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Evidence of cognitive visual problems in children with hydrocephalus: a structured clinical history-taking strategy

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Originally appeared in in Developmental Medicine & Neurology 1999, 41: 298 -306.

Damage to the occipital cortex in children can result in many complex disorders of cognitive visual function. A series of clinical questions, developed from the specific problems of a cohort of children with cortical visual impairment, was asked of the parents of 200 children with no history of cerebral pathology, aged 5 to 12 years. One hundred and ninety-two parents gave reliable consistent responses. The results show a progressive improvement in performance with age, culminating in few 11 and 12-year olds having frequent problems, apart from 8% having frequent difficulty with orientation in new surroundings and 2% having problems with simultaneous perception tasks. The parents of-52 children (aged 5 to 17 years) with shunted hydrocephalus were then asked the same set of questions. Evidence of cognitive visual problems was identified in 27 of these children of whom 16 manifested multiple difficulties. The disabilities identified by our study comprised problems with: shape recognition, simultaneous perception, perception of movement, colour perception, orientation, object recognition, and face recognition. The range, nature, and combinations of these disorders are presented in this paper.

A large proportion of brain function is devoted to visual tasks, ranging from discerning the presence of an object to imagining three-dimensional imagery from different viewpoints. While much is understood about the basic aspects of this hierarchy of visual function, such as visual acuity, visual fields, and colour vision, the nature and development of higher visual function and its disorders are more enigmatic, particularly in the developing child.

Disorders of cognitive visual function in adults are known as visual agnosias (for reviews see Grusser and Landis 1991, Rizzo and Nawrot 1993, Milner and Goodale 1995). There are few studies concerning analogous disorders in children with cerebral pathology (Miller and Lovleen 1971, Rabinowicz 1974, Mancini et al. 1994, Dutton et al. 1996).

Detailed history taking from parents of children with occipital cerebral damage has revealed visual difficulties analogous to those described in adults (Ahmed and Dutton 1996, Dutton et al. (1996). These include problems with recognition, orientation, depth analysis, simultaneous perception, detection of movement, and difficulties recognizing and describing colour.

Classical descriptions of visual agnosias in adults comprise detailed investigation of patients with focal brain damage. Such an approach cannot easily be adopted for children with brain pathology for a number of reasons: impairment of overall cognitive function in children with brain damage can render objective evidence of additional specific cognitive visual problems difficult to obtain; brain growth and development influenced by appropriate interventional strategies may lead to recovery from, or circumvention of problems of analyzing the visual world, so that they are less evident or only manifest when visual tasks are more difficult; classical methods of assessment commonly require more time than is available for a young child with a short attention span and other time commitments; parents often describe sporadic problems of visual function which may not be detected by structured assessment.

Hydrocephalus is associated with many ophthalmic complications, the incidence, severity, and nature of which are well described (Goddard 1965, Lorber 1967, Harcourt 1968, Rabinowicz 1974, Humphrey et al. 1982, Ghose 1983, Arroyo et al. 1985, Gaston 1985, Corbett 1986, Cedzich et al. 1990). Biglan (1990) has reported on a series of 298 children with spina bifida followed up for 15 years. Of these, 278 had shunted hydrocephalus, and ophthalmic complications of varying severity were found in 80%.

Few reports describe visual problems due to cognitive dysfunction in these children (Miller and Lovleen 1971, Rabinowicz 1974), and to our knowledge no systematic studies have been carried out. Problems described include difficulty with figure-ground discrimination, shape recognition, and figure completion, and a tendency to reverse designs, letters, and shapes. Rabinowicz (1974) indicated that such defects are underestimated in children with hydrocephalus.

More recently, Donders and colleagues (1991) found subtle deficits of some complex visuospatial measures in children with hydrocephalus aged between

5 and 8 years. They felt that these findings were due to more than just ocular or fine motor dysfunction. Other authors have found a discrepancy between verbal and non-verbal cognitive skills (Fletcher et al. 1992b). In a MRI study by Fletcher and coworkers (1992a), it was concluded that non-verbal cognitive skill development requires intact white matter in both hemispheres.

For the present study we identified the principal visual problems previously seen in our series of 20 children with disorders of cognitive vision (Dutton et al. 1996). From this a standardized clinical history-taking strategy was devised and administered to 200 children without disorders of cognitive vision, aged between 5 and 12 years. The same strategy was applied to a group of 52 children with hydrocephalus in a similar age range.

Method

The clinical questions asked are as follows: what age is your child? Has your child ever worn glasses? Does your child have a squint? Is your child right- or left-handed?

In addition parents were asked to rate on a scale of 1 to 4 (1 being 'never' and 4 being 'always') how often their children experienced difficulties with recognizing people, shapes, and colours; whether they had trouble with finding their way around; and whether they had problems distinguishing objects from their background and from one another. Appendix 1 lists the questions which were asked. The first 20 of these questions considered the principal problems described by parents of children in our previous study (Dutton et al. 1996). The last two questions (21 and 22) sought evidence of homonymous visual-field defects or visual inattention in the context of a symmetrical environment. The questions were asked by the author (AT - an ophthalmologist) in the form of a structured clinical history, giving parents the opportunity to clarify any questions and to discuss complex issues.

Control Subjects

The control group comprised 200 children attending the day surgery unit, orthopaedic and ENT clinics at the Royal Hospital for Sick Children in Glasgow, Scotland. The children did not have any neurological or ocular disorders, other than squint or refractive error. The group was classified into four age groups: 5 to 6 years, 7 to 8 years, 9 to 10 years, and 11 to 12 years. The percentages of children in each age group with a particular response are given as descriptive statistics.

Patients

A cohort of patients who have hydrocephalus and who satisfied the following criteria was selected: binocular visual acuity at least 6/60, 5 years of age or older, previous treatment with a ventricular shunting procedure, recent high-quality CT scans, and intellectual and physical ability allowing cooperation with clinical assessment.

Patients were identified from the In-patient Disease Index, the radiology department, the vision assessment clinic, eye outpatient clinics, and surgical outpatient clinics. Information was collected from the clinical notes on the patients including demographic data, underlying cause of hydrocephalus, surgery carried out, complications related to the ventricular shunt, and the number of procedures for revision or replacement of the ventricular shunt. Eye examination was carried out and the following were recorded: visual acuity, colour vision; visual field to confrontation, eye movements, and slit lamp and fundus examination. The patient group was coded by numbers 1 to 52 in the order they attended. Data from the questionnaires, examinations, and case notes were recorded on a spreadsheet.

The responses to the questions were compared with those for their age-matched control groups. Owing to the small numbers of control children with frequent or constant difficulties, a cognitive visual-dysfunction was considered significant when it was reported in the patient group but was entirely absent in the appropriate control groups. To compare control and patient groups, the X2 test was employed when applicable for those with occasional difficulties when these occurred with sufficient frequency for the test to be applied.

Results

Table 1 summarizes the results for the control group. Inconsistent responses with contradictory answers which rendered the results unreliable were obtained in eight cases. The total number of children with consistent results was 192, of whom 59 were aged 5 to 6 years, 49 were aged 7 to 8, 36 were aged 9 to 10, and 48 were aged 11 to 12.

The patient group comprised 29 males and 23 females. Table II illustrates their age distribution and the numbers with evidence of constant or frequent cognitive visual problems. Twenty-five (48%) children manifested no visual problems. Table III summarizes the details of the 27 (52%) patients with evidence of cognitive visual dysfunction. They can be subdivided into three groups: those involving one cognitive function (11 of 27), those with two or three cognitive functions affected (14 of 27), and those with more than three functions affected (two of 27).

Face Recognition

None of the control children in any age group was described as having frequent or constant difficulty recognizing parents, family members, or friends. However, 5% of the youngest age group were said to have occasional difficulty doing so. Occasional difficulty recognizing people from photographs was more common in the three youngest groups. Children of 11 to 12 years of age showed no evidence of difficulty recognizing faces in any category.

Frequent difficulty with recognizing faces was described for three patients (5, 21, and 22), all of whom manifested additional cognitive visual problems (see

Table III). All three had evidence of primarily right-sided brain damage. Patient 5 had a left hemiplegia, patient 21 left homonymous hemianopia, and patient 22 left homonymous inferior quadrantanopia.

Shape and Object Recognition

None of the children of any age group was described as having frequent or constant difficulty recognizing shapes or objects. However, for the 5- and 6-year olds, occasional difficulty was described for shape and object recognition in 5%. For the 7- and 8-year olds, no such difficulty was reported, but 6% still had occasional problems with object recognition. This declines to 3% in the 9- and 10-year olds, and was not described for the oldest group.

Twelve patients (23%) in the study group were reported to manifest constant or frequent difficulty with shape recognition. Constant difficulty was reported in two patients (1 and 25) and frequent difficulty in ten (4, 5, 6, 29, 30, 32, 35, 39, 43, and 50). In one patient, number 30, this was an isolated finding. In nine (1, 4, 6, 25, 29, 32, 35, 39, and 50) it occurred in association with one or two other defects, and in two (5 and 43), with multiple defects.

Four patients (8%) had frequent difficulty with object recognition, and this was accompanied by impaired shape recognition in each case. The incidence of occasional difficulty with object recognition was not significantly higher than controls for any age group (P=0.45).

Colour Naming and Matching

None of the control children in any age group had frequent or constant difficulty in naming and matching colours. However, 2% of children in the two youngest groups had occasional difficulty in this task. Neither problem was described for the older age groups.

Difficulty with colour naming occurred either constantly or frequently in seven patients (14%). Two patients (25 and 43) had constant difficulty, while five (5, 15, 21, 40, and 42) had frequent difficulty with colour naming. Two patients (40 and 41) had no other associated cognitive visual problems. Three had one or two associated problems (15, 21, and 25), and two had multiple problems (5 and 43). In three (5, 15, and 43) difficulties in both colour naming and matching were described. The prevalence of occasional difficulty with colour naming was not significant when compared with the control group (P=0.125).

Two patients (25 and 43) were reported to have lost the ability to name colours following ventriculoperitoneal shunt insertion in the left occipital area.

Orientation

None of the control children of any age group had problems with orientation at home. Question 11 was not contributory because it was interpreted as referring to losing things around the home. However, orientation in new

surroundings was more problematic, although constant difficulty was not reported for any age group.

Five patients (9%) had constant problems with orientation (4, 20, 34, 43, and 47). One patient, number 43, had difficulty finding his way in familiar places such as at home, and had multiple other cognitive visual problems. Patient 20 was described as having frequent difficulty with orientation both at home and in new surroundings. This was his only visual problem, which had been constant a year earlier, and it was improving with time. Three patients showed constant difficulty with orientation in new surroundings (34, 43, and 47). Patient 34 had additional simultaneous perception problems, but for patient 47 this was an isolated phenomenon. Occasional and frequent difficulties in new surroundings were not significantly more common in the patients (P=0.90) than in the controls.

Depth Perception

Constant difficulty with depth perception was described in three control children of the 5- and 6-year-old group but none of the older groups. Both aspects of depth perception as addressed by the two relevant questions (15 and 16) were impaired. Two of these children had a history of squint.

The questions on depth perception were not applicable in 32 of the study group due to associated mild or moderate degrees of motor incoordination. It was not, therefore, possible to distinguish such visual problems from motor difficulties by means of the questions asked.

Motion Perception

The control results for motion perception varied depending on whether the child was stationary or moving. For moving objects, none of the children in any age group had constant difficulty. Frequent and occasional difficulty was, however, occasionally described for the two youngest groups. For the children aged 9 and older no difficulty was reported. For when the child was moving, difficulties were observed more frequently in all age groups.

Constant problems with perception of moving objects were described for seven patients (14%), (2, 15, 19, 22, 29, 50, and 52). In two children these were isolated findings (19 and 52). None of the control group manifested constant difficulty. However, the incidence of occasional and frequent difficulty when compared with the control group was not significant (P=0.975).

[REFER TO TABLE I]

Table I: Percentages for results obtained for each question asked of control population

		5 to 6-yr. olds				7 to 8-yr. olds				9 to 10-yr. olds				11
	Question	**A**	**B**	**C**	**D**	**A**	**B**	**C**	**D**	**A**	**B**	**C**	**D**	**A**
1.	Parent recognition	95	5	0	0	100	0	0	0	100	0	0	0	100
2.	Family recognition	95	5	0	0	96	4	0	0	100	0	0	0	100
3.	Friend recognition	85	15	0	0	94	6	0	0	100	0	0	0	100
4.	Photograph recognition	80	20	0	0	94	6	0	0	92	8	0	0	100
5.	Self-photograph recognition	90	10	0	0	94	6	0	0	95	5	0	0	100
6.	Shape recognition	100	0	0	0	100	0	0	0	100	0	0	0	100
7.	Object recognition	95	5	0	0	94	6	0	0	97	3	0	0	100
8.	Colour naming	98	2	0	0	98	2	0	0	100	0	0	0	100
9.	Colour matching	98	2	0	0	98	2	0	0	100	0	0	0	100
10.	Finding way in home	100	0	0	0	100	0	0	0	100	0	0	0	100
11.	Asking way in home	45	35	20	0	50	25	25	0	40	50	10	0	65
12.	Losing objects at home	90	2	2	6	80	17	3	0	95	5	0	0	100
13.	Finding way in new places	80	16	0	4	90	10	0	0	85	15	0	0	95
14.	Asking way in new places	85	10	0	5	90	10	0	0	100	0	0	0	98
15.	Reaching and grasping objects	90	3	0	7	88	12	0	0	95	0	5	0	100
16.	Distinguishing line from step	90	7	0	3	88	12	0	0	92	4	4	0	100
17.	Seeing moving objects	90	5	5	0	93	5	2	0	100	0	0	0	100
18.	Seeing objects while moving	55	30	10	5	85	10	5	0	90	10	0	0	85

19.	Finding objects while moving	70	25	5	0	90	7	3	0	95	5	0	0	95
20.	Finding objects in complex pictures	65	25	7	3	80	17	3	0	80	15	5	0	94
21.	Eating food from part of plate	73	17	7	3	90	7	3	0	86	10	2	2	90
22.	Misjudging doorways/corridors	70	17	10	3	90	7	3	0	90	6	4	0	95

Problems identified: A, never; B, occasionally; C, most of the time; D, always.

Visuospatial Perception

None of the control children in any age group was described as having constant difficulty distinguishing an object from a patterned background. However, frequent difficulty was encountered in small proportions of the two younger groups. Occasional difficulty was described in 25% of the 5- to 6-year-old group, but in smaller proportions of older children. Apart from 3% of the 5- and 6-year olds, none of the control children in any other age group had constant difficulty with tasks requiring simultaneous perception. Nevertheless, frequent difficulty was described in a small proportion of each age group. Occasional difficulty in doing such tasks was common in the three youngest groups but was rare in the oldest group.

Eleven patients (21%) were said to have constant difficulty in performing simultaneous perception tasks both when distinguishing an object from a patterned background or extracting information from a complex picture. In four cases this was isolated (3, 8, 14 and 38), five had one or two other problems (2, 6, 22, 34, and 35), and two (5 and 43) had three or more. The prevalence of constant problems in extracting information from complex pictures in the patients aged between 5 and 6 was found not to be significant (P=0.975) when compared to the age-matched controls. For the whole patient group, the prevalence of occasional and frequent difficulty either distinguishing objects from a patterned background or looking at complex pictures also did not reach significance (P=0.250 and P=0.100 respectively).

Visual-Field Defects

Question 21 gave many false-positive responses. However, one child in the youngest control group tended to ignore one side of the visual field persistently. This child also had problems with depth perception but no strabismus.

In the patient group the questionnaire responses provided evidence of possible field neglect in three patients (1, 21, and 34). On confrontation testing of the visual field, two had homonymous hemianopia, while the third

manifested left-field neglect. All three had associated cognitive visual problems.

Table II: Age distribution of study group and numbers with constant or frequent cognitive visual problems

Age (y)	Nr in group	Nr with constant or frequent cognitive visual problems
5	9	4
6	8	6
7	3	3
8	6	2
9	11	6
10	5	2
11	3	2
12	1	0
13	4	2
>13	2	0
Total	52	27

Table III: Patients with significantly* affected visual cognitive function N=27)

Patient Nr	Age (y)	Significantly* affected visual cognitive function
One function affected		
3	9	Simultaneous perception
8	13	Simultaneous perception
14	5	Simultaneous perception
19	9	Motion perception
20	8	Orientation-home and new surroundings
30	5	Shape recognition
38	9	Simultaneous perception

40	7	Colour naming
42	6	Colour naming
47	6	Orientation-new surroundings
52	11	Motion perception

Two or three functions affected

1	11	Shape recognition, object recognition, field neglect
2	9	Motion perception, simultaneous perception
4	6	Shape recognition, orientation- new surroundings
6	5	Shape recognition, simultaneous perception
15	6	Colour naming, colour matching, motion perception
21	13	Face recognition, colour naming
22	6	Face/motion perception, simultaneous perception
25	5	Shape recognition, colour naming
29	10	Shape recognition, motion perception
32	7	Shape recognition, motion perception
34	9	Orientation-new surroundings, simultaneous perception, field neglect
35	10	Shape recognition, simultaneous perception
39	9	Shape recognition, object recognition
50	8	Shape recognition, motion perception

Multiple cognitive visual problems

5	7	Face/shape/object recognition, colour naming/matching, simultaneous perception
43	6	Shape recognition, colour naming, colour matching, simultaneous perception, orientation - home and new surroundings

* Significantly affected cognitive visual function refers to those children with a frequency of difficulty which was not encountered in any child in their age-matched control group.

Handedness

Sixteen of the control group were found to be left-handed, giving a prevalence of 8%. The prevalence of left-handedness among the patient group was 18 of 52 (35%). The incidence of left-handedness did not differ significantly between patients with and without visual cognitive dysfunction.

Ocular Examination

General ocular examination was carried out on 51 patients. (One patient was unable to attend the clinic, but her parents completed a questionnaire by telephone interview.) A comparison of the ophthalmic complications in subjects with and without cognitive visual dysfunction revealed a similar prevalence for both groups.

Visual Acuity

A visual acuity of 6/60 in both eyes was found in two patients (4%). In 20% the visual acuities ranged from 6/9 to 6/36, but in most (64%) they were 6/5 or 6/6 in both eyes. Anisometropic amblyopia was found in eight children (16%) with a difference of acuity between the two eyes between 2 to 6 lines on the Snellen chart.

Refractive Error

Refractive error was identified in 30 control subjects. These children did not show any obvious differences in their questionnaire responses in comparison to those without a refractive error. Nineteen of the patients wore a spectacle correction.

Squint

A history of squint was given for nine of the control children. Two of these children had constant difficulty with all aspects of depth perception. Squint was present in 31 patients (61 %). Of these, 22 were esotropic and nine exotropic. In almost one-third of the cases the squint alternated.

Visual Fields

Visual-field defects were identified in eight patients (16%). Homonymous hemianopia was found in six (three right sided and three left sided). In three cases this had not been apparent to the parents and was not identified by the questionnaire. Homonymous inferior quadrantic field loss was found in two patients (one right sided and the other left).

Nystagmus

Nystagmus was evident in 20 patients (39%), and was manifest in the primary position in 11, gaze evoked in six, and latent in three. Two of the patients had visual acuities of 6/60 in both eyes with bilateral optic atrophy.

Abnormal Ocular Movements

Saccadic movement in both up-gaze and down-gaze was deficient in one patient, and reduced in another. In neither of these cases nor in the rest of the group was any other evidence of the dorsal midbrain syndrome found. No horizontal gaze disorders were identified in this group. All of the squints were concomitant.

Pupillary Reactions

Pupillary light responses were bilaterally sluggish in seven patients and a relative afferent pupil defect was seen in two patients.

Optic-Nerve Appearance

Optic atrophy was seen in 15 patients (31%), was bilateral in 10 and unilateral in five. The degree of optic atrophy was assessed subjectively and deemed to be mild in 11 cases, moderate in two, and severe in another two cases. The severity of optic atrophy was related to visual acuity. In patients with mild optic atrophy the visual acuity ranged from 6/5 to 6/9. In those with moderate changes, visual acuity was 6/60 in both eyes in one patient, and 6/9 in the one eye and 6/36 in the other eye of the second patient. Papilloedema was not detected in any patient.

Information from Case Notes

Details of the medical and surgical aspects of the patients' histories which might influence the occurrence of ocular or visual cognitive disorders were sought from the patients' records. When comparisons were made between the incidence of the main ophthalmic complications in the study group (i.e. squint, nystagmus, and optic atrophy) and the medical and surgical variables (i.e. perinatal factors, type of hydrocephalus, shunt complications, and associated neurological complications), no relation was identified (P=0.975).

To assess the role of these medical and surgical factors in the development of visual cognitive dysfunction, they were compared in children with and without visual cognitive dysfunction in the study group, and amongst patients with various degrees of cognitive dysfunction. No statistically significant relation was identified (P=0.975).

Discussion

Normative data were sought for children from the age of 5 years because in our experience disorders of the cognitive aspects of vision are rarely presented before the age of 5, as it is difficult to know whether apparent paradoxes in visual behaviour are simply due to youth. The results for the control population show very similar patterns of evolution for each aspect of visual behaviour. 'Normality' is established in all 11- and 12-year olds for most of the functions described, except orientation in new surroundings and simultaneous perception, which may take longer to develop in a few children.

It can be argued that the visual problems identified in the children with hydrocephalus reflect a reduction in mental age. However, the age distribution of the children with hydrocephalus with evidence of cognitive visual dysfunction was the same as those with no dysfunction. The visual deficits observed were focal in nature, while other visual functions were age appropriate.

The terms used to describe visual agnosias in adults have not been used in this paper because they refer to the loss of a function, rather than the failure to gain it. Therefore, it seems more appropriate to adopt descriptive terminology. In this study we have found evidence of frequent or constant specific visual cognitive difficulties in 27 of 52 patients (52%) with no age-matched controls being similarly affected. This implies that such difficulties occur in an 'all or none' fashion, and do not manifest as intermittent phenomena.

Face Recognition

Recognition of faces is a complex process involving a large cerebral volume (Sergent et al. 1992). Face data are encoded, then matched with the features of those already stored (recognition) (Carey 1992). In this study a small proportion of younger control children had occasional difficulty recognizing parents and family members. Occasional difficulty was described for recognition of people from photographs, but this diminished with increasing age so that none of the 11- and 12-year olds was described as having such problems. Carey's study (1992) of recognition of unfamiliar faces from photographs showed progressive improvement with age, except at age 12 when a worse performance was found. The reason for this is not known, but has also been noted for recognition of flags, houses, voice and tonal memory (Ellis 1992). Children's problems in face recognition probably concern the encoding of new faces. When faces differ in expression, angle of view, direction of lighting, hairstyle, clothing, size of photograph, or background, then children perform less well.

In adults, difficulty recognizing faces with intact understanding of facial expression is known as prosopagnosia (Tranel et al. 1988,Young et al. 1993). In children, difficulty in recognizing people has been reported by Dutton and colleagues (1996) in 15 of 90 children with cerebral cortical damage, due to various causes, including hydrocephalus. Various degrees of severity were

evident, ranging from complete inability to recognize people, through inability to recognize first-degree relatives, to inability to recognize people from photographs but not real life. Mancini and coworkers (1994) have also reported children with early brain damage who have a variety of disorders of face recognition. Young and Ellis (1989) have reported a case of childhood prosopagnosia in a patient with shunted hydrocephalus following meningitis. Developmental prosopagnosia has been reported both in an otherwise healthy individual (McConachie 1976) with 15-year follow up (De Haan and Campbell 1991), and in association with Asperger syndrome (Kracke 1994).

MRI and PET scan studies suggest that processing of the face is carried out in the ventromedial aspect of the right cerebral hemisphere (Sergent and Signoret 1992, Sergent et al. 1992, De Renzi et al. 1994). Lesions in this territory can result in prosopagnosia, especially with associated corpus callosum involvement (Damasio 1982). Diffuse damage to the white matter caused by hydrocephalus and corpus callosum abnormalities could, therefore, cause problems of this nature. Three children in the study group had obvious difficulties in recognizing faces associated with other visual cognitive difficulties. All three manifested left homonymous visual-field deficits, a feature which has been described previously in adults and children (Grusser and Landis 1991, Boeri and Salmaggi 1994, Dutton et al. 1996)

Shape and Object Recognition

Recognition of shapes is probably necessary before object identification can occur. The process of visual object recognition is thought to occur in hierarchical stages, with the first stage being shape coding (Humphrey and Riddoch 1993). The control group in this study showed few problems with both shape and object recognition with the development of shape recognition appearing to precede the development of object recognition.

In adults, difficulty in recognizing objects is known as object agnosia. It is a sequel to a range of disorders involving the ventral aspects of the occipital and temporal lobes (Grusser and Landis 1991), with a number of different forms of object agnosia being related to particular regions of the cortex (Humphrey and Riddoch 1993). Object-recognition disability has been reported in children with cerebral cortical damage due to various causes including hydrocephalus (Chin et al. 1992). The prevalence in the present study was 8%. Shape recognition was affected in all of these patients, and in one, face recognition and simultaneous perception were also impaired. This reinforces the suggestion that both object and face recognition are complex functions which can be affected by extensive brain damage.

Difficulty in recognizing shapes has been previously described in children with hydrocephalus (Miller and Lovleen 1971, Rabinowicz 1974). Significant difficulty was described in 12 patients in the present study (23%) and was the most common cognitive visual dysfunction. In only one patient was this an isolated finding.

Naming and Matching of Colours

Colour naming and matching seemed to be well developed in all the control age groups surveyed. The ability to match colours is said to be better than the ability to name them at 6 years of age probably because further maturation is required to link visual recognition of colours to verbal expression for them (Bornstein 1985). However, in our control group, occasional problems for matching and naming of colours were reported with equal frequencies.

In the children with hydrocephalus, problems with colour matching and naming were described in isolation and with other cognitive problems. The three patients who were unable to match colours were also unable to name them. Impairment of colour naming with sparing of matching indicates the existence of separate territories for each function. This is exemplified by case number 25 who lost colour-naming ability immediately after shunt insertion on the left, but who could still match colours. This accords with descriptions in adults who develop colour anomia after damage to the junction of the occipital and temporal lobes in the left hemisphere, resulting in right homonymous hemianopia and colour anomia (Oxbury et al. 1969).

Orientation

Orientation at home or in familiar surroundings develops at an early age and did not cause any problems for the control children. In new surroundings, many children get lost but this improves as the child grows up. Problems with orientation in new surroundings became more common in the 9- and 10-year olds. This may reflect the increased opportunities that older children have for independent exploration.

In adult patients with topographic agnosia, the right medial occipitotemporal area (sometimes bilaterally) can be involved (Landis et al. 1986, Hublet and Demeurisse 1992). The right posterior parietal lobe has also been implicated and is thought to be involved in spatial memory. Loss of environmental familiarity is the inability to recognize familiar surroundings in spite of relatively intact verbal memory, cognition, and perception (Landis et al. 1986).

In the children with hydrocephalus, constant difficulty with orientation both at home and in new surroundings was described for five patients and could not be accounted for by general reduction in cognition. Orientation difficulties occurred both as an isolated phenomenon and in association with other cognitive visual problems. Left homonymous hemianopia was found in two-thirds of affected patients in Dutton and colleagues' (1996) clinical study. All 16 of the patients with acquired topographic agnosia reported by Landis and coworkers (1986) had left-sided field defects. In the present study, however, none of the affected children was found to have visual-field defects or other lesions suggestive of right-brain dysfunction.

Depth Perception

The ability to perceive images in three dimensions appears to develop during the early years of life and continues to improve in later childhood. Two of the control children with constant difficulty in depth perception had a history of squint reported. No problems with depth perception were reported for the other seven children with a history of squint.

We have found that impaired appreciation of depth, for example difficulty identifying steps, can be a major problem for some children with occipital pathology (Ahmed and Dutton 1996, Dutton et al. 1996), but in the present study, questions 15 and 16 did not adequately discriminate which children with hydrocephalus had such problems because 32 had mild or moderate motor incoordination. More specific clinical questions warrant consideration.

Motion Perception

There was a marked difference in responses for the control group depending upon whether the child was stationary or moving. When the child is stationary, problems are less frequent than when the child is moving. The parents of seven of the patients described profound difficulties in the perception of movement to a degree not seen in the control group. In two control children this was the only abnormality reported. Impaired perception of movement has major implications for safety, as affected individuals may not be aware of moving traffic. When affected individuals are moving quickly themselves, stationary objects may become less 'visible'.

The part of the brain that primarily responds to visual motion is V5, located in the lateral prestriate cortex at the junction of the occipital and temporal lobes (Zeki 1993). Disturbance of cerebral motion perception is known as akinetopsia. This condition was first described by Zihl and colleagues in 1983 in a patient who had sustained bilateral cerebral lesions affecting the lateral temporooccipital cortex and the underlying white matter. Similar disorders follow surgical removal of brain tissue (Regan et al. 1992). Different mechanisms subserve spatial discrimination of motion-defined and luminance -defined form (Regan 1995). Shunts for hydrocephalus are commonly placed close to this area, thus this topic warrants further study.

Visuospatial Perception

Although figure-ground and simultaneous perception improved steadily with age in the control subjects, problems were still reported for a small proportion of the oldest age group. For such tasks, visual short-term memory is required to store the image data, while attention moves to another location (Coslett and Saffran 1991). Simultanagnosia describes a condition in which the affected person is unable to interpret complex visual arrays despite preserved recognition of single objects (Coslett and Saffran 1991). This condition is known to occur in adults with bilateral lesions affecting the superior occipitoparietal region (Grusser and Landis 1991), and has also been

reported in cases with unilateral parietooccipital lesions (Coslett and Saffran 1991).

Damage to white matter is thought to be the cause of such defects, and may contribute to impaired development of non-verbal cognitive skills in children with hydrocephalus (Donders et al. 1991, Fletcher et al. 1992b). There appears to be impairment of the processes linking the structural description of objects to their locations and there may be an improved ability to identify simultaneously presented stimuli when they were thematically related (Coslett and Saffran 1991). In the patient group, impaired simultaneous perception was found in 219, and was the most common isolated problem (four patients). This has important implications for education of children with hydrocephalus. In the presence of normal or near-normal visual acuity and intellectual status, children with hydrocephalus may have a simultaneous perception problem, which can easily be misinterpreted as an indication of poor intelligence. Parents and teachers need to be made aware that complex illustrations may lead to information overload and that sequential presentation of clear images against a plain background can be very helpful.

Visual-Field Neglect

Hemispatial neglect comprises partial or complete unawareness of the existence of stimuli in the affected hemifield, despite a full visual field. This is usually seen in patients with a right parietal lesion, but has also been described in association with frontal, lenticular, and thalamic lesions (Heilman et al. 1993). Hemispatial neglect was detected by examination in one child in this study and in 4% in the Rabinowicz study (N=4). As with visual-field loss, this type of field defect may present difficulties at play, in learning, and may prove hazardous when crossing roads.

Rehabilitation

The adult mammalian nervous system shows some plasticity following damage (Steele Russell 1992).The studies reviewed by Gouvier and Cubic (1991) showed that acquired visuoperceptual disorders are sometimes amenable to restructuring of the environment, teaching compensatory methods, and using residual strengths to remediate deficits. They state that improvement is often maintained and can extend to improvements in other areas of daily life. A study to assess the effects of a remedial programme on visual-motor perception in children with spina bifida, showed that dysfunction was reduced when reassessed 2 months after administration of the programme (Gluckman and Barling 1980).

Conclusion

This study shows that a wide range of cognitive visual disorders are probably present in this selected population of children with shunted hydrocephalus. Most affected children had more than one area of cognitive visual dysfunction (16 of 27). The findings cannot be attributed to the known ocular sequelae of

hydrocephalus, as the prevalence of such disorders in those with and those without cognitive visual dysfunction was no different.

We recommend that these cognitive visual disorders should be actively sought by structured history taking. Once they have been identified, assessment by an interdisciplinary team to characterize the problems and to plan for the special educational needs is indicated. Standardization of age-corrected investigations of cognitive visual function warrant development as many visual-cortical syndromes have not been well studied, and are poorly understood (Rizzo and Nawrot 1993). The development of novel strategies is needed to help parents and teachers circumvent these problems. With the child's potential for growth and development, it may be that these deficits can be overcome or diminish with training. The earlier that deficits can be detected, and measures implemented to circumvent them, the greater the potential opportunity for functional improvement.

Accepted for publication 23rd December 1998.

Appendix 1

Parent Questionnaire

For the following questions, which number best describes how your child responds in the following situations:

Never	1
Occasionally	2
Most of the time	3
Always	4

(A temporal classification was employed rather than a qualitative evaluation so that the meaning of the responses would be common to all parties.)

1. Does your child recognize you before you speak?
2. Does your child recognize other family members?
3. Does your child recognize friends?
4. Does your child recognize people from photographs?
5. Can your child identify himself/herself from photographs?
6. Can your child recognize shapes?
7. Can your child recognize objects?
8. Can your child name colours?
9. Can your child match colours?
10. Can your child find his/her way around the house?
11. How often does he/she ask for directions at home?
12. Does he/she lose objects around the house?
13. Can your child find his/her way around new surroundings?
14. How often does he/she ask for directions when in new surroundings?

15. Does he/she have difficulty reaching out for and grasping objects?
16. Does your child have difficulty distinguishing a step from a line on the ground?
17. Is your child able to see moving objects or are they seen only when they are stationary, e.g. pets, traffic.
18. Does your child have difficulty seeing objects when he/she is moving quickly him/herself?
19. Can your child find objects on a patterned carpet?
20. Can your child find objects in complex pictures?
21. Does your child eat food from only one part of the plate and ignore the rest?
22. Does he/she misjudge going through doorways or along corridors?

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