



Strabismus in hydrocephalus patients

Bengu Ekinici Koktekir¹ · E. Koktekir² · G. Ozcan¹ · H. Karabagli²

Received: 5 April 2019 / Accepted: 9 May 2019 / Published online: 21 May 2019
© Springer-Verlag GmbH Germany, part of Springer Nature 2019

Abstract

Purpose To evaluate the characteristics and outcomes of strabismus in patients with hydrocephalus.

Methods A retrospective chart review of patients with strabismus and hydrocephalus is performed in the Department of Pediatric Ophthalmology between 2012 and 2018.

Results Seventeen patients between the ages of 6 months and 13 years met the criteria of strabismus and hydrocephalus. Although all had developmental delay, five patients out of 17 were premature (lower than 36 weeks of gestation). All patients had ventriculoperitoneal shunt placement for congenital hydrocephalus. Three patients had exotropia, whereas 14 had esotropia. Glasses were prescribed to 13 patients: hyperopic correction in 12 and myopic correction in one patient. Surgical correction with bimedral recession was performed in five patients. Four of them achieved successful ocular alignment.

Conclusions Children with hydrocephalus most likely have esotropia. Although good ocular alignment is achieved with surgical correction in some patients, some patients may benefit from glasses.

Keywords Ventriculoperitoneal shunt · Acquired strabismus · Congenital hydrocephalus · Esotropia

Introduction

Hydrocephalus is characterized by enlargement of cerebral ventricles caused by abnormality in absorption, production, or circulation of cerebrospinal fluid. Although a wide range of neuro-ophthalmologic conditions may occur, strabismus is the most commonly seen complication of hydrocephalus [1–4]. Compared with the general population, these patients are more likely to develop esotropia; however, they may present with exotropia, superior oblique overaction, and dissociated vertical deviation [1–3, 5, 6].

This study was performed to demonstrate the characteristics and clinical outcomes of patients with hydrocephalus and strabismus.

Methods

We retrospectively reviewed the charts of patients with congenital hydrocephalus who were referred to the pediatric ophthalmology department between 2012 and 2018 in this study. The inclusion criterion was having shunt surgery. Ethics committee at the Medical Faculty of Selcuk University approved this study. Informed consent was not required due to de-identification of patients.

The examinations of patients were performed by a single pediatric ophthalmologist (BEK). All patients underwent complete ophthalmological examination including visual acuity testing if possible, evaluation of ocular motility and alignment, dynamic retinoscopy, cycloplegic refraction with retinoscopy, and autorefractometry when possible; also, anterior segment examination with biomicroscopy and dilated fundus examination were performed. In preverbal or uncooperative children, visual acuity was measured using fixation preferences in distance and near, using a fixation target. In the case of cooperated patients, age-appropriate recognition optotypes like Snellen letters, E chart, or Allen pictures were used to obtain data. Ocular alignment was evaluated by Hirschberg test, Krimsky test, and alternate prism cover test in near and distance using a fixation target when possible. The presence of any esotropia or exotropia was noted. Esotropia was

✉ Bengu Ekinici Koktekir
benguekinici@hotmail.com

¹ Department of Ophthalmology, Selcuk University Faculty of Medicine, Konya, Turkey

² Department of Neurosurgery, Selcuk University Faculty of Medicine, Konya, Turkey

considered as congenital if presented before 6 months of age and acquired esotropia if occurred after 6 months of age.

For ocular motility assessment, ductions and versions were evaluated using a penlight or a fixation target at near. The presence of nystagmus was noted. Cycloplegic refraction was measured by using a streak retinoscope in a semi-dark room 30 min after two drops of cyclopentolate 1% administration 10 min apart to each eye. After noting the refractive status of the patient, indirect ophthalmoscopic for the evaluation of lens, vitreous, and fundus was performed. Any abnormality especially in the optic disc, such as the presence of optic atrophy or papilledema, was noted.

Descriptive statistical analysis was performed using SPSS 20.0 Inc.

Results

The mean age of patients was 26.8 ± 22.5 months (range between 10 months and 8 years) at the initial visit. There were 9 (53%) boys and 8 (47%) girls. The follow-up period ranged from 2 to 6 years with an average of 4.1 ± 1.4 years. All patients had developmental delay; 5 (29%) patients were premature. Only one of the premature patients needed treatment for retinopathy of prematurity, as peripheral retinal laser photocoagulation.

All patients had congenital hydrocephalus, one patient had Dandy-Walker malformation, and one had desmoplastic infantile astrocytoma in addition.

Fourteen out of 17 patients had esotropia. Among the patients with esotropia, four had congenital esotropia while 10 children had acquired esotropia. One of the patients with acquired esotropia had inferior oblique muscle overaction. Three out of 17 patients had exotropia but all was acquired. One of the esotropia patients had limitation in abduction due to associated sixth nerve palsy. In the initial visit, mean deviation in children with esotropia was 33.5 ± 13.1 prism diopters (range between 14 and 60), whereas the patients with exotropia had 40 ± 20 prism diopter deviation (range between 20 and 60). Nine of the 14 patients with esotropia needed prescription of glasses with hyperopic correction. One of the patients with exotropia was premature and treated for ROP and was prescribed glasses with myopic astigmatism correction (Table 1). The acquired type of esotropia had an average of 3.42 ± 1.32 (1.25–5D) diopters, while the patients with congenital esotropia had low to moderate hyperopia with a mean of 0.25 ± 0.50 (0–1D) diopters. One out of three patients with exotropia had low myopic refractive error ($-2.5/-3.0$ D).

Five of our patients (29%) had optic atrophy of any degree and one had rotatory nystagmus. Three of the five patients with optic atrophy had a history of shunt revision.

Visual acuity examination in 10 children younger than 2 years of age revealed that they could fixate and follow the

light and fixation target. In 7 children over 2 years of age, 5 was uncooperative to any visual acuity test; however, only two children had visual acuity over 0.7 that was measured by E optotypes.

Five of the esotropia patients (4 congenital esotropia, 1 acquired esotropia) needed surgery. The bimedial recession was performed as all had alternative esotropia. Four of these five patients had successful ocular alignment. One patient needed additional surgery for residual esotropia; however, parents of the patient did not accept.

Six out of 17 patients needed revision of the shunt procedure. The mean final deviation was greater in these patients ($p = 0.04$). The amount of final deviation is also correlated with the number of revision surgeries ($\rho, 0.581$; $p = 0.03$).

Discussion

The compression and hypoxic brain injury were proposed to be reasons for having ophthalmological abnormalities in association with hydrocephalus [7, 8]. Strabismus is the most commonly observed ophthalmic complication of hydrocephalus. Increased intracranial pressure in hydrocephalus was reported to affect the visual sensory system and oculomotor nerves; moreover, it can be the cause of optic atrophy and/or bilateral abducens palsy [2, 9]. Varying degrees of esotropia (ET) and exotropia (XT) ratio as 0.75:1 to 3.5:1 have been reported [1, 2, 10, 11]. Esotropia, as the convergent form of strabismus, might be initiated by the abducens palsy and later might become concomitant [9]. We have observed esotropia more frequently; our study population had a 4.6:1 ET:XT ratio, higher than in the literature. The majority of esotropia was in acquired type and observed after 6 months of age.

Sixth nerve palsy was the cause of limitation of abduction in one patient. This patient had esotropia probably related to increased intracranial pressure and also underwent shunt revision surgery. Nystagmus was seen in one patient out of 17; it was in rotatory fashion.

All patients with esotropia were found to have some degrees of hyperopia. The acquired type of esotropia had an average of 3.42 diopters, while the patients with congenital esotropia had low to moderate hyperopia. One of the patients with acquired esotropia underwent surgery for strabismus. But nine out of 10 patients with acquired esotropia gained orthophoria only with glasses. All four patients with congenital esotropia had strabismus surgery; only one had residual esotropia of 25 prism diopters.

All our patients had a history of ventriculoperitoneal shunt surgery for congenital hydrocephalus; only one patient with additional Dandy-Walker malformation and one with desmoplastic infantile astrocytoma.

Prematurity low gestational birth weight was blamed for strabismus [12]. Our study population included five premature

Table 1 Data of patients with strabismus and hydrocephalus

Patient	Age (month)	Type of strabismus	Initial deviation (D)	Final deviation (D)	Refraction (spherical equivalent) –OD/OS	Strabismus surgery	Shunt failure (times)
1	12	Esotropia	45	15 PD with glasses	+ 5.00/+ 5.25	No	Yes
2	23	Esotropia	60	Ortho with glasses	+ 2.25/+ 3.25	Yes	No
3	35	Esotropia	35	Ortho with glasses	+ 4.5/+ 5.00	No	No
4	14	Esotropia	35	Ortho with surgery	0	Yes	No
5	24	Esotropia	60	30PD with glasses	+ 3.5/+ 4/00	No	No
6	12	Esotropia	45	18PD with glasses	+ 4.5/+ 4.5	No	No
7	6	Esotropia	15	10PD with glasses	+ 1.00/+ 1.25	No	No
8	34	Exotropia	20	20PD	0	No	Yes (2)
9	33	Esotropia	30	Ortho with surgery	0	Yes	No
10	62	Esotropia	40	Ortho with surgery	+ 2.00/+ 1.75	Yes	No
11	16	Esotropia	40	5PD with glasses	+ 5.00/+ 4.50	No	No
12	10	Exotropia	60	15PD with glasses	– 3.5/– 4.00	No	No
13	37	Esotropia	40	25PD with glasses	+ 2.50/+ 3.5	Yes	Yes (2)
14	95	Esotropia	15	15PD	0	No	Yes (2)
15	11	Esotropia	30	35PD with glasses	+ 3.00/+ 2.50	No	Yes (1)
16	22	Esotropia	30	20PD with glasses	+ 1.50/+ 1.25	No	Yes (1)
17	10	Exotropia	40	12PD with glasses	+ 4.00/+ 3.75	No	No

D, diopters; OD, right eye; OS, left eye

children and 12 patients, normal gestational age. The mean amount of deviation was similar between mature and premature children. There were four cases with esotropia and one with exotropia in the premature group.

Cerebrospinal shunt failures are frequently seen in congenital hydrocephalus. A high number of shunt failures were shown to have positive ophthalmological evidence that intracranial pressure is high. Change in visual acuity, reversible congruous homonymous hemianopsia, change in the angle of strabismus, or significant refractive errors are some reported findings regarding shunt failure; moreover, these findings might be a previous sign before ventriculomegaly [2, 8]. Ophthalmic complications are also reported to be more frequently observed in revisions of cerebrospinal fluid shunts [13, 14]. We have six patients who underwent shunt revision procedures. The mean amount of deviation in those patients was higher than in the patients without shunt failure. Five of our patients had some degrees of optic atrophy. Three of these five optic atrophy cases had two shunt revision procedures. A complete ophthalmic examination is mandatory in hydrocephalus patients to be sure about the function of shunts. Strabismus, visual acuity, and optic nerve function assessment should be monitored and abnormality should be managed before the visual system becomes mature and more resistant to any treatment.

In conclusion, esotropia of acquired type is more commonly observed in our study group of congenital hydrocephalus. Most of the patients benefit refractive correction with glasses, although some of them, mostly the congenital type esotropia,

needed surgery to align the eyes and relieve strabismus. Successful ocular alignment might be obtained despite the developmental delay and comorbid conditions in congenital hydrocephalus patients with strabismus.

Compliance with ethical standards

Ethics committee at the Medical Faculty of Selcuk University approved this study. Informed consent was not required due to de-identification of patients.

Conflict of interest On behalf of all authors, the corresponding author states that there is no conflict of interest.

References

1. Mankinen-Heikkinen A, Mustonen E (1987) Ophthalmic changes in hydrocephalus. A follow-up examination of 50 patients treated with shunts. *Acta Ophthalmol* 65:81–86
2. Altintas O, Etus V, Etus H, Ceylan S, Caglar Y (2005) Risk of strabismus and amblyopia in children with hydrocephalus. *Graefes Arch Clin Exp Ophthalmol* 243:1213–1217
3. Aring E, Andersson S, Hård AL, Hellström A, Persson EK, Uvebrant P, Ygge J, Hellström A (2007) Strabismus, binocular functions and ocular motility in children with hydrocephalus. *Strabismus* 15:79–88
4. Rudolph D, Sterker I, Graefe G, Till H, Ulrich A, Geyer C (2010) Visual field constriction in children with shunt-treated hydrocephalus. *J Neurosurg Pediatr* 6:481–485
5. Anderson HA, Stuebing KK, Buncic R, Mazow M, Fletcher JM (2012) Factors associated with strabismus in spina bifida myelomeningocele. *J Pediatr Ophthalmol Strabismus* 49:284–289

6. DeBenedictis CN, Rubin SE, Kodosi SR (2015) Esotropia in children with ventricular-peritoneal shunts. *Strabismus* 23:117–120
7. Constantini S, Umansky F, Nesher R, Shalit M (1987) Transient blindness following intracranial pressure changes in a hydrocephalic child with a V-P shunt. *Childs Nerv Syst* 3:379–381
8. Chou SY, Digre KB (1999) Neuro-ophthalmic complications of raised intracranial pressure, hydrocephalus, and shunt malfunction. *Neurosurg Clin N Am* 10:587–608
9. Houtman WA, Meihuizen-de Regt MJ, Rutgers C (1981) Strabismus and meningocele. *Doc Ophthalmol* 50:255–261
10. Biglan AW (1995) Strabismus associated with meningocele. *J Pediatr Ophthalmol Strabismus* 32:309–314
11. Gaston H (1991) Ophthalmic complications of spina bifida and hydrocephalus. *Eye* 5:279–290
12. Hennein L, Koo E, Robbins J, de Alba Campomanes AG (2019) Amblyopia risk factors in premature children in the first 3 years of life. *J Pediatr Ophthalmol Strabismus* 56:88–94
13. Sainte-Rose C, Piatt JH, Renier D, Pierre-Kahn A, Hirsch JF, Hoffman HJ, Humphreys RP, Hendrick EB (1991-1992) Mechanical complications in shunts. *Pediatr Neurosurg* 17:2–9
14. Katz DM, Trobe JD, Muraszko KM, Dauser RC (1994) Shunt failure without ventriculomegaly proclaimed by ophthalmic findings. *J Neurosurg* 81:721–725

Publisher's note Springer Nature remains neutral with regard to jurisdictional claims in published maps and institutional affiliations.