Research Article

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Visual disorders in children with shunted hydrocephalus.

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ABSTRACT

The introduction of shunt operations in neurosurgical practice gave a chance for life to these patients and significantly improved its quality. Along with its positive effect, surgical treatment is associated with a number of complications that, together with hydrocephalus itself, affect the subsequent development of shunted patients in the short and long term. In the long term, problems observed in shunted patients are mental retardation, cerebral palsy (CP), epilepsy, hearing and vision disorders. Visual disturbances are common in children with shunted hydrocephalus. These include reduced visual acuity, eye movement disorders, refractive anomalies, visual field defects, reduced color vision and strabismus, optic atrophy. The mechanism of visual disturbances is associated with damage to the oculomotor pathways, optic nerves and optic radiation, dilatation of the lateral ventricles, and damage to the periventricular white matter. Age at onset of hydrocephalus and ventricular dimension are important in the development of visual disturbances and placement of a shunting system can improve visual function. At the same time ophthalmic disturbances may be the first and only sign of shunt dysfunction and rapid normalization of elevated ICP preserves vision. Visual disturbances in patients with shunted hydrocephalus correlate with higher incidence of CP and epilepsy. They determine the children's social integration and quality of life.

Keywords : shunted hydrocephalus, reduced vision, strabismus, nystagmus, optic atrophy.

INTRODUCTION

Hydrocephalus is an active expansion of the ventricular system resulting from inadequate passage of cerebrospinal fluid (CSF) from its point of production in the cerebral ventricle to the point of absorption in the systemic circulation (1).The introduction of shunt operations in neurosurgical practice gave a chance for life to these patients and significantly improved its quality. Along with its positive effect, surgical treatment is associated with a number of complications that, together with hydrocephalus itself, affect the subsequent development of shunted patients in the short and long term. The most common immediate complications of placement of shunt systems are mechanical (obstruction of the valve mechanism, ventricular or peritoneal catheter end), functional (inadequate immediate or permanent drainage of the shunt system), and inflammatory (ventriculitis, sepsis, and shunt nephritis). In the long term, problems observed in shunted patients are mental retardation, cerebral palsy (CP), epilepsy, hearing and vision disorders, frequent and/or prolonged hospitalizations. Each complication impairs the quality of life of patients and their families. Visual disturbances in shunted patients are due to damage to the oculomotor pathways, optic nerves, and optic radiation. These include reduced visual acuity, eye movement disorders, refractive anomalies, visual field defects, reduced color vision and strabismus, optic atrophy (2-8).

Papilledema has been described as a sign of shunting system dysfunction. Arnell et al. (9) found papilledema in six older children with shunted hydrocephalus (8-14.5 years) without or with very mild symptoms of increased ICN. In five of the children, this happened during control medical examinations, including ophthalmoscopy, and in one, on the occasion of a mild headache.Five of the children had no new neurological symptoms and only one had ataxia. CNS CT showed mild ventricular dilatation in three of the children, and intraoperatively increased ICP was demonstrated in all. The authors emphasize the importance of papilledema as a sign of dysfunction and the need for control ophthalmoscopies in the follow-up of shunted patients.

The incidence of visual disturbances in shunted hydrocephalus is high. Anderson et al. (10) found ophthalmological abnormalities in 83% of examined children with shunted hydrocephalus. Only 27% of the examined children with with shunted hydrocephalus and myelomeningocele (MMC) in Gaston's (11) study had normal visual function. Rabinovich (7) found a high incidence of reduced vision, reaching 24% of observed children with shunted hydrocephalus. About 3 decades later, Heinsbergen et al. (12) found severe visual

impairment (reduced visual acuity below 0.3) in 13% of the children studied. Similar results were also reported by Anderson et al. (10) and Person et al. (13). The lower incidence of reduced visual acuity observed in the later studies is explained by earlier diagnosis and improved treatment. Children with hydrocephalus and MMC have better vision. Bilgan (2) reported reduced vision in only 5% of children with hydrocephalus with MMC, which is consistent with the results of Anderson et al. (10). Both Heinsbergen et al.(12) and Anderson et al.(10) found no correlation between visual acuity and number of shunt revisions. The authors emphasize the importance of timely treatment of increased ICN for the prevention of visual acuity. Optic atrophy can result from optic nerve ischemia, optic nerve or chiasm traction, chiasmal compression, and transsynaptic neuronal degeneration (14). Optic atrophy (OA) in hydrocephalus may result from optic nerve ischaemia, optic nerve or chiasmal traction, chiasmal compression, and transsynaptic neuronal degeneration (14). Due to the proximity of the posterior visual pathways to the lateral ventricles, they can be damaged by ventricular dilatation. Optic atrophy is found in varying frequency (14-22%) in scientific reports (15-17). The incidence is significantly higher (28-30%) in shunted patients born with very low birth weight who developed posthemorrhagic hydrocephalus (18). Anderson and Hellstrom (15), examining the optic disc and retinal vessels in children with surgically treated hydrocephalus found a significantly reduced optic disc with an abnormal vascular pattern. The authors conclude that hydrocephalus is characterized by a subnormal optic disc, which is associated with pre- and perinatal disorders in the development of these structures. The lower incidence of optic atrophy is probably related to better perinatal care and regulation of ICN (15).

Refractive disorders were found in 67% of children with shunted hydrocephalus studied by Anderson et al. (10) with significant hyperopia in 46%. According to Saunders et al. (19), hyperopia is the most common finding in children with brain pathology in general. The reason for this remains unclear. Contrary to this reading, Makkinen-Heikkinen and Mustonen (20) found astigmatism as the most common refractive error (25.5%), hyperopia in 20% of children, myopia in 13%. In Bilgan's study (2), astigmatism was significantly lower (13.8%). Frequency of strabismus was highest (69%) in the study by Aring et al.(21) and Anderson et al.(10) compared to other studies by BIlgan (2), Fernell et al.(16), Gaston (17), Makkinen-Heikkinen and Mustonen (20) and Rabinowich (7) where its frequency is between 39% and 61%. Gaston (11) found a high rate (42%) of strabismus in children with hydrocephalus with MMC, finding it to be the most common abnormality in the ophthalmic status of these patients. Koktekir et al. (22) studied the characteristics of strabismus and its evolution in seventeen patients between the ages of 6 months and 13 years. All had developmental delay, five patients out of 17 were premature. The most frequent finding is esotropia in 14 children and exotropia is found in only three. Glasses were prescribed to 13 patients: hyperopic correction in 12 and myopic correction in one patient. Surgical correction was performed in five patients. Four of them achieved successful ocular alignment. AlObaisi SS et al. (23) followed 190 children with shunted hydrocephalus, of which 63 (33.1%) were diagnosed with strabismus. Exotropia was diagnosed in 26 (13.6%) of the patients at the initial assessment, and in 7 (3.6%) of the patients at the final assessment. The authors found a statistically significant association between VP shunt and strabismus.

The frequency of nystagmus is also high (44-48%) among shunted patients of Anderson et al. (10) and Aring et al. (21). Its frequency was lower (29%) in children with hydrocephalus with MMC in Gaston's (17) study, where it was the second most common visual abnormality after strabismus.

Several studies have been devoted to visual perceptual problems (24, 25, 7). Anderson et al. (10) found such problems in 59% of the examined children and confirmed the data of Houliston et al. (24). These disorders are found in various combinations and include impaired ability to plan movements through depth, impaired simultaneous perception, impaired movement perception, impaired orientation, and impaired cognition. Normal vision does not exclude visual-perceptual problems. According to the data of Anderson et al. (10), 36% of their patients with normal vision have visual-perceptual disorders. Damage to the occipital cortex in children can result in many complex disorders of cognitive visual function. (24) The parents of 52 children (ages 5 to 17) with shunted hydrocephalus were then asked the same set of questions about the specific problems).

Evidence of cognitive visual problems was identified in 27 of these children of whom 16 manifested multiple difficulties. The disabilities identified by this study comprised problems with: shape recognition, simultaneous perception, perception of movement, colour perception, orientation, object recognition, and face recognition. Cognitive visual dysfunction was identified in 59% (38/64 от пациентите в проучването на Andersson и съавт.(10). Neuro-ophthalmological examination is important for the diagnosis and therapy of elevated IOP (26). Gaston (11) states that acute manifestations of squint, other eye motor disturbances, and papilledema are usually indicative of uncontrolled hydrocephalus. According to Tzekov et al.(27), a large part of the ophthalmological manifestations of shunt dysfunction precede the manifestation of CT changes, which makes them important for early diagnosis and treatment. Ophthalmic disturbances may be the first and only sign of shunt dysfunction (28, 29, 11). Rapid pressure normalization is required to preserve vision (26, 11).

In their study, Arring et al. (21) found that the number of

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shunt operations, ventricular dimensions and the etiology of hydrocephalus have less importance for the development of eye complications compared to the age of onset of hydrocephalus. These conclusions are based on the fact that children with hydrocephalus detected at birth have significantly more heterophoria, esotropia, abnormal head position, and eye motility defects than children who develop hydrocephalus after the first year. At the same time (28, 29, 11) indicate that shunt implantation in children with hydrocephalus can improve previously impaired visual function Additional diagnoses such as cerebral palsy and epilepsy correlate with a higher frequency of eye disorders: lower visual acuity, presence of nystagmus and strabismus. Anderson et al. (10) found a higher frequency of impaired vision, nystagmus, and strabismus in children with shunting hydrocephalus and an IQ below 70. Visual disorders were most frequent in those with epilepsy, cerebral palsy, and/or cognitive disability (10).

The presented literature data demonstrate that disturbances in the visual function are often observed in children with shunted hydrocephalus. The mechanism of visual disturbances is associated with damage to the oculomotor pathways, optic nerves and optic radiation, dilatation of the lateral ventricles, and damage to the periventricular white matter. Literature sources indicate that ventricular dimensions, the etiology of hydrocephalus and the number of shunt operations have less importance for the development of eye complications compared to the age of onset of hydrocephalus, and the placement of a shunting system can improve visual function. Literature data confirm that ophthalmic disturbances may be the first and only sign of shunt dysfunction and rapid normalization of elevated IOP preserves vision. Vision is fundamental to child development, allowing access to information, social integration and movement, and this determines its extreme importance for the general outcome and quality of life of shunted patients. The role of visual disturbances as an aggravating factor, worsening intellectual development and the quality of life of shunted patients is confirmed.

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