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CLINICAL STUDY

A comparison of visual function scores in hydrocephalic infants with and without lumbosacral myelomeningocoele

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Abstract

Purpose **The cerebrum is frequently malformed in children with myelomeningocoele. This anomaly renders them potentially susceptible to cerebral visual impairment. In these patients, hydrocephalus is an important and frequent complicating lesion which compromises intellectual function and may also cause cerebral visual impairment. In this study, we determined whether hydrocephalic patients with lumbar myelomeningocoele (HLM) are at a greater risk of visual impairment than hydrocephalic patients without this lesion (H).**

Methods **In this prospective study, we assessed five parameters of visual function in 20 hydrocephalic children with lumbar myelomeningocoele and compared the total visual function scores (TVFS) obtained with those from hydrocephalic children without overt spinal dysraphism, but similar in age, sex and ventricular size. The parameters, which were assessed with the aid of a quantitative grading scale, were pupillary size and reaction, optic atrophy, visual fixation and tracking.**

Results **The age and sex distributions of the patients in the two groups were similar. The anterior and posterior dimensions of the lateral ventricles were also similar. The mean (SD) of the TVFS were 24.25 (3.63) and 24.20 (3.47) respectively for the two groups (P = 0.90).**

Conclusions **The results suggest that, in hydrocephalic infants with lumbar myelomeningocoele, visual function is not further diminished by the associated dysraphism and that ventricular dilatation is the major determinant of visual impairment.**

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Introduction

Myelomeningocoele, a neural tube defect[1] is one of the most disabling developmental defects of infancy, childhood and adult years.[2,3] It represents the commonest form of spina bifida and is associated with several anomalies of the brain and spinal cord, cranial, vertebral and appendicular skeleton, and genitourinary system.[3] The incidence of hydrocephalus in patients with this disorder is 80–90%,[3] resulting usually from hind brain deformity and aqueductal obstruction.[2] The hydrocephalic process represents a risk factor for impairment of intellectual development.[4]

Disorders of the visual system are common in patients with this condition. These include abducens paresis, secondary optic atrophy and skew deviation due to Chiari malformation.[2] Although cataracts and corneal ulcerations are the common causes of blindness in African communities,[5] the contribution of lesions like hydrocephalus in these communities is probably unknown. Recently, we showed that in infants with hydrocephalus, vision is impaired in direct proportion to the degree of ventricular enlargement measured at the levels of anterior horns and collateral trigones.[6] The purpose of this study is to determine whether hydrocephalic children with lumbosacral myelomeningocoele are at a greater risk of visual impairment than those without this dysraphic state.

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Materials and methods

We prospectively evaluated 20 hydrocephalic children with lumbosacral myelomeningocoele (HLM) and 20 non-dysraphic hydrocephalic (H) children aged 18 months and below, who attended the neurosurgical service of our institution. The latter served as a control group. Both groups were similar in their distributions of age, sex and ventricular sizes (Table 1). We excluded children with closed fontanelles, visual dysfunction traceable to a clinically obvious disease of the eye and occipital encephalocoele. Patients who had ultrasound evidence of ventriculitis, multiloculated cysts or cerebral abscess were also excluded. The diagnosis of hydrocephalus was made by clinical examination and confirmed by ultrasonography.

We scanned each patient on the Accouson, ATL (Advanced Technology Laboratories, USA) ultrasound scanning machine, a B-mode real-time scan with gray scale imaging. The patients were scanned in supine position with or without sedation, employing all possible views, with a 7.5 MHZ sector transducer—moved to different positions on the scalp or tilted at different angles to obtain corresponding views.[3] An aqueous gel was used in all instances for good skull contact. We scanned the patients through the anterior fontanelle and less often through the posterior fontanelle. We measured ventricular size transversely at: (a) the levels of the anterior horns; and (b) collateral trigones and calculated the 'sonographic' Evans' ratio. This is a modification of Evans' original description of ventricular sizes based on computerized tomography (Figure 1).[3] The greatest transverse dimensions of the lateral ventricle at these reference points divided by the greatest transverse diameter of the cerebrum are the modified anterior (mAER) and trigonal (mTER) Evans' ratios respectively (Figure 1).

We evaluated visual function by a quantitative grading scale which we designed for a larger on-going study in the neurosurgical unit, of visual function in hydrocephalic children. The scale consists of an assessment and grading of the following parameters: visual acuity, pupillary diameter and reaction to direct light, and optic disc palor (Table 2). We assessed visual acuity by using an optokinetic tape and drum for the fixation target, as the children aged 18 months and below could not be tested quantitatively. For the

Table 1 Total visual function scores in 20 hydrocephalic patients with lumbar myelomeningocoele and in 20 non-dysraphic hydrocephalic children similar in age, sex and ventricular dimensions

Patients	*Hydrocephalus and myelomeningocoele*					*Hydrocephalus only*				
	Age (mo)	*Sex*	*Vent ratio*		*TVFS*	*Age (mo)*	*Sex*	*Vent ratio*		*TVFS*
			mAER	*mTER*				*mAER*	*mTER*	
1	2.0	M	0.50	0.70	20	2.0	M	0.54	0.86	24
2	15.0	M	0.62	0.83	21	1.7	M	0.40	0.50	18
3	3.0	F	0.72	0.78	20	3.0	F	0.70	0.80	22
4	2.0	M	0.54	0.86	24	2.0	M	0.59	0.82	24
5	1.0	M	0.45	0.52	28	1.5	M	0.35	0.54	28
6	1.0	F	0.65	0.77	26	1.0	F	0.66	0.68	24
7	4.0	F	0.55	0.70	28	3.0	F	0.72	0.78	25
8	5.0	F	0.59	0.66	23	5.0	F	0.46	0.50	25
9	2.5	M	0.38	0.39	28	3.0	M	0.42	0.47	26
10	2.0	M	0.46	0.49	24	2.5	M	0.59	0.82	24
11	7.0	M	0.50	0.54	28	6.0	M	0.68	0.85	25
12	4.0	F	0.39	0.44	28	5.0	F	0.38	0.46	26
13	0.50	M	0.62	0.70	22	0.75	M	0.64	0.73	23
14	0.75	M	0.65	0.79	20	1.0	M	0.63	0.75	24
15	1.50	F	0.33	0.37	29	1.25	F	0.40	0.42	30
16	1.50	F	0.29	0.34	30	1.0	F	0.36	0.44	28
17	0.50	M	0.65	0.72	22	1.0	F	0.64	0.70	23
18	0.50	M	0.90	0.80	22	0.25	M	0.74	0.79	24
19	5.0	M	0.64	0.79	24	4.50	M	0.43	0.46	26
20	9.0	F	0.74	0.85	18	8.0	F	0.63	0.84	14
Mean	3.39		0.56	0.65	24.3	3.44		0.52	0.66	24.2
SD	3.57		0.15	0.17	3.63	3.79		0.13	0.16	3.47

TVFS = total visual function score.
Vent ratio = ventricular ratio.
mAER = modified anterior Evans' ratio.
mTER = modified trigonal Evans' ratio.

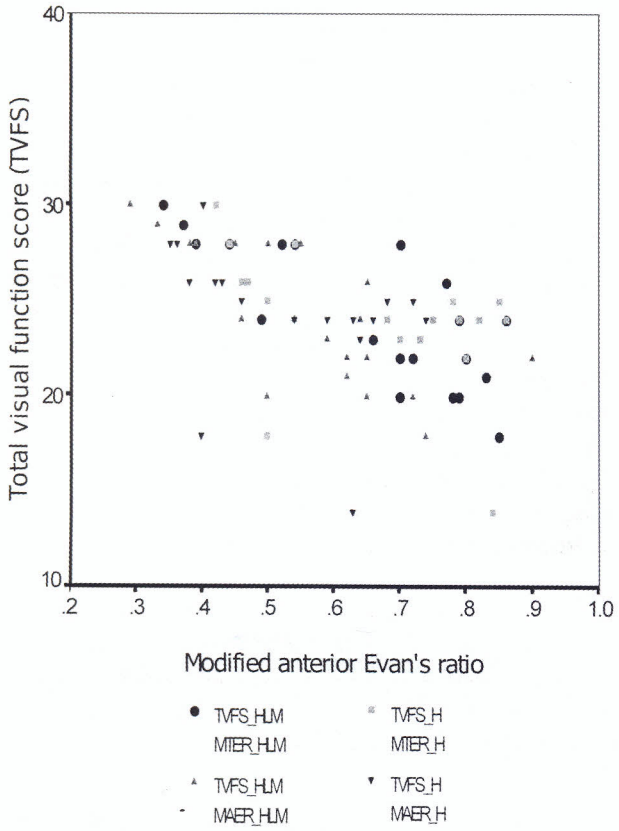

Figure 1 Scatter diagram illustrating the correlation, in the control group between total visual function score (TVFS_H) and modified anterior Evans' ratio (MAER_H, ▼), (TVFS_H) and modified trigonal Evans' ratio (MTER_H, ■); and in the myelomeningocoele group (HLM) between total visual function score (TVFS_HLM) and modified anterior Evans' ratio (MAER_HLM, ▲) and TVFS_HLM and modified trigonal Evans' ratio (MTER_HLM, ●).

assessment of pupillary size and reaction, the light source was a Finoff transluminator connected to a battery powered Welch–Allyn handle. The optic discs were finally examined after dilating the pupils with Tropicamide (0.5%). We did not have the facilities for electrophysiological evaluation of vision. To eliminate interobserver error, the same examiner evaluated all the patients for visual function. Each eye was scored a maximum of 15 and a minimum of 5 points. The total visual function score (TVFS) is the sum of the scores for both eyes. The scale has a range of 10–30 points. The scores for the two groups were analysed with SPSS for Windows version 7.0, using the Student's *t*-test for unpaired comparison. A *P* value less than 0.05 was taken as significant.

Table 2 Visual Function Scale (VFS)

Visual function	*Score*
Visual fixation and tracking	
Immediate	3
Slow	2
Absent	1
Pupillary size and reaction	
Size	
<4 mm	2
≥ 4 mm	1
Reaction	
Brisk	3
Sluggish	2
None	1
Optic disc pallor	
Severity	
Nil	3
Moderate	2
Severe	1
Extent	
<25% of disc surface	4
25–50% of disc surface	3
50–75% of disc surface	2
>75% of disc surface	1

The visual function score for each eye is the sum of the scores for all parameters.
The total visual function score (TVFS) for the patient is the sum of the scores for both eyes.
Minimum score = 10.
Maximum score = 30.

Results

Age and sex distribution (Table 1)

In the control group, the mean age at presentation was 3.4 months with a range of 0.25–8 months. There were 11 males and nine females. In the HLM group, there were 12 males and eight females. The mean age at presentation was 3.4 with a range of 0.5–15 months. The mean (SD) age, 3.4 (3.6) months, of patients in the HLM group, did not differ significantly from that of the control group (3.4 (3.8) months, $P = 0.97$).

Ventricular size and total visual function (Tables 1 and 2; Figure 1)

The mean (SD) values for the mAER were 0.52 (0.13) and 0.56 (0.15) for the control and HLM groups respectively, while the corresponding values for the mTER were 0.66 (0.16) and 0.65 (0.17) respectively. The two ratios did not differ significantly between the two groups ($P = 0.82$ and 0.87 respectively).

The mean TVFS (SD) in the control group was 24.2 (3.5) with a range of 14–30. The mean TVFS (SD) in the HLM group was 24.3 (3.63) with a range of 18–30.

There was no significant difference between the mean values for both groups ($t = 0.094$, $P = 0.93$).

In the control group, the anterior and trigonal Evans' ratios were inversely correlated with the total visual function scores. However, only the correlation between the trigonal ratio and the TVFS was statistically significant. The correlation coefficient (r) and the P values for the anterior Evans' ratio were -0.40 and 0.08 respectively (Figure 1), while the corresponding values for the trigonal Evans' ratio were -0.47 and 0.04 respectively (Figure 1).

In the HLM group, both anterior and trigonal Evans' ratios were significantly inversely correlated with the total visual function score. The correlation coefficient (r) and the P value for the anterior ratio were -0.75 and 0.01 respectively (Figure 1). The corresponding values for the trigonal ratio were -0.78 and 0.01, respectively (Figure 1).

The head was large at birth in 15 of the dysraphic infants. In the remaining five, the heads enlarged within 2–4 months of birth.

Discussion

Recently, we showed that ventricular size (measured at the levels of the anterior horn and collateral trigone) correlates inversely with visual function in hydrocephalic children.[6] In the present investigation, we have confirmed this relationship by demonstrating that visual function is impaired in direct proportion to the degree of ventricular enlargement in both control and HLM groups of patients. These results suggest a structural or functional damage to the visual pathways due to ventricular distension.

Hydrocephalus occurs frequently in children with myelomeningocole[3] in whom it predisposes to an unfavourable intellectual outcome, if untreated.[4]

In the present study, the age and sex distributions of the control and HLM groups were similar. Furthermore, the ventricular dimensions at the anterior and posterior portions of the lateral ventricular were closely matched in both groups. There was no significant difference in visual status as measured by the total visual function score. Indeed, the relationships between ventricular distension and visual function score are similar in the two groups. This suggests that hydrocephalus is the major factor in the genesis of cerebral visual impairment in myelodysplastic infants. Whereas multiple brain malformations involving the visual pathways may occur in these patients, it appears that the important factor for visual morbidity is ventricular dilatation. Infants with generalized lesions of the brain are at a high risk of visual impairment, even if visual acuity is unaffected.[7,8] Furthermore, in patients with cerebral visual impairment secondary to non-focal cerebral lesions, ultrasonographic appearance of the brain, visual, motor and cognitive functions are closely correlated.[9–11] and visual function can be an important predictor of cognitive development. The development of perceptual and motor skills also depends on visual function.[12]

There is therefore, a clear implication for the management strategies for improving the quality of life of these patients. We propose that early detection (including prenatal) and monitoring of ventricular enlargement by the best available neuroimaging technique and prompt treatment will contribute substantially to the preservation of vision and enhance the learning capacity and intellectual development of affected children. Serial postnatal assessment of visual function is of particular value in this regard.

The benefit of prenatal and postnatal serial ultrasonographic screening to detect ventriculomegaly before the onset of obvious head enlargement cannot be over-emphasized. Even in our earliest presenters (2 weeks old), the head circumference already exceeded the 97th percentile and the visual function score was 22 (maximum of 30). Nevertheless, head circumference measurement remains a useful tool for follow-up clinical evaluation of these patients since it shows a strong inverse correlation to visual function score.[6]

The visual system in children is relatively immature and sensitive to insults.[13] Furthermore, the acquisition of several aspects of visual function in children is age-dependent.[14] Given the intimate relationship between the visual pathways and the ventricular system, both extant vision and eventual visual function are predictably jeopardized by ventricular dilatation. The evaluation of visual function in infants remains difficult despite the availability of tests designed to circumvent lack of cooperation and verbal ability.[10,11] It is therefore customary to assess multiple parameters of visual function in studies in this age group.[11,14] In accordance with this principle we have assessed visual function by evaluating visual fixation and tracking, pupillary size and reaction, and assessment of the optic disc and integrated the results into a numerical scoring scale for ease of analysis and interpretation. Evidently, the composition of the parameters in the scoring system emphasizes optic nerve damage and may underestimate the contribution of cortical lesions to visual impairment. A number of studies have addressed several aspects of vision in patients with myelomeningocoele and have shown that vision is compromised frequently and in diverse ways. These include visuoperceptual disturbance,[15] strabismus and amblyopia, papilloedema, impaired visual fields and optic atrophy.[16] In these studies, the independent effect

of myelomeningocoele was not clearly addressed and separated from that of hydrocephalus which was frequently present in the subjects. However, there are reports[17,18] of patients with myelomeningocoele and shunted hydrocephalus who suffer visual deterioration in association with shunt malfunction, and recover following shunt revision. Our results provide evidence in support of the notion that in these patients, the major factor in visual morbidity is the hydrocephalic process rather than the multi-focal brain changes of spinal dysraphism.

In conclusion, we have confirmed that visual function score varies in inverse relationship to ventricular size in hydrocephalic infants. We have also demonstrated that this relationship is faithfully retained in those of them with lumbar myelomeningocoele. Furthermore, visual function, as measured by the total visual function score did not significantly differ between the two groups of hydrocephalic children, suggesting that in this dysraphic condition, the major determinant of visual morbidity is the extent of ventricular distension. These findings underscore the need for careful evaluation and monitoring of vision in all hydrocephalic patients and in particular those who have the additional disability arising from a dysraphic state.

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