original article

The prevalence of strabismus and visual outcomes in children with hydrocephalus and a ventriculoperitoneal shunt: medical record review

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BACKGROUND: Many ophthalmological complications have been associated with hydrocephalus (HC), including ocular motility disorders, visual field defects, optic atrophy, and loss of visual acuity. No studies have investigated the prevalence of strabismus and visual outcomes of children with congenital HC after ventriculoperitoneal (VP) shunt in Saudi Arabia.

OBJECTIVES: Estimate the frequency of strabismic children diagnosed with HC who underwent a VP shunt procedure.

DESIGN: Medical record review

SETTING: Tertiary care center

PATIENTS AND METHODS: We reviewed the files of all pediatric patients diagnosed with hydrocephalus before the age of 2 years and treated with shunts during the period 2010 to 2020 at our institution.

MAIN OUTCOME MEASURES: Strabismus types and ophthalmic assessment (visual state, outcomes, and ocular motility state).

SAMPLE SIZE AND CHARACTERISTICS: 190 children; 98 (51.5%) males.

RESULTS: Eighty-nine (46.8%) had congenital HC followed by intraventricular hemorrhage 36 (18.9%); 74 (38.9%) patients had regular follow-ups in ophthalmology. Sixty-five (34.2%) patients had no ophthalmic assessment or fundus examination records, while 63 (33.1%) were diagnosed with strabismus. At the initial assessment, 26 (13.6%) patients had exotropia (XT). At the final assessment, 7 (3.6%) patients had XT. The association between VP shunt and strabismus was statistically significant, (χ^2=6.534, df=1, P<.01).

CONCLUSION: More than one-third of children diagnosed with HC who had surgical treatment in a tertiary hospital did not have any records of ophthalmic assessment, which highlights the need to implement a specific ophthalmological examination protocol in patients with HC. Further studies are needed to analyze the association between VP shunt and strabismus.

LIMITATIONS: Evaluating the ocular state and visual function before and after VP shunt was not possible.

CONFLICT OF INTEREST: None.

Hydrocephalus (HC) is an enlargement of the head caused by an accumulation of cerebrospinal fluid (CSF). This accumulation leads to a harmful increase in ventricular pressure and volume, requiring surgical intervention to release the pressure.[1] The ventriculoperitoneal (VP) shunt is the medical device used to drain excess CSF and restore normal pressure and flow.[1] HC in children can result from various etiologies: congenital, or acquired disorders such as malformations, tumors, infections, and intraventricular hemorrhage due to premature birth (post-hemorrhagic hydrocephalus).[2]

Many ophthalmological complications have been associated with HC,[3-6] including ocular motility disorders, visual field defects, optic atrophy, and loss of visual acuity (VA).[7] Additionally, there are ocular motility manifestations that can indicate VP shunt dysfunction and increased intracranial pressure, such as convergent squints, lateral rectus palsy, and divergence palsies.[8,9] In children with HC, strabismus is the most common ocular condition.[3]

Esotropia (ET), exotropia (XT), superior oblique overaction, and dissociated vertical deviation are types of strabismus that have been associated with HC.[3-5] Many studies have found that ET is highly associated with HC in children.[3,10-12] Although it has been reported that the shunting procedure will usually improve the angle of strabismus,[12,13] the revisions of the shunt may be a risk factor for strabismus and amblyopia.[5,12,13] It has also been suggested that the age of onset of HC has a significant effect on ocular motility disorders and visual function more than HC etiology, the number of shunt revisions, and ventricle size.[5] Many studies have concluded that children with HC benefit from ocular intervention, and successful ocular alignment can be achieved.[5,10,11,14] Understanding the association between HC and VP shunt with strabismus and the ocular state may help in the normal development of the ocular state. Moreover, regular follow-ups for ocular complications could be used as an indicator of VP shunt dysfunction, and this would provide an appropriate treatment plan for children with HC and strabismus.

In Saudi Arabia, the prevalence of HC is 1.1 per 1000 live births.[15] King Abdullah Specialist Children's Hospital (KASCH) is considered the main center that accepts HC in Riyadh. To our knowledge, no studies have investigated the prevalence of strabismus and visual outcomes of children with congenital HC and VP shunt at a tertiary care center, in Saudi Arabia. Thus, this study was conducted to evaluate the visual state in general, the characteristics, prevalence and outcomes of strabismus, and the ocular motility state in children diagnosed with HC who underwent a VP shunt procedure at KASCH.

PATIENTS AND METHODS

This study was a retrospective cohort descriptive chart review conducted at King Abdullah Specialist Children's Hospital (KASCH) in Riyadh, Saudi Arabia. The study included children who were diagnosed with HC and surgically treated before the age of 2 years. All pediatric patients who were diagnosed with HC before the age of 2 years in KASCH or who had been diagnosed before the age of 2 years and then referred to KASCH from 2010 to 2020 were included. Children with traumatic HC, who were surgically treated after the age of 2 or passed away after the surgical intervention were excluded. The full ophthalmic examination, any previous history of strabismus surgery, etiology of HC, number of follow-ups in ophthalmology, number of VP shunts, and the age of diagnosis were obtained from patient files. Detailed ophthalmic examinations were collected from multiple visits. However, there were some variabilities in ocular measurements between these visits. These included measurements of visual acuity testing, extraocular motility, and alignment, cycloplegic refraction to determine the refractive error, and a dilated fundus examination.

The best visual acuity at distance was measured with a Snellen chart if possible. If the patient was uncooperative, visual acuity was measured using fixation preferences in distance and near, a fixation target, and recorded as fix and follow. Ocular alignment was evaluated by alternate prism cover test at a near distance if possible and the measurements of ET, XT, or hypertropia were recorded. In uncooperative patients, ocular alignment was evaluated by the Hirshberg test or Krimsky test for near and distance viewing conditions. The extraocular motilities were also evaluated using a penlight and any presence of nystagmus was recorded. Cycloplegic refraction was obtained by using a retinoscope to determine the refractive error in a semi-dark room, 30 minutes after instillation of two drops of cyclopentolate 1% for both eyes to each eye. Cycloplegic refraction was recorded as a spherical equivalent (SE). Then, an indirect ophthalmoscope was used for the evaluation of the lens, vitreous, fundus, and any abnormality in the optic disc, such as the presence of optic atrophy or papilledema.

Refractive errors were calculated as the SE (sphere + (0.5 × cylinder) and subjects were subdivided into three refractive groups. Emmetropia was classified as the SE between 0.00 to <+3.00. Hyperopia was classified as the SE ≥+3.00 and was divided into the following

categories: +3.00 to <+8.00, and high hyperopia ≥+8.00. Myopia was classified as the SE ≥-0.50 and divided into the following categories: from -0.50 to <-6.00, and high myopia ≥ -6.00.16

Descriptive statistical analysis was performed using GraphPad Prism version 6 (California, USA) and IBM SPSS Statistics version 28. Data were checked for normal distribution using the Shapiro Wilk test. The discrete data were expressed as number and percentages, and the continuous data as median with minimum and maximum values. The chi-square test and Spearman correlation tests were conducted to determine relationships between categorical and continuous variables, respectively. *P*<.05 was considered statistically significant.

RESULTS

This study included 218 children who were diagnosed with HC and surgically treated before the age of 2 years. Their current ages range from 2-16 years with a median (IQR) of 11.0 (8) and range of 1.5 to 27.0 years. Twenty-eight passed away, leaving 190 children; 98 (51.5%) males and 92 (48.5%) females. Fifty children were born full-term (26.3%), 63 (33.1%) were born prematurely; birth term was unspecified in 77 (40.5%). Of 190 children, 105 (55.2%) were born and diagnosed at KASCH; 73 patients (38.4%) were referred to KASCH for surgical intervention. Of those, the record did not specify whether 12 children (6.3%) were born and diagnosed in KASCH or not. Of the 190, 152 (80%) were diagnosed and surgically treated at birth, 9 (4.7%) before the age of 6 months, and 10 (5.2%) after 6 months and before the age of 2 years. In 19 of the children (10%), the exact age at diagnosis was unspecified, but they were diagnosed and surgically treated before the age of 2 years. Most of our subjects had congenital HC (n=89, 46.8%) and intraventricular hemorrhage was the second most common etiology in 36 (18.9%), followed by meningitis in 18 (9.4%) (**Table 1**).

Table 1. Etiology in patients diagnosed with hydrocephalus.

Congenital hydrocephalus	89 (46.8)
Intraventricular hemorrhage	36 (18.9)
Meningitis	18 (9.5)
Spina bifida	9 (4.7)
Dandy-Walker syndrome	7 (3.7)
Chiari malformation	5 (2.6)
Microcephaly	3 (1.6)
Multiple congenital anomalies	15 (7.9)
Unspecified	9 (4.7)

Data are n (%).

All patients were surgically treated with VP shunts except two patients who did not need surgical intervention. Only 14 (7.3%) were treated with an Ommaya reservoir; 174 (91.6%) were treated with a VP shunt, and 76 (40%) had VP shunt once and did not need revisions. Of those needing revisions, 45 (23.6%) had one revision, 38 (20%) had two, 9 (4.7%) had three, 4 (2.1%) had four, and 2 (1%) had five revisions.

Less than half of our subjects (n=74, 38.9%) had regular follow-ups in ophthalmology clinics; 25 patients (13.1%) had one visit, which included a full ophthalmic assessment, and 26 patients (13.6%) were referred for fundus examination only. Sixty-five patients (34.2%) had no records of an ophthalmic assessment or fundus examination (**Table 2**). For the visual acuity test, 16 patients (8.4%) were able to see ≥ 20/22. Only 1 patient (0.5%) had counting fingers vision; in 46 (24.2%) visual acuity was recorded as follow and fixate, and 25 patients (13.1%) had light perception. Seventy-five patients (39.4%) had no records of VA. Three patients had cataracts, and one patient had 6th nerve palsy. (**Table 2**)

SE results showed that the majority of the patients were emmetrope, 46 patients (24.2%) ranged from 0.00 to <+3.00, followed by 26 patients (13.6%) who had hyperopia ≥+3.00 up to +8.00 and only one patient (0.5%) had high hyperopia of more than +8.00; 22 patients (11.5%) had myopia ≥-0.75 up to -6.00 and 1 patient (0.5%) had myopia of >-6.00. Nearly half of the subjects, 94 patients (49.7%) had no record of cycloplegic refraction (**Table 2**).

In our data set, 73 patients (38.4%) had normal fundus exam results; 16 patients (8.4%) had optic atrophy, 7 patients (3.6%) had retinopathy of prematurity, and 5 patients (2.6%) had chronic papilledema. One patient (0.5%) had a macular scar, and another one (0.5%) had a retinal detachment (**Table 2**).

Out of 190 subjects in this study, 73 patients (38.4%) had full extraocular motility and 63 (33.1%) patients were diagnosed with strabismus (**Table 3**). The initial median of the deviation measurement was 15.0 and the range was 0.0 to 65.0. An "A" pattern was presented in one patient, and three had a "V" pattern and one abnormal head position. Out of 63 subjects with strabismus, 32 (50.8%) did not need surgical intervention. Only 9 (14.3%) patients needed botox injections to correct their strabismus, and 2 (3.1%) patients needed only optical correction (**Table 4**). Only 15 (23.8%) patients underwent strabismus surgery, and 2 (3.1%) patients had

Table 2. Ocular findings in patients diagnosed with hydrocephalus (n=190).

Follow-ups	
Regular follow-ups	74 (38.9)
One visit only	25 (13.1)
Referred for fundus examination only	26 (13.6)
No records of an ophthalmic assessment or fundus examination	65 (34.2)
Visual acuity	
≥ 20/22	16 (8.4)
≥ 20/40	16 (8.4)
≥ 20/400	80 (4.2)
Counting fingers vision	1 (0.5)
Follow and fixate	46 (24.2)
Light perception	25 (13.2)
No record of VA	75 (39.5)
Other ocular conditions	
Nystagmus	15 (7.9)
Amblyopia	3 (1.9)
Cataract	3 (1.9)
6th nerve palsy	1 (0.5)
Spherical equivalent	
0.00 to < +3.00	46 (24.2)
≥ +3.00 up to +8.00	26 (13.6)
> +8.00	1 (0.5)
≥ -0.75 up to -6.00	22 (11.5)
> -6.00	1 (0.5)
No records of cycloplegic refraction	94 (49.7)
Fundus examination	
Normal fundus	73 (38.4)
Optic atrophy	16 (8.4)
Retinopathy of prematurity	7 (3.6)
Chronic papilledema	5 (2.6)
Retinal detachment	1 (0.5)
Macular scar	1 (0.5)

Data are n (%).

surgery with botox injections. At the final assessment, 10 patients (5.2%) had ET, 7 patients (3.6%) had XT, and only one patient had hypertropia. The median of the final deviation measurements was reduced to 0 with a range up to 40, but most values were missing (NA=131) (**Figure 1**).

A chi-square test was performed to study the relationship between several variables. For VP shunt and strabismus, the relation between these variables was significant, (χ^2=6.534, df=1, P<.01). Also, use of an Ommaya reservoir and strabismus were significantly related (χ^2=97.12, df=1, P<.0001). VP shunt and type of deviation at initial and final assessment were significantly related (P< .01). VP shunt and measurement of deviation at initial and final assessment were not significantly related (P= .15 and P=.98, respectively).

DISCUSSION

The results of our study found that the prevalence of strabismus in children with HC who underwent surgical intervention was 33.1%. VP shunt and strabismus were statistically associated. VP shunt and the deviation at initial and final assessment were statistically associated. Moreover, the data showed that 65 (34.2%) of the patients who had HC did not have any records of ophthalmic assessment or fundus examination.

Strabismus is the most common ocular condition that children with HC develop.[3] In our study, 63 patients (33%) of children who had HC and were surgically treated were diagnosed with strabismus. These findings resemble another study that examined 251 HC patients; 30% of their subjects had strabismus.[3,14] The previous studies have shown methodological variations, as there are some differences in methods and analyses which can make it difficult to compare results between studies. However, a higher frequency of strabismus has been reported in another study. They found that 69% of their subjects had strabismus compared to the normal population P≤.01.[17] The results of our study found that 12.1% had ET, 13.6% had XT at the initial assessment, and 5.26% had ET and 3.68% had XT at the final assessment (**Table 3**). Even though more children had XT than ET at the initial assessment, the difference was insignificant. Nevertheless, other studies have found that ET was more common than XT in their study group.[3,5,11]

Strabismus and VP shunt were statistically associated, and the deviation at initial and final assessments was also statistically associated with VP shunt. These findings are comparable to previous studies, which suggested that the shunting procedure is related to strabismus and may improve the angle of

strabismus.[12,13] However, due to the missing data in our study, it was not possible to obtain further statistical assessments to understand whether the number of VP shunt revisions affects the angle of strabismus or not. Contrarily, a recent study found that the shunting procedure and the number of revisions had minor importance regarding strabismus.[5] Additionally, they found that the onset of HC was more important as a risk factor for ocular motility disorders.[5]

The prevalence of strabismus is higher in children with HC compared to a normal population.[3,14,17] Additionally, visual disorder is considered the first and may be the only sign of VP shunt malfunction or raised CSP pressure.[12,18,19] Many studies have suggested that regular visual assessment is a necessity for HC patients, and it can be a valuable source of information.[18-22] Less than half of our subjects (38.9%) had regular follow-ups in ophthalmology, and 34.2% had no ophthalmic assessment or fundus examination records. As discussed earlier in previous studies, HC has a marked effect on the visual system and ocular motility.[2-6] Nevertheless, with regular visual assessment or surgical intervention, good ocular alignment can be achieved in these patients.[3,4,11] These findings highlight the importance of implementing an examination protocol in Saudi Arabia's health system that includes visual assessment for all children with HC. This would help to understand the effect of the shunting procedure on the ocular motility and visual state. Correspondingly, it may also provide an appropriate treatment plan for children with HC to improve the development of their visual function.

Several limitations need to be acknowledged in this study. The study was a retrospective and descriptive study. Missing ophthalmic findings for more than one-third of the patients limited further statistical analysis. Even though this study highlights some significant findings, further prospective studies are needed. However, to our knowledge, this is the first study in Saudi Arabia that has investigated the relationship between HC and VP shunt with strabismus and ocular state. In conclusion, 33.1% of children who had HC and were surgically treated with VP shunt had strabismus. Strabismus and VP shunt were associated. We recommend further prospective studies to understand this association between VP shunt and strabismus. More than one-third of children diagnosed with HC and who had surgical treatment in a tertiary hospital did not have any records of ophthalmic assessment. This highlights that it is essential to have a specified examination protocol that includes visual assessment for all children with HC in Saudi Arabia.

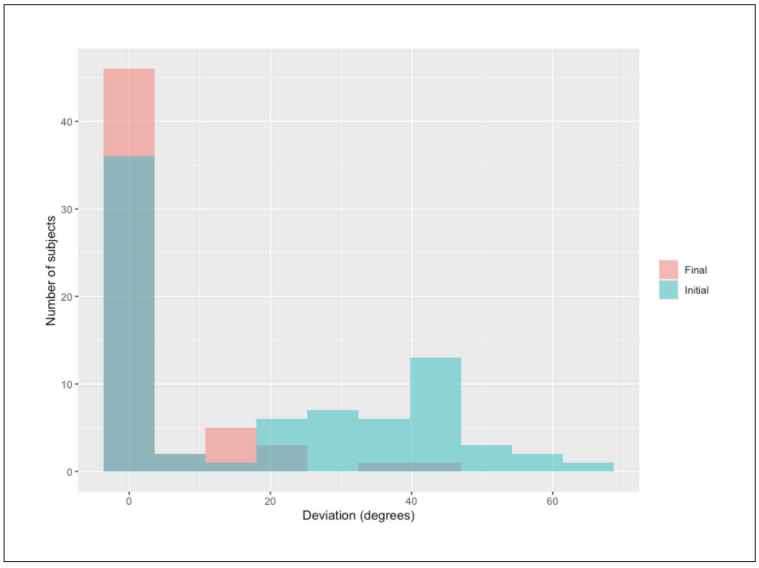

Figure 1. Initial and final measurements of ocular deviation in patients with extraocular motility.

Table 3. Strabismus at the initial assessment and the final assessment.

Type of deviation	Initial assessment (n=56)	Final assessment (n=18)
Esotropia	23 (12.1)	10 (5.3)
Exotropia	26 (13.7)	7 (3.7)
Intermittent exotropia	2 (1.1)	0
Hypertropia	3 (1.6)	1 (0.5)
Exotropia with hypertropia	2 (1.1)	0

Data are n (%).

Table 4. Treatment plan in patients with strabismus (n=63).

Did not need surgical intervention	32 (50.8)
Only botox injections	9 (14.3)
Needed only optical correction	2 (3.1)
Strabismus surgery	15 (23.8)
Surgery with Botox injections	2 (3.1)

Data are n (%). Data not available for 3 patients.

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