF ruler</td>
<td>Near point of convergence (NPC)</td>
<td>The mean of three measurements recorded in cm</td>
</tr>
<tr>
<td>7a</td>
<td>aCT at 0.33 m with best correction, with and without +3D lens</td>
<td>Accommodative convergence to accommodation ratio (AC/A); gradient method (near deviation in PD−near deviation in PD with addition of +3 D lens)/3 D</td>
<td>Accommodative convergence in PD per dioptre (D) of accommodation</td>
</tr>
<tr>
<td>7b</td>
<td>Interpupillary distance, aCT at 3 m and 0.33 m with best correction</td>
<td>AC/A; heterophoria method; interpupillary distance* in cm + (near deviation in PD−distance deviation in PD)/3 D</td>
<td>Accommodative convergence in PD per D of accommodation</td>
</tr>
</tbody>
</table>

* The interpupillary distance was measured using an Autorefractometer (Topcon A6300) simultaneously with the measurement of the refraction.
Subjects and methods

**Test 2b:** If no heterotropia was found an alternate cover test (aCT) (Ansons and Davis 2001) was performed to investigate heterophoria. We used the aCT because it dissociates the two eyes more effectively than does the uncover test. Fixation distances and gaze positions were used in the same way as for the CT.

**Test 2c:** A prism and cover test (pCT) (Rainey et al. 1998) was used in case of any deviation to quantify the size of the deviation. The pCT is known to achieve good quantitative data of the detected deviation, as it is a reliable and repeatable test with good inter-examiner reliability (Ansons and Davis 2001). The pCT also gives accurate results to within 2 PD (Ansons and Davis 2001; von Noorden and Campos 2002). Heterophoria was defined as a deviation of $\geq 2$ PD found with the pCT. At 0.33 m, the pCT was also performed with best correction and with addition of a $+3.0$ D lens, for calculation of the AC/A.

**Test 3:** Anomalous head postures and the presence of nystagmus were noted during the Hirschberg test when the child was fixating the penlight. In addition, abnormal head posture was also looked for during binocular vision testing.

**Test 4:** Two clinically widely used stereo acuity tests were employed; TNO random-dot test and, if the child was unable to identify the TNO figures, the Titmus stereo test (Marsh et al. 1980; Ohlsson et al. 2001; von Noorden and Campos 2002). For the TNO test, the stereo acuity was defined as the smallest level of disparity for which both test figures were correctly identified. In Paper II the Lang 1 test was added, as one child did not wish to use the spectacles needed for the TNO and Titmus tests. Children with heterotropia considered clinically large were not tested with any of the stereo tests. In addition, it should be pointed out that the TNO test and Titmus tests are not strictly comparable as the TNO test is a random-dot test and the Titmus test is a Polaroid contour test (von Noorden and Campos 2002).

All children were examined with best optical correction or with their habitual glasses.

**Test 5 a and 5b:** For investigation of versions, a penlight was used for the child to fixate and follow with both eyes together upward, downward, right and left gaze.

For investigating ductions, again a pen light was used which the child was asked to fixate and follow in the principal direction of action of the extra ocular muscles (EOMs). If there were uncertainties regarding under action- or overaction of single muscles, an aCT was performed.

**Test 6:** For NPC evaluation, the RAF ruler (Ansons and Davis 2001) was used. This is an objective method with a high reproducibility, although the near limit is only 6 cm (which makes it impossible to measure closer convergence).
Subjects and methods

Test 7: The pCT was used to obtain values of the deviation for calculating the AC/A with the gradient and heterophoria methods, both of which are often used in clinical practice (Ansons and Davis 2001; von Noorden and Campos 2002).

Visual fixation recordings in paper III and IV

Binocular eye positions (simultaneous horizontal and vertical) were recorded using the Orbit infrared (IR) system (IOTA Inc., Timrå, Sweden) (Ygge et al. 1999) (Bolzani et al. 1998). In this IR device, pulsed infrared light, emitted inside a pair of goggles is reflected against the ocular surface and detected by eight detectors also harboured within the goggles. Eye position signals are conducted via a sound card to a computer where they are recorded. The maximum temporal resolution of our system is 100 Hz, the spatial resolution under optimal conditions is 0.01°, and the linearity is 10% (manufacturer’s data).

During recording, the child sat comfortably in a chair, with his/her head restrained, using a chin and a forehead rest (Fig 4a), 53 cm from a computer screen (Fig 4b); the position of the goggles was secured with a strap extending the back of the head. The child’s head was adjusted so that the eyes were level with the centre of the computer screen. No eyeglasses were used during the recordings, as they interfered with the functioning of the recorder and the uncorrected visual acuity was adequate for seeing the stimuli in all cases. Ambient illumination was mesopic. The placement of the IR goggles is important for adequate recordings and to avoid crosstalk between the horizontal and vertical channels. Considerable crosstalk in the recordings from the pre-test paradigm led to readjustment of the goggles and this procedure was repeated until a satisfactory result was obtained. After the goggles were in a satisfactory position the position of the goggles was not changed. A monocular, two point (horizontal/vertical; 10 deg) calibration paradigm was performed before recording the fixation; the calibration duration was 25 s. After the calibration paradigm, a stable fixation stimulus (a yellow dot subtending a visual angle of 0.3°) was displayed for 20 s centrally on the otherwise black computer screen: the child was asked and encouraged to fixate on the stimulus and keep the eyes as steady as possible.

Visual fixation analysis in paper III and IV

Off-line data calibration of all four channels of recordings (right and left eye, horizontal and vertical) was performed using the JR program (Bolzani et al. 1998; Ygge et al. 1999): the calibrated data was then transferred to a computer program (Origin 7.0; Microcal Inc., Northampton, MA, USA) for further analysis and plotting.

During the total 20s of fixation recording, the longest continuous fixation time without interruption by blinks, saccades (defined as the rapid change of eye position of more than 3 ° amplitude), or drifts (defined as the slow change of eye posi-
Figure 4a. Child wearing recording goggles, and are restrained in the chin and forehead rest.

Figure 4b. Child sitting comfortably 53 cm from the computer screen during the recording procedure.
tion of more than 3 °) was noted. Blinks were defined as the large rapid change of eye position back and forth in all channels, without an intersaccadic interval (the normal characteristics of blinks with the present system). During the 10s with the least intruding movements, intruding saccades, drifts, and blinks were counted (Fig 5). When analysing nystagmus the calibration period was included, as the calibration was performed with one eye occluded and thus latent nystagmus may be found.

During each recording of eye position the X and Y coordinates for each fixation point during the entire recording was saved on a computer hard drive for subsequent off-line analyse. The mean of the X and Y coordinates in each subject were calculated to be used as “centre of gravity” (Källmark and Ygge 2005).

Methods used in the present multidisciplinary study

Visual acuity

Visual acuity was measured with the KM letter-matching chart (Moutakis et al. 2004). All children in the comparison group were evaluated with the KM test,
as were all but ten of the children in the HC group; one child could not fixate the pen light, six could fixate the pen light and the three others were tested with the HVOT, LH, or Cardiff tests.

Visual impairment (VI) was defined as visual acuity <0.3 (>0.5 logMar; World Health Organization 1977).

**Refraction**

Refraction under cycloplegia, was performed with an autorefractometer (Topcon A6 300) after installation of a mixture of cyclopentolate (0.85%) and phenylephrine (1.5%). Simultaneously with the refraction, the interpupillary distance was measured. Hyperopia was defined as a spherical equivalent (SE) of >+2 D, myopia as SE >-0.5 D, and astigmatism as >0.75 D.

**Associated neurological impairments (CP, EP, and learning disabilities)**

Information about CP and EP was obtained from records from the paediatric and neurological departments. Information about cognition was gathered from clinical records, and from neuropsychological test in the form of the Wechsler Intelligence Scale for Children-III (WISC-III), developed for ages six to sixteen years, and the Wechsler Preschool and Primary Scale of Intelligence-Revised (WIPPSI-R), for children aged three to seven years. The Griffiths Developmental Scales were used for children with a developmental aged below three years. Learning disabilities was defined as a full-scale IQ < 70. Results presented elsewhere (Lindquist 2007).

**Neuroradiological examination (ventricular width)**

The most recent postoperative neuroradiological examination record was evaluated by a neuroradiologist. These records were based on MRI in 13 children, CT scan in 58 children, and ultrasound in 1 child. Information was missing in three children.

**Ophthalmological characteristics**

A summery of the ophthalmological findings in children with HC and in the age- and sex-matched comparison group, are present in *table 2*.

**Paediatric characteristics of children with hydrocephalus**

Information about time of onset and the aetiology of HC, and associated impairments were obtained from medical records. In study II, overt HC at birth was seen in 30 children and 45 children developed HC during first year of life, *table*
33

Table 2. Ophthalmological findings (paper II).

<table>
<thead>
<tr>
<th>Ophthalmological findings</th>
<th>Comparison group (n=140) n (%)</th>
<th>Children with hydrocephalus (n=75) n (%)</th>
<th>p-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Binocular visual acuity</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>European decimal (logMar)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>&lt;0.3 (&gt;0.5)</td>
<td>None</td>
<td>11/73* (15)</td>
<td>&lt;0.01</td>
</tr>
<tr>
<td>0.3-0.8 (0.5-0.1)</td>
<td>13 (9)</td>
<td>25/73* (34)</td>
<td>&lt;0.01</td>
</tr>
<tr>
<td>&gt;0.8 (&lt;0.1)</td>
<td>127 (91)</td>
<td>37/73* (51)</td>
<td>&lt;0.01</td>
</tr>
<tr>
<td>Refraction</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>myopia &gt;0.5 D SE</td>
<td>7 (5)</td>
<td>7/70* (10)</td>
<td>n.s.</td>
</tr>
<tr>
<td>hyperopia &gt;2.0 D SE</td>
<td>13 (9)</td>
<td>32/70* (46)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>astigmatism &gt;0.75 D</td>
<td>31 (22)</td>
<td>35/70* (50)</td>
<td>&lt;0.001</td>
</tr>
</tbody>
</table>

*Some children did not participate.

3 shows the time of onset in relation to the aetiology of HC. Thirty seven children in study II had additional neurological impairments to the HC, such as CP and/or EP and/or learning disabilities; table 4 shows the proportion of associated neurological impairments. A summary of the paediatric results are presented elsewhere (Persson 2007).

Cognitive outcome

A third of the children had learning disabilities (IQ<70), and the mean IQ of the whole group was 75 and as a group, children with HC had better verbal IQ

Table 3. Time of onset of HC in relation to aetiology of HC (paper II).

<table>
<thead>
<tr>
<th>Aetiology</th>
<th>Overt hydrocephalus at birth (n=30)</th>
<th>Hydrocephalus developing during 1st year of life (n=45)</th>
</tr>
</thead>
<tbody>
<tr>
<td>MMC (n=28)</td>
<td>10</td>
<td>18</td>
</tr>
<tr>
<td>Other CNS malformations* (n=20)</td>
<td>15</td>
<td>5</td>
</tr>
<tr>
<td>Post-haemorrhagic hydrocephalus (n=18)</td>
<td>4</td>
<td>14</td>
</tr>
<tr>
<td>Infection (n=4)</td>
<td>0</td>
<td>4</td>
</tr>
<tr>
<td>Unknown (n=5)</td>
<td>1</td>
<td>4</td>
</tr>
</tbody>
</table>

*These findings include aqueduct stenosis (n=2), Dandy-Walker syndrome (n=2), Arnold Chiari (n=2) tumours, etc.
CNS = central nervous system.
than performance IQ (Lindquist et al. 2005). Additional impairments, such as cerebral palsy or epilepsy, were associated with an increased risk of cognitive or behavioural problems. Children with MMC were clustered around IQ 70-85, while children without MMC had a wider IQ range; however, the median IQ did not differ between the two groups (Lindquist et al. 2005).

**Ventricular width**

Normal ventricle width was found in 28 children, enlarged in 29, slit in 15, and in three children information was missing.

**Statistical analysis**

Means, medians, and ranges were calculated for descriptive purposes. As normal distribution could not be assumed, non-parametric tests were used. For comparison between two groups, Wilcoxon-Mann-Whitney’s test was used for ordered and continuous variables; and Fisher’s exact test for dichotomous variables. For comparison between three groups, the Mantel-Haenszel Chi2 test was used. The correlation analysis in paper III was performed with the Spearman correlation test. A significance level of 5% was applied.

The age- and sex-matched comparison groups in papers II and IV were selected individual by individual by minimizing the maximal t-values between the group of children with HC and the study cohort of Swedish children aged 4-15 years (n=143) from Paper I, over the variables of age and sex.
Results

Paper I
Among Swedish children aged 4-15 years (Fig 3) five children (3.5%) had heterotropia, four had esotropia and one had exotropia. Heterophoria at near and/or distant fixation was found in 37 (26%) and orthophoria at both near and distant fixation was noted in 101 children (71%). One child with esotropia had abnormal ocular motility, a slight overaction of both inferior oblique muscles, while 97% had stereo acuity of 60" or better. NPC was ≤6 cm in 97% of the children. In the whole group, the median AC/A was 5.6/1 prism dioptres/dioptre (PD/D) calculated with the heterophoria method and 1.3/1 PD/D calculated with the gradient method. No anomalous head postures or nystagmus were observed and all children had normal versions (table 5).

Paper II
Children with early surgically treated HC differed significantly in all orthoptic variables in comparison with the age- and sex-matched comparison group (n=140) selected from paper I (Fig 3). Heterotropia was more common among

Table 5. Orthoptic findings (paper II).

<table>
<thead>
<tr>
<th>Orthoptic findings</th>
<th>Comparison group (n=140) n (%)</th>
<th>Children with hydrocephalus (n=75) n (%)</th>
<th>p-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Strabismus</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Heterotropia</td>
<td>5/140 (4)</td>
<td>51/74* (69)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>esotropia (ET)</td>
<td>4/140 (3)</td>
<td>26/74* (35)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>exotropia (XT)</td>
<td>1/140 (1)</td>
<td>21/74* (28)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>varying ET/XT</td>
<td>None</td>
<td>4/74* (5)</td>
<td>0.014</td>
</tr>
<tr>
<td>Heterophoria (Xph and Eph)</td>
<td>37/140 (26)</td>
<td>10/74* (13)</td>
<td>0.045</td>
</tr>
<tr>
<td>Orthophoria</td>
<td>101/140 (72)</td>
<td>13/74* (18)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Abnormal head posture</td>
<td>None</td>
<td>31/75 (41)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Nystagmus</td>
<td>None</td>
<td>33/75 (44)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Stereo acuity</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>60&quot; or better</td>
<td>135/140 (96)</td>
<td>22/67* (34)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>120&quot;-480&quot;</td>
<td>None</td>
<td>5/67 (7)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Negative</td>
<td>4/140 (3)</td>
<td>40/67 (60)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Ocular motility defects</td>
<td>1/140 (1)</td>
<td>41/68* (60)</td>
<td>&lt;0.001</td>
</tr>
</tbody>
</table>

*Not all children participated.
HC children (p<0.001), and stereo acuity of 60” or better was found in 34%, compared to 96% in the comparison group (p<0.001). Abnormal ocular motility (i.e. ductions or versions), abnormal head posture, and nystagmus were also found more frequently among HC children than among the comparison group (p<0.001) (table 5). Children with overt HC at birth had significantly higher frequency of heterotropia (p=0.0006), abnormal head posture (p=0.02), and motility defects (p=0.003), compared with children with HC developing during the first year of life. The aetiology of HC and the ventricular width seems to be of minor importance compared with the age of onset of HC regarding the risk for orthoptic abnormalities.

**Paper III**

Among Swedish children aged 4-15 years (Fig 3), we found that fixation behaviour and fixation stability were age-dependent. The group was divided in four age groups (4-6; 7-9; 10-12; and 13-15 years) and an increase with increasing age, was seen in both the fixation time (p=0.02) (Fig 6), and the fixation density around centre of gravity of fixation (p<0.01). Intruding saccades decreased with increasing age (p<0.01) (Fig 7), while blinks, and drifts showed no age correlation. There were no significant differences with regard to gender or laterality in any of the investigated variables. No nystagmus was observed.

A more centred fixation was seen during age, note the difference between a four year old child (Fig 8a) and a 14 year old child (Fig 8b).

![Figure 6. Box and whisker plot showing the longest continuous fixation time without intruding saccades, drifts or blinks during the full 20 seconds of recordings in different age groups. The box showing one standard deviation (SD), whiskers showing 95% and 5 % interval, x showing the 99% and 1% interval and □ shows the mean value.](image-url)
Figure 7. Box and whisker plot showing the number of intruding saccades during 10 seconds of recordings, in different age groups. The box showing one standard deviation (SD), whiskers showing 95% and 5% interval, x showing the 99% and 1% interval and □ shows the mean value.

Figure 8a, 8b. Figures showing XY-plots (horizontal and vertical eye position) during 10 seconds of recording of two different children in the comparison group (4 and 14 years).
**Paper IV**

Children with HC had shorter fixation time (p<0.01), had higher frequency of drifts (p<0.01) and more nystagmus (p<0.01) than the age- and sex- matched comparison group (n=117), selected from paper III (Fig 3). Children with MMC did not differ from the comparison group in any of the investigated variables compared, while children with aetiologies other than MMC had shorter fixation time (p=0.02) (Fig 9) and displayed more drifts (p=0.048) (Fig 10). A lower frequency of blinks was seen in children with overt HC at birth compared to children developing HC during the first year of life (p=0.03), and shorter fixation time was seen in children with epilepsy compared to children without epilepsy (p=0.046), and children with CP displayed more nystagmus compared to children without CP (p<0.01).

![Box and whisker plot showing the longest continuous fixation time without intruding saccades, drifts or blinks, during the full 20 seconds of recordings, in children with MMC (n=14), in children with aetiologies other than MMC (n=21), and in the comparison group (n=117). The box showing one standard deviation (SD), whiskers showing 95% and 5%, x= 99% and 1% and □ shows the mean value.](image)

**Orthoptic findings and visual fixation, in relation to aetiology, and onset of HC, and associated impairments**

The frequency of orthoptic abnormalities, for example heterotropia are equal in the different aetiological groups, while fixation abnormalities and associated impairments (except learning disabilities) are more frequent among children with aetiologies other than MMC (table 6).

Children with overt HC at birth had more orthoptic abnormalities than children who developed HC during the first year of life, while fixation behaviour and associated impairments (except learning disabilities) did not differ (table 7).
Table 6. Orthoptic findings, visual fixation, and associated impairments in relation to aetiology of HC.

<table>
<thead>
<tr>
<th></th>
<th>IH (n=47)</th>
<th>MMC (n=28)</th>
<th>p-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Paper II</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Heterotropia</td>
<td>34 (72)</td>
<td>17 (61)</td>
<td>n.s</td>
</tr>
<tr>
<td>Nystagmus</td>
<td>20 (42)</td>
<td>13 (46)</td>
<td>n.s</td>
</tr>
<tr>
<td>Paper IV</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Fixation time sec (mean)</td>
<td>2.6</td>
<td>5.1</td>
<td>&lt;0.05</td>
</tr>
<tr>
<td>Drifts n (mean)</td>
<td>1.3</td>
<td>0.5</td>
<td>&lt;0.05</td>
</tr>
<tr>
<td>Visual impairment</td>
<td>11 (23)</td>
<td>0</td>
<td>&lt;0.01</td>
</tr>
<tr>
<td>CP</td>
<td>13 (28)</td>
<td>0</td>
<td>&lt;0.01</td>
</tr>
<tr>
<td>EP</td>
<td>17 (36)</td>
<td>2 (7)</td>
<td>&lt;0.01</td>
</tr>
<tr>
<td>Learning disabilities</td>
<td>20 (34)</td>
<td>7 (25)</td>
<td>n.s</td>
</tr>
</tbody>
</table>

Heterotropia occurred more frequent in children with associated impairments (except in children with CP), than in those without impairments. In paper IV the subgroups of children with different associated impairments were few, although a shorter fixation time could be seen in children with EP, compared to those without EP (table 8).

No association with the ventricular width was found in any of the investigated variables.
Table 7. Orthoptic findings, visual fixation, and associated impairments in relation to time of onset of HC.

<table>
<thead>
<tr>
<th></th>
<th>HC overt at birth n(%)</th>
<th>HC dev. during first year of life n(%)</th>
<th>p-value</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Paper II (n=70)</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Heterotropia</td>
<td>29 (97)</td>
<td>22 (49)</td>
<td>&lt;0.01</td>
</tr>
<tr>
<td>Nystagmus</td>
<td>16 (53)</td>
<td>17 (38)</td>
<td>n.s</td>
</tr>
<tr>
<td><strong>Paper IV (n=12)</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Fixation time sec (mean)</td>
<td>3.8</td>
<td>3.6</td>
<td>n.s</td>
</tr>
<tr>
<td>Drifts n (mean)</td>
<td>0.9</td>
<td>0.9</td>
<td>n.s</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Visual Impairment</th>
<th>Paper II (n=70)</th>
<th>Paper IV (n=35)</th>
</tr>
</thead>
<tbody>
<tr>
<td>CP</td>
<td>4 (13)</td>
<td>1 (2)</td>
</tr>
<tr>
<td>EP</td>
<td>11 (37)</td>
<td>6 (16)</td>
</tr>
<tr>
<td>Learning disabilities</td>
<td>13 (43)</td>
<td>10 (31)</td>
</tr>
</tbody>
</table>

Table 8. Orthoptic findings and visual fixation in relation to associated impairments.

<table>
<thead>
<tr>
<th></th>
<th>Visual Impairment</th>
<th>CP</th>
<th>EP</th>
<th>Learning disabilities</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Paper II (n=70)</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Heterotropia (n=51)</td>
<td>11/11**</td>
<td>12/13</td>
<td>18/19*</td>
<td>20/21*</td>
</tr>
<tr>
<td>Nystagmus (n=23)</td>
<td>10/11**</td>
<td>10/11*</td>
<td>12/16</td>
<td>14/19*</td>
</tr>
<tr>
<td><strong>Paper IV (n=35)</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Fixation time sec (mean) (n=35)</td>
<td>1.4 and 1.4</td>
<td>1.0</td>
<td>1.9*</td>
<td>3.7</td>
</tr>
<tr>
<td>Drifts n (mean)</td>
<td>2.0 and 0</td>
<td>1.0</td>
<td>0.8</td>
<td>1.3</td>
</tr>
</tbody>
</table>

* p<0.05; **p<0.001

P-values indicating relation to children without associated impairments. In all groups the p-value is indicating more abnormal findings in children with associated impairments.
Discussion and speculations

Heterotropia was one of the most common abnormalities found in this multidisciplinary investigation of children with HC where ophthalmological, neurological, and psychological evaluations were performed. In addition, these children exhibited increased frequency of ocular motor abnormalities, and abnormal fixation behaviour compared to an age and sex matched comparison group. Our findings indicate a vulnerability of the binocular system in children with early developing HC.

One problem in this study was to find normative data for the different orthoptic tests and outcomes, as different studies use different tests in different ways, use different tests for the same outcome and use different classifications for normative outcomes. Furthermore, many studies fail to provide information on how each test was used, or indeed which test was used. In addition, most studies present the results of one or a few tests. Therefore, in paper I, 143 children were investigated with a battery of accurately described tests, commonly used in clinical practice. Care was taken to select accurate tests and definitions suitable for use in clinical praxis in children in general as well as in children with different diseases.

When diagnosing strabismus, the cover test is the most accurate, and preferred, test but it requires an ability to fixate a target for some seconds. In children with poor cooperation, the Hirschberg test is commonly used. However, it is important to note that with the Hirschberg test many cases of heterotropia < 5 degrees could have been missed (Choi 1998). In this study, among children with HC we would have missed 33% of all heterotropias that were identified using cover test if we had used only the Hirschberg test.

We chose 3m for distant fixation for the cover test because the visual acuity test is constructed for that distance, and a longer distance might have reduced attention. A disadvantage with this short distance might be that some exodeviations may have been missed. When measuring the secondary positions at 3m, stationary fixation targets were placed on the wall, at 1m distance from straight a head position (up, down, right and left), as in general the child’s eye position, when sitting in the chair corresponds to 1m above the floor. Our aim was to keep the child’s head stable while measuring the secondary position: this makes it easier to compare the angles between objects compared to letting the child move her/his head.

In classifying strabismus, some types are defined in terms dependent on the AC/A calculations. Normal AC/A is considered to be between 2/1 and to 4/1 (Ansons and Davis 2001; von Noorden and Campos 2002; Rowe 2004). We found a large variation in AC/A with both methods used in the present study including nega-
tive values (1/1 to 15/1 and –6/1 to 9/1). In the calculation of AC/A, it is assumed that the amount of accommodation exerted is known. In reality, it is difficult to control accommodation during the measurement although an accommodative fixation object is used. In our opinion, these tests are unreliable when used in children in the two ways we chose. However, to our knowledge; no study has evaluated their usefulness in children. Consequently, we were not able to make a detailed classification of strabismus type according to AC/A.

We chose to study the frequency of heterophoria >2PD, and found a higher prevalence (26%) in the comparison group than that of “significant” heterophoria (17%), defined as 5 PD or more including, intermittent strabismus in an old study (Fletcher and Silverman 1966). Our findings were slightly lower than the frequency of heterophoria at distance (28%) reported in an Australian study (Junghans et al. 2002) where heterophoria was diagnosed with the Howell phoria card; and significantly different from the 0.2% reported in a screening study from Turkey in which deviations were “checked” with a method that the authors fail to describe (Turacli 1995). Again, these other studies used different diagnostic methods and different criteria than the present study, which could explain the differences.

However most of our results from paper I are in agreement with those of other studies examining one or few variables in larger populations and therefore we believe that our results may be used for comparisons with different patient groups.

The ophthalmologic/orthoptic evaluation of children with HC is often difficult due to neurological and cognitive deficits. In the present, multidisciplinary study neurological and psychological examinations were also performed which gave us valuable information about the abilities/disabilities of each child.

A number of authors have described an angle difference between up and down gaze in A-and V-syndrome; however, to our knowledge, no report has described which fixation distance to use, or specified a distance for measuring the secondary positions (Ansons and Davis 2001; von Noorden and Campos 2002; Rowe 2004). The wide range of frequencies of A-and V-syndromes reported among children with HC (4-61%) (Harcourt 1968; Houtman et al. 1981; Hamed et al. 1993; Folk 1997; Altintas et al. 2005; Hoyt 2005) may indicate methodological problems.

Interestingly, there was a high frequency of abnormal head postures among children with HC, while abnormal head posture was completely absent in the comparison group. The main ophthalmologic reasons for abnormal head posture are efforts to maintain binocular single vision and maximal VA. In Paper II, all except one child with abnormal head posture had heterotropia in all directions of gaze, and binocular single vision could not be obtained. A large proportion (81%) of the children with abnormal head posture also had nystagmus and/or ocular motility defects. In some of these children, the nystagmus may have been dampened by the head posture.
Regarding stereoacuity, different authors and different screening studies have different pass fail criteria for “normal” stereoacuity (Ohlsson et al. 2001; Jung-hans et al. 2002; Marsh et al. 1980). In the present study, mean stereoacuity in the comparison group was better than 60" in all four age groups and no child had subnormal values (120"-480"). Among children with HC, there was a high rate of heterotropia with negative stereoacuity (60%), and five (7%) of all the children (n=67) who could perform the stereo test had subnormal results (120” to 480”), one of these five children had orthophoria and the rest had heterophoria.

Stereoacuity is perception of the relative depth of images presented to each eye. In the TNO and Lang test used in the present study, the child is assumed to extract a visual scene of depth by assigning relative depth values from random dots, and creating one image for each eye, with slight differences, based on binocular disparity. These are complex functions probably requiring an amount of perceptual processing which might be affected in children with HC.

The motility investigation of ductions and versions was something of a challenge in the children with HC, mainly due to poor cooperation, and difficulties with fixating and following. Despite these limiting factors in the examination, we found that more than 60% (41/68) had defect motility i.e. over- and/or under-actions of the extra ocular muscles.

Few studies on fixation behaviour with objective measurements have been performed in children. In one study an increase in time of maintained central gaze by suppressing reflex saccades, while fixating nonsense geometric patterns, and suddenly appearing shapes like in TV-games, was described in boys between 8 and 10 years of age (Paus et al 1990). In addition, Fischer and Hartnegg used the infrared reflexion technique in children with and without dyslexia (Fischer and Hartnegg 2000), and described a decrease of intruding saccades during fixation while fixating a stationary point. However, children with dyslexia performed more intruding saccades compared to children without.

Children with HC (paper IV) of other aetiologies than MMC had shorter fixation time and performed more drifts than children in the comparison group do. Contrary to our expectations, no difference was found between children with HC and the comparison group in frequency of intruding saccades. As a high frequency of intruding saccades, have previously been described, in studies of adults with cerebral pathology (Sharpe et al. 1982; Abadi 2004) we expected the frequency of intruding saccades to be increased in children with HC. It may be speculated that reason for our finding is that we chose the 10 best seconds in the analysis and thus rejected the part of the registration with the highest frequency of intruding saccades.

An increase in spontaneous blinking during childhood was speculated to reflect brain maturation in a study by Zametkin and co-workers, which also discussed the involve-
ment of blinking in sensory motor processing and attention (Zametkin et al. 1979). In the comparison group, a significant increase in number of blinks was seen during development. Children with overt HC at birth performed fewer blinks than did children with HC developing during first year of life. These findings might indicate abnormal development and maturation of the blink behaviour in children with HC. Interestingly, only one child in the HC group showed manifest nystagmus in both eyes during the 10-second evaluation, while intermittent and latent nystagmus could be detected in an additional 16 children in other parts of the registration or in the calibration session. In eight of the seventeen children with nystagmus (47%) detected by the eye tracker the nystagmus was not detected using gross observation or slitlamp examination: one reason for this may be that they had an intermittent type of nystagmus with extended foveation periods. The majority of children with nystagmus in the present study had intermittent jerk nystagmus (16/17) 94% and nystagmus occurred more often in children with CP than in those without (p<0.01). There was no difference in VA between children with or without nystagmus in the present study. This may be explained by extended foveation periods, which have previously been described in children with nystagmus (Dorn et al. 2005; Jacobs and Dell’Osso 2004). Another explanatory factor for this finding is most likely that the most visually impaired children were excluded from the Orbit analysis, as the recordings require ability to fixate an object.

Regarding aetiology in relation to visual impairment and fixation behaviour, differences did occur between children with MMC and children with aetiologies other than MMC: the latter group which was dominated by children with generalised parenchyma lesions (Persson et al. 2006, accepted for publication) had more visual impairment, performed more drifts, and had shorter fixation time. In addition, this group had more associated neurological impairments than the MMC group.

One interesting finding was that heterotropia was equally common independent of aetiology (MMC/not MMC), while fixation abnormalities, visual impairment, CP, and EP were more frequently seen in children with aetiologies other than MMC to their HC. Although heterotropia is a less severe condition it is possible that the cerebral systems responsible for binocular functions and those responsible for the fixation behaviour and other associated neurological diagnoses have different vulnerability to the increase in intracerebral pressure and dilatation of the ventricles in HC early in life.

Over the years, the morbidity of HC has changed. The incidence of MMC has decreased, and the survival of very/extremely pre term children has increased (Persson 2007). Thus, the HC group of children is deviating towards a population with more associated neurological impairments and more fixation abnormalities. In addition, strabismus has been a frequent finding over the years and the present study shows no trend of decrease. Our study group of children with HC was heterogeneous, with adverse events occurring during different episodes, and differ-
ent frequencies of episodes during development. Naturally, the outcome differs depending on the maturity of the system, and the location, type, and degree of the adverse event. Obvious abnormalities like wide-angle heterotropia in acute high ICP are seldom missed, while subtle defects in children may be neglected. Subtle fixation abnormalities may have an influence on different activities in everyday life, such as school performance. Children in the present study have low average IQ, learning disabilities, attention problems and autistic spectrum behaviour. However, cause and effect relationship of the different function abnormalities are yet unknown.

Children with HC and associated neurological impairments, which make the evaluation of visual functions difficult, are very likely to have strabismus and fixation abnormalities. Therefore, to gain insight into the child’s abilities and disabilities, a multidisciplinary assessment including an orthoptic evaluation is recomended as a basis for habilitation.

Considerations of the study

Children in the comparison group were selected for their suitability as references in clinical studies. We chose children aged 4 to 15 years because it is difficult to investigate children younger than four years of age properly. This comparison group was not population-based, and it would have been preferable to include a larger group of children however, the group was representative of the general population regarding gestational age and birth size. Neither living conditions nor socio-economic group differed from those in other communities in Sweden or the Swedish standard, when we compared information regarding socio-economic background obtained from the parents, with data from Västra Götaland, other communities in Sweden, and the total Swedish population (SCB)– (Statistiska Centralbyrån) (Grönlund, Andersson et al. 2006). The orthoptic findings in the comparison group are in accordance with that of other studies examining one or few variables in larger populations and thus we conclude that our results may be used for comparisons with different patient groups. All children in both groups were investigated with the same test, the same investigator (EA), in the same order and mostly on weekends when they were free from school to optimise the concentration.

Fixation behaviour was recorded for 20 seconds. During that time, the longest continuous fixation time of the right eye without intruding drifts, blinks and saccades was chosen and the fixation time was calculated. Drifts, blinks and intruding saccades were counted during the ten seconds of the registration that the analyser considered the best. This may have caused a bias since it might be difficult to decide with precision exactly which period was the best. An objective selection by the computer would have been preferable. More information about the quality of the child’s fixation might have been gained by calculating the total
time the child was fixating the target during the 20-second registration, and even a greater difference might have been found between the groups. The reason why we chose to select the ten best second was that it was supposed to be fairer to the children as a fixation time of 20 seconds is a long time for children to keep the attention. The analyses of continuous fixation time were done by the same analyst in both groups one of the authors (EA) while the 10-second analyses, were performed by two analysers, one for the comparison group and one of the authors (EA) for the HC group.
Conclusions

Results from the orthoptic examination in our comparison group of children 4-15 years of age from four different schools and pre-schools were in accordance with the results of other studies investigating one or more orthoptic variables.

Orthoptic abnormalities such as heterotropia, motility abnormalities, abnormal head posture and nystagmus were much more frequent in children with HC than in an age- and sex-matched comparison group.

Children born with HC had a higher risk of orthoptic abnormalities as compared to those who develop hydrocephalus during first year of life. A higher frequency of orthoptic abnormalities were seen among children with associated neurological impairments than in those without. There was no correlation between frequency of orthoptic abnormalities and the aetiology of HC or ventricular width.

In the comparison group of children, 4-15 years of age, fixation density around centre of gravity of fixations and fixation time increased with increasing age, while intruding saccades decreased with increasing age. Blinks and drifts could not be shown to be age dependent.

Children with hydrocephalus had shorter fixation time and had a higher frequency of drifts, than an age- and sex-matched comparison group.

Children with MMC in combination with HC did not differ from the comparison group regarding fixation behaviour, while children with aetiologies other than MMC had more fixation abnormalities. Children born with HC had lower frequency of blinks compared to those who develop hydrocephalus during first year of life. Children with associated neurological impairments, regardless of aetiology, had more fixation abnormalities. There was no correlation between ventricular width and visual fixation.
Människans synsystem är ett komplicerat nätverk av olika hjärnfunktioner. Detta system styr de båda ögonen och håller deras rörelser stadigt tillsammans, samtidigt som det förmedlar synintryck som hjärnan bearbetar, så att vi kan tolka och förstå det vi ser. Olika medfödda eller förvärvade hjärnskador kan påverka ögonens rörelseförmåga och samarbete vilket kan orsaka skelning, påverka för-mågan att hålla blicken stadig och/eller ge annan neurologisk påverkan. Orsaken till skelning är okänd men man vet att ärfighet, för tidig födelse, hjärnskador och kromosomavvikelser medför ökad risk för skelning. Fördjupande studier av skelning och samsyn mellan ögonen används oroptiska undersökningsmetoder och vid undersökning av blickstabilitet kan man använda mätutrustning för registrering av ögonrörelser. Syftet med mina studier har varit att kartlägga förekomsten av skelning, samsynsfunktioner och fixationsbeteende hos barn i allmänhet och hos en grupp barn med känd hjärnskada (barn opererade för vattenskalle (hydrocefalus)). Hos barn med hydrocefalus har vi har även jämfört våra resultat med orsak och tid för uppkomsten av hydrocefalusen och om våra fynd skiljer sig mellan barn med tilläggshandikapp så som CP, epilepsi, inlärningssvårigheter och synskada och hos dem utan tilläggshandikapp.

Eftersom det saknats referensvärden till många av de testmetoder som vi ortoptister använder för att diagnostisera skelning och samsynsfunktioner undersökte vi 143 barn mellan 4 och 15 år, från fyra olika skolor och förskolor i Göteborg med omnejd (studie I). Denna grupp är tänkt att användas som jämförelsegrupp till olika patientgrupper i framtiden. I studie I fann vi att manifest skelning förekom hos 3.5 % av barnen och dold skelning förekom hos 26 % av barnen. Endast ett barn hade onormala ögonrörelser, 97 % hade utmärkt samsyn och lika många, 97 % kunde föra ögonen tillsammans mot näsan (konvergera) 6 cm från ansiktet eller närmare. Inget barn hade nystagmus (ögondarr) eller tvångshuvudhållning. Våra resultat stämde väl överens med tidigare studier där man undersökt en eller flera av dessa ortoptiska funktioner.

I studie II ingick 75 barn opererade för hydrocefalus, dessa barn undersöktes även av barnneurolog, barnneuropsykolog och barnögonläkare. Resultaten jämfördes med resultaterna från en ålders- och könsmatchad grupp från studie I. Av barnen med hydrocefalus skelade nästan 20 ggr så många (69 %) som barnen i jämförelsegruppen och 60 % hade onormala ögonrörelser (ett barn i jämförelsegruppen). Bland barnen med hydrocefalus var nystagmus vanligt (44 %), liksom förekomsten av tvångshuvudhållning (41 %). Att födas med hydrocefalus medförde ökad risk för ortoptiska problem jämfört med att utveckla hydrocefalus under första levnadsåret. Skelning och nystagmus var vanligare bland barn med tilläggshandikapp (CP, EP och inlärningssvårigheter) än bland dem utan. I studie III undersökes 117 barn från studie I, med en infraröd ögonrörelsemätare.
Svensk sammanfattning

(Orbit). Med ökande ålder uppvisade barnen i jämförelsegruppen längre sammanhängande fixationstid och fixationen blev mer sammanhållen kring fixationspunkten. Antalet ofrivilliga sackader (snabba ofrivilliga förflyttningar av blicken bort från det fixerade objektet och tillbaka) minskade med ökande ålder. Antal blinkningar och drifter (långsamma ofrivilliga förflyttningar av blicken bort från det fixerade objektet) var oförändrade mellan 4 och 15 år. I studie IV fann vi att fixationsbeteendet skiljde sig mellan barnen med hydrocefalus och de i jämförelsegruppen (Studie III). Barnen med hydrocefalus hade kortare sammanhållna fixationstid, gjorde fler drifter och hade mer nystagmus än barn ur den ålders och könsmatchade jämförelsegruppen.

Barn men ryggmärgsbråck i kombination med hydrocefalus skiljde sig inte ifrån jämförelsegruppen i sitt fixationsbeteende medan de barn som hade annan orsak till sin hydrocefalus, hade kortare fixationstid och gjorde fler drifter. De barn som hade tilläggshandikapp hade mer avvikande fixationsbeteende (hade kortare fixationstid, gjorde fler drifter och hade mer nystagmus) än de utan. Inget sammanband sågs mellan de studerande variablerna och storleken på hjärnans hållum.

Sammanfattningsvis så visar våra fynd att bland barn med hydrocefalus är samsynfunktionen en av de oftast påverkade funktionerna i hela denna multidisplinar undersökning. Fixationsbeteende skiljer sig mellan barn med hydrocefalus av annan orsak än ryggmärgsbråck och jämförelsegruppen. De med tilläggshandikapp har oftare skelning vilket kan bidra till de svårigheter dessa barn har i det dagliga livet. Det är av stor vikt att ha kunskap om hela barnet för att få en optimal uppfattning om barnets styrkor och svagheter för att kunna erbjuda bästa möjliga habilitering.
Acknowledgements

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