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Non-epileptic clinical diagnoses in children referred for an outpatient EEG using video monitoring

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ABSTRACT – Simultaneous video (closed circuit television [CCTV]) and EEG recordings are important in the differentiation of epileptic and non-epileptic paroxysmal episodes and in the classification of epilepsy syndromes. An additional benefit from the observation of the child on CCTV is the possible identification of specific clinical, including genetic, conditions. This three-year prospective study of 2780 consecutive children undergoing routine EEG investigations identified 17 conditions that had not previously been diagnosed by the clinicians who had requested the EEG.

Keywords: clinical diagnosis, video-EEG recording, child

The role and importance of simultaneous video (closed circuit television [CCTV]) and electroencephalographic (EEG) recordings is well-established in the investigation and classification of the epilepsies (Chen *et al.* 1995, Connolly *et al.* 1994, Saravanan *et al.* 2001, Shihabuddin *et al.* 1999). Previous video-EEG studies have understandably focused on the correlation between a patient's clinical behaviour and electrical cerebral activity (Chen *et al.* 1995, Connolly *et al.* 1994). An obvious benefit from reviewing the patient on CCTV is the opportunity of seeing the patient's appearance and noting any abnormal, including dysmorphic, features. This may be particularly relevant where the EEGs are reported by a clinician, and specifically, a paediatric neurologist who often has considerable clinical knowledge and experience of children's diseases and genetic syndromes, in contrast to an adult neurophysiologist, psychiatrist or general practitioner, the other doctors who commonly report EEGs in Great Britain (Ganesan *et al.* In press).

The purpose of this study was to prospectively identify all children in whom a clinical (including genetic) diagnosis was considered on the basis of the clinical history and video (CCTV) recordings of children undergoing routine electroencephalography and which was subsequently confirmed by appropriate genetic, biochemical or radiological investigations. The specific question that was being asked in designing this study was "in how many children who attend the EEG department for a routine outpatient EEG, can the video-footage (CCTV) suggest a specific clinical, including genetic, diagnosis that has not previously been made or considered by the clinician requesting the EEG".

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Patients and methods

The EEG department at Alder Hey provides a secondary and tertiary service for all children (up to 17 years of age) in a total population of approximately two million. All children were referred for an EEG for the following reasons: to confirm a clinical diagnosis of epilepsy; to confirm or exclude a diagnosis of non-convulsive status epilepticus; to classify the specific epilepsy syndrome or as part of a comprehensive evaluation of children with severe, non-progressive developmental delay or developmental regression. Details of the clinical information provided by the doctor requesting the EEG varied, from "Does this child have epilepsy?" to simply enclosing a copy of an outpatient clinic letter. Additional clinical information on the description of the child's paroxysmal episodes and any relevant family history (including of epilepsy) was also routinely obtained by the EEG technicians at the time of the EEG (Beirne *et al.* 1996). All children underwent digital EEG and simultaneous video (CCTV) recordings using a Micromed Brain Quick System. Electroencephalograms undertaken on all consecutive children attending the department for an outpatient EEG over a three year period were included in this study. Inpatient EEGs (including those undertaken on the paediatric intensive care unit) were excluded from this analysis. All EEGs were recorded by two paediatric EEG technicians and reported by a single paediatric neurologist (RA) in conjunction with the technicians. All EEG reports provide an electro-clinical summary and are based on the clinical information provided on the EEG request, together with additional information provided by the technicians and the findings on the child's EEG. The paediatric neurologist provides a supplemental comment on the report where there is a clinical suspicion of a specific diagnosis (e.g. genetic syndrome, chromosomal abnormality or other condition), based on the clinical information provided and the clinical appearance of the child with or without supportive evidence from the EEG (e.g. as in Rett and Angelman syndromes). Whenever the possibility of a clinical/genetic diagnosis was included in the EEG report, the referring clinician was asked to contact the EEG department with information that might confirm or refute the suspected genetic/chromosomal disorder or other condition. If this information was not provided, the authors did not attempt to contact the clinicians.

It was not the purpose of this study to evaluate EEG patterns in isolation when identifying focal structural or inflammatory lesions (e.g. cerebral tumours, encephalitis) or cerebral dysgenesis (e.g. lissencephaly).

Results

Three thousand and twelve EEGs were performed in 2780 children over the three years. The reasons why the EEGs were requested and the clinical diagnoses (including genetic/chromosomal conditions) suspected and subsequently confirmed in 17 of these 2780 children are shown in *table 1*. In two children, the clinicians who had requested the EEG had themselves suggested the possibility of the actual clinical diagnosis; one of the two children with Rett syndrome and one of the three children with fetal

Table 1. Genetic syndromes, chromosomal abnormalities and other conditions diagnosed in 17 of the 2780 children (numbers of children in brackets)[a].

Clinical diagnosis (numbers of children)	Features suggesting the diagnosis when reporting the EEG
Fetal valproate syndrome (3)	Facial appearance; family history of epilepsy on the EEG request
Rett syndrome (2)	Facial appearance; hand stereotypies; microcephaly (identified on EEG; EEG appearance [1 patient])
Tuberous sclerosis (2)	Large forehead - fibromatous plaque (1 patient); subtle facial rash (1 patient)
Angelman syndrome (1)	Facial appearance and behaviour
Freeman-Sheldon syndrome (1)	Facial appearance
Mucopolysaccharidosis type III (1)	Facial and body appearance
Myotonic dystrophy (1)	Facial appearance (child and child's mother)
Neurofibromatosis type 1 (1)	Macrocephaly; *café-au-lait* lesions on arms (seen when the child had the EEG)
Prader-Willi syndrome (1)	Facial appearance; small hands and feet
Williams' syndrome (1)	Facial appearance; clinical history of developmental delay and a "heart murmur" on the EEG request
Wolf-Hirschhorn syndrome (1)	Facial appearance; mannerisms
Miscellaneous chromosomal disorders: (trisomy 9p [1], mosaic trisomy 21 [1])	Facial and body appearance

[a] 15 of the 17 children underwent an EEG to either "confirm a clinical diagnosis of epilepsy" or to "classify the epilepsy/epilepsy syndrome"; the EEGs in the remaining two patients were requested to "confirm or exclude non-convulsive status epilepticus" (one of the two patients with Rett syndrome) and as part of an evaluation into possible developmental regression (the child with Wolf-Hirschhorn syndrome).

valproate syndrome. The clinicians who had requested the EEG in the remaining 15 patients had not suggested the underlying clinical diagnosis when submitting the EEG request.
Another 10 children were suspected of having a specific clinical (including genetic or chromosomal) disorder, but the suspected diagnosis was neither confirmed nor refuted as the clinician requesting the EEG did not respond; it is unclear how many of these children were either not investigated or in whom the investigations did not confirm the suspected disorder. The possible diagnoses in these 10 children included Angelman syndrome (two children), Rett syndrome (two children), trisomy 21 (one patient), Prader-Willi syndrome (one patient), mucopolysaccharidosis (one patient), fetal valproate syndrome (one patient) and a possible, but non-specific genetic or chromosomal disorder (based on the facial appearance; two patients).
There were no particular clinical features, in any of the EEGs of the remaining 2753 patents, either alone or in conjunction with the EEG patterns that confidently suggested a specific clinical or genetic/chromosomal disorder.

Discussion

One of the major benefits of digital video (CCTV)-EEG is the ability to record and document a child's clinical and behavioural activity whilst simultaneously recording their electrical cerebral activity. Previous studies have identified the importance of video-EEG in differentiating epileptic from non-epileptic paroxysmal events, the classification of seizure types, the identification of a specific epilepsy syndrome and non-convulsive status epilepticus and the localization of possible epileptogenic foci (Chen *et al.* 1995, Connolly *et al.* 1994, Saravanan *et al.* 2001, Shihabuddin *et al.* 1999). The additional benefits of video-EEG in the recognition and diagnosis of clinical (including genetic) conditions would appear to have been either not considered or unreported (Chen *et al.* 1995, Connolly *et al.* 1994). This is likely to reflect the fact that the focus of video-EEG reporting is on the EEG and its correlation with possible clinical epileptic (or non-epileptic) seizure activity. However, another explanation is that the majority of medical staff whose responsibility it is to report EEGs will be adult neurophysiologists, adult neurologists, psychiatrists or general practitioners (Ganesan *et al.* In press), who are unlikely to have had any clinical paediatric experience. In contrast, paediatric neurologists who routinely report EEGs (as in our hospital) are in a better position to recognize specific clinical (and genetic or chromosomal) disorders.
Clearly, this is very much an observational study and the yield of specific clinical disorders diagnosed on viewing the child on CCTV is, predictably, very low (17 of 2780 children [only 0.6%]). In addition, there are no data on the 10 children in whom a disorder was suspected, but neither confirmed nor refuted. It would have been useful to have contacted the clinicians who had requested the EEGs in these patents to provide this information.
It is very likely that the diagnoses of these 17 children would also have been established had one or both authors been asked to assess the children in the outpatient clinic, particularly as the EEG appearance was helpful in supporting the clinical diagnosis in only one patient (the child with Rett syndrome; *table 1*). One could reasonably argue that the 17 children in this study were provided with a 'two for the price of one' diagnostic report.
Finally, it must be acknowledged that the practice of suspecting specific clinical or genetic disorders on the basis of often limited clinical information and limited patient observation (on CCTV) may not necessarily be appropriate and could potentially lead to inappropriate and unnecessary investigations. However, the feedback from the referring clinicians has been very positive and has certainly led to a number of clinical diagnoses with consequent management and genetic benefits. □

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