Evaluation of the electrodiagnostic investigation of children using the Greenwich Grading System

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CLINICAL STUDY

Abstract

Purpose To evaluate the contribution of electrodiagnostic testing (EDT) to the management of children in a paediatric ophthalmology service using the Greenwich Grading System (GGS).

Methods A retrospective analysis was performed of the case notes of 105 of the 113 paediatric patients referred from the Strabismus and Paediatric Service at Moorfields Eye Hospital for electrophysiological testing over a 1-year period. The GGS was used to quantify the contribution of EDT to the diagnosis, overall investigation, and treatment of each patient. Patients were further subdivided into different diagnostic groups to allow comparison of the value of EDT in different conditions.

Results EDT was found to be of value in 91% of the children tested and was considered an essential investigation in 71%. EDT made a new diagnosis in 7% of patients, changed it in 5%, and confirmed or excluded a diagnosis in 79%. EDT made a useful contribution to the overall investigation of 89% of the patients and was considered the only test that could provide the required information in 71%. The results of EDT allowed reassurance and/or explanation with regard to the diagnosis, prognosis, and treatment in 91% of children. In one patient, treatment was changed as a result of EDT. The clinical outcome was not adversely affected in any patient.

Conclusions EDT was of value to the clinical management of most of the children reviewed, mainly by confirming or excluding a clinical diagnosis and allowing explanation and reassurance to children and parents. Electrodiagnostic information gave a new or changed diagnosis in 12% of the children.

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Introduction

Clinical investigation should only be undertaken if it is likely to assist in the overall care of the patient.^{1,2} Benefits of the investigation must outweigh any risks and the investigation must be cost effective. Having elected to investigate, the clinician must select the most appropriate test, balancing risks^{3,4} and expense^{5,6} against availability and the probability of obtaining useful results.⁴ The Greenwich Grading System (GGS) was developed to quantify the contribution of an investigation to the overall management of a patient⁷ and was initially used to assess the role of electrodiagnostic testing (EDT) in adults. Children present different diagnostic problems because the range of diseases encountered is different, they may be less cooperative to examine and investigate and may provide less clinical information on which to base a diagnosis. Electrophysiology provides objective evaluation of retinal and intracranial visual pathway function to help localise dysfunction, confirm or exclude diagnoses, and give information on disease progression and prognosis.^{8–10} The role of EDT is potentially more important than in adults, but reliable results may also be more difficult to obtain.

The contribution of EDT to the management of children with visual problems has been assessed by reviewing consecutive, unselected paediatric referrals to the electrophysiology

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department at Moorfields Eye Hospital from the Strabismus and Paediatric Service over a 1-year period. The GGS was used to assess the contribution of electrodiagnostic testing to the diagnosis, overall investigation, and management of the patients.

Methods

A retrospective analysis was performed of the notes of 105 (93%) of the 113 paediatric referrals to the Electrophysiology Department at Moorfields Eye Hospital from the four consultants of the Strabismus and Paediatric Service over a 1-year period. The analysis was performed by a specialist registrar and one of the referring consultant paediatric ophthalmologists with experience of electrophysiology. The GGS was used to quantify the contribution of EDT to the diagnosis, investigation, and treatment in each case (Table 1). From this, an overall value of EDT in the management of each patient was established by taking the highest score in the above three categories. The tests performed were selected from flash and pattern electroretinograms, electro-oculogram and flash and pattern appearance, or reversal visual-evoked potentials as appropriate. Most children had an electroretinogram and either a flash or pattern visual-evoked potential depending on age and co-operation. Electro-oculograms were only performed if specifically indicated. No attempt was made to evaluate the contribution of the different tests separately because accurate interpretation of test results involves an overall evaluation of the results of the different tests performed. For example, the interpretation of an abnormal cortical visual-evoked potential depends on knowing whether retinal function is normal. All testing was performed by a technician experienced in recording from a paediatric population and interpreted by a consultant

electrophysiologist experienced in interpreting paediatric records. Full details of the scoring system used by the GGS have been given elsewhere.⁷

Results

The ages of the patients undergoing testing ranged from 3 months to 15 years 11 months, with a median of 6 years (Figure 1). Testing was evaluated under the three GGS headings of effect on diagnosis, contribution to the overall investigation of the patient, and effect on management. An overall score was then derived as the best score in the above three categories.

Overall, EDT was found to be of value in 91% of the 105 patients reviewed, being considered to be 'essential' according to the GGS in 71%, 'important' in 16%, and 'helpful' in 5% of children. EDT did not help in 9% (in 5% this was because of limited co-operation and in the others because it did not increase diagnostic certainty). The clinical outcome was not adversely affected in any patient.

A new diagnosis was made by EDT in 7% of patients, the diagnosis was changed in 5%, and diagnostic certainty was increased in 79% of patients by either confirming or excluding a clinical diagnosis. In four of the patients where the diagnosis was changed, EDT demonstrated a previously undiagnosed retinal dystrophy in a patient with a retina of normal appearance; the provisional clinical diagnoses had been of cortical visual loss, myopia with reduced vision, optic atrophy, and amblyopia. In the fifth patient, electrophysiological changes typical of albinism were demonstrated in a child with nystagmus, but without cutaneous signs or iris transillumination.

EDT was considered to have made a useful contribution to the overall investigation in 89% of the patients and it was considered to be the only test that

Table 1 GGS for assessing the value of an investigation in the management of a patient

Value of the investigation (grade)	Value of the investigation to overall management	Role of the investigation in the different management categories		
		Diagnosis	Investigation	Treatment
3	Essential	Changed	Only test able to give required information	Initiated
2	Important	Made	Replaced others	Altered or Stopped
1	Helpful	Confirmed or excluded	Suggested more	Reassurance Explanation
0	Unnecessary	No help	Not altered Others more use	No change
-1	Adverse effect	Incorrect	Delayed others	Incorrect

If -1 is scored in either the diagnosis, investigation, or treatment categories, the overall grade is -1. Otherwise, the overall grade is the highest scored in any of the three categories.

Figure 1 Histogram showing the age at which children underwent electrophysiological examination.

could provide the required information in 71%. EDT replaced other investigations in 16% and in 2% the results of EDT indicated the need for further investigations.

EDT allowed better information to be given to patients and parents with regard to diagnosis, prognosis, and treatment in 91% of cases. In one child who had a longstanding traumatic cataract in an eye that was believed to have poor visual potential due to posterior segment damage, EDT showed good retinal and optic nerve function. The management was changed as a result of EDT and surgery was performed with a successful outcome. In no case did EDT result in an incorrect treatment being advised.

The GGS ascribes a grade to the certainty of diagnosis before and after EDT. EDT was very useful in the 16 cases in which the pretest diagnosis was most uncertain, giving a 'certain' diagnosis in six (38%) and 'probable' diagnosis in eight (50%) of these patients. After EDT, eight were considered to be normal and four to have retinal disease.

Analysis by post-EDT diagnosis

The diagnoses for all cases after EDT are given in Table 2. The three commonest diagnoses were of retinal dystrophy, amblyopia, and electrophysiological normality. These are presented separately below.

Retinal disease (23 patients)

EDT was found to be of value in all patients and deemed essential in 20 (87%), important in one (4%), and helpful in two (9%). The diagnosis was changed to a retinal dystrophy after testing in four patients (22%) and a new diagnosis made in two (9%). The investigation of the

Table 2 Diagnoses for all the patients following electrodiagnostic testing

Normal electrophysiology ^a	31	30%
Retinal pathology	23	22%
Amblyopia	21	20%
Albinism	9	9%
Cortical visual loss	7	7%
Oculomotor nystagmus	5	5%
Functional visual loss	2	2%
Optic neuropathy	1	1%
Delayed visual maturation	1	1%
Inconclusive	5	5%

^aThe five patients with oculomotor nystagmus also had normal electrophysiology.

Table 3 Classification of retinal pathology

Cone dystrophy	5
Cone-rod dystrophy	1
Rod-cone dystrophy	1
Leber congenital amaurosis	1
Retinitis pigmentosa	2
Inner retinal dystrophy	3
CSNB	2
X-linked retinoschisis	2
Macular dystrophy	4
Retinal coloboma	1
Unilateral retinal dysfunction	1

patient was considered to have been assisted by EDT in 21 (91%) and considered the only test that could provide the required information in 20 (87%).

The results allowed reassurance and/or explanation with regard to the diagnosis, prognosis, and treatment in all patients. Ten (45%) of the patients with retinal dystrophy were recorded as having either normal fundal appearances or only myopic changes.

The features of the electrophysiological changes found allowed the nature of the retinal pathology to be elucidated in all patients. Details of the retinal conditions diagnosed following EDT are shown in Table 3.

Amblyopia (21 patients)

In general, amblyopic children were referred for electrodiagnostic investigation to exclude other pathology when their visual acuity did not improve with patching in the presence of normal fundi, particularly if the acuity of the fellow eye was subnormal. Overall EDT was considered to be of value in 19 (91%) and deemed to be 'essential' in 18 (86%), 'important' in one (5%), and of no help in two (10%).

A diagnosis of amblyopia was made in one patient with reduced vision of previously uncertain aetiology

and was confirmed, or other causes of reduced vision excluded, in 15 (71%). Diagnostic certainty was not increased in six patients (29%) either because of technical problems (one patient) or because electrophysiological findings could not exclude optic nerve or other pathology.

The overall investigation of the patient was considered to have been improved by EDT in 19 patients (91%) and deemed the only test that could provide the required information in 18 (86%).

The results of EDT in amblyopia allowed reassurance/explanation with regard to the diagnosis, prognosis and treatment in 18 patients (91%). EDT was of no value in the management of two patients (9%).

Normal (36 patients)

The results of EDT were considered to be definitely normal in 22 patients (61%) and probably normal in 14 (39%) of this group. Those considered probably normal showed no clearcut abnormality, but were generally either slightly outside normal limits or were technically unsatisfactory on a particular test. Five of these 14 had developmental delay. In 18 (50%) referral had been made because of reduced acuity without definite cause, including one patient considered on clinical grounds to have functional visual loss. Five patients with nystagmus and five with developmental delay were found to have normal electrophysiology. Others with normal electrophysiology had been referred because of abnormal fundal appearances (two patients), a family history of genetic eye disease (three patients), photophobia suggestive of possible retinal dystrophy (two patients), or opaque media (one patient). Nine of these patients were referred within the first year of life.

Analysis by indications for referral

In addition to classifying patients by their eventual diagnosis, the following groups were analysed by features at referral.

Developmental delay

Of 13 patients known to have developmental delay, useful results were obtained in 11. Of these, six were found to have evidence of cortical visual impairment and five had normal results.

Myopia and reduced best-corrected acuity

Of eight children referred with myopia, reduced best-corrected acuity and normal or myopic fundal

appearances, four were found to have retinal dystrophies.

Amblyopia

Of 23 patients investigated for amblyopia, the diagnosis was confirmed by successful exclusion of other pathology in 16 patients. This was by demonstration of a normal electroretinogram and by the presence in the cortical visual-evoked potential only of changes compatible with amblyopia. One patient referred with probable bilateral amblyopia and normal fundal appearances was found to have a retinal dystrophy and one suspected amblyope was found to be normal. In three patients optic nerve dysfunction could not be excluded with confidence and further investigation was recommended.

Discussion

Evaluation using the GGS has shown that electrodiagnostic investigation made a useful contribution to the diagnosis, overall investigation, and management of most of the children reviewed. In the majority of patients, a probable diagnosis made on clinical grounds was confirmed or a possible diagnosis excluded. However, EDT was of greatest value in the 12% of cases where a new or altered diagnosis was made. In particular, retinal dystrophies were found in a number of children with normal fundal appearances who had been referred because of subnormal best-corrected visual acuities, either of unknown cause or where a different reason for reduced vision was suspected clinically. Of myopic children with a reduced best-corrected acuity and normal or myopic fundal appearance, half were found to have a retinal dystrophy. In only 5% of children were no useful results obtained because of inadequate patient cooperation.

As a result of the nature of the tests, it is not surprising that EDT was of most value where a retinal dystrophy was diagnosed. This applied not only overall, but also in the diagnosis, investigation, and treatment subcategories of management; it was especially useful in myopic patients whose vision did not improve fully with correction. In addition to making the diagnosis, it also allowed the nature of the dystrophy to be elucidated, which was of particular value in giving the patient and parents a prognosis and allowing for better advice and planning for the child's education and future employment prospects. It also facilitated accurate genetic counselling of both parents and children.

In amblyopic patients, EDT mainly excluded other causes of reduced vision. This was of particular help in cases with reduced visual acuity in both eyes. In the

majority of children, responses were characteristic enough for amblyopia to be diagnosed with confidence; neuroimaging to exclude structural pathology affecting the optic nerve or other parts of the central visual pathways was recommended in only a few children.

EDT was very useful in establishing the normality of the visual pathways either in children referred because of an unexplained reduction in visual acuity or with a condition such as nystagmus or developmental delay which may be associated with retinal or visual pathway pathology. In these patients, EDT played a particularly valuable role in allowing reassurance of both children and parents.

As in the earlier study, which assessed the value of EDT in adults,⁷ the GGS was used retrospectively. It proved straightforward to use and provided a useful framework with which to assess the value of EDT in paediatric ophthalmology. In comparison with adults, EDT was considered essential in a much greater proportion of patients (71 cf 25%) and not helpful in fewer (9 cf 20%). Similar numbers of changed and new diagnoses were made in both groups, but treatment was initiated or changed in fewer children (1 cf 14%). The overall differences reflect particularly the greater contribution of EDT to the overall investigation of children compared to adults. In part, this was because of the different range of diagnoses found in children; in particular, amblyopia and genetic eye conditions are usually diagnosed before adulthood. It also reflects the greater difficulty of using the two main alternative investigations of fluorescein angiography and neuroimaging in children. EDT was able to demonstrate good optic nerve function in most amblyopic children who did not respond to patching and only two were recorded as having imaging because central visual pathway pathology could not be excluded with confidence. Equally, the demonstration of a retinal

dystrophy in children with horizontal nystagmus and no other neurological signs precluded the need for imaging.

This study has demonstrated that in experienced hands electrophysiological investigation makes a valuable contribution to the diagnosis, investigation, and management of children. This contribution differs from that in adults because of the different spectrum of diseases presenting in childhood and the different availability of alternative investigations.

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