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Visual field constriction in children with shunt-treated hydrocephalus

Clinical article

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Object. Many ophthalmological abnormalities are described in conjunction with hydrocephalus. The results of visual field diagnosis remain a matter of further discussion. The aim of this study was to investigate visual field deficits in children with shunt-treated hydrocephalus.

Methods. All children over 6 years of age treated for hydrocephalus at the authors' institute between December 2007 and December 2008 were included in the study. The children underwent an ophthalmological investigation for strabismus and binocular function, ophthalmoscopy, visual acuity, and refraction.

The special focus was the visual field diagnosis, which the authors established in all children with cognitive conditions. The investigation was made by using the Goldmann visual field examination (kinetic perimetry). Children with and without visual field defects were compared concerning age at the time of ophthalmological examination, genesis of hydrocephalus, and frontooccipital horn ratio measured on current CT or MR images.

Results. Complete investigations were undertaken in 56 children (24 girls and 32 boys, mean age 15.1 years). The following orthoptic pathological entities were diagnosed: 29 children have a strabismus in 29 cases, 17 of these have an exotropia, 12 an esotropia, 4 children a hypotropia, 2 a hypertropia and 3 children a heterophoria. A nystagmus was found in 10 children. The ocular fundus investigation showed 13 children with an optic nerve atrophy.

A visual field diagnosis was possible in 44 of the 56 patients and was incomplete in 12 patients with cognitive deficits or inadequate compliance. In 24 of 42 children there was a concentric visual field constriction between 10° and 50° out of the center. Children with visual field deficits were older than those with a normal visual field ($p = 0.051$). Nine of 10 children with postmenigitic hydrocephalus had a visual field defect ($p = 0.025$). In children with visual field defects the frontooccipital horn ratio was significantly higher ($p = 0.013$).

Conclusions. The results suggest that children with shunt-treated hydrocephalus have a higher risk of having ophthalmological abnormalities. Visual field deficits are often a problem in these patients. A diagnostic visual field examination can complete the ophthalmological monitoring in patients with hydrocephalus, especially in patients with large ventricles. Children with postmeningitic hydrocephalus should be ophthalmologically monitored more frequently and intensively. *(DOI: 10.3171/2010.8.PEDS1042)*

Key Words • hydrocephalus • shunt • children • visual field • ophthalmological outcome

Hydrocephalus is characterized by an enlargement of the cerebral ventricles caused by any pathological condition in production, resorption, or circulation of CSF. Due to the close anatomical relationship between the ventricular system and the visual pathway, this ventricular dilation associated with intraventricular and intracranial pressure carries the risk of neuroophthalmological disorders. Optic atrophy may result from optic nerve ischemia, optic nerve or chiasmal traction, chiasmal compression, and transsynaptic neuronal degeneration.

Many ophthalmological abnormilities are described in conjunction with hydrocephalus, especially strabismus and amblyopia, reduction of visual acuity, refractive errors, and ocular motility disorders.[1–4,10–12,14,15] Compared with other neuroophthalmic complications, the results of visual field examination are less frequently described. Humphrey et al.[13] and Molia et al.[18] reported several cases involving visual field defects. Within an extended clinical study Houliston et al.[12] identified visual field defects in 8 patients. It is our impression that the results of visual field defects remain a matter of further discussion. The purpose of the present study was to perform an extended ophthalmological examination in children with shunt-treated hydrocephalus, focusing on the visual field examination.

Abbreviation used in this paper: FOH = frontooccipital horn.

Methods

We conducted a prospective, nonrandomized, nonblinded study. All children attending the hydrocephalus consultation in pediatric surgery at the University of Leipzig were included. The minimum age of participation was 6 years. All children suffered from a hydrocephalus of different genesis. We only included children in whom a shunt system was implanted. Children with myelomeningocele, acute or chronic inflammation of the eye, and injuries or tumors of the eye were excluded. Between December 2007 and December 2008, data obtained in 56 patients were evaluated. All patients underwent examinations at the Department of Ophthalmology of the University of Leipzig. In all cases an extensive ophthalmological examination was performed, including diagnosis of strabismus and binocular function, ophthalmoscopy, visual acuity, refraction, and visual field diagnosis. The aim was to examine the visual field in all children. The visual field diagnosis was performed using the Goldmann examination. We tested 24 dots per eye using Goldmann mark III/4. Mark V/4 was used in children who could not see Mark III/4. We preferred the kinetic perimetry, which is much easier to handle for children than the static perimetry. The perimetry was done manually because the manually operated perimetry enables the examiner to have an optimal control over the patient. All examinations were done twice and only reproducible results were included in this study. We considered different results of both visual field examinations to be unreliable. These results were excluded.

Age at the time of examination, age at the time of first shunt insertion, genesis of hydrocephalus, week of gestation, birth weight, head circumference before shunt operation, number of revisions, and current CT or MR image were reviewed in all cases.

To compare the ventricular size before shunt insertion to the current ventricular size, the ventricular index before shunt insertion and the FOH ratio on current CT or MR images were reviewed in all cases. Because in most patients ultrasonography through the fontanel was performed before shunt insertion, the ventricular index is the only index of the ventricular size before shunt surgery that could be reviewed. Although the FOH ratio is a different index, each allows a proportion of the ventricular size.

The FOH ratio was measured by adding the distance of the lateral limits of the frontal horn and the occipital horn and dividing it by the double internal diameter of the cranium. The ventricular index was calculated sonographically by measuring the distance between the cerebral falx and the lateral limit of the lateral ventricle in the amount of the foramen of Monroe and dividing it by the internal diameter of the cranium.

Results

A total of 56 ophthalmological examinations were performed between December 2007 and December 2008. Of these examinations 32 were performed in boys and 24 were performed in girls, all ranging from 6 to 27 years of age. The mean age was 15.1 years. Causes of hydrocephalus were related to intracranial hemmorrhage in 16 (28.6%), meningitis in 10 (17.9%), occlusion in 7 (12.5%), and other causes in 23 (41.1%). There were 54 VP shunts and 2 ventriculoatrial shunts.

Strabismus and Nystagmus

Twenty-nine (51.8%) of the 56 children had a strabismus. Esotropia was found in 12 cases (21.4%), exotropia in 17 cases (30.4%), hypotropia in 4 cases (7.1%) and hypertropia in 2 cases (3.6%). Nystagmus of different intensity was evident in 10 patients (17.9%).

Ocular Fundus

Total optic nerve atrophy was seen in 4 patients (7.1%) and was bilateral in all 4. Partial optic nerve atrophy was documented in 9 patients (16.1%) and was bilateral in 8 and unilateral in 1.

Visual Acuity

Visual acuity was examined with a Snellen Eye Chart with best correction monoculary. A visual acuity score of less than 0.3 was found in 15 cases (26.7%), a visual acuity score of 0.3–0.8 was seen in 24 cases (43%), and a visual acuity score higher than 0.8 was documented in 17 cases (30.4%).

Refraction and Anisometropia

Refractive errors were identified in 31 patients (55.4%). Fifteen children (26.8%) had myopia of greater than –1.0 diopter. A hyperopia of higher than +1.0 diopter was determined in 13 cases (23.2%). Astigmatism of higher than 1.5 diopter was found in 11 patients (19.6%). Anisometropia was found in 7 children (12.5%) with a difference of acuity between both eyes between 2.5 and 3.0 diopters.

Visual Field

The visual field examination was undertaken in all 56 patients. Results that were not certainly reproducible were excluded. The examination was incomplete in 12 patients. Reasons for not having an analyzable result in these cases were cognitive deficits on the one hand and inadequate compliance on the other. Thus, there were 44 visual fields for analysis.

Visual field deficits were documented in 24 patients (54.5%). The results obtained in each visual field were analyzed individually. In 5 patients the visual field examination was possible in one eye only. One of these patients had an anophthalmus and in the other 4 cases the visual acuity of one eye was not adequate. Finally 83 eyes were analyzed. Of the 83 results, 43 concentric visual field constrictions (52%) were noted. There were visual field constrictions with a mean distance of 10° (and up to 50°) from center. A visual field constriction of 10° distance from center was found in one eye (1.2%), a constriction of 20° distance from center in 5 eyes (6%), a constriction of 30° distance from center in 17 eyes (20.5%), a constriction of 40° distance from center in 7 eyes (8.4%), and a constriction of 50° distance from center in 13 eyes (15.7%). The time of occurrence of the visual field defects in our patients cannot be stated because the visual field testing did not have any relation to the shunt revision procedure and the analysis of the ophthalmological disorders was not longitudinal in this study.

Statistical Analysis

We compared data obtained in patients with normal visual fields and those with visual field defects. The variable included age and ventricular index at the time of first shunt surgery, week of gestation, birth weight, head circumference before shunt operation, age at the time of examination, FOH index on current CT or MR images, genesis of hydrocephalus, and number of shunt revisions. The comparison of both groups concerning age and ventricular index at the time of first shunt operation showed no difference between those with and without visual field defects. Consequently, children with visual field defects did not statistically have larger ventricles before shunt insertion than children with a normal visual field. Therefore, the visual field constrictions did not occur before shunting. Moreover, when comparing week of gestation, birth weight, and head circumference before shunt surgery no significant difference could be shown. These results demonstrate that all children statistically had the same preconditions before shunt insertion.

By contrast, the comparison of both groups at the present time indicates significant differences. Patients with visual field defects are older than those with a normal visual field. This difference is not statistically significant, but it shows a trend ($p = 0.051$, Fig. 1).

Whereas the ventricular index before shunt surgery was statistically identical in both groups ($p = 0.538$), the FOH ratio on current CT or MR images indicates a significant difference (Fig. 2). Patients with visual field defects have a higher FOH ratio than those with a normal visual field ($p = 0.013$).

Concerning the genesis of hydrocephalus there were significantly more children with hydrocephalus after meningitis who had visual field defects ($p = 0.025$ (Fig. 3). To evaluate possible changes of the cortex due to bacterial meningitis, we reanalyzed all current CT or MR images in these children, but we found no greater ischemic damage to the striate cortex or radiations in the majority of these cases. In merely 2 children were degenerative changes of the striate cortex demonstrated.

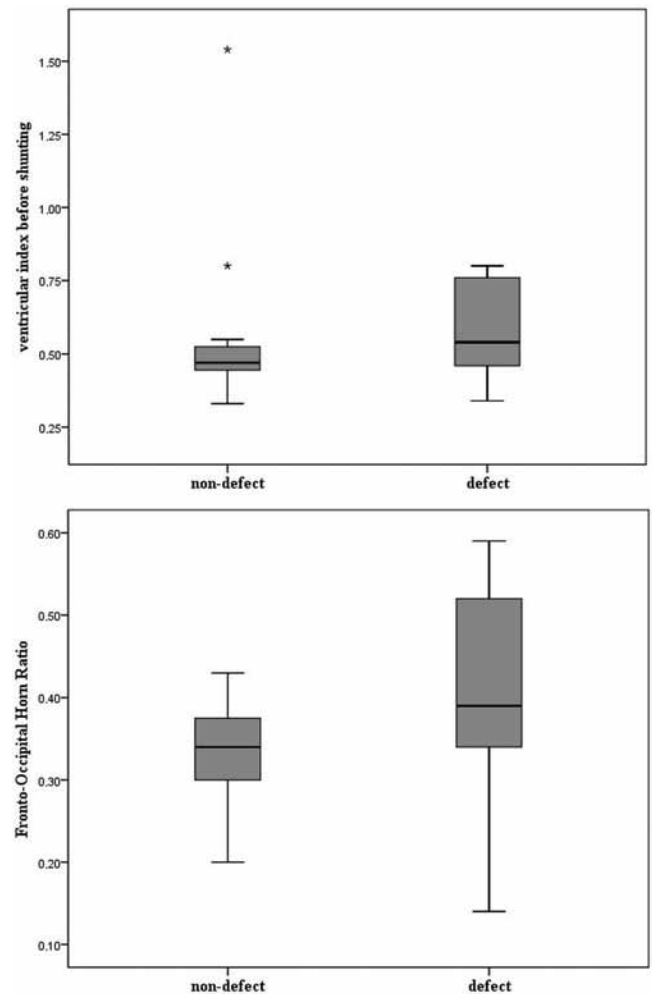

Fig. 2. Upper: Bar graph showing the ventricular index before shunt insertion. No significant difference could be shown ($p = 0.538$). **Lower:** Bar graph demonstrating a significant difference in patients with and without visual field defects in terms of FOH index on current CT or MR images ($p = 0.013$).

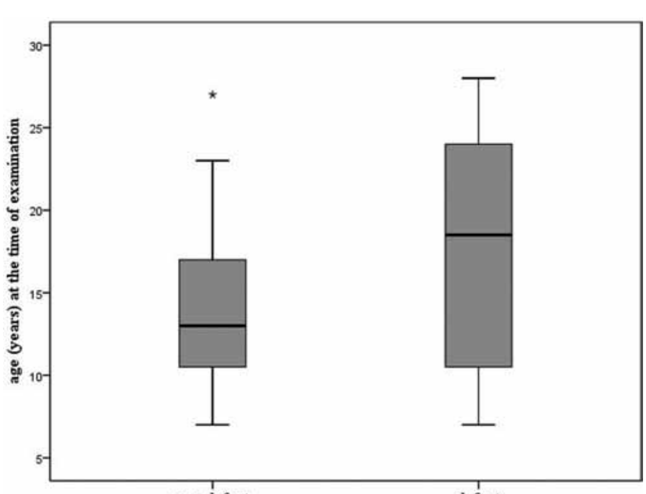

Fig. 1. Bar graph depicting age at the time of examination. The intergroup difference is not significant but it shows a trend ($p = 0.051$).

In terms of the number of revisions, no significant difference can be shown.

Children with visual field defects have a significantly higher prevalence of strabismus ($p = 0.035$) and refractive errors ($p = 0.014$). By contrast, concerning nystagmus ($p = 0.054$) and the occurrence of optic nerve atrophy ($p = 0.477$) no significant difference can be shown. The comparison of both groups indicates a significant difference of the visual acuity ($p = 0.001$). However, the visual acuity only correlates slightly positively with the extent of the visual field constriction, which is statistically not significant ($r = 0.279$, $p = 0.070$) (Fig. 4). Thus, children with visual field defects have a lower visual acuity, but the assumption that a low visual acuity automatically leads to a greater extent of the visual field constriction cannot be proved.

Discussion

More than half (51.8%) of our examined patients

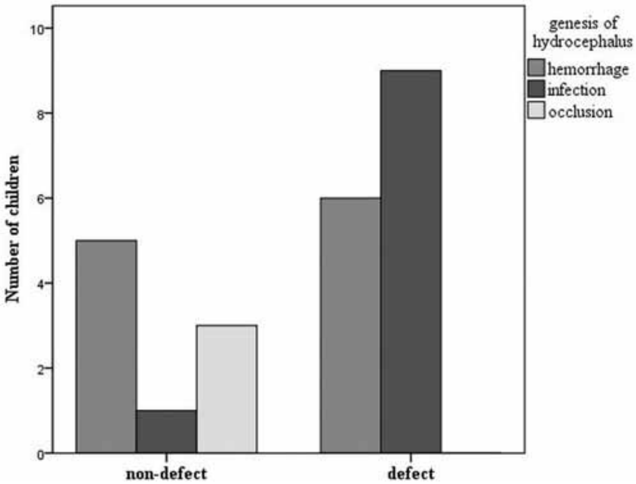

Fig. 3. Graph demonstrating genesis of hydrocephalus. Significantly more children with postmeningitic hydrocephalus presented visual field defects (p = 0.025).

exhibited a strabismus, which is comparable to previous studies. Zeiner et al.[21] reported on 8 of 18 patients with a history of strabismus, whereas Altintas et al.[1] reported that 40% of their patients manifested a squint. Compared with patients (69%) in a study by Andersson et al.,[2] the prevalence of strabismus was lower in our patients.

A nystagmus was found in 17.9% of our patients, an incidence that is similar to that (18%) reported by Mankinen-Heikkinen and Mustonen.[16] By contrast, Andersson et al.,[2] as well as Aring et al.,[3] showed a higher prevalance of nystagmus (48% and 44%, respectively).

The frequency of optic nerve atrophy has been reported to be between 14% and 31%,[2,4,8,9,10,12] which is in agreement with our results (23%).

Compared with Andersson et al.[2] and Heinsbergen et al.,[11] we found more children with visual impairment. Whereas Andersson et al.[2] reported on 11 (15%) of 74 children having a visual acuity of lower than 0.3, which was similar to that (13%) cited by Heinsbergen et al.,[11] 15 (26.7%) of our 56 patients had a visual acuity of lower than 0.3. A visual acuity between 0.3 and 0.8 was found in 24 (43%) of 56 children, which is also a higher incidence than that (34%) reported by Andersson et al.[2]

Significant refractive errors were present in 55% of our patients, which is in contrast to the results of Andersson et al.,[2] who identified refractive errors in 67% of their patients; the results are also in contrast to those of Altintas et al.,[1] who reported that approximately 20% of their patients had refractive measurements indicating amblyogenetic refractive errors.

Our results of the visual field examination are in absolute contrast to previous studies. Molia et al.[18] reported on 2 cases of visual field defects. One of these patients had a right incongruous homonymous hemianopia and superior constriction, and the other patient a right homonymous hemianopia defect. Humphrey et al.[13] described 4 cases of visual field defects in obstructive hydrocephalus. One patient was afflicted with a bilateral central scotoma, another with an unilateral central scotoma, one with a temporal loss, and one with a temporal hemianopia of the left

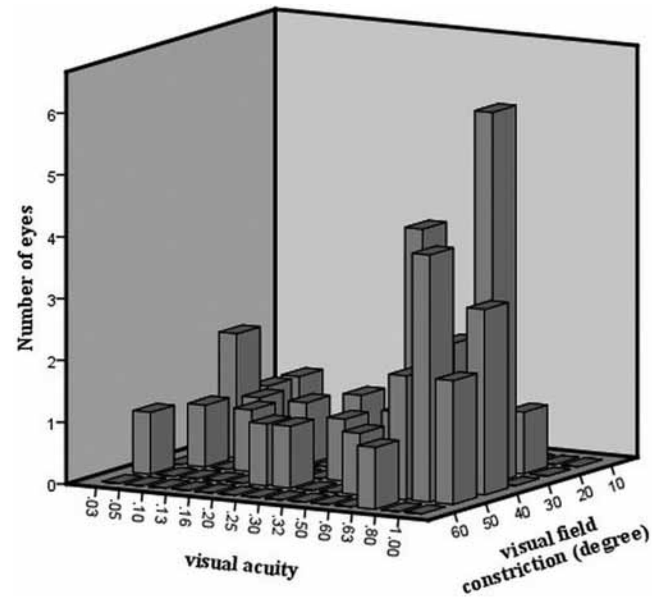

Fig. 4. Graph demonstrating the low correlation between visual acuity and the extent of visual field constriction (r = 0.279, p = 0.070).

eye as well as a temporal field loss of the right eye. The study by Mankinen-Heikkinen and Mustonen[16] showed a normal Goldmann visual field in 37 (84%) of the 44 eyes. Two of these patients had homonymous hemianopia. In the study by Houliston et al.,[12] visual field defects were identified in 8 patients (16%). On confrontation testing, only 5% of the examined children in the study by Gaston[9] had visual field defects. Although it is difficult to compare our results with those in previous studies because of the varying number of cases, we found a much higher incidence of visual field defects (52%). Furthermore, we only found concentric visual field constrictions, which is also in absolute contrast to previous studies.

The different kinds of visual field deficits may be traced back to different localizations of the damage of the visual pathway. It has already been stated that compression and distortion of the visual pathway may produce ophthalmological disorders and visual field defects.[5–7,17,20] An exclusive constriction of the visual field might result from an impairment of the optic nerve or the postgeniculate sensory pathway.[19] Most likely, in our opinion, this is the hypoxic-ischemic damage of the optic radiation within the brain injuries caused by hydrocephalus. As all visual field constrictions in our patients are bilateral, we considered the compressive optic neuropathy to be implausible. Thus, the compression and hyoxic-ischemic brain injuries of the optic pathway, especially of the optic nerve, chiasma, and the optic radiation may be one reason for having ophthalmological abnormalities in association with hydrocephalus. The effect of vigilance, concentration, and understanding of the examination, however, must be also discussed. A lack of vigilance and concentration can worsen the results of a visual field examination, especially in working with young children. We attempted to reduce the influence of these factors as much as possible by taking the necessary steps for examination. Nevertheless, disorders of concentration in children suffering from brain injuries might be one reason for having this high incidence of visual field defects.

Conclusions

In this study, we noted that children with shunt-treated hydrocephalus have an increased risk of ophthalmological abnormalities, both orthoptic abnormalities and ocular fundus pathologies. In addition, a visual field defect is often a problem in these patients: its diagnosis in young children is difficult and might produce unreliable results, especially in those with cognitive deficits and a low level of concentration. Nevertheless, all possible defects in the visual system related to hydrocephalus should be closely monitored as intensively as possible to discern any developing ophthalmological abnormalities. The visual field diagnosis is not adequate to diagnose acute increase of intracranial pressure but should be included in the ophthalmological investigation, especially in older children. Children with postmeningitic hydrocephalus should be monitored more frequently and intensively.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author contributions to the study and manuscript preparation include the following. Conception and design: Geyer, Rudolph, Sterker, Graefe. Acquisition of data: Rudolph, Sterker. Analysis and interpretation of data: Geyer, Rudolph, Sterker. Critically revising the article: Geyer, Sterker, Graefe, Till. Reviewed final version of the manuscript and approved it for submission: all authors. Statistical analysis: Rudolph, Ulrich. Administrative/technical/material support: Sterker, Graefe. Study supervision: Geyer, Sterker, Graefe.

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