Abuse or metabolic disorder?

EDITOR,—Hoffman and Naughton rightly raise awareness for the potential misdiagnosis of the inherited metabolic disorder glutaric aciduria type I (GA-I) for the shaken baby syndrome, with its attendant psychosocial trauma to the family.1

When considering the diagnosis, particularly when there is no convincing history of significant trauma, they advocate the evaluation of urinary organic acids and serum carnitine to exclude GA-I.2 The typical urinary metabolites glutaric and 3-hydroxyglutaric acids may be present in the urine only intermittently, or not at all in non-excretors, irrespective of residual enzyme activity, even at times of decompensation.3 If strongly suspected, glutaryl-CoA dehydrogenase activity in cultured skin fibroblasts should be measured to avoid missing this important diagnosis.3

Misdiagnosis might not only expose the family to the risk of false accusation of child abuse, but may have disastrous consequences for the child. GA-I is a potentially treatable condition if diagnosed when neurodevelopment is normal, before metabolic decompensation, which characteristically leaves a severe, irreversible, dystonic movement disorder. Aggressive treatment of intercurrent infections, the institution of hyperalimentation during catabolic crises, with long term dietary protein restriction and carnitine supplementation may avoid decompensation4 and therefore avoid striatal damage. Confirmation of the diagnosis additionally allows the possibility of prenatal diagnosis and early treatment of affected siblings. The true potential for prevention of neurological sequelae requires evaluation following the introduction of national neonatal screening for this condition by tandem mass spectrometry.5

The typical presentation every effort should be made to obtain the diagnosis, and must include enzymology on cultured skin fibroblasts.

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Teaching paediatrics for the developing world

EDITOR,—I agree with many of the sentiments expressed by Williams3 in his helpful article. I qualified in a developing country and started to practise as a part time general practitioner in “the capital”. I must confess that it did not take me long to realise that I was not prepared to recognise or handle the problems that were presented to an emergency surgery. I became confused and disillusioned. I thought I needed to go to the West to learn, as my medical school had failed to prepare me properly.

In England in the late 1960s I soon started to work in hospitals. Due to the language barrier, I had no difficulties in recognising or managing problems. However, it took me several years to realise that the initial problems that brought me to the West were universal—namely, that teaching hospitals throughout the world do not prepare doctors to function in their communities. Patients in hospital are not representative of the population as a whole, yet almost 100% of medical teaching takes place in these centres of excellence. Graduates are ignorant about the sociomedical problems of the society. The introduction of vocational training schemes for general practitioners training in the UK is a welcome, specialised career.

Medical schools worldwide still ignore this need or at best pay only lip service. If the native medical schools fail to impart appropriate knowledge to enable graduates to work and function in their own communities, how much more difficult will it be for a doctor from an entirely different cultural background? Even a well qualified community physician with great expertise in one country cannot assume that his or her skills will be appropriate in a completely different culture.

I have no doubt that both developed and less developed countries can learn from each other, but extreme care must be exercised not to have fragile and immature systems overtaken by purely philanthropic enthusiasm or self interest.

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Significance of the EEG after the first afebrile seizure

EDITOR,—Although Panayiotopoulos in his personal practice article and Cross in her accompanying commentary partly reject the significance of the EEG after the first afebrile seizure, I think that their arguments are not correct.

First, the EEG is not necessarily a “harmless” investigation. Although it may be relatively non-invasive and therefore physically harmless, it may be harmful in terms of its interpretation. An unsatisfactorily recorded EEG, undertaken by technical staff who have never been trained to perform such investigations in children, which is reported by a clinician who has not been taught the normal maturational as well as abnormal appearances of children’s EEGs, may result in inaccurate diagnoses of both epilepsy and the specific epilepsy syndrome. Clearly, this may have serious medical, psychological, and social consequences. Unfortunately, appropriately trained technical staff and paediatric neurologists or clinical neurophysiologists are not ubiquitous, particularly within the UK.

Second, epilepsy is a clinical diagnosis and is defined on the basis of not one but recurrent seizures, as clearly emphasised by Cross. The finding of an abnormal EEG, including the demonstration of characteristic features of a specific epilepsy syndrome, does not imply that the child will inevitably have a further seizure. Most parents are likely to elect not to give their child antiepileptic medication after a first seizure (even if it was a tonic–clonic convulsion), if they are told truthfully that it is possible that there will be no recurrence. Understanding, much depends on how parents are given this information. In my experience, their decision is usually made irrespective of the finding of an abnormal even “syndrome diagnostic” EEG, and the potentially high predictive risk of a further seizure. If there is any chance, no matter how small, that their child may not have a second seizure, most parents would prefer to adopt a wait and see approach.

Third, much of the discussion is clearly adult oriented. Importantly, the implications of a diagnosis of epilepsy and even an epileptiform EEG (as intimated by Panayiotopoulos’s argument) may be very different for a child and an adult, specifically in career/employment and leisure/driving. Epilepsy in children is not, and never should be, regarded as simply a downward extrapolation of epilepsy in adulthood.

These issues must be considered when interpreting EEG findings in relation not just to the seizure history but to the overall clinical situation. It must also be remembered that most children with epilepsy can be seen outside teaching hospitals or tertiary epilepsy centres and do not necessarily have access to the technical and clinical personnel who should be undertaking and reporting their EEGs.

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1 Panayiotopoulos CP. Significance of the EEG after the first afebrile seizure [see comments]. Arch Dis Child 1998;78:575–7.

Dr Panayiotopoulos comments:

Dr Cross partly accepts and Dr Appleton partly rejects the significance of the EEG after the first afebrile seizure. My reaction to their views is clinical, not EEG, matters. They rely on an unhelpful definition of epilepsy—two or more recurrent seizures—instead of emphasising that there are hundreds of seizure disorders with different and often diverse causes, manifestations, and prognoses that frequently mandate different short and long term management. Any further discussion would be pointless if our aim was to diagnose epilepsy and not a specific epileptic condition.

The inclusive term “epilepsy” or the simplistic rules “to treat or not to treat”, “good and bad”, “start with sodium valproate or carbamazepine after the second seizure” are detrimental to the diagnosis and management of epilepsies. It is because of these problems that in a recent report from London nearly half of children with typical absence
seizures are inappropriately treated with carbamazepine and vigabatrin.

Regarding EEG after the first afebrile seizure none of my four arguments was debated:
- It is possible to recognise children with features of specific epileptic syndromes.
- Minor seizures such as absences and myoclonic jerks that often escape clinical detection may be recorded.
- EEG is imperative in establishing seizure precipitating factors.
- An EEG in an untreated stage of an epileptic syndrome is imperative.

Instead, the responders insist on the significance of to treat or not to treat, with J. L. emphasised "is not a convincing argument: the prime aim in medicine is the diagnosis that determines prognosis and treatment strategies".

Appleton has four arguments that I reply to:
- Anything in medicine, clinical or laboratory, may be harmful if misinterpreted—raising standards, not abandoning the practice, is the answer.
- Clinical diagnosis may often be incorrect without EEG—see, for example absences versus complex partial seizures, visual seizures versus migraine or unreported myoclonic jerks.
- There is nothing adult oriented in my text.
- It is because most children are managed outside teaching hospitals that awareness of the various aspects of epilepsies should be improved and standards raised above "epilepsy is more than two seizures" or "the treatment of epilepsy is with drug A or drug B".

The message of my paper was that, as in all other fields of paediatrics, children with epilepsies are entitled to a diagnostic, prognosis, and management that is specific and precise. I should add that this is possible for most of the children with one or more seizures based on skillful clinical and EEG evaluation.

Systematic review of the school entry medical examination

EDITOR,—This important review of the school entry medical (SEM) examination is so flawed that the key messages cannot be supported. The School Health Service was introduced as a therapeutic, not just an epidemiological tool, when the Boer War revealed the magnitude of untreated disease for which neither treatment nor care was available. In 1976, the Court Report recommended that the SEM be a statutory examination: not to identify missed disease, but to support needy children, because of the profound impact of indifferent health, disadvantage, and developmental delay on educational progress. The Polnary Report, Health needs of young age children (1995), clearly advised that the health care assessment by school nurses at entry should also advance health promotion.

School health varies dramatically between inner city, urban, and rural schools. In our 'Trust serial audits' 7 to ascertain the decisions made about care and support offered to children at school entry, have demonstrated a skew in need with an eightfold increase in schools serving disadvantaged areas. The content of the work has changed dramatically over the past 30 years: consider child protection work, the integration of special needs children in mainstream school, the effects of serial parenting on young children. Medical time in school is spent with children with significant problems, not necessarily healthy children nor on perusing letters and reports. Numerical comparisons of pick up rates of defects over time (1962–96) and place are not meaningful.

Let us move the argument from the value of screening to the broader issue of how to provide effective health care for the many children who start school with health and developmental disadvantages.

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Dr J Barlow and Stewart- Brown comment:

We agree with Dr Leff that the most important role of the SEM is the provision of effective health care for children who start school with health and developmental disadvantages, through the identification of unmet health needs. It is, however, unclear how it is possible to assess the effectiveness of the SEM in meeting these health needs without assessing the effectiveness and efficiency of the screening procedures used in their identification.

We have stated that the evidence shows that large numbers of children are identified as having a problem at school entry and that many of these problems are newly identified as a result of the SEM. Furthermore, a large proportion of these problems result in referral for further investigation. However, the failure of most studies to follow up referrals to assess the number of false positive cases, or indeed to follow up the cohort to identify false negative cases, precludes the possibility of establishing the extent to which the SEM is actually successful in identifying and meeting children’s health needs. Neither is there any evidence available to show the success of either routine or selective SEMs in providing “care and support” to “needy children” as an integral part of the SEM, or in the positive promotion of health and the maintenance of a body of knowledge in the community regarding child health and development. We agree that the “numerical comparison of pick up rates” is unsatisfactory. However, this is all the literature appears to provide for most of the children with one or more seizures based on skillful clinical and EEG evaluation.


Chinese motor paralysis

EDITOR,—Chinese motor paralysis, or acute motor axonal neuropathy, is a re-emerging acute inflammatory neuropathy that differs from classic Guillain–Barré syndrome in clinical, neurophysiological, and pathological features.1 Cases have been reported from China, Japan, India, and South America,2–7 but not previously in northern Europe.

A 14 year old boy presented with a five day history of poor grip and falling from his bicycle, followed by progressive symmetrical mainly proximal weakness affecting both
upper and lower limbs equally, without an ascending picture. He had been constipated for two days and had developed hoarseness and a weak cough. There was no other relevant history. Examination showed mild weakness of jaw opening and facial musculature; complete inability to flex his neck against gravity (MRC 2) with normal neck extension (MRC 5), and predominantly proximal weakness (MRC 3) with less severe distal weakness (MRC 4). Distal tendon jerks were absent, but proximal jerks could be elicited. His peak flow rate was 30% of predicted. He received intravenous immunoglobulin (IVIG) 0.4 g/kg daily for five days. On day 2 he had a day 19, and walked alone on day 5. He was able to write clumsily. By day 5 he began to improve; thereafter he showed recovery of function persisting until day 4 when an enema was given. Despite the severe axial and proximal weakness, distal power was less affected to the extent that he was still able to write clumsily. By day 5 he began to improve; he was extubated on day 14, walked with support on day 19, and walked alone on day 37. However, only by 22 months was power completely normal. He also developed a unilateral neuropathic tremor. Investigations showed an initial lymphopenia (normal cerebrospinal fluid at 48 hours with a protein of 0.4 g/l, a rise in serum creatinine, normal cerebrospinal fluid at 48 hours and 15 days later, and upgoing plantar responses. Constipation persisted until day 4 when an enema was given. Despite the severe axial and proximal weakness, distal power was less affected to the extent that he was still able to write clumsily. By day 5 he began to improve; he was extubated on day 14, walked with support on day 19, and walked alone on day 37. However, only by 22 months was power completely normal. He also developed a unilateral neuropathic tremor. Investigations showed an initial lymphopenia (normal cerebrospinal fluid at 48 hours with a protein of 0.4 g/l, a rise in serum creatinine to seven times the upper limit of normal by day 3, which fell to normal by day 8, and normal serum IgM anti-GM, and IgM anti-Mag (tumour necrosis factor) (untypeable) was isolated from rectal swabs. Muscle biopsy on day 4 showed some central nuclei and two small angulated fibres only. Neuropathological investigations at seven and 15 days post-onset showed pronounced reduction of the amplitudes of the compound muscle action potentials, but normal distal latencies, motor nerve conduction velocities, and sensory nerve conduction studies. This is the first report of Chinese motor paralysis acquired in northern Europe. Our patient resembled previous reports from Asia in that he had severe involvement of facial, bulbar, and nerves, with only minimal inflammation and demyelination and relative sparing of dorso-lateral roots even in advanced cases. In China annual epidemics occur associated with Campylobacter jejuni infection and anticytoplasmic antibodies. Brain 1995;118:597–605. Griffin JW, Li CY, Ho TW, et al. Guillain-Barre syndrome during the 1992–93 filmches of neuropathological changes in clinically defined cases. Brain 1995;118:577–95.

Current means of obtaining a PhD in the UK

EDITOR,—Following my study into MD degrees I investigated the current means of obtaining a PhD in the UK. The same questionnaire was sent to the 10 doctors whose PhD was quoted in the paediatric/perinatal section of the Index to theses with abstracts journal in 1986–95. Nine replies were obtained. Though this was a small study one can make some comparisons between MDs and PhDs. All candidates pursuing a PhD, unlike all those doing an MD, had research posts, the shortest being for 30 months. The median length of their research period, compared to two years for the MD candidates. Surprisingly, none of the PhD students in contrast to 29% of the MD candidates, were able to submit their PhD within their research period. PhD students spend a smaller part of their time (median 10% compared to 20%) on non-research related activities. It would seem that almost all higher degree students have clinical commitments and demands unrelated to their research and there may be a case for adding six months to the research period to allow for this inescapable fact. All of the PhD supervisors had a higher degree, whereas only 78% of the MD supervisors had an MD or PhD. PhD students also had more meetings with their supervisors. PhD students received their theses back from examiners more quickly that the MD students (Mado C, three compared with six months). Only 22% of PhD candidates had to make revisions to the theses compared with 46% of MD candidates. All PhD candidates had an oral examination compared to 47% of MD students. The experience of PhD candidates who needed to make revisions to their theses may reflect the fact that their research projects were superior or possibly that they were better supervised. Alternately, the oral examination may have given candidates the opportunity to explain or elaborate on certain points, thus decreasing the number who needed to make revisions. Interestingly, the length of time from the start of the higher degree to confirmation that it had been obtained was equal for PhD and MD students (median 54 months); however, given that a PhD generally requires a longer research period and is a more detailed study, this reflects better on PhD candidates.


Depth of insertion of a lumbar puncture needle

EDITOR,—We read with interest the article on the depth of insertion of a lumbar puncture needle. There are various factors that one might like to make. First, the height of the child has to be recorded before applying the quoted formula or referring to the graph produced by the study. In our experience, having worked at busy casualty and acute paediatric units, height is probably more difficult than weight to measure in an acutely unwell child, who is distressed and extremely unwilling to be separated from their parents. Second, in most cases of failed attempts, it is because the needle is off midline rather than not being inserted to an adequate depth. So if one could ensure correct needle orientation and proper positioning of the patient, a reasonably high success rate in obtaining an uncontaminated cerebrospinal fluid sample should be achievable.

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BOOK REVIEWS


Edward Brett’s textbook of paediatric neurology remains a standard reference for this growing specialty. When first published in 1983 it was one of a relatively small number of books on the subject but now has to compete with many other quality reference books. The knowledge base in paediatric neurology is expanding, as predicted by Brett in the preface to his first edition. There was a significant increase in information between the first and second edition, published in 1989. The third edition shows a further, although less dramatic expansion. The third edition is more of a multiauthor text, which does slightly detract from the very personal approach so characteristic of the first edition.
The new information is important, including data on the incidence of Haemophilus influenzae vaccination in the United Kingdom, expanded discussion of central nervous system involvement in HIV and AIDS, and some discussion of the prion diseases. The third edition has lost the important chapter on neurogenetics (presumably an encouragement for us to purchase Dr Baraitser's neurogenetics database).

The strength of Brett's book remains his personal style, with information drawn from a breadth and depth of personal experience few of us will achieve in our lifetimes. For those of us who have been privileged to work with the author the pages come alive with his presence, with emphasis on the basis of all good medicine—that is, taking a history, careful examination with appropriate investigation to confirm the clinical diagnosis. The practising clinician needs to have a good understanding of the important rare conditions. This is not a book of lists but its contents are enriched with historical background and other information derived from long personal experience. I often refer patients to this book, including families who have a child with one of the more distressing conditions, because of the thorough and sympathetic manner in which the disorders are discussed. Paediatric neurology remains one of the great clinical specialties. This text is written by a clinician with the clinical approach in mind and will remain an important source reference for me for many years to come.

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There is a continuing need for a book of manageable size that can be used as an introduction and resource by trainees and professionals whose work brings them into contact with issues concerned with child mental health. Most texts in English that aim to perform this function are out of print so there is an important gap to be filled. Although Goodman and Scott have gone a long way to achieve their goal there are a number of deficiencies and disappointments.

The audience explicitly includes a wide range of disciplines from education, social work, nursing, and psychology to paediatrics, psychiatry, and general practice, yet there is remarkably little mention of the contributions of disciplines other than child psychiatry to the assessment and treatment of children with psychological and psychiatric disorders. For example, there is no significant account of psychological assessment nor the manner in which teachers can be included in treatment programmes.

The ordering of certain chapters and sections seems unsatisfactory so that assessment precedes classification and epidemiology, and risk factors follow detailed accounts of a range of specific disorders. Incidentally, maltreatment of children is included as a specific disorder presentation when strictly it is a risk factor.

There is more emphasis on factors than processes with one or two notable exceptions—for example, in the section on psychosomatisms, where the authors there are chapters on preschool problems and disorders of adolescence, a developmental perspective is not strongly represented.

Assessment is not put in context so that how different components of assessment fit together is not discussed. There appears to be a misunderstanding about what constitutes good semi-structured interviewing, and the suggested scheme for history taking is full of closed questions. There is a notable weakness in proposed methods of learning about relationships. Formulation is referred to much later in the book but is not developed either in the assessment or classification chapters. The manner in which the chapters on specific disorders and presentations relate to classification is not explained, and there is considerable variation in their organisation. One consequence is that information is not systematic about all disorders—for example, there is nothing on the treatment of substance abuse and use. Comorbidity is barely discussed. This is an important issue. The authors raise the question of whether conduct disorders are appropriately dealt with by child psychiatric services when those disorders are “clearly, socially determined”. As the authors demonstrate, there are a wide range of factors and other disorders that are commonly associated with conduct disorders. These factors and their attendant processes are often missed in assessment, pointing to the need for medical and psychological input. Related to this, only school refusal is given a chapter, not truancy or the wider range of school attendance difficulties.

Related to the last point there is a tendency to give less weight to the contribution of social factors; this is exemplified in the discussion of models for the link between mental retardation and psychiatric disorders. The model of a common genesis of low IQ and psychiatric problems from social factors is dismissed, yet four pages later when considering brain disorders it is acknowledged that there is at least continuing controversy as to whether children with brain disorders are “more vulnerable to ordinary risk factors or simply as vulnerable”. In other words the notion that there can be an interaction between brain dysfunction and social factors just as there could be between social factors and IQ when treating psychiatric problems, is overlooked under mental retardation.

Parenting skills gets a good deal of mention but ‘parent-child’ relationships are very little. There is a chapter on attachment in the risk factors section but no full discussion of parenting and the parent-child relationship in other respects.

The chapter on preschool problems is particularly disappointing with little acknowledgement of the importance of prevention.

The lack of emphasis on broader social factors is sustained in that these are not included in the risk factors section even though there is good evidence for the impact of wider social factors, such as community and housing, both by their effect on parenting and more directly on the older child. Incidentally only peer popularity and unpopularity are referred to and not other peer influences.

The treatment section understandably gives greater weight to evaluated treatments but this leads to what appears to be a rather positive account of behavioural approaches, although the quoted outcome studies have only palpable effect sizes for problems, many of which fall outside the domain of child psychiatry, such as antisocial behaviour in pubertal children, and wetting. There is nothing on inpatient treatment and no section on broader aspects of management such as working with other agencies including schools.

Finally the different sections are not clearly related to each other so that it becomes a little strange to see that there is more on treatment of enuresis than of almost any other condition in the specific disorders section. This becomes understandable when it is appreciated that the treatment section aims to address treatment issues related to a number of different conditions.

Despite the criticisms there is much to commend the authors’ achievement in succinctly summarising so much information in a readable fashion, although there are occasions when terms are used without adequate explanation at the first usage. For example, the reference to attachment relationships on page 6 when the attachment chapter starts on page 199, and the undefined reference to significant harm on page 166 in the maltreatment chapter.

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Abuse or metabolic disorder?

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