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24

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The relationship between ventricular size and visual function in children with hydrocephalus

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Summary

We examined the relationship between ventricular size and visual function in 50 children (36 males and 14 females) with hydrocephalus. The third and lateral ventricular sizes and the visual function scores did not significantly differ between the sexes. Ventricular enlargement was most pronounced at the trigones and least at the level of the foraminae of Monro. The visual function score correlated inversely with the size of the lateral ventricle measured at the levels of the anterior horn and the trigone and expressed as coronal diameter and "Modified" Evans' ratio ($r = 0.49$; $P = 0.001$ and $r = -0.38$, $P = 0.01$ for the anterior horn; $r = 0.48$, $P = 0.001$ and $r = -0.35$, $P = 0.001$ for the trigone). The size of the third ventricle did not correlate with visual function score. A "modified" anterior Evans ratio of 0.60 and a trigonal Evans ratio of 0.73 were associated with very low visual function score. Furthermore, there was significant inverse correlation between occipitofrontal circumference (OFC) and visual function ($r = -0.6379$, $P = 0.001$), but OFC was not valuable for predicting visual function before the onset of head enlargement.

Keywords: *Hydrocephalus; vision; ventricular size; visual function score; children.*

Résumé

Nous avons examinè la relation entre la taille ventriculaire et lan fonction visuelle chez 50 enfants (36 males et 14 femelles) ayant une hydrocephalus. Les tailles des troisiemes et des ventricules lateraux et la fonction visuellen n'on pas differè de manière significative dans les 2 senes.

L'elargisement ventriculaire n'a pasètè prononcè aux triefones visuelle a ètè inversement proportinelle à la tao;;e des vemtroci;es ;ateraix ,esires, au nivaux de la coerneèeanterieure et du trigone. Les valeurs ont ètèe exprimes en fonction du diametre coronal et du ratio Modifiè d'Evans (r = 0,49;)P=0, 001 et r = 0.38; P=0,01) pour la cornes anterioure; r = -0;48, p=0,001 et r=-0,35, p=0,001 pout la trigone). La taille du troisieme ventricule n'a pas correlè avec la fonction visuelle.Une modification du ration anterieur d'Evans (ration de 0,60) et du trigonal d'Evans (ration de 0,73) ont ètè associès 'a une faible fonctin visuelle.

De plus, il y a eu une correlation inversement significative entre la circonfference occipito-frontale (OFC) et la fonctin visuelle (r= -0,6379, P=0,001) mais l'OFC n'a pas ètVe valable pour la prediction de la fonction visuelle, avant de debut de l'elargisesement de la tète.

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Introduction

Hydrocephalus, the commonest congenital malformationof the central nervous system (CNS) seen in Ibadan constitutes 40.6% of all CNS anomalies [1]. Fifty-three percent of affected children present in advanced stages with gross head enlargement and neurological disability including seizures and loss of vision [2]. Visual loss is commonly attributed to optic atrophy resulting from a variety of causes such as papiloedema, chiasmal traction, papilomacular bundle atrophy, developmental abnormalities and third ventricular dilatation acting either singly or in combination [3,4].

One of the consequences of hydrocephalus is that it impairs tissue metabolism by tampering both globally and focally with the cerebral microcirculation [5]. Cortical maturation and myelination, two developmental pre requisites for normal neurological function, are highly dependent on a normal circulation. It is therefore not surprising that a variety of neurobehavioural deficits accompany hydrocephalus. An inverse relationship between ventricular size and intellectual development has been described in infants with myelomenigocoee [6]. Furthermore, the neurologic outcome following shunting procedures in advanced hydrocephalus is poor [2].

The precise relationship between ventricular dilatation and visual deterioration in hydrocephalic children is not known. The purpose of this study is to determine first, whether there are any relationship between the extent to which visual function is impaired, and the degree of ventricular dilatation; and , second, to predict the ventricular size below which shunting procedures will produce the best outcome for vision.

Patients and methods

This was a prospective study of fifty consecutive hydrocephalic young children aged 18 months or less, evaluated at our Neurosurgical Clinic between January 1995 and June 1996. Only those who met the study criteria were selected. Infants with closed fontanelles, associated encephalocoele or visual impairment traceable to an existing eye disease, were excluded.

The initial evaluation consisted of history and examination to obtain a clinical diagnosis. This was followed by trnafontanelle ultrasound scanning using the ACCUOUSON scanner (ATL, Advanced Technology Laboratories Ltd. USA) a B-Mode real-time scan with gray scale imaging. Each patient was 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 [6]. An aqueous gel was used in all instances for good skull contact and the scanning was done through the anterior fontanelle. Three examiners scanned all patients. The value for each patient was taken as the mean of the three values obtained. Coronal widths of the third and lateral ventricles were measured. Lateral ventricular width was measured at three levels, i.e., anterior horn, Foramen or Monro and the trigones. Lateral ventricular size was also assessed at the levels of anterior horns and trigones employing the Evans' ratio method [7,8] (described in fig. 1). modified for ultrasonography. The original description of the method was based on computerized tomography [6,8].

Visual function was scored by assessing the following parameters;

a) Visual acuity, expressed by visual tracking and fixation, and optokinetic eye movements.
b) Pupillary size and reaction to direct light; and
c) The severity and extent of optic disc pallor.

Each eye was examined in turn, the other eye having been covered with an occlusive patch. The optic disc was examined after pupillary dilation with 0.5% tropicamide eye drops. Each parameter was scored as shown in table 2. The visual function score for each eye ranged between a minimum of 5 and a maximum of 15 units. The patient's total visual function score (TVFS) was the sum of the scores for both eyes. The range of scores was 10 – 30. The relationship between ventricular size and visual function score was assessed by correlation analysis and standard test of significance. A *P* value less than 0.05 was taken as significant.

Results

Patients' characteristics

Of the 50 infants studied, 36 were males and 14 were females. At presentation, they ranged in age from one day to eighteen months with a mean of 4.5 ± 2.6 months. Table 1 summarizes their age and sex distribution. More than half of these infants (54%) presented before or at three months of age. Progressive head enlargements, the commonest mode of presentation , occurred in 39 (78%) patients. The remaining infants presented primarily with lumbar myelomeningocoele. In this latter group, hydrocephalus was manifest at presentation or was detected by ultrasonography.

Table 1: Age and sex distribution of hydrocephalic children

Age range (mo)*	Males (%)	Females (%)	Total (%)
0 – 3.0	5 (10)	7 (14)	27 (54)
3.1 – 6.0	5 (10)	4 (8)	9 (18)
6.1 – 9.0	7 (14)	3 (6)	10 (20)
9.1 – 12.0	2 (4)	0 (0)	2 (4)
12.1 – 15.0	1 (2)	0 (0)	1 (2)
15.1 – 18.0	1 (2)	0 (0)	1 (2)
Total	36 (72)	14 (28)	50 (100)

Mnths. = months

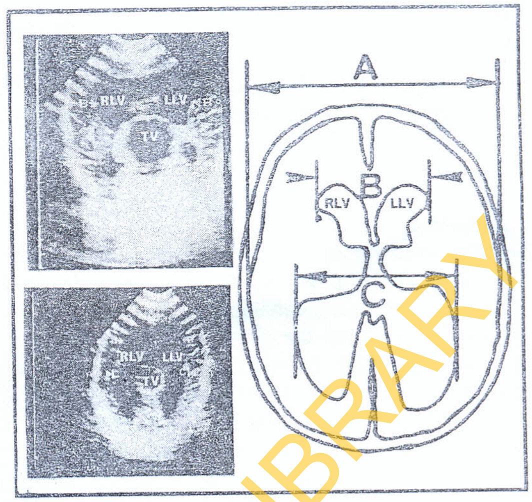

Fig. 1: The lateral and third ventricles as visualised by a coronal Transfontanellar ultrasound scan; and a line diagram of the horizontal section of the brain (showing the lateral and third ventricles) as used for the estimation of Evan's ratio. A = greatest transverse diameter of the cerebrum; B = greatest distance between the anterior horns; C = greatest distance between the trigones; RLV = right lateral ventricle; LLV = left lateral ventricle. Anterior Evan's ratio = B/A; trigonal Evan's ratio = C/A.

Total visual function score (TVFS)

Each of the patient's eyes was scored for visual acuity, pupillary size and reaction to direct light as well as optic disc paor (Table 2). The sum of the scores for both eyes is taken as the total visual function score (TVFS) for the child. The mean ± SD TVFS for the entire sample was 23.6 ± 5.7. There was no significant difference between the mean TVFS of males, 23.6 ± 6.0 and females 24.6 ± 4.7, (*P* > 0.05).

Table 2: Visual function scale (TVFS)

Visual function	Score
Visual fixation and tracking	
Immediate	3
Slow	2
Absent	1
Pupillary size and reaction	
Size less than 4 mm	2
Size 4 mm and above	1
Brisk reaction	3
Sluggish reaction	2
No reaction	1
Optic disk pallor	
Nil	3
Moderate	2
Severe	1
< 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 is 10
Maximum score is 30.

Ventricular size (transverse diameters) and visual function

Mean lateral ventricular widths (i.e. transverse diameters) were measured at the level of foramen of Monro, the anterior horns and the trigones and their relationship to visual function was investigated. A significant inverse correlation was found between TVFS and the ventricular diameter at the anterior horns as well as the trigones (r = 0.49 at P = 0.0003 and r = -0.48 at P = 0.0004, figures 2a and 2d, respectively).

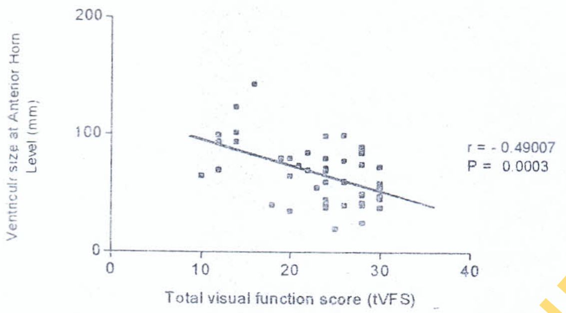

Fig. 2a: Scatter diagram to illustrate the correlation between TVFS and the transverse diameter of the lateral ventricle at the level of the anterior horn.

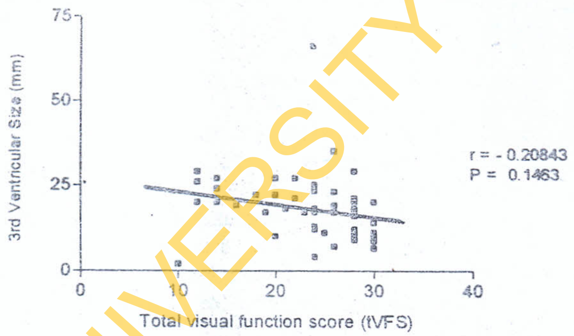

Fig. 2b: Scatter diagram to illustrate the correlation between TVFS and the size of the third ventricle.

There were a poor correlation between TVFS on the one hand and the third ventricular width and the size of the lateral ventricle at the level of foramen of Monro (LVZ) on the other (r = 0.2084 at P = 0.1463 and r = -0.1411 at P = 0.3283; Fig. 2b and 2c respectively).

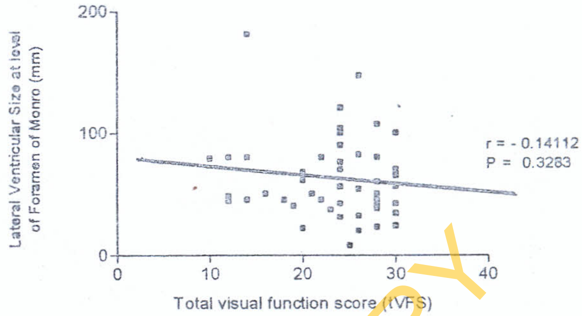

Fig. 2c: Scatter diagram to illustrate the correlation between TVFS and the transverse diameter of the lateral ventricle at the level of the foramen of Monro.

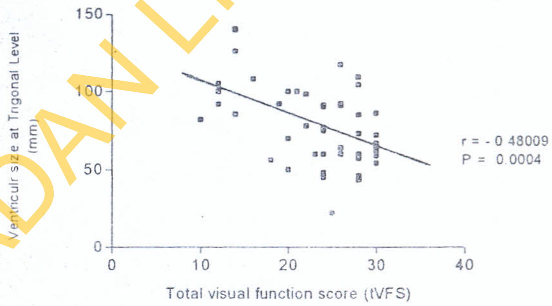

Fig. 2d: Scatter diagram to illustrate the correlation between TVFS and the transverse diameter of the lateral ventricle at the level of the trigone.

Ventricular size (modified Evans' ratio) and visual function

a. Modified Anterior Evans' Ratio (mAER) and Visual Function
The mean value of the mAER obtained for the sample was 0.5968 ± 0.1775. The mean value for males and females were 0.6036 ± 0.1858 and 0.5664 ± 0.1561, respectively. There was no significant sex difference (P > 0.05). The total visual function score (TVFS) declined steadily with increasing mAER. These two parameters, TVFS and mAER, were significantly inversely correlated (r = -0.3893 and P = 0.0052, Fig. 3a)

b. Modified Trigonal Evans' Ratio (mTER) and Visual Function
The mean lateral ventricular size measured by Evans' ratio at the level of the trigones was 0.7402 ± 0.1697 for the entire sample, 0.7097 ± 0.1544 for males and 0.6900 ± 0.2101 for females. There was no significant sex difference (P > 0.05). However, there was a significant inverse correlation total visual function score between and mTER (r = -0.34631 and P = 0.0138; Fig. 3b)

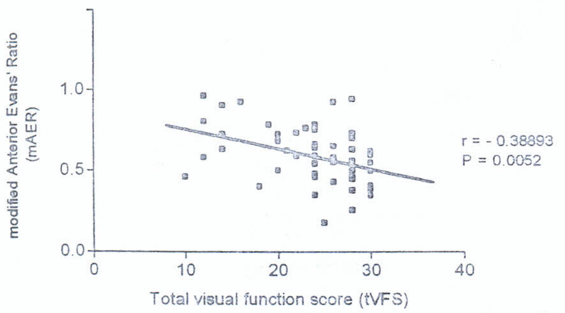

Fig. 3a: Scatter diagram to illustrate the correlation between TVFS and the modified anterior Evan's ratio.

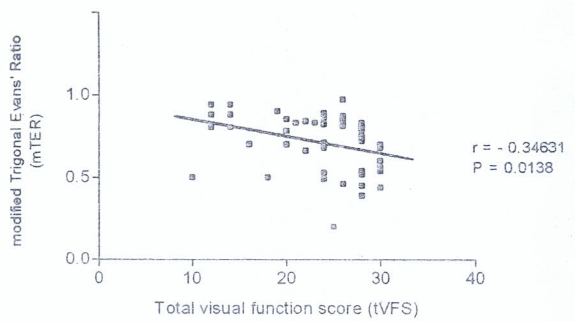

Fig. 3b: Scatter diagram to illustrate the correlation between TVFS and modified trigonal Evan's ratio.

Ventricular size and severity of visual impairement

Impairment of visual function was empirically categorized into severe, moderate and mild forms based on visual score ranges of 10 – 17, 18 – 26 and 27 – 30 units, respectively. Mean values of mAER and mTER for patients in each category was estimated.

Table 3 shows that 48% of the infants had visual impairement of moderate degree. Infants manifesting severe visual impairment comprised 16% of the group while a mild loss occurred in 36% of the sample. Table 3 also reveals that mean Evans' ratio decrease with decreasing severity of visual impaîrement and that a mean mAER greater than 0.60 and mTER higher than 0.73 appear to be predictive of imminent severe visual deterioration.

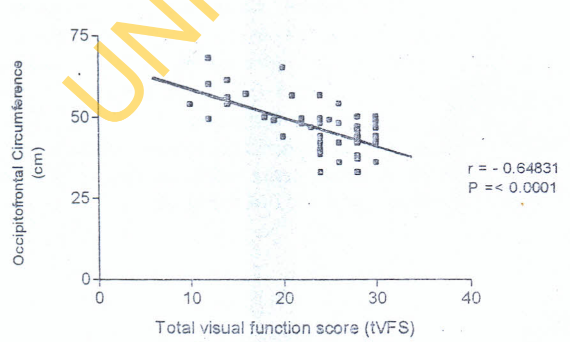

Fig. 3c: Scatter diagram to illustrate the correlation between TVFS and the occipito-frontal circumference.

Occipito-frontal circumference (OFC) and visual function

The maximum measurable occipito frontal circumference (OFC) was obtained using a tape measure in all infants at initial evaluation. Values varied between 36 cm and 68 cm, (mean = 47.06 ± 7.69 cm). OFC greater than 97th percentile for age and sex (macrocephaly) was obtained in 39 (78%) of the 50 infants. In the remaining patients (22%), ventriculomegaly was detected by transfontanelle ultrasound performed because of pre-existing spinal dyspraphism. A high inverse correlation was drawn between (OFC) and total visual function score among these infants (Fig. 3c). Comparing other parameters related to visual function in this work, OFC correlated best with visual function ($r = -0.65$ at $P = 0.001$).

Discussion

In cases of hydrocaphalus, ventricular dilation does not merely represent a progressive accumulation of cerebrospinal fluid due to inbalance between production and absorption, but the absorptive defect sets off a complex interplay between altered intracranial pressure and the biochmeical properties of the brain itself. Whether the ventricular dilatation exerts direct influence on cerebral functions has been the subject of numerous previous investigations. The effect of ventricular dilatation on visual function is the subject of the present study.

The study sample consisted of 36 males and 14 females. The male predominance (ratio 2.57:1) confirmed earlier reports of higher incidence of congenital hydrocephalus among males [12,18], although in Binitie's series from northern Nigeria, the sexes were equally affected [13]. Males are known to be at a greater risk than females of developing congenital hydrocephalus [9] and in 2 out of 3 patients with congenital hydrocephalus a lesion obstructing the aqueduct can be demonstrated [14], suggesting that aqueductal stenosis is common among males.

Our results corroborate this finding. Sixty-five percent of the 20 children who had definitive ultrasound diagnosis demonstrated features of aqueductal obstruction, and 85% of these were males. It has also been suggested that in hydrocephalus, males are prone to greater ventricular dilatation than females [15]. Our results do not support this assertion. It is not surprising that ventricular dilatation was most pronounced at the trigones.

Anatomically, the collateral trigones constitute the largest and probably the most distensible portion of the lateral ventricular system [16].

A semiquantitative system of assessment of visual function in pre-literation hydrocephalic infants was pre-literate designed for this study. This system grades the three visual parameters: visual acuity, optic disc palor and pupillary size and reaction to direct light and scores each eye 5-15 units. It's repeatability, reproducibility, rate of agreement by two observes as well as limitations have been subjected to statistical tests (unpublished data).

When total visual function scores obtained using this system were correlated with ventricular size measured at two levels (anterior horns and trigones) using two techniques (coronal diameters and modified Evans' ratio), significant inverse correlations were obtained as follows, in descending order of correlation (r): anteriror coronal width r = -0.4900; trigonal coronal width r = -0.4801; modified anterior Evans' ratio (mAER) r = -0.3889; and modified trigonal Evans' ratio (mTER) r = -0.3463.

The third ventricular size did not correlate with visual function score, although correlations between visual function score and Evans' ratio are lower than similar values for coronal diameters, the former is still considered a better indirect predictor of cerebral function than the latter because it incorporates both ventricular and brain parachyma sizes and reflects not only the extent of ventricular enlargement but also that of cerebral compression when present.

The correlation obtained between ventricular sizes and visual function scores cannot be explained entirely on the basis of anatomic proximity. There was no correlation between the size of the third ventricle (a close relation of the optic chiasma) and visual function. There was a good inverse correlation of visual function score with the anterior horns which maintain no direct contact with the visual pathways. It is easier to explain trigonal correlation based on proximity of the trigones to the optic radiation and close relation between its extension into adjacent horns and the lateral geniculate bodies as well as the calcarine cortex [16].

Clearly, other factors are involved. Cortical histology, cerebral fluid and electrolyte content, intraventircular pressure, cerebral blood flow, oxygen and glucose metabolism over extensive areas of the brain including the visual cortices, and cerebral vascular architecture are altered in hydrocephalus. Their relationship to visual function await further investigation [5, 17, 18, 19].

We graded group means of Evans' ratio and compared them to visual loss graded as severe, moderate and mild. We found mAER greater than 0.60 mTER greater than 0.73 to be indicative of severe visual impairment. It is suggested that infants with such or greater ratios be shunted promptly even when the head size is normal. This is of particular importance in preterm infants and infants with myelomeningocoele undergoing serial sonographic screening for follow up of hydrocephalus. It is expected that such prompt intervention will produce a good outcome for visual function.

Excessive enlargement of the head, especially an excessive rate of cranial enlargement, is usually the first and most reliable feature of hydrocephalus in infancy [20]. An occipito-frontal circumference (OFC) greater than 97 percentile for age and sex is highly suggestive of hydrocephalus [21]. This range of OFC was found in 78% of our patients, the largest being 68 cm and smallest 33 cm. Visual scoring revealed a high inverse correlation r = 0.6379, P = 0.001) between OFC and visual function. Compared with other measured parameters employed in this work, OFC correlated best with visual function.

However, since increase in OFC (i.e., onset of head enlargement) is a late event for hydrocephalus, OFC is considered to be of limited value for predicting early visual deterioration. Although Milhorat (1979) expressed the view that no definite correlation exists between the degree of ventriculomegaly, the size of the head and the eventual development of mental and motor skills [20], our study suggests that OFC is a good but late predictor of visual function after the onset of head enlargement. OFC is the simplest measurement that predicts visual status even at initial clinical assessment.

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