Fortnightly review

Epilepsy in childhood

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Childhood epilepsy requires integrated medical, educational, and community services, and its treatment spans acute and disability medicine.

There are many childhood epilepsies, and seizures are the commonest paediatric neurological symptom. Epilepsy—that is, susceptibility to continuing seizures—occurs in 0.5-1.0% of the population and is intractable to current antiepileptic drug treatment in 20-25%. Epileptic seizures, including febrile convulsions, occur in 3-5% of children. Epilepsy starts in childhood in 60% of cases, and most of the clinically significant aspects of the disease occur during childhood.¹

Method

I chose the studies on which I have based my review because they support the development of clinical guidelines. The six crucial clinical issues are:

- Identifying non-epileptic seizure syndromes
- Identifying benign epilepsy syndromes with a comparatively good outcome
- Identifying malignant syndromes in which continuing and increasing disabilities occur and need detailed investigation, aggressive treatment, and research
- Identifying, assessing, and managing the four major disabilities that result from early brain damage or dysfunction and commonly coexist—epilepsy, motor disorder (cerebral palsies), cognitive disability, and psychiatric disability—particularly for the many epilepsy syndromes that start in the first two years of life
- Ensuring that antiepileptic drugs are used in an ordered fashion with audit of results
- Anticipating and managing status epilepticus.

Non-epileptic attacks

Non-epileptic attacks include simple blue breath holding attacks, white reflex asystolic attacks, reflex syncope, benign paroxysmal vertigo, benign myoclonus of infancy, vasovagal attacks, sleep phenomena, pseudo-seizures, and Munchausen syndrome by proxy. There are more than 20 such syndromes, identified by specific clinical characteristics² and the setting in which they occur, particularly the provoking factors.

Simple blue breath holding attacks, usually after the age of 6 months, provoked by frustration or other upset, consist of a positive Valsava manoeuvre, cyanosis, stiffening, and coma. White reflex asystolic attacks may start earlier, usually follow minor injury,

Summary points

Identify non-epileptic attacks

Identify benign and malignant syndromes

Use predicted prognosis to guide management

Identify and manage coexisting disabilities

Ensure multidisciplinary and multiagency coordination of treatment

Select cases amenable to surgery as soon as possible

and consist of vagal asystole, pallor, rapid coma, stiffening, and opisthotonos. Faints while standing or sometimes from sitting are usually well recognised. All three conditions may be accompanied by late true epileptic phenomena of jerking secondary to cerebral hypoxia or ischaemia, but their natural history remains self limiting and harmless like that of the first episode. Such episodes are, however, frightening for carers.

Non-epileptic attacks may be misdiagnosed and treated with antiepileptic drugs, the attacks tending to persist despite treatment and good prognostic features (see table 1).

Management of epilepsy

The principles of management of epilepsy fall easily into a problem oriented approach:

- Take detailed description of seizures
- Draw up a problem list and include some assessment of cognition
- Clarify the problem list by doing investigations
- Decide on the need for treatment
- Predict an outcome
- Review predicted outcome at a suitable time and the problem list annually, including a description of a typical day in the child's life.

A description of the seizures is the main diagnostic evidence, and a systemised approach to collecting clinical data is required (box on next page). Several classifications of epileptic seizures, including that of the International League Against Epilepsy, exist but

Classification of seizures

Partial with early motor phenomena (described)—for example, tonic-clonic seizures and automatic movements

Partial with early aphasic phenomena (described)

Partial with early sensory phenomena (described)—visual, limb tingling, auditory, gustatory

Partial with early psychological phenomena (described)—for example, fear

Partial with loss of awareness

Partial with postictal phenomena (described)

Partial with secondary generalisation (described)

Generalised tonic-clonic:

- With focal features
- Without focal features

Generalised tonic

Infantile spasms:

- With focal features
- Without focal features

Myoclonic:

- Focal (described)
- Axial

Atonic

Absence seizures

- Simple
- With additional or atypical features

contain hybrid data on both seizure types and epilepsy syndromes. The box shows a classification of seizures in which judgment about the degree of disturbance of consciousness is left to a second stage without initial separation into simple and complex partial seizures.

The main advance in understanding childhood epilepsies and their logical investigation and treatment has been the delineation of epilepsy syndromes and their separation into benign and malignant prognoses. In my examples, however, I emphasise that benign may be relative. Many children's seizure disorders do not fit a described syndrome, but the prognostic factors in table 1 may be used to predict an outcome.

The genetic localisation of seizure syndromes is separable into pure lesionless conditions that have epilepsy as their only initial manifestation and conditions with lesions that commonly cause epilepsy but which have additional primary manifestations. The primary pure genetic epilepsies with simple mendelian inheritance and gene localisation³ are benign familial neonatal convulsions, benign familial infantile convulsions, partial epilepsy with auditory symptoms, autosomal dominant nocturnal frontal lobe epilepsy, and progres-

Description of seizures

Setting or provocation

Warning or early phase ("aura")

Motor aspects:

- Type of movement
- Distribution

Sensory aspects:

- Type
- Distribution

Responsiveness

Length

Colour change

Recovery phase

What was it like for child and carer?

Table 1 Good and adverse prognostic features for outcome of epilepsy

Good outcome	Adverse outcome
Single seizure type	Multiple seizure types
No additional impairment	Additional neurological impairment (especially in cognitive function)
Late age of onset	Early age of onset (for the syndrome)
Episode is related to illness with full recovery or was provoked	Spontaneous seizures
Short seizures	Status epilepticus
Low rate of seizures	High rate of seizures
(Good response to antiepileptic drugs)	(Poor response to antiepileptic drugs)

sive epilepsy with mental retardation (Northern epilepsy). Possible localisations are proposed for three epilepsies with complex inheritance: febrile convulsions, juvenile myoclonic epilepsy, idiopathic generalised epilepsy.

Neonatal seizures

Benign and malignant epilepsy syndromes are both seen in the neonatal period. Despite an overall poor prognosis for both survival and future impairments (largely cognitive and motor), separating the syndromes into those with a comparatively good outcome and those with a poor outcome is useful. Those with a good outcome include benign familial neonatal seizures, fifth day seizures,⁴ and seizures secondary to hypocalcaemia and subarachnoid haemorrhage. Those with a poor outcome include seizures secondary to hypoxic ischaemic encephalopathy and developmental brain defects. Thus if neonatal seizures are age limited without persisting neurological abnormality, the prognosis is excellent and further investigation and long term antiepileptic treatment are not required.

Two rare treatable recessively inherited disorders—pyridoxine dependent epilepsy and biotinidase deficiency—may cause spontaneous seizures in the first two years of life. All babies with unexplained epilepsy should receive pyridoxine 100 mg/day orally, the initial dose being given in hospital in case of a hypotonic reaction,⁵ and a trial of biotin or measurement of biotinidase.

Benign syndromes of epilepsy

Benign syndromes are age limited, stereotyped, and often provoked or provokable; they occur in otherwise normal children and are commonly genetic.

Febrile convulsions

Febrile convulsions are epileptic seizures provoked by fever of extracranial infective origin that occur in at least 3% of children, mainly between the age of 6 months and 5 years. Intracranial infection must be excluded, particularly meningitis, which is indicated by drowsiness and irritability, and fever may show underlying epileptogenic disease. The seizures are usually brief, bilateral clonic, or tonic-clonic attacks. About 10% of children have complex, prolonged, or focal seizures requiring emergency treatment for status epilepticus. Severe sequelae such as hemiplegia are now uncommon, but there has been much dispute about the risks of continuing epilepsy. The collaborative perinatal project showed that the risk of subsequent epilepsy and non-febrile seizures was 10% even with all

three adverse factors (family history of epilepsy, pre-existing neurological abnormality, and complex initial seizure).^{6,7} Surgical⁸ and magnetic resonance imaging studies,⁹ however, both showed a high rate of previous febrile status epilepticus in mesial temporal sclerosis, and a causative pathogenic sequence is supported by animal studies.¹⁰ The low individual risk from epidemiological studies¹¹ could nevertheless, in this common disorder, produce 100-200 cases of mesial temporal sclerosis a year in the United Kingdom. There have been several reviews of these data.¹² The practical conclusion was that febrile status should if possible be prevented and vigorously treated.

Febrile status epilepticus is, however, commonly the first seizure,⁶ and subsequent preventive measures may have little effect. Both phenobarbitone and sodium valproate reduce the rate and severity of febrile seizures; sodium valproate is preferred because phenobarbitone has behavioural side effects. The choice is between continuous prophylaxis and the contingent use—that is, at onset of seizures—of rectal diazepam for those with a history of complex or frequent febrile convulsions. Rectal diazepam is the commonest treatment. Prophylactic management of fever may be effective. Attending to why a child has many febrile illnesses is also important.

Benign epilepsy with Rolandic foci

Benign epilepsy with Rolandic (centrotemporal interictal) foci has an onset between 2 and 12 years, is maximal between 7 and 10 years, and stops by 13 years. The attacks are clonic, partially sensorimotor affecting the face, bulbar (tongue and pharyngeal) muscles, hand, and arm, and they occur particularly on waking. An electroencephalogram shows characteristic patterns. The children are free of major impairments but may have neuropsychiatric abnormalities.¹³ Carbamazepine is commonly used if seizures are unpleasant. When adverse features are present (table 1), imaging is required to exclude a Rolandic lesion.

Benign occipital epilepsy

Benign occipital epilepsy with sharp and slow wave activity suppressed by eye opening is problematic. Some children with typical partial seizures with visual phenomena, postictal headache, and migrainous characteristics¹⁴ have a benign prognosis, but others have a poorer cognitive and seizure outcome, and some have lesions. The rules on the importance of cognitive function in predicting the outcome of seizures apply (table 1).

Early myoclonic epilepsies

Separation of early myoclonic epilepsies into benign and malignant outcomes may not be possible early.

Absence seizures

Typical absence seizures are uncommon. Their separation from daydreaming is clinical. Absence seizures totally interrupt activities in which the child is engaged for several seconds. The outcome, mainly the rate of tonic-clonic seizures in adult life, can be predicted by the expected adverse factors: history of tonic-clonic seizures, an intelligence quotient (IQ) of <90, and a family history of seizures. Seizures continue in about 10% of cases with no adverse factors or only one, and

in all cases with three adverse factors. Atypical and complex absence seizures may have in addition clonic, tonic, or automatic movements and an altered sensorium before or after. Electroencephalograms help diagnosis and treatment. Phenytoin and carbamazepine are unhelpful or aggravate the condition, particularly myoclonic absence seizures. Absence seizures range from a benign self limiting condition to part of a malignant syndrome of multiple seizure types, particularly the Lennox-Gastaut syndrome.

Malignant syndromes of epilepsy

Although developmental arrest and regression as a consequence of epilepsy are common in malignant syndromes of epilepsy, investigations for metabolic, degenerative, and structural brain disease are essential. The most severe example is early myoclonic encephalopathy, which usually presents neonatally with fragmentary myoclonus and other seizure types, with later massive myoclonic and tonic seizures. Electroencephalography shows suppression or burst activity, and one cause is non-ketotic hyperglycaemia. Mortality is 50%, and surviving children are severely disabled.

Infantile spasms

Infantile spasms, or West's syndrome, consist of runs of tonic spasms occurring every 5-10 seconds. The commonest spasms are flexion, but extensor and mixed types are seen and may show lateralisation. Parents may notice loss of visual and social interaction rather than spasms. West's syndrome is symptomatic of many static cortical diseases, and with modern imaging and a careful history, only 10% remain without antecedent problems.¹⁵ Attacks start in the first year with a peak onset at around 4 months, the outcome being worse with early onset. A grossly disorganised epileptic electroencephalogram (hypsarrhythmia) is characteristic but not always seen.

The mainstays of treatment have been corticosteroids (tetracosactrin or prednisolone), but recent studies have shown vigabatrin to be effective,¹⁶ although a comparative study has yet to be performed. West's syndrome is sometimes a secondary generalised epilepsy with obvious lateralised lesions; some studies using positron emission tomography have shown cold (hypometabolic) lesions, which have occasionally been excised with relief of seizures.¹⁷ The occurrence of the syndrome is no bar to the surgical treatment of focal epilepsy, but the developmental outcome will be adversely affected if the syndrome is present—for example, the generally poor cognitive outcome and autistic features of children with tuberous sclerosis and other diseases with early onset seizures, including West's syndrome.¹⁸ A death rate of about 20% and the occurrence of cerebral palsy in 30-50% and cognitive disability in up to 85% should prompt early ascertainment, energetic treatment, and further research.

Chevrie and Aicardi showed the poor developmental outcome for most children with spontaneous seizures—that is, seizures that are not provoked by fever, infection, trauma, or metabolic disturbance—if they start in the first year of life, with or without West's syndrome.¹⁹ Thus all children with early onset spontaneous seizures should be urgently referred. The

non-spontaneous group includes non-accidental head injury, meningitis, and hypernatraemic dehydration, so only simple febrile convulsions may be managed in primary care.

Severe myoclonic epilepsy of childhood

Severe myoclonic or polymorphic epilepsy begins at 4-10 months, often with a prolonged partial clonic seizure associated with moderate fever. Later attacks include multiple seizure types, including complex absences, myoclonias, complex partial seizures, and apnoeic attacks. With the onset of multiple types of seizure, developmental regression, often severe, occurs.²⁰

Lennox-Gastaut syndrome

In the Lennox-Gastaut syndrome, multiple seizures occur in the first eight years of life. A single seizure type including infantile spasms may occur beforehand. The seizures include tonic attacks (particularly in sleep), atypical absence seizures, atonic and myoclonic attacks, and episodes of non-convulsive status, with long periods of impairment of motor and cognitive function. Half of the children affected have primary developmental delay, and all have learning problems after five or more years of the condition; only up to 10% have a reasonable outcome. Characteristic electroencephalograms show bilateral variable slow (1-2 Hz) spike-waves.

Seizures are often intractable to antiepileptic drugs, but benzodiazepines, sodium valproate, lamotrigine, corticosteroids, and a ketogenic diet are regularly used. Drug intoxication may contribute to disability. Felbamate has been used with good effect, but its use is restricted because of side effects. The underlying pathology is either unknown or closely overlaps that of infantile spasms. Occasionally resective surgical treatment can be offered, and the palliative procedure of callosotomy may reduce severe drop attacks (axial myoclonic or atonic seizures that cause sudden falls and head injury). The Lennox-Gastaut syndrome illustrates the apparently insurmountable problems of providing a coherent medical, social, and education programme for the children and families who face dealing with severe epilepsy and continuously varying levels of function. Many of these children do not attend education unless special multiagency help is available.

Partial seizures of lesional origin

Partial seizures of lesional origin are diverse, and surgical treatment may be offered. The definitive study of the natural history of partial seizures of temporal lobe origin predates modern imaging.^{21 22} A mortality of at least 0.5% per annum and an outcome of a third with full recovery, a third with continuing problems, and a third being totally dependent was reported. The predictors of poor outcome were similar to those in table 1 and are apparent early, giving strong justification for surgical intervention in childhood rather than later in adult life, as is still often the case. Younger children, however, present with a wide range of clinical syndromes, including early cognitive and autistic regression with developmental tumours of the temporal lobe, which may constitute a surgical emergency. Clinically significant recovery from autistic features may be achieved.²³

Landau-Kleffner syndrome

The Landau-Kleffner syndrome is uncommon. Typically, partial or generalised seizures (including atypical absence seizures) begin and language comprehension and speech are lost after two or more years of normal development. Aphasia is not related to frequency or severity of seizures, and clinical seizures may be absent in about 20% of children, but they will show typical language regression and electroencephalographic changes (variable temporal or temporoparietal multifocal spikes or spike-wave complexes intensified in sleep, including non-convulsive status epilepticus).²⁴ The range of impairments when early aphasia, epilepsy, and typical electroencephalographic abnormalities are present includes behavioural disorders and global cognitive, motor,²⁵ and social regression with many autistic features. The contribution of this epileptic mechanism to autism is not resolved. Structural imaging usually gives normal results, and lesional variants are mentioned earlier. Antiepileptic drugs often have no effect on the cognitive deficit, but high doses of sodium valproate, often in combination with lamotrigine, sometimes help. A clinically significant, sometimes dramatic, response to corticosteroids makes them essential, but side effects and long term dependency may be a problem. Preliminary results of surgical treatment using multiple subpial transections suggest that about half of those with clear evidence of a driving hemisphere can be helped.²⁶

The Landau-Kleffner syndrome is a good model for cognitive arrest and deterioration in epilepsy. One aspect of its pathophysiology is that part of the temporal lobe is chronically hypoperfused and hypometabolic.²⁷ Thus two pathogenic mechanisms seem to exist, one fast and electrical, producing clinical seizures, and the other with a slower and vascular or metabolic component, related to non-convulsive status in sleep and producing prolonged cognitive deficits.

General features

Several of these conditions (infantile spasms, polymorphic epilepsy, the Lennox-Gastaut syndrome, and the Landau-Kleffner syndrome) illustrate the degenerative effects of severe epilepsy with loss of cognitive and social functions but without change in magnetic resonance images or rate of head growth. The general designation of an epileptic encephalopathy seems justified and prompts further research into these potentially remediable but currently severely disabling conditions. The question posed by Sadzot's recent editorial, "Do seizures beget seizures?"²⁸ is perhaps not the main issue in childhood when cognitive arrest and dementia are higher research priorities.

The problems faced by families dealing with these syndromes of epilepsy has promoted the development of several excellent parent support groups.

Drug treatment

There are few well controlled studies and no placebo controlled studies to justify much of how childhood epilepsy is managed with antiepileptic drugs. Thus arguments about whether to treat after the first, second, or third seizure are based on a general approach rather than evidence. The case for not necessarily treating mild seizures and for inpatient withdrawal of all drugs

Table 2 Drugs of use in different types of childhood epilepsy

	Dose (increments, when applicable)	Usual maximum daily dose (mg/kg/day)	Blood reference range
Generalised tonic-clonic and focal seizures			
Carbamazepine	4 (4) mg/kg/day	20	Up to 50 µmol/l (epoxide metabolite) up to 9 µmol/l
Sodium valproate	10 (5) mg/kg/day	40	Up to 700 µmol/l
Phenytoin	5 (2) mg/kg/day	10	Up to 80 µmol/l
Vigabatrin	30 (10) mg/kg/day	80	40-270 µmol/l
Phenobarbitone	3 (2) mg/kg/day	5	Up to 170 µmol/l
Primidone	10 (5) mg/kg/day	20	Up to 60 µmol/l
Lamotrigine:			
Alone	1 (2) mg/kg/day	10	4-16 µmol/l*
With sodium valproate	0.1 (0.1) mg/kg/day	1.5	4-16 µmol/l*
Gabapentin†	5 mg/kg/day (5 daily)	70	Up to 120 µmol/l
Topiramate†	0.5-1 (1) mg/kg/day	4-5	6-74 µmol/l
Prednisolone‡	2 mg/kg/day	NA	NA
Ketogenic diet‡	NA	NA	NA
Chlormethiazole	10-20 mg/kg/day	NA	3-18 mg/l
Absence seizures			
Ethosuximide	10 (5) mg/kg/day	30	700 µmol/l
Sodium valproate	10 (5) mg/kg/day	40	Up to 700 µmol/l
Lamotrigine	1 (2) mg/kg/day	10	4-16 µmol/l*
Myoclonic seizures§			
Nitrazepam	0.1-0.5 mg/kg/day	0.5	50-150 µg/l
Clonazepam	0.05-0.2 mg/kg/day	0.2	25-85 µg/l
Clobazam	0.25-1.0 mg/kg/day	1.0	Up to 200 µg/l (desmethyl clobazam (metabolite) up to 2000 µmol/l)
Sodium valproate	10 (5) mg/kg/day	40	Up to 700 µmol/l
Lamotrigine	1 (2) mg/kg/day	10	4-16 µmol/l*
Febrile seizures			
Intermittent diazepam			
Sodium valproate	10 (5) mg/kg/day	40	Up to 700 µmol/l
Phenobarbitone	3 (2) mg/kg/day	5	Up to 170 µmol/l
Antibiotics			
Status epilepticus			
Diazepam	Intravenous or rectal 0.15-0.3 mg/kg	NA	1 mg/l (desmethyl diazepam (metabolite) up to 1.5 mg/l)
Lorazepam	Intravenous 0.03-0.05 mg/kg	NA	1.5 mg/l
Phenytoin	Intravenous 15-20 mg/kg (slowly)	NA	Up to 80 µmol/l
Chlormethiazole	Up to 0.7 g/h (adult dose), 300-800 mg in 100 ml (4 ml/min) (5-18 mg/kg/h)	NA	3-18 mg/l
Thiopentone	4-8 mg/kg (induction), 10 mg/kg/h (ventilation)¶	NA	NA
Infantile spasms			
Tetracosactrin depot	0.5 mg	1 mg	NA
Prednisolone	2 mg/kg/day for 2 weeks	NA	NA
Vigabatrin	Up to 150 mg/kg/day	150	40-270 µmol/l
Nitrazepam	0.1-0.5 mg/kg/day	0.5	50-150 µg/l
Sodium valproate	10 (5) mg/kg/day	40	Up to 170 µmol/l

NA=not applicable. *To be revised upwards. †Not licensed for use in children. ‡Also for intractable myoclonic epilepsy and absence seizures. §Do not use carbamazepine, phenytoin, or vigabatrin.

¶Not licensed for use in children under 2 years.

in poorly controlled epilepsy has been strongly argued. The suggested approach is to use the predicted natural history as the main guide to treatment. A randomised, controlled study of single drug treatment in children aged 3 years and over with partial and generalised tonic-clonic seizures showed similar rates of control and side effects with phenytoin, carbamazepine, and sodium valproate.²⁹ The study used slowly increasing increments of drugs and blood concentrations up to the top half of a defined therapeutic range. Phenobarbitone was removed early because over half the children had unacceptable behavioural side effects. Because of the predictive importance of previous

neurological, particularly cognitive impairments, children were stratified for neurodisability, and this proved to be a significant predictor of seizure control and a crucial element of such paediatric studies.

Table 2 shows the drugs used in childhood epilepsy. Drug licensing does not cover all of the current uses in children, and parents should be counselled appropriately. My approach to treatment is:

- To decide on the need for treatment on the basis of prognosis
- To select a single drug and increase the dosage slowly, giving the family clear information on how to manage status epilepticus
- To maximise the drug dosage if seizures continue
- To add a second drug if epilepsy is not controlled, with combined treatment lasting for sufficient time to assess the second drug as a possible line of treatment before withdrawal of the first
- To try further drugs and instigate detailed investigations if the epilepsy proves intractable to two drugs. If adverse factors are absent the diagnosis of epilepsy should be questioned
- Not to react to a brief cluster of seizures if the epilepsy is generally well controlled
- To investigate urgently deterioration in any domain of function.

The use of new antiepileptic drugs in children has been recently reviewed by Pellock.³⁰ Ketogenic diets have been in use in epilepsy for 75 years and come in two forms: using animal fat and, more recently, medium chain triglycerides. Several reports support their use,³¹ but well controlled studies of comparative (with antiepileptic drugs) and selective (which type of epilepsy) efficacy are not available.

Although the concomitant use of psychotropic drugs with antiepileptic drugs in epilepsy needs careful monitoring, behavioural and drug treatment of hyperactivity and attention deficit can be successful, but this issue requires further study.

Surgery

The surgical treatment of childhood epilepsy is growing rapidly, justified by studies of the natural history of the disease, which show that waiting for remission in most children with intractable epilepsy is a false hope. If a surgical solution is possible it should

Indications for and types of surgery

Indications for assessment

Intractable partial seizures
Intractable hemiepilepsy
Intractable, apparently generalised, epilepsy with focal clinical electroencephalographic or radiological features
Regression in epilepsy

Procedures

Lesionectomy
Focal resections:
Temporal
Extratemporal
Hemispherectomy
Functional procedures:
Callosotomy
Subpial transections

therefore be used early in the child's decline. The main advances allowing these developments have been magnetic resonance imaging,⁹ T2 mapping,³² proton spectroscopy,³³ single photon emission tomography between and during seizures,³⁴ and video telemetry. The use of these magnetic resonance techniques in children requires sedation, anaesthesia, and age appropriate sequences and normative values. The complexity of these children's problems requires a team of paediatric neurologists, neurosurgeons, anaesthetists, neuropsychologists, psychologists, psychiatrists, speech therapists, nurses, and neuropathologists.

The use of surgery for epilepsy is developing rapidly.³⁵ Part of paediatric practice is the young end of adult series, which is dominated by temporal lobe resections for mesial temporal sclerosis (fig 1). In one study of surgery in children, half of the subjects were 6 and younger, a group that usually has behavioural and cognitive impairments.²⁴ Most would not enter adult surgical studies because mortality and morbidity are high in children. The younger children's problems include:

- Unilateral congenital hemispheric defects (particularly developmental migration defects) (fig 2) and ischaemic intrauterine events with hemiplegia and a preserved hemisphere with early onset epilepsies, including infantile spasms. Affected children may have total arrest of development, which may be related as much to subclinical seizures as obvious epilepsy
- Acquired damage due to injury and infection, which suggests the possibility of damage more widespread than the source of the seizure
- The Sturge-Weber syndrome, in which an acute, combined epileptic and vascular encephalopathy leads

Fig 3 Gadolinium enhanced magnetic resonance scan showing left cerebral atrophy and pial enhancement pathognomic of Sturge-Weber syndrome

Fig 1 Right hippocampal sclerosis

Fig 2 Left hemimegalencephaly

to acquired unilateral damage but global cognitive regression (fig 3)

- Focal areas of dysplasia, including dysembryoplastic neuroepithelioma, which in the temporal lobe may be associated with massive regression and autistic features (fig 4)
- Acquired unilateral inflammatory disease (Rasmussen's syndrome)
- Acquired aphasia (Landau-Kleffner syndrome).

The clinical indications for surgery and the types of procedures are given in the box. A non-invasive package of investigations has allowed most children to be evaluated without invasive monitoring. Invasive monitoring is required, however, when surgery might encroach on eloquent cortex or the extent and site of epileptogenesis is unclear.³⁶

The wide range of disease, epilepsy syndromes, and additional disabilities in childhood epilepsy make it

Fig 4 Right cortical dysplasia

essential that a scheme of categorisation of predicted outcome is used. Such an approach, proposed by Taylor et al, uses three categories: expected cure, predicted clinically significant amelioration, and procedures of less certain outcome.³⁷ Taylor et al also develop a contract with the parents and patient when appropriate, exploring their aims for surgery and deciding whether these are achievable and with what risks. This is of course a basic requirement of all medical interventions.

Several relevant parent support groups are part of Contact-a-Family, 170 Tottenham Court Road, London W1P 0HA (tel: 0171 383 3555).

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- 1 Aicardi J. *Epilepsy in children*. 2nd ed. New York: Raven, 1994.
- 2 Stephenson JBP. *Fits and faints*. London: MacKeith Press with Blackwell Scientific, 1990. (Clinics in developmental medicine No 9.)
- 3 Elmslie F, Gardiner M. The epilepsies. In: Rimoin DL, Connor JM, Pyeritz RE, eds. *Emery and Rimoin's principles and practice of medical genetics*. New York: Churchill Livingstone, 1996:2177-96.
- 4 Pryor DS, Don N, Macourt DC. Fifth day fits: a syndrome of neonatal convulsion. *Arch Dis Child* 1981;56:753-8.
- 5 Kroll JS. Pyridoxine for neonatal seizures: an unexpected danger. *Dev Med Child Neurol* 1985;27:377-9.
- 6 Nelson KB, Ellenberg JH, eds. *Febrile seizures*. New York: Raven, 1981.
- 7 Nelson KB, Ellenberg JH. Prenatal and perinatal antecedents of febrile seizures. *Ann Neurol* 1990;27:127-31.
- 8 Falconer MA, Taylor DC. Surgical treatment of drug resistant epilepsy due to mesial temporal sclerosis. *Arch Neurol* 1968;19:353.
- 9 Cross JH, Jackson GD, Neville BGR, Connelly A, Kirkham FJ, Boyd SG, et al. Magnetic resonance imaging in intractable epilepsy of childhood. *Arch Dis Child* 1993;69:104-9.
- 10 Meldrum BS, Horton RW, Brierley JB. Epileptic brain damage in adolescent baboons following seizures induced by allylglycine. *Brain* 1974;97:407.
- 11 Verity C, Ross E, Golding J. Outcome of childhood status epilepticus and lengthy febrile convulsions: findings of a national cohort study. *BMJ* 1993;307:225-8.
- 12 Liu Z, Mikati M, Holmes G. Mesial temporal sclerosis: pathogenesis and significance. *Paediatric Neurol* 1995;12:5-16.
- 13 Beaussart M. Benign epilepsy of children with Rolandic (centro-temporal) paroxysmal foci: a clinical entity. Study of 221 cases. *Epilepsia* 1972;13:795-811.
- 14 Gastaut H. Benign epilepsy of childhood with occipital paroxysms. In: Roger J, Bureau M, Dravet C, Dreifuss FE, Perret A, Wolf P, eds. *Epileptic syndromes in infancy, childhood and adolescence*. 2nd ed. London: Libbey, 1992:201-17.

- 15 Hrachovy RA, Frost JD. Infantile spasms. *Pediatr Clin North Am* 1989;36:311-30.
- 16 Chiron C, Dulac O, Beaumont D, Palacios L, Pajot N, Mumford J. Therapeutic trial of vigabatrin in refractory infantile spasms. *J Child Neurol* 1991;6:2S52-6.
- 17 Chugani HT, Shields WD, Shewmon DA, Olson DM, Phelps ME, Peacock WJ. Infantile spasms. I. PET identifies focal cortical dysgenesis in cryptogenic cases for surgical treatment. *Ann Neurol* 1990;27:406-13.
- 18 Gomez MR, ed. *Tuberous sclerosis*. 2nd ed. New York: Raven, 1988.
- 19 Chevrie JJ, Aicardi J. Convulsive disorders in the first year of life: neurological and mental outcome and mortality. *Epilepsia* 1978;19:67-74.
- 20 Cavazzuti GB, Ferrari P, Lalla P. Follow-up of 482 cases with convulsive disorders in the first year of life. *Dev Med Child Neurol* 1984;26:425-37.
- 21 Ounsted C, Lindsay J, Richards P. *Temporal lobe epilepsy 1948-1986: a biographical study*. Oxford: MacKeith Press with Blackwell Scientific, 1987.
- 22 Taylor DC. Mental state and temporal lobe epilepsy: a correlative account of 100 patients treated surgically. *Epilepsia* 1972;13:727-76.
- 23 Neville BGR, Harkness WFJ, Cross JH, Cass HC, Burch VC, Lees J, et al. Surgical treatment of autistic regression. *Paediatric Neurol* 1997;16:137-40.
- 24 Beaumanoir A. The Landau-Kleffner syndrome. In: Roger J, Dravet C, Bureau M, Dreifuss FE, Perret A, Wolf P, eds. *Epileptic syndromes in infancy, childhood and adolescence*. London: Libbey, 1993:231-43.
- 25 Neville BGR, Boyd SG. Selective epileptic gait disorder. *J Neurol Neurosurg Psychiatry* 1995;58:371-3.
- 26 Morrell F, Whisler WW, Smith MC, Hoepfner TJ, de-Toledo-Morrell L, Pierre-Lewis SJ, et al. Landau-Kleffner syndrome. treatment with subpial intracortical transection. *Brain* 1995;118:1529-46.
- 27 Maquet P, Hirsch E, Metz-Lutz MN, Motte J, Dive D, Marescaux C, et al. Regional cerebral glucose metabolism in children with deterioration of one or more cognitive functions and continuous spike-and-wave discharges during sleep. *Brain* 1995;118:1497-520.
- 28 Sadzot B. Epilepsy: a progressive disease? *BMJ* 1997;314:391.
- 29 de Silva M, MacArdle B, McGowan M, Hughes E, Stewart J, Neville BGR, et al. Randomised comparative monotherapy trial of phenobarbitone, phenytoin, carbamazepine, or sodium valproate for newly diagnosed childhood epilepsy. *Lancet* 1996;347:709-13.
- 30 Pellock JM. Utilization of new antiepileptic drugs in children. *Epilepsia* 1996;37(suppl 1):S66-73.
- 31 Prasad AN, Stafstrom CE, Holmes GL. Alternative epilepsy therapies: the ketogenic diet, immunoglobulins and steroids. *Epilepsia* 1996;37(suppl 1):S81-95.
- 32 Jackson GD, Connelly A, Duncan JS, Gadian GD, Grunewald R. MRI detection of hippocampal pathology in temporal lobe epilepsy: increased sensitivity using quantitative T₂ relaxometry. *Neurology* 1993;43:1973-99.
- 33 Gadian DG, Connelly A, Duncan JS, Cross JH, Kirkham FJ, Johnson CL, et al. ¹H magnetic resonance spectroscopy in the investigation of "intractable epilepsy." *Acta Neurol Scand* 1994;89 (suppl):116-22.
- 34 Cross JH, Gordon I, Jackson GD, Boyd SG, Todd-Pokrapek A, Anderson PJ, et al. Children with intractable focal epilepsy: ictal and interictal ^{99m}Tc HMPAO single photon emission computed tomography. *Dev Med Child Neurol* 1995;37:673-81.
- 35 Engel J Jr, ed. *Surgical treatment of the epilepsies*. 2nd ed. London: Raven, 1993.
- 36 Duchowny M. Epilepsy surgery in children. *Curr Opin Neurol* 1995;8:112-6.
- 37 Taylor DC, Cross JH, Neville BGR. Mental states of childhood epilepsy surgery candidates. *Epilepsia* 1996;37(suppl 4):51.

When I use a word ...

Fingerprints

Fond though doctors are of using long words of Greek and Latin origins, in some cases we use ordinary English words. We say hiccup (not singultus), yawn (not pandiculation), and fingerprints (not dermatoglyphics).

The recent discovery that koala bears have fingerprints that more closely resemble those of humans than chimpanzees' fingerprints do is said to support the hypothesis that fingerprints evolved as an aid to climbing. Whether or not that is so, fingerprints have been used for centuries as means of identification. Greek vases, for instance, bear examples as signatures, and in India a fingerprint used in this way by an illiterate person was known as tipsahi. But fingerprints were first systematically described by Johannes Evangelista Purkenje (sic) in his thesis, *Commentatis de examine physiologico organi visus et systematis cutanei*, published in Breslau in 1823. And they were really put on the map by the astronomer William Herschel (the Younger), who first devised a method for printing them in 1858, and by Francis Galton the geneticist, who in 1892 wrote a book, *Fingerprints*, about the differences in skin creases in different individuals.

The word dermatoglyphics was invented in 1926 by Harold Cummins and Charles Midlo and was used for the first time in a

paper on what they called "epidermal ridge configurations" (*Am J Phys Anthropol* 1926;9:471-502), where they restricted its use to ridges and their arrangements, excluding flexion creases and other secondary folds. Its origin is simple: from δέρμα (derma), the skin, and γλῦφω (glupho), I sculpt.

In everyday speech we may prefer to say "fingerprints," but "dermatoglyphics" is used much more often in publications, partly no doubt because it sounds more scientific, but also because it can be used to describe not only the prints themselves but also the study of them. Thus, although "fingerprint" and its derivatives were used in 3825 titles or abstracts of bioscience papers published between 1966 and 1997, in only 197 of those was dermatoglyphics meant, the other cases all being to do with fingerprinting as a molecular biological tool. In contrast, 1475 papers about proper fingerprints used the word dermatoglyphics.

Dermatoglyphics also leaves a distinctive fingerprint in the dictionary: it is the longest word in the English language (15 letters) that uses no letter twice.

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