

## Ocular features seen in children with hydrocephalus at University of Nigeria Teaching Hospital Enugu: a case series

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### ABSTRACT

**Background:** Hydrocephalus has been known since antiquity. It has been defined as an increase in size of the ventricular system of the brain, resulting from raised cerebrospinal fluid volume.

**Objective:** The purpose of this study is to determine the various ocular disorders that may be associated with hydrocephalus in children seen in the University of Nigeria Teaching Hospital (UNTH) Enugu.

**Patients and Methods:** This study was carried out on 15 patients attending the neurosurgical outpatient clinic of the University of Nigeria Teaching Hospital Enugu. All patients who met the criteria for inclusion in the study of hydrocephalus, aged less than two years and attended the neurosurgical outpatient clinic between January and December 1998 were included. The diagnosis of hydrocephalus was made based on the history, clinical examination and brain ultrasonography. The data was analysed with a scientific calculator Casio FX-82 Lb and Epi info version 6 software.

**Results:** Fifteen children with un-operated hydrocephalus confirmed by transfontanelle ultrasonography were examined at presentation for ocular complications of hydrocephalus. The mean age at presentation was 29.8 weeks. The peak age group of presentation was 11-20 weeks. The male/female ratio was 1:1. Ocular complications of hydrocephalus included setting sun appearance, nystagmus, optic disc palor and hyperaemia.

**Conclusion:** The Ophthalmologist has a well established role to play in assisting the paediatric neurosurgeon in the long term management of children with hydrocephalus if the risk of blindness is not to be added to their other physical disabilities.

**Keywords:** Children, features, hydrocephalus, ocular

### INTRODUCTION

Hydrocephalus has been defined as an increase in the size of the ventricular system of the brain, resulting from raised cerebrospinal fluid pressure.<sup>1</sup> It may be caused by obstruction to cerebrospinal fluid (CSF) circulation, over-production or failure of absorption of CSF. Hydrocephalus has been studied since the time of Hippocrates<sup>1</sup> but association of hydrocephalus with the eye and central nervous system (CNS) anomalies was first noted by Walker<sup>2</sup> in 1942. Walker described a case of early hydrocephalus and ocular complications. Third ventricular dilatation may stretch the optic chiasma and tracts leading to gross visual impairment, bitemporal visual field defects and hypothalamic disorders.<sup>3</sup> Clinical features which may present with hydrocephalus

include massive head size, with enlarged fontanelle and palpably separated sutures, thinned scalp, prominent scalp veins, eyelid retraction, strabismus, spastic paraplegia, triangular appearance of the face and setting sun appearance of the eyes (Fig 1). Visual loss in children is commonly due to optic atrophy.

It is a common observation that ocular movements may be abnormal in patients with uncontrolled hydrocephalus.<sup>4</sup> Strabismus is a frequent feature in these children. It is most commonly an incomitant convergent squint of the alternating type due to sixth nerve paresis<sup>1,5</sup> due to a rise in intracranial pressure. An increased angle of squint is repeatedly seen as a sign of raised intracranial pressure<sup>5</sup>, and is sometimes the first unequivocal sign of raised pressure.<sup>6</sup> Fully accommodative squints are uncommon<sup>6</sup> but refractive errors could be

found more commonly in hydrocephalic patients.<sup>7</sup> But, most squints in these children appear to be based on lateral rectus palsy rather than hypermetropia.

Nystagmus in hydrocephalic children has been reported in the literature and includes pendular, jerk, micronystagmus and musculoparetic nystagmus. According to Goddard<sup>8</sup>, hydrocephalic patients are found with horizontal and rotary types of nystagmus.

The midbrain eye signs in hydrocephalus include upgaze palsy, upbeat nystagmus, lid retraction, convergence retraction nystagmus, forced down gaze or setting sun appearance, unilateral and bilateral fourth nerve palsies and pupillary light near dissociation.<sup>5</sup>

Until recently, the outlook for life in these children has been extremely poor because the progressive hydrocephalus was unlikely to be spontaneously arrested so that surviving infants were mentally retarded.<sup>3</sup> The rewards of expert and energetic treatment are great for although many of the afflicted children require special residential care and education, the majority are of normal intelligence.

It is important to study ocular complications of hydrocephalus in children so as to be able to make prompt and appropriate diagnosis and offer early treatment which will improve the quality of life of the patient. The ophthalmologist will occasionally be of vital importance to the management of these children by diagnosing for the first time blockage of cerebrospinal fluid flow by noting increasing squint or papilloedema. This is a study of the pattern of ocular complications of hydrocephalus in children seen at University of Nigeria Teaching Hospital (UNTH) Enugu.

#### **MATERIALS AND METHODS**

This study was carried out on patients attending the neurosurgical outpatient clinic of the University of Nigeria Teaching Hospital (UNTH), Enugu, after obtaining the approval of the Ethical Committee of the hospital.

All patients with hydrocephalus who were aged less than two years who attended the neurosurgical outpatient clinic over a period

of one year from January to December 1998 were included. All patients above two years of age and all patients whose anterior fontanelle had fused and who presented no obvious portal for ultrasound study were excluded.

The patients were examined after informed consent was obtained from their parents. All the patients were examined by the same person. The name, age and sex were obtained. The diagnosis of hydrocephalus was based on the history, clinical examination, and brain ultrasonography.

The measuring tape was used in measuring the occipitofrontal head circumference by passing the tape over the most prominent part of the occiput and just above the supraorbital ridges. The pen torch was used to assess the pupillary light response, following of light and the optical blink reflex. The surgical loupe was used to assess the eyelids, the size, position and symmetry of both eyes, the presence of abnormal eye movements such as nystagmus and strabismus, the conjunctiva, the clarity of the cornea, anterior chamber depth and the iris.

The pupils were dilated by the use of 0.5% Tropicamide administered two times, thirty minutes before examination. The fundi were examined with the direct ophthalmoscope with repeated efforts as an out patient without anaesthesia or sedatives. The indirect ophthalmoscope was used in cases without nystagmus. Lenticular opacity, vitreous haziness, optic nerve head abnormalities like coloboma, hypoplasia, pallor or papilloedema were searched for. Retinal abnormalities such as detachment or dysplasia were also looked for.

Finally, thorough general examination of the patient was performed for other abnormalities. The patients were sent for brain ultrasonography. The ultrasound was performed by the same person in the Radiology Department of UNTH Enugu. This was done by a transfontanelle real time B-Scan with a sector scan probe at a frequency of 7megaHertz. Simple data analysis was done using a scientific calculator Casio FX-82Lb. Relevant statistical test of significance

(Chi-square) was done using the Epi info version 6 software.

## RESULTS

Fifteen patients met the criteria for inclusion in the study. The age distribution at presentation was from birth to 76 weeks. The mean age at presentation was 29.8 weeks. The peak age group at presentation was 11-20 weeks (Table 1). The age at onset as determined from patients history was from birth to 40 weeks. The male/female ratio was 1:1 (Table 1). The difference in male/female ratio was not statistically significant. ( $\chi^2 = 8.304$ ,  $df = 6$ ,  $p = 0.2167$ ).

In all the patients, the occipito-frontal head circumference exceeded the 95<sup>th</sup> percentile. Visual assessment showed that 6(40%) could follow light while 9(60%) had menace reflex.

Eye movement abnormalities included nystagmus which occurred in 13 patients with 9(60%) having horizontal pendular nystagmus, 3(20%) with upbeat nystagmus, 1(6.7%) with horizontal jerk nystagmus. Two (13.3%) had no nystagmus, but all the patients had setting sun appearance. Dilated fundus examination in all the patients showed optic disc pallor in 5(33.3%), normal disc in 2(13.3%) and hyperaemic disc in 8(53.4%).

## DISCUSSION

**Setting Sun Appearance:** This was the commonest disorder found in all the cases. Setting sun appearance is a classic sign of hydrocephalus and kernicterus but may also occur in premature babies and transiently in normal full term babies.<sup>9</sup> The setting sun appearance is the result of a pressure effect on the brain stem in the region of the posterior end of the third ventricle.<sup>9</sup> Christoff<sup>4</sup> in 1974 concluded that upward gaze paralysis occurs mainly with bilateral lesions involving the pretectum, posterior commissure or dorsal midbrain tegmentum.

In Gaston's study,<sup>9</sup> 83% of the children had setting sun appearance in a six-year study and in Olurin-Aina's study,<sup>10</sup> 76% of the children had setting sun appearance. In this study, 100% of the children had it as well, and this

100% may be related to grossly dilated ventricles in all the children with the occipito-frontal circumference exceeding the 95<sup>th</sup> percentile.

**Nystagmus:** Hydrocephalic patients are often found to have disturbances of eye movement.<sup>11</sup> Nystagmus from hydrocephalus results from brain stem disorder.<sup>12</sup> In this study, 13(86.7%) children had nystagmus while 2(13.3%) had no nystagmus. The types of nystagmus observed in this series included horizontal pendular nystagmus in 9(60%) children, upbeat nystagmus 3(20%), horizontal Jerk nystagmus 1(6.7%) while 2(13.3%) did not have any form of nystagmus.

Gaston<sup>9</sup> observed horizontal pendular nystagmus in 10(3%) children, correlating the report of Rabinowicz (4%).<sup>5</sup> The higher frequency 9(60%) of horizontal pendular nystagmus observed in this study is probably due to the gross lesions seen in all the children.

Olurin-Aina<sup>10</sup> observed 24% with horizontal nystagmus, 2% upbeat, 6% downbeat, 2% gaze evoked and 1% rotary nystagmus. The lower figure may be explained by the fact that her study included patients with spina bifida, as well.

**Visual Function:** The diagnosis of visual loss can be a difficult task in the preverbal child.<sup>3</sup> Primary optic atrophy in congenital hydrocephalus is well known<sup>13</sup> and commonly is a cause of visual loss.<sup>3</sup> Dilated third ventricle in acquired obstructive hydrocephalus can occasionally cause visual field defects.<sup>13</sup>

In this study, visual assessment showed that 6(40%) children could follow light while 9(60%) had menace reflex. The ability to fixate, follow and respond to visual stimulus is evidence of an intact visual system. As in Olurin-Aina's<sup>10</sup> series, this study could not determine any specific visual function abnormality. In Gaston's<sup>9</sup> series, only 86(27%) had normal visual function. The visual assessment was possible because Gaston examined children of up to eighteen

years. Mankinen-Heikkinen and Eila<sup>11</sup> reported that the most common neuro-ophthamological complications of hydrocephalus were reduced visual acuity, pupillary abnormalities and optic atrophy. Rabinowicz<sup>5</sup> also found defective vision to be a particular difficulty encountered by hydrocephalic children.

**Papilloedema/Disc Hyperaemia:** There was no case of established papilloedema in this study. Eight (53.4%) children showed disc hyperaemia. It is widely believed that congenital hydrocephalus is only very rarely associated with papilloedema.<sup>14</sup> Goddard<sup>8</sup> attributed the rarity of papilloedema in congenital hydrocephalus to the ease with which the infant's skull expands. This would indicate that the generalized intracranial pressure in congenital hydrocephalus is seldom raised.

In Ghose's<sup>14</sup> study, papilloedema was seen in 24 out of 165 cases (14.5%) of congenital hydrocephalus. Harcourt<sup>5</sup> also reported two cases of papilloedema. The cases in which papilloedema was found was hydrocephalus associated with meningocele.

Papilloedema in older patients is the most common cause of visual loss in patients with hydrocephalus<sup>15</sup>. Mankinen-Heikkinen and Eila<sup>11</sup> stated that it is a general opinion that congenital hydrocephalus is very seldom connected with papilloedema except as a relatively late phenomenon when compensation fails. The absence of papilloedema in this series supports this view.

**Disc Pallor:** Optic disc pallor was found in 5(33%) of cases and 2(13.3%) had normal pink disc. In Ghose's study<sup>14</sup>, the optic disc was normal in the majority of cases (56.5%) and the most common change was primary optic atrophy (17%) followed by temporal pallor (11.5%). Mankinen-Heikkinen and Eila<sup>11</sup> cited Walsh's study in which he examined 25 hydrocephalic children and found them to have pale optic disc or optic atrophy.

According to Kirkham<sup>16</sup>, one cause of optic atrophy is stretching of the chiasma, or optic nerves by the expanded third brain ventricle. Primary optic atrophy in congenital hydrocephalus is well known.<sup>13</sup> A rise in intracranial pressure may cause papilloedema leading to secondary optic atrophy or dilatation of the third ventricle or a shift in the position of the brainstem giving rise to a primary optic atrophy from stretching of the optic nerves or optic chiasma. Visual loss in children with hydrocephalus is commonly due to optic atrophy<sup>3</sup>.

**Retinal Dysplasia:** No case of retinal dysplasia was seen. Descriptions have appeared in the literature of several patients with early hydrocephalus associated with ocular abnormalities especially retinal dysplasia, retinal detachment and severe developmental defects of the central nervous system not resulting from any recognizable cause.<sup>17</sup>

Various names have been proposed for this association including those of Walker's lissencephaly, encephalo-ophthalmic dysplasia, oculo-cerebral malformation syndrome, retinal dysplasia with hydrocephalus and HARD ± E Syndrome (Hydrocephalus, Agyria and Retinal Dysplasia with or without Encephalocele)<sup>17</sup>. Giles<sup>18</sup> in 1963 described advanced idiopathic hydrocephalus and concomitant marked chorioretinitis.

The lids were normal in all the cases in this study. There were no pupillary abnormalities, and no strabismus. The corneal sensations were also normal.

#### CONCLUSION

Setting sun appearance and nystagmus are the most common complications. This can be alleviated with surgery, for the preservation of the patient's sight.

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Table 1: Gender distribution of children with hydrocephalus

Age (weeks)	Male	Female	Total
0 – 10	1 (7%)	2 (13%)	3 (20%)
11 – 20	2 (13%)	2 (13%)	4 (26%)
21 – 30	2 (13%)	0 (0%)	2 (13%)
31 – 40	0 (0%)	1 (8%)	1 (8%)
41 – 50	0 (0%)	2 (13%)	2 (13%)
51 – 60	1 (7%)	0 (0%)	1 (7%)
Over 61	2 (13%)	0 (0%)	2 (13%)
<b>Total</b>	<b>8 (53%)</b>	<b>7 (47%)</b>	<b>15 (100%)</b>

Figure 1: Antero-posterior view of a child with hydrocephalus showing enlarged head, dilatedveins and setting sun appearance

