Guidelines for the Diagnosis and Management of EPILEPSY in General Practice

The Irish College of General Practitioners

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GUIDELINES FOR THE DIAGNOSIS AND MANAGEMENT OF EPILEPSY IN GENERAL PRACTICE

Dr Ray O'Connor, Dr John Cox, Dr Michael Coughlan

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Foreword

The subject of epilepsy proved to be a very formidable challenge to the Clinical Review Committee of The Irish College of General Practitioners. Commencing in September, 1990, the task force on epilepsy spent 18 months patiently searching the literature, reviewing appropriate publications and consulting experts in the field.

Much credit is due to Dr Ray O'Connor, leader of the task force, who carried out the bulk of the work and repeatedly presented draft documents to the general committee before final agreement was reached.

This document addresses the important issues in epilepsy and should provide the family doctor with a useful guide when coming to a decision on subjects such as fitness to drive, which drug to prescribe, when to withdraw treatment, management of febrile convulsions, pregnancy and many other common everyday problems.

The issue of childhood immunisation proved to be a very difficult and contentious one, but consensus was eventually reached and the current global position is stated.

This document should lend to a uniformity of approach among general practitioners in the management and treatment of epilepsy and consequently to improved quality care for patients.

Dr Michael V Coughlan
Chairman
Clinical Review Committee
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Dr Raymond O'Connor
May 1992

Produced with the assistance of the Irish Epilepsy Association
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What to do if a fit occurs

- Do not restrain but protect patient from hurting him/herself, for example, from falling
- Place patient in semi-prone recovery position
- Do not try to force object between teeth
- Administer diazepam as follows:
  - Adult: By slow IV injection as a 0.5% solution of injectable diazepam emulsion (Diazemuls) 5 mg/ml, 10-20 mg (1-2 vials) at a rate of 0.5 ml (2.5 mg) per 30 seconds; repeat if necessary after 30-60 minutes.
  - or By rectum: Adults and children over three years diazepam enema (Stesolid) 10 mg; Elderly 5 mg; repeat after five minutes if necessary.
  - Child: By slow IV injection in a dose of 250 micrograms/kg of injectable diazepam emulsion (Diazemuls).
  - or By rectum: Dose of 0.5 mg/kg. Practically this means diazepam for: Age one to three years diazepam enema (Stesolid) 5 mg. Age over three years diazepam enema (Stesolid) 10 mg. Repeat after five minutes if necessary.

Indications for hospital admission after febrile convolution

- Admission is usually advised for all children after first febrile convolution
- Children under 18 months old
- A child whose convolution is complex, that is:
  - where the seizure lasts more than 20 minutes
  - has focal features
  - there are repeated seizures within 24 hours
  - there is incomplete recovery after one hour
- Where there are complicating factors, eg suspicion of intracranial disease, social factors or where early review by a doctor is not possible.

Notes on Diazepam

In practical terms either of the following routes is equally efficacious:

**Intravenous Diazepam:** Injectable Diazepam Emulsion (Diazemuls 10 mg/2 ml vial) is used in preference to Diazepam injection because it is less likely to cause vein damage or thrombosis.

**Rectal Diazepam:** In an emergency injectable Diazepam eg Valium or Diazemuls may be given using 2 ml syringe (minus needle). Stesolid diazepam enema is more convenient for GP's bag and for relatives to use.
**Definition**

> **TWO OR MORE PAROXYSMAL DISCHARGES OF CEREBRAL NEURONES**

Epilepsy is defined as a continuing tendency to seizures which are caused by paroxysmal discharge of cerebral neurones, resulting in a clinical event apparent to the subject, or an observer, or both.  

A **convulsion** is an episode of paroxysmal activity which is uncontrolled and unpredictable. It is the physical manifestation of a seizure but may or may not accompany it.

**Diagnostic formulation**

The diagnosis should take into account some (or all) of the following features.

- Age of onset (75% of cases occur before the age of 20)
- Type(s) of seizure suffered
- Present frequency of seizures
- Presumed cause
- Associated features, for example, mental retardation
- The patient’s present social and economic position.

**Incidence and prevalence**

- **Annual incidence** = 0.2/1000
- **Annual prevalence** = 5/1000

In a list of 2,000 patients one would expect four children and seven adults with active epilepsy (that is, either on treatment or getting seizures off treatment) and 35 patients who have had epilepsy at some stage in their life and are now in remission.

**Aetiology**

- **70% IDIOPATHIC**

Some 70% of epileptic disorders are idiopathic. A number of known causes of seizures are listed in Table 1.

**Table 1**

**Known causes of seizures**

**Local causes:**
- Congenital, eg tuberose sclerosis
- Traumatic, eg perinatal head injury, surgical
- Inflammatory, eg meningitis, acute and subacute encephalitis, neosyphilis
- Neoplastic, eg tumour, abscess, hematoma, aneurysm, angioma
- Degenerative, eg Alzheimers, Pick's, lipidoses, thrombosis, hypertension, cerebrovascular disease

**General causes:**
- Exogenous poisons: eg alcohol, drugs, eg theophyllines, drug withdrawal (eg anticonvulsants)
- Anoxia: eg respiratory failure, cardiac arrest
- Disordered metabolism: eg hypoglycaemia, hypocalcaemia, uraemia, hepatic encephalopathy

* Denotes commonest causes

**Genetic factors in epilepsy**

There are no clear guidelines as to the genetics of epilepsy. Genetic factors may act by lowering the convulsive threshold and by causing the transmission of diseases of which fits may be a symptom, eg tuberose sclerosis.

In febrile convulsions and petit mal, genetic factors are of paramount importance, but they play a much less significant role in partial epilepsy and in some of the serious epileptic syndromes of infancy and early childhood, eg infantile spasms.

The overall risk to a child of developing epilepsy when one parent has epilepsy is 2%-3%. Where both parents have epilepsy it is approximately 25%.

The risk appears to be highest when epilepsy occurs at an early age in patients with idiopathic epilepsy, absence seizures or previous family history, particularly in the mother. (Dr N Callaghan, personal communication).

**Types of seizures**

Seizures are divided into two main categories depending on whether the initial seizure is generalised or partial (localisation related or focal). More than one seizure type may occur in the same patient. A third category (unclassified) is
used for seizure types that don’t fall into these two categories.

Generalised seizures

- **Grand Mal (commonest type of seizure)**
- **Absence**
  - Typical (Petit Mal)
  - Atypical
- **Myoclonic (uncommon)**

**Grand mal seizures:** Neuronal discharge is associated with a generalised powerful muscular contraction (tonic). Breathing ceases; cyanosis may occur. Generalised movements follow (clonic), lasting two to three minutes. A period of unconsciousness is followed by postictal confusion lasting 20-60 minutes. This is the commonest of all seizure types.\(^7\)

**Absence seizures:** These fall into two categories:
- Typical (petit mal) seizure: Sudden onset and finish; altered consciousness with or without automatisms or jerks.
- Atypical: Less abrupt onset and less abrupt cessation of consciousness.

**Myoclonic seizures:** These are uncommon. Sudden uncontrollable jerks often of upper limbs. Commonly associated with typical absences.

Partial seizures

(Second commonest type of seizure)

- **Simple**
- **Complex**
- **Secondary generalised**

Partial seizures are also known as focal or localisation related and are the second most common form of seizures.\(^8\) They can be:
- Simple; where consciousness is not impaired
  - Motor (Jacksonian)
  - Sensory
  - Rolandic – commonest epilepsy of childhood
- Complex; where consciousness is impaired
  - Temporal lobe epilepsy
- Secondary generalised; this starts off as a focal seizure but spreads to become generalised.

Differential diagnosis of a seizure

- Syncope (fainting). Loss of consciousness is gradual and usually preceded by nausea and sweating. Once fallen, consciousness is usually recovered quickly without confusion. Syncope often occurs in response to strong emotional stimuli, prolonged standing still or standing after prolonged sitting.
- Cardiac dysrhythmia e.g. complete heart block.
- Transient Ischaemic Attack (TIA) – usually consciousness is maintained.
- Hyperventilation\(^9\)
- Breath holding – often in response to insult in a stubborn child.
- Reflex Anoxic Seizures: occur in childhood. A sudden unpleasant stimulus may cause transient cardiac asystole due to excessive vagal tone. When longer than eight seconds, anoxic convulsive movements may ensue.\(^2\)
- Night terrors – characteristic history.
- Migraine
- Behaviour disorder (simulated fits).

Establishing a cause

- **History**
- **Examination, usually normal**
- **Consider investigations pre-referral**
- **Hospital referral indicated in all new cases**

**History:** Careful history from both patient and witness(es) is essential

**Examination:** Physical exam: Neurological signs, eg retardation or mild hemiparesis may be found but most have no physical signs

**Investigation:** Consider investigation of seizures: Simple biochemistry such as urea and electrolytes, calcium, magnesium, glucose, liver function tests may be checked before referral. Chest x-ray may help rule out a pulmonary source of a cerebral metastasis, eg bronchogenic carcinoma. Skull x-ray provides no useful information.\(^3\) Some neurology centres may prefer to do their own work up however.

**Referral:**

- **Serious cause more likely in neonates and over 40 years old**

Hospital referral is indicated for further investigation eg EEG, CT Scan.

EEG: Used to confirm or identify seizure type. Normal in 30% of those with undoubted epilepsy.

CT Scanning: Used to rule out expanding intra...
cranial lesion, e.g., tumours, angiomas and structural lesions, e.g., atrophy. Not indicated in all cases.

In general, serious causes are more likely in the neonatal period and in those whose attacks begin after age 40 years.

Medical management of seizures

This is considered under the following headings:
1) Counselling
2) Irish Epilepsy Association
3) Drug therapy
4) Follow up.

Objectives of management:
- Total abolition of seizures except where patient wishes otherwise. (e.g., patients with occasional partial seizures which they can conceal may prefer these to an effective but unpleasantly large dose of anticonvulsants).
- To maintain as full and active a life as possible.

Counselling

What to do if a fit occurs:
- Do not restrain but protect patient from hurting him/herself, e.g., from falling
- Place patient in semi-prone recovery position
- Do not try to force object between teeth
- Consider rectal diazepam if fit lasts more than 10 minutes. Dose: Under three years: 5 mg; Over three years: 10 mg. May be repeated after five minutes if fitting continues.

Driving

"In the case of an applicant who suffers or has suffered in the past from epilepsy, fitness to drive may be certified for a limited period in relation to vehicles of category A, AB, EB, or W (that is motorcycles, tractors, cars with a capacity of eight persons or less and with a design gross vehicle weight of under 3,500 kg) where the applicant has not suffered an epileptic attack during the previous two years". It is left to the examining medical officer to decide the duration of the licensing period, i.e., one, three or 10 years.

In the case of single fits and following a change in drug dosage, certifying fitness is also left to the discretion of the examining doctor. Legally, it would be prudent in such cases to seek the opinion of a specialist.

However, fitness shall not be certified in relation to vehicles of category C, CD, D, ECI, EC, EDI, or ED (that is all other vehicles, mainly heavy goods and public service vehicles).

Other points
- Avoid precipitating factors, e.g., flashing lights, TV, alcohol and stress
- Avoid potentially lethal hazards, e.g., swimming unsupervised, rock climbing; safe sports can be encouraged
- Free prescription entitlement
- Ensure some identification is carried in case of fit. Epilepsy bracelets supplied by the Irish Epilepsy Association are ideal (See Appendix 1)
- Contraception: There is interaction between the pill and enzyme-inducing anticonvulsants, e.g., phenytoin, reducing the efficacy of the pill
- Starting a family: Consider rationalisation or withdrawal of drugs pre-conception and folic acid supplements.
- Employment and rehabilitation. Brainwave provides advice and information and organises training courses in conjunction with FAS, (Irish Training and Employment Authority). Close liaison is maintained with the National Rehabilitation Board.

Irish Epilepsy Association (Brainwave)

The Irish Epilepsy Association was established in 1967 to:
- Help and advise people with epilepsy, their families and friends. Professional social workers provide information, advice and counselling.
- Improve public understanding and elimi-
nate prejudice. The association provides a range of information leaflets and literature about epilepsy and organises regular meetings, talks and seminars on different aspects of the condition. An Epilepsy Newsletter is provided for members of the association to keep them up to date on developments and the work of the association. The association also aims to represent the interests of people with epilepsy on specific legal and welfare issues.

Encourage and assist research into epilepsy.
Promote an awareness of the need for education, rehabilitation and employment.

See Appendix 1 for addresses and telephone numbers.

**Drug therapy**

- **Drug choice influenced most by age and seizure type**
- **Treat early**
- **Use a single drug**
- **Seizure diary**
- **Increase dose with care**
- **Serum levels**
- **Avoid drug combinations if possible**
- **Beware toxicity and teratogenicity**

**General:** Age and seizure type are the most important considerations when choosing a drug. The possible importance of early treatment is emphasised by recent studies suggesting that the long-term prognosis is influenced by how many seizures have occurred before treatment. The prognosis seems poorer as the number of pre-treatment seizures increases.

- With a progressive cerebral disorder or clearly epileptic EEG, treatment may be started after a single seizure
- Seizures widely separated in time (more than 1 year)
- Identified precipitating factors, eg drugs, alcohol
- Probability of poor compliance, eg psychotic patients
- Attitude of patient or parents

### Table 2

**Factors in deciding to commence therapy**

<table>
<thead>
<tr>
<th>Problem</th>
<th>Usual practice</th>
<th>Factors modifying usual practice</th>
</tr>
</thead>
<tbody>
<tr>
<td>1 Prospective risk of epilepsy (eg post surgery)</td>
<td>No treatment</td>
<td>With a progressive cerebral disorder or clearly epileptic EEG, treatment may be started after a single seizure</td>
</tr>
<tr>
<td>2 Single seizure (clinically diagnosed)</td>
<td>No treatment</td>
<td>Seizures widely separated in time (more than 1 year)</td>
</tr>
<tr>
<td>3 Two or more (clinically diagnosed seizures)</td>
<td>Monotherapy</td>
<td>Identified precipitating factors, eg drugs, alcohol</td>
</tr>
</tbody>
</table>

**Irish College of General Practitioners**

**Section 1**

**Epilepsy — General**
Table 3

<table>
<thead>
<tr>
<th>Specific syndromes</th>
<th>Type of seizure or convulsion</th>
<th>Commonest age of onset (years)</th>
<th>First line drugs</th>
</tr>
</thead>
<tbody>
<tr>
<td>Petit mal</td>
<td>Simple absence with or without tonic-clonic</td>
<td>5 to 15</td>
<td>Valproate, Ethosuximide</td>
</tr>
<tr>
<td>Tonic-clonic</td>
<td>Tonic-clonic</td>
<td>5 to 25</td>
<td>Phenytoin, Valproate, Carbamazepine</td>
</tr>
<tr>
<td>Grand mal</td>
<td>Sensory</td>
<td>Any</td>
<td>Carbamazepine, Primidone, Phenytoin, Valproate</td>
</tr>
<tr>
<td>Infantile spasms</td>
<td>Salaam spasms</td>
<td>First year</td>
<td>Benzodiazepines, Steroids, ACTH</td>
</tr>
<tr>
<td>Simple</td>
<td>Motor (Jacksonian)</td>
<td>Any</td>
<td>Carbamazepine, Primidone, Phenytoin, Valproate</td>
</tr>
<tr>
<td></td>
<td>Sensory</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Complex</td>
<td>Focal with loss of consciousness, eg psychomotor (temporal lobe epilepsy) usually but not always arises in Temporal Lobe</td>
<td>Any</td>
<td>Phenytoin, Carbamazepine, Valproate</td>
</tr>
<tr>
<td></td>
<td>Persistent seizure activity for more than 30 minutes</td>
<td>Any</td>
<td>Diazepam, IV or PR</td>
</tr>
</tbody>
</table>

**Some syndromes of epilepsy**

Some of the commonest syndromes of epilepsy are summarised in Table 3.

Up to 70% to 80% of patients quickly enter a prolonged remission of their seizures.

**Choice of drugs:**

- **No difference in efficacy between phenobarbitone, phenytoin, carbamazepine, and valproate for the treatment of specific seizure types in newly diagnosed adults or children**
- **Phenobarbitone not recommended**

While many clinicians have been persuaded that one or other drug is likely to be most effective against particular seizure types and syndromes in adults, it is difficult to identify satisfactory clinical trials that support this.\(^7\)

It is doubtful whether there is any difference in efficacy between carbamazepine, phenytoin barbiturates and valproate in treatment of newly diagnosed epilepsy in adults or children.\(^2,12\)

While phenobarbitone is of benefit in treatment of some seizure types, it is no longer recommended in the overall management of epilepsy.

**Drugs:**

Individual doses, contraindications, adverse drug reactions and interactions are detailed in MIMS, British National Formulary and NDAB information book 33.\(^9\) Some general points are made here.

**Acute toxicity:**

- **Non specific encephalopathy**
- **Seizures may increase with high drug levels**
- **Newer agents have fewer adverse effects**

**Acute Dose Related Toxicity:**\(^7\) Most anticonvulsants including phenytoin, carbamazepine, barbiturates and benzodiazepines give rise to a non-specific encephalopathy associated with high blood concentrations.

Patients exhibit sedation, nystagmus and with increasing blood levels, ataxia, dysarthria and ultimately confusion and drowsiness. Seizure frequency may increase with high levels.

All anticonvulsants have adverse effects that can be detected at therapeutic concentrations and which become more apparent both with polytherapy and increased blood levels of individual drugs.
The fact that newer agents such as carbamazepine and valproate have fewer adverse effects on cognitive function and behaviour is one of the most powerful arguments for preferring these to longer established antiepileptic drugs.

**Teratogenicity**

> All anticonvulsants are potentially teratogenic

All antiepileptic drugs must be regarded as potentially teratogenic. Carbamazepine was reputed to be less so than other drugs. However, recent studies have not substantiated this view. The most common malformations are cleft lip and palate and cardiovascular anomalies. There seems to be an association between neural tube defects and sodium valproate. Overall, in pregnancy, if a drug is necessary, monotherapy with carbamazepine is probably the best choice.

<table>
<thead>
<tr>
<th>Table 5</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Drug</strong></td>
</tr>
<tr>
<td>Carbamazepine (Tegretol)</td>
</tr>
<tr>
<td>Phenytoin (Epanutin)</td>
</tr>
<tr>
<td>Sodium Valproate (Epilem)</td>
</tr>
<tr>
<td>Primidone (Mysoilene)</td>
</tr>
</tbody>
</table>

Cost

The cost of 20 days treatment at the ‘average’ doses is shown in Table 5.

**Long term management of drug therapy**

The majority (approximately 70%) of patients developing epilepsy achieve long lasting remission that begins very quickly after the start of therapy.

For these patients drug withdrawal may be

<table>
<thead>
<tr>
<th>Common or important side effects of the major anticonvulsant drugs</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Drugs</strong></td>
</tr>
<tr>
<td>-----------</td>
</tr>
<tr>
<td>Carbamazepine</td>
</tr>
<tr>
<td>Ethosuximide</td>
</tr>
<tr>
<td>Phenobarbitone</td>
</tr>
<tr>
<td>Phenytoin</td>
</tr>
<tr>
<td>Primidone</td>
</tr>
<tr>
<td>Sodium Valproate</td>
</tr>
</tbody>
</table>
considered after two, three or more years. Up to 60% of such patients may remain seizure free on no medication.

Drug withdrawal is considered in more detail below.

Follow up of patients with epilepsy

This should be planned and regular and should ideally be shared with a consultant neurologist at a seizure clinic or neurology OPD while on long term anticonvulsants.

- Planned and regular shared review
- See frequently initially
- Assess problems and understanding
- Ensure easy access to health care team
- Disease register
- Regular review
- Call/recall system
- Assess problems, control, side-effects
- Blood tests
- Use flow charts and co-op cards next review
- Plan to the next review

- Patients with epilepsy and their families should be seen frequently after the diagnosis has been made. The patients' understanding of their problems, physical, psychological and social should be addressed. Easy access to the doctor must be assured, but much of the counselling could be carried out by others, eg practice nurse or a trained social worker with the Irish Epilepsy Association.

- Patients with epilepsy should be entered on a disease register (Appendix 3).

- Regular review should be ensured at intervals appropriate to the patients needs. A recall system and strict control of the repeat prescribing of anticonvulsants would help to ensure this.

- At the review: Listen to any problems; assess control, compliance and side effects (Table 4); blood tests:

  1. Routine monitoring of serum levels should be restricted to the following:
     - Phenytoin or multiple drug treatment where dose adjustment is necessary
     - Where toxicity is suspected
     - In cases of poor control and to check compliance

  2. Biochemistry (Calcium, Phosphate, Alkaline phosphatase, urea and electrolytes), full blood count and liver function tests should be checked at six monthly intervals. While some patients develop abnormal liver function tests (LFT) based on idiosyncratic reaction to anticonvulsants, very few of these patients actually proceed to develop clinical evidence of hepatocellular damage. Clinical monitoring should always take precedence, as when liver failure occurs it is usually an acute event. (Dr N Callaghan)

Consistent care is made easier by using flow charts and possibly patient co-op cards. A proposed clinical review card is included with this document (Appendix 2).

- Try to devise a plan to cover the period up to the next assessment. Appropriate advice for early return should be given, eg symptoms of toxicity. Problems that have come to light in an Irish context are:
  - Poor control due to poor compliance and alcohol abuse
  - Inadequate counselling at the time of diagnosis
  - Fear of the disease and of stigmatisation among patients with epilepsy
  - Private patients are more likely to attend hospital only and not to have been seen by their GP

Prognostic factors

- Relapse commoner when adult onset, mental retardation, cerebral pathology demonstrable

Relapse is more likely in adult onset epilepsy and epilepsy in those with mental retardation, neurological deficit or demonstrable cerebral pathology.

Withdrawal of anti-epileptic drugs

- 70% remission from seizures on therapy
- With minimum of two years seizure freedom drug withdrawal is considered
## Factors Influencing Drug Withdrawal

<table>
<thead>
<tr>
<th>Favourable</th>
<th>Adverse</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Primary generalised seizures</td>
<td>1. Partial seizures (especially complex partial with secondary generalisation)&lt;sup&gt;27&lt;/sup&gt;</td>
</tr>
<tr>
<td>2. Childhood onset after age of one year</td>
<td>2. Adult onset</td>
</tr>
<tr>
<td>3. Short duration of epilepsy</td>
<td>3. Long duration of epilepsy</td>
</tr>
<tr>
<td>4. No cerebral disorder</td>
<td>4. Underlying cerebral disorder</td>
</tr>
<tr>
<td>5. Few seizures</td>
<td>5. Many seizures</td>
</tr>
<tr>
<td>7. Carbamazepine treatment</td>
<td>7. Abnormal EEG before treatment and unchanged before withdrawal (Class 4 EEG)</td>
</tr>
</tbody>
</table>

**Factors influencing drug withdrawal**<sup>24,27</sup>

- **Potential withdrawal effects**
  - Seizures
  - Anxiety
  - Agitation
  - Most relapses occur during or within one year of withdrawal

Some seizures may be prompted by drug withdrawal.

Other reactions including anxiety, agitation and insomnia are a problem with phenobarbitone and the benzodiazepines.

These reactions do not occur in withdrawal of phenytoin, carbamazepine or sodium valproate. It is not known whether patients on more than one drug should have them tailed off sequentially or together.<sup>29</sup>

Gradual dose reduction under supervision is essential over a minimum of six months. In adults each step should not usually exceed carbamazepine 200mg.<sup>25</sup>

Patients should be told to report any seizure immediately.

Most relapses occur during or within 12 months of drug withdrawal. The first six months after drug withdrawal is the most hazardous period.

Up to 60% of patients will remain seizure free on no medication.<sup>27</sup>
Prevalence

The prevalence is approximately 8/1,000 in school children in the UK, but this is widely regarded as being an underestimate. Of these 65% attend normal school classes, 35% are multi-handicapped and have a poor prognosis for intellectual development. 

The Epileptic Syndromes are usually considered in chronological order as follows:

Generalised epileptic seizures

Babies: (Fig 1 and Fig 2)

- Atypical seizures in babies
- Prognosis poor
- Distinguish from jitteriness

Seizures in the newborn and in the early months of life are usually partial and fragmented and frequently signify serious brain abnormality and dysfunction. They carry a poor prognosis. Immediate referral is indicated. Infantile spasms present around five to six months. In a typical case the child makes sudden flexion spasms involving the trunk while looking acutely anxious.

A structural brain disorder can be demonstrated in approximately 50%, eg cerebral birth injury. The prognosis is poor. Referral for diagnosis and treatment (usually corticoterphin) is indicated. An important differential diagnosis is jitteriness. Unlike seizures this is stimulus sensitive and is tremor dominant, without chronic jerking.

Young children:

- Generalised myoclonic epilepsy one to six years
- Prognosis poor
- Primary genetic epilepsy: generalised and partial

The generalised myoclonic epilepsies which occur between the ages of one to six years form another age related group of epilepsies with many similarities to infantile spasms.

There is frequently an associated variable degree of mental handicap. The long term outlook is uncertain and recovery from the more severe cases is rare.

Older children: Between the ages of five and 10 years, primary epilepsies predominantly due to genetic factors are common and may be either generalised or partial in their seizure manifestations. Generalised seizures include:

- Grand mal: Tonic-clonic convulsive episodes occur in 75% of all children with epilepsy either as the only evidence of their epilepsy, or in combination with other seizure types. Clinically there are two phases:
  - Tonic-clonic seizure
  - Subsequently a period of deep sleep
An aura is not experienced in the classical grand mal attack in childhood. (Professor NV O'Donohoe, personal communication).

Petit mal: This is characterised by episodes of "absence attacks" or unconsciousness which, though brief, may be very frequent. Structural brain abnormality is rare. The prognosis is good with most resolving by puberty.

Partial epileptic seizures

These may be simple or complex depending on whether consciousness is unaltered or impaired. One of the best known complex partial seizure types is temporal lobe epilepsy.

The seizures here may take the form of absence attacks, usually preceded by an aura and lasting longer than 10 seconds. Full consciousness returns slowly.

The lesions causing focal seizures in childhood are usually non progressive and have a good chance of being controlled by anticonvulsants.

Rolandic epilepsy (Benign focal epilepsy of childhood):

ROLANDIC IS THE COMMONEST EPILEPSY OF CHILDHOOD

This is a simple partial seizure, often with secondary generalisation. It is probably the commonest epilepsy of childhood and seems to be caused by a temporary functional disturbance in the brain.

The usual age of onset is between seven and 10 years of age. The seizures typically occur during sleep, often becoming secondarily generalised producing a grand mal seizure. The prognosis is excellent with or without drug treatment.

Clinical Assessment:

HISTORY, EXAMINATION, DEVELOPMENTAL ASSESSMENT, REFERRAL

The most important steps are history and detailed examination. Developmental assessment is also important. Skin signs may indicate a specific lesion, eg port wine stain of Sturge-Weber Syndrome.

Height, weight and head circumference are important. Investigations are probably best done after referral which is indicated in all cases.

Management

The patient's ideas, concerns and expectations are dealt with. Early hospital outpatient appointment or admission to a paediatric neurology department is arranged to confirm diagnosis, assess for abnormality and possibly to commence drug therapy.

Figure 2:
Chronology of epilepsy in infancy and early childhood

Figure 1 and 2 are reproduced by kind permission from O'Donohoe NV, Epilepsies of Childhood, Second Edition, 1985. Butterworths, London Boston.
Treatment

Drug therapy: 20

- **GENERALLY POOR COMPLIANCE**

Only about 50% of children take anticonvulsants as prescribed. 21

Sodium valproate:

- **GOOD DRUG**
- **TWICE DAILY DOSE**
- **BEWARE HEPATOTOXICITY BY MEASURING LFT**
- **DRUG LEVEL ESTIMATION UNHELPFUL**

Sodium Valproate is a good general purpose anticonvulsant especially for tonic-clonic fits, absences and myoclonus. Efficacy against partial seizures is similar to that of carbamazepine and phenytoin. Cognitive impairment is uncommon. Hepatotoxicity seems to be largely confined to handicapped children under three years of age. It is most commonly prescribed twice daily.

There is no useful relation between serum concentrations and anticonvulsant efficacy or toxicity so routine drug levels are unnecessary.

Carbamazepine:

- **GOOD DRUG**
- **SKIN RASHES**

Regarded as valuable in most types of childhood seizure except for petit mal and myoclonic epilepsy. Skin rashes limit its usefulness in 5%-10% of patients. There are few long term difficulties with it. Drug interactions are common.

Phenytoin:

- **TOXIC SIDE EFFECTS**

Phenytoin is effective for the prophylaxis of partial and generalised tonic-clonic seizures.

Because of an impressive array of toxic effects, including cosmetic changes (e.g. gum hypertrophy, acne and hirsutism) and psychosocial disorders (e.g. aggression, sedation, depression) it is now used less often in children. Drug interactions are also a problem.

Phenobarbitone:

- **NOT RECOMMENDED EXCEPT IN NEWBORN**

Phenobarbitone is not regarded as a good drug for children because of insomnia, hyperkinesia and aggression. Still used in the newborn as there is less experience of other drugs at this age.

Ethosuximide:

- **USED IN INFANTS AND ABSENCE SEIZURES**

Ethosuximide is used widely for treatment of absence seizures in childhood and adolescence. It is the drug of choice in infants because of the risk of fatal hepatotoxicity with sodium valproate.

Emergencies 20

- **IV OR PR DIAZEPAM**

A few children develop status epilepticus (i.e. persistent seizure activity for more than 30 minutes).

Intravenous diazepam 0.25 mg/kg slowly over two to three minutes is the standard treatment, or rectal diazepam (e.g. Stesolid) at 0.5 mg/kg.

In practice this means 5 mg for a one to three year old and 10 mg for a child over three years of age.

The family will be frightened and explanation and support is needed. Once seizures are controlled admission to hospital and/or review of medication and search for a precipitating cause can be made.

Prognosis

- **GENERALLY GOOD PROGNOSIS**
- **AVOID OVER RESTRICTION**

In general, patients who are normal, except for their epilepsy, tend to have a good prognosis, especially if they present with grand mal, petit mal or Rolandic epilepsy. 20

Complete control of seizures is possible in 60% to 70%; 15% to 20% have considerably reduced seizure frequency. 20

Over 70% of children will stay in remission if drugs are withdrawn after two years without seizures. 20

Withdrawal of therapy after two years free of seizures is the commonest approach and should be undertaken in agreement with the parents and in conjunction with a neurologist or paediatrician.

Needless restrictions must not be placed on the child’s activities. Most should be taught to swim, but a capable supervisor must always be available in the pool.
**Epilepsy and pregnancy**

- **ANNUAL INCIDENCE IN PREGNANCY**
  - 40-50/100,000
- **PLAN PREGNANCY**
- **ANTICONVULSANT INTERACTION WITH ORAL CONTRACEPTION**

The age specific annual incidence of epilepsy is roughly constant at 40-50 cases per 100,000 women throughout the childbearing period.

We have already discussed the advisability of women with epilepsy in this age group to attempt to defer pregnancy until they are seizure free for two to three years.

Then anticonvulsant medication could be gradually withdrawn before a planned pregnancy. Up to 60% will remain seizure free on no anticonvulsant medication.

However, some women require anticonvulsant drugs continuously and for these the dose should be as low as possible.

Monotherapy with carbamazepine is probably the best choice.

Overall, children of epileptic mothers taking anticonvulsants have roughly twice as many significant malformations (6%) as children of mothers in the population as a whole.

There appears to be no data documenting an increase in risk to the foetus due to seizures compared with teratogenic defects occurring in patients taking anticonvulsant medication (Dr N Callaghan, personal communication).

Some women with epilepsy may become pregnant unexpectedly while taking oral contraceptives if they are also taking enzyme inducing anticonvulsants, eg phenytoin.

This is more likely to happen if a low dose contraceptive pill is used, but may also occur with the 50 microgram oestrogen contraceptive.

Women must be advised of this risk and use either an alternative method of contraception or change their anticonvulsant if they do not wish to become pregnant.

**First seizures in pregnancy**

- **RULE OUT ECLAMPSIA**

First rule out Eclampsia! There is no statistical evidence that pregnancy is likely to precipitate epilepsy. A patient who starts to have seizures during pregnancy should be referred to a neurologist.

**Frequency of seizures during pregnancy**

The evidence suggests that untreated epileptic pregnant women are about equally likely to experience fewer seizures as more seizures in pregnancy.

**Epilepsy and foetal abnormalities**

- **INCREASED INCIDENCE OF FOETAL ABNORMALITIES DUE TO:**
  - Genetic factors
  - Convulsions
  - Anticonvulsants

Apart from the possibility of a genetic association between epilepsy and the risk of foetal abnormality, seizures could cause foetal damage through hypoxia or associated trauma.

Facial clefts and cleft palate are among the most common foetal abnormalities associated with epilepsy and its treatment. But studies suggest that the rate is higher if the foetus is exposed to anticonvulsant drugs. If these drugs are necessary in early pregnancy, the smallest dose compatible with control of seizures should be used.

There is good evidence of a genetic predisposition to congenital abnormalities induced by phenytoin. There is an association between sodium valproate and neural tube defects. Recently reports of spina bifida in infants of mothers who used carbamazepine in pregnancy have appeared, making the choice of an appropriate anticonvulsant in pregnancy a very difficult one. Monotherapy with carbamazepine is probably the best choice.

**Other metabolic disturbances induced by anticonvulsants**

- **LOW FOLATE. USE SUPPLEMENTS PERI-CONCEPTUALLY AND DURING PREGNANCY**
- **RICKETS AND OSTEOMALACIA**
- **NEONATAL HAEMORRHAGE RISK**
- **NEONATAL WITHDRAWAL SYMPTOMS**

**Serum Folate Concentration:** This may be reduced by anticonvulsants. A low dose of folate...
supplement seems to be sufficient to prevent this.
Low folate levels at the time of conception may increase the risk of neural tube defects. Folic acid supplements started before pregnancy are recommended.

Vitamin D Metabolism: Long term treatment with phenytoin carbamazepine and phenobarbitone has been shown to induce clinical osteomalacia and rickets. Asian epileptic mothers may be more at risk, as may their babies, but this problem has also been shown to occur in an Irish context.

Vitamin K Metabolism: Neonates born to mothers with epilepsy who have been treated with anticonvulsants are at an increased risk of haemorrhage.
This can be prevented by giving women vitamin K during pregnancy.

Neonatal withdrawal symptoms and signs may appear at this time in the children of mothers who have been on anticonvulsants during pregnancy. Expectant supervision is usually adequate. If tremulousness or seizures develop, 3 to 5 mg/kg/day of phenobarbitone may be given.

Breast feeding:

- Mothers may breast feed
Mothers taking anticonvulsant drugs may safely breast feed their children.

Motherhood

If the mother of a young child has seizures which are not fully controlled, it is wise for her to take precautions to protect the child from danger.
- The baby should be fed on the floor.
- The baby should be washed on a mat on the floor and not in a baby bath.
- The baby should be left to play in a playpen.
- The stairs should be guarded with a stair guard.
- A fire guard should always be in place.
- All medicines should be out of reach.
- Tasks such as ironing should be delayed until the baby is resting or in a playpen.
Febrile convulsions

Definition:

-Occurs between age of six months and five years
-Two thirds single and brief
-Not epilepsy

A febrile convolution is an epileptic seizure occurring in a child from six months to five years, precipitated by fever arising from infection outside the nervous system in a child who is otherwise neurologically normal. In two thirds of cases the seizure is single and brief (lasting up to a few minutes); it terminates spontaneously and a complete recovery occurs.

Febrile convulsions are distinguished from epilepsy which is usually characterised by recurrent non-febrile seizures at any age.

Incidence:

-3% of children
-90% due to viral URTI

Febrile convulsions occur in about 3% of children. The highest incidence occurs between the ages of 10 months and two years. Ninety percent of febrile convulsions are due to viral upper respiratory infections.

Aetiology:

Any pyrexia. The seizure tends to occur at the height of the pyrexia.

Management

-Telephone advice
-Rectal diazepam
-Antipyretic measures
-Explanation and support

Brief telephone advice is given about laying the child prone and keeping him/her cool. A visit is then arranged as soon as possible. About 5% of seizures last more than 30 minutes.

Rectal diazepam is used to terminate a convolution in a dose of 0.5mg/kg. A 5mg rectal tube is used for a child aged one to three years and 10mg tube for a child over three years. This should be given as soon as possible after the onset of convolution. Do not give it if the convolution has stopped.

Antipyretic measures are taken including stripping off clothes and paracetamol. Physical methods of temperature reduction such as fans and tepid sponging are no longer recommended. Family support and a full explanation are important.

Admission to hospital

Any febrile illness can provoke a convolution. Meningitis and encephalitis are difficult to exclude in children under two years of age. In addition, hypotonia following a fit and the use of diazepam may mask a meningal irritation. Parental anxiety is often at its maximum at this time.

Therefore, admission to hospital after the first febrile convolution is usually advised. If possible, antibiotics should be withheld initially for at least 24 hours as they may mask meningitis. Subsequent febrile convulsions may safely be managed at home.

Indications for hospital admission:

-Admission to hospital advised if:
  -First febrile convolution
  -Age less than 18 months
  -Complex convolution
  -Social factors

-Admission is usually advised after first febrile convolution.
-Children under 18 months of age
-A child whose convolution is complex, that is:
  -lasting more than 20 minutes
  -has focal features
  -repeated convulsions occur within 24 hours
  -there is incomplete recovery after one hour
-Complicating factors, eg suspicion of intracranial disease, social factors or where early review by a doctor is not possible.

Advice to parents

The family will need considerable support. Parents tend to feel guilty. Was it their fault? Did they seek help in time? How can they tell friends and relatives or babysitters?

Coping with subsequent febrile illness:

-Use temperature indicator
-Paracetamol
-Antipyretic measures

The use of a temperature indicator on the forehead and tympanic membrane has shown that children do not develop febrile convulsions.
head is useful. If parents are in doubt, it may confirm that the child is febrile. Paracetamol 25 mg/kg/day (usually 120 mg four hourly) keep clothes to a minimum (and only a sheet at night) encourage fluids.

Other physical methods such as fanning, tepid sponging are likely to cause discomfort and are not now recommended.\(^2,^3\)

While these methods may lower temperature, their effect is transient. When stopped, the temperature rises to pre sponging levels within within 30 minutes.\(^3\)

**Subsequent convulsions**

- 5% RISK OF SUBSEQUENT CONVULSIONS
- KEEP CHILD COOL
- PRONE POSITION
- CONSIDER PR DIAZEPAM
- GP ASSESSMENT
- RECURRENCE MORE LIKELY IF AGED LESS THAN ONE YEAR AND POSITIVE FAMILY HISTORY

One third of febrile convulsions occur before the parents realise that the child has a fever. The risk of recurrence of febrile convulsion in subsequent febrile illness is also about a third. Unfortunately, temperature control has failed to prevent recurrent febrile convulsions.\(^2,^3\)

Keep the child cool and place in a prone position away from hard surfaces. Call emergency assistance if the convulsion lasts more than five minutes.

Some parents can be taught how to use a rectal solution of diazepam 5 mg during a fit. (The level of fever inducing a febrile convulsion is usually in excess of 39°C). Even if the convulsion is brief the general practitioner should be called to determine the cause of the fever.

**Prognosis**

The average recurrence rate of febrile convulsions is 33%. The only two factors closely linked to possibility of recurrence are:

- Age of child less than one year.
- Family history of febrile convulsions in first degree relatives, ie siblings and/or parents.

There is no evidence to suggest a long term risk of impaired cognitive development following febrile seizures.\(^2,^3\)

**Risk of subsequent epilepsy**

In the case of a single uncomplicated febrile fit (ie lasts less than 15 minutes and no neurological sequelae) the risk of subsequent epilepsy is increased minimally from 0.8% to 1.04%.

The risk is higher than this (up to 6.3%) if: \(^5\)
- The fit was prolonged (longer than 15 minutes)
- Focal features present
- More than one febrile fit occurs within the same period of illness
- There is a family history of non febrile seizures of a genetic origin

**Prophylactic anticonvulsant therapy**

- ANTICONVULSANTS DO NOT PREVENT SUBSEQUENT DEVELOPMENT OR NEUROLOGICAL DEFICIT

Anticonvulsants prevent further febrile seizures, but there is no evidence that prophylaxis will prevent the subsequent development of epilepsy or neurological deficit.\(^7\) They are now rarely indicated.\(^8\)

Prophylaxis for febrile convulsions usually continues for one to two years after the last convulsion or until the child is five years old.

**Drugs used in prophylaxis**

All drugs used cause toxic effects in regular use. Phenytoin and carbamazepine have no proven prophylactic value.

**Intermittent rectal diazepam:**

- RECTAL DIAZEPAM DRUG OF FIRST CHOICE
- DURING FIT
- MAY REPEAT IN FIVE MINUTES
- DO NOT GIVE IF FIT HAS STOPPED

Intermittent rectal diazepam is now the first choice in prophylaxis. It may be given at the start of or during a fit. Its routine use at the onset of fever is not now recommended.\(^9\)

Dose is 0.5 mg/kg. Practically this means 5 mg diazepam enema (Stesolid) in a child aged one to three years and 10 mg diazepam enema (Stesolid) in a child over three years. If the child...
continues to fit, the dose may be repeated after five minutes.

Sodium valproate:
Sodium valproate is given at a dose of 20-40 mg/kg/day.

It is prudent to monitor liver function before and during therapy.

Many people are now reluctant to use this treatment for prophylaxis in children under two years unless the recurrence risk is very high and compliance assured.  

Phenobarbitone:
Phenobarbitone is given in a dose of 4 to 6 mg/kg/day. Serum levels should be checked after three weeks.

Over 25% of children have to stop taking phenobarbitone because of side effects including rashes and behavioural changes. It seems difficult to justify phenobarbitone prophylaxis unless the recurrence risk is very high and compliance assured.

Immunisation

> FULL IMMUNISATION IS IMPORTANT

It is important for all children to be fully immunised.

Measles and pertussis can both provoke a febrile convolution but, if there is a danger of febrile response following immunisation, preventive measures can be taken.

Pertussis vaccine:

> NOT CONTRA-INDICATED

Pertussis vaccine is not contra-indicated for a child with a personal history of febrile convulsions. Children who have febrile convulsions before immunisation against diphtheria, pertussis and tetanus because the immunisation has been delayed, should be immunised after their parents have been instructed about the management of fever and the use of rectal diazepam.

For children with a documented history of non-febrile convulsions or neonatal cerebral damage, the risk of the vaccine should be weighed against the risk of damage from pertussis.

The advice of a paediatrician or consultant in public health should be sought before deciding not to immunise. The evidence is that pertussis vaccine has no causative role in brain damage whatsoever.

Measles Mumps Rubella vaccine:

> GIVE MMR BUT BEWARE FEBRILE RESPONSE

Children with a personal or close family history of convulsions should be given MMR vaccine provided that the parents understand that there may be a febrile response and have a knowledge of how to cope with same.

Immunoglobulin must not be given with MMR vaccine since the immune response to rubella and mumps may be inhibited. Doctors should seek specialist paediatric advice rather than refuse vaccination.
Appendix 1

Head office:

Irish Epilepsy Association (Brainwave),
249 Crumlin Road,
Dublin 12.
Tel: (01) 557500

Regional offices:

Cork: 20A Washington Street,
Cork.
Tel: (021) 274774

Galway: Ozanam House,
St Augustine’s Street,
Galway.
Tel: (091) 68180

Limerick: Social Services Centre,
Henry Street,
Limerick.
Tel: (061) 314111

Waterford: 125 The Quay,
Waterford.
Tel: (051) 54262

Epi-Alert bracelets are available from Dublin Office of Irish Epilepsy Association.

Appendix 3

Contra indications, special precautions, interactions and adverse drug reactions of commonly used anticonvulsants (MIMS)

<table>
<thead>
<tr>
<th>Drug: C/I or S/P</th>
<th>Interactions</th>
<th>Adverse drug reactions</th>
</tr>
</thead>
<tbody>
<tr>
<td>Phenytoin: Liver dysfunction, pregnancy/lactation withdraw slowly</td>
<td>Coumarin Anticoagulants Isoniazid Chloramphenicol Oral contraceptives</td>
<td>Gastric distress, allergic reactions, sleeplessness, unsteadiness, hirsutism, gingival hypertrophy, blood dyscrasias, lymphadenopathy nystagmus</td>
</tr>
<tr>
<td>Sodium valproate: Hepatic dysfunction; children with severe epilepsy and mental retardation or congenital metabolic defects, in the first 6/12 check LFT in those most at risk and monitor all for clinical symptoms of hepatic failure</td>
<td>Antidepressants Other anti-convulsants</td>
<td>Weight gain, hair loss, oedema, pancreatitis, hepatic failure, hyperammonemia, thrombocytopenia, neurological effects</td>
</tr>
<tr>
<td>Phenobarbital: Respiratory depression, chronic psychosis, phobic or obsessive states, chronic renal or hepatic disease, elderly, pregnancy and lactation; judgement may be impaired</td>
<td>Alcohol and other CNS depressants Anticonvulsants</td>
<td>Drowsiness, ataxia, confusion vertigo, GI disturbances, abnormal psychological reactions, hypotension, visual disturbances, skin rashes, urinary retention rarely blood disorders and jaundice</td>
</tr>
<tr>
<td>Carbamazepine: AV conduction abnormalities unless paced; renal or hepatic impairment, blood dyscrasias, pregnancy, lactation, test blood regularly</td>
<td>MAOI, Anticoagulants, macrolide antibiotics, lidocain, oral contraceptives</td>
<td>Gastric upset, diplopia, dry mouth, drowsiness, oedema and hypotremia, blood dyscrasias, rashes, acute renal failure and cholestatic jaundice</td>
</tr>
</tbody>
</table>
Appendix 4

How to construct and use a disease register for epilepsy

1. A filing box and cards are obtained.

2. Each time a prescription for anticonvulsants is written or the notes of a patient with epilepsy are seen:
   (a) The notes are marked with a colour coded sticker (yellow in the RCGP coding system).
   (b) A card is made out with the patient's name, date of birth and address and put in the file.
   (c) An ‘R’ is put through the yellow sticker to confirm the patient is on the register.

3. After three months, when it is hoped that the file will be largely complete, a recall system is instituted.

4. All cards are divided into two by a vertical pencil line.

5. The cards are divided into six equal groups and filed in slots corresponding to the next six months.

6. Each month the cards filed in the respective slot are withdrawn and the patients contacted either by letter or telephone.

7. They are sent forms for FBC, calcium, phosphate, Alk Phos and serum levels of the drug they are taking and asked to attend for venesection as soon as possible.

8. They are then given an appointment two weeks* ahead to allow time for the results to come back to surgery.

9. When the patients attend they are reviewed according to the flow sheet, the date is put on the file card and this is filed in the disease register six months ahead.

10. At the end of the month, the cards of the patients who have not attended are withdrawn from the file and they are sent for again, if necessary, by a visit to their home.

* In some areas the time the laboratory requires to do the tests may exceed this, in which case the interval between venesection and clinical review should be adjusted accordingly.

Source: RCGP Epilepsy Information folder
1.1 Definitions:
A seizure is a paroxysmal discharge of cerebral neurones.
A convulsion is an episode of paroxysmal activity which is uncontrolled and unpredictable.
It is the physical manifestation of a seizure but may or may not accompany it.
Epilepsy refers to two or more non febrile seizures occurring in a patient.

1.2 The diagnosis should take account of the following features:
- Age of onset
- Type(s) of seizure
- Present frequency of seizures
- Presumed cause
- Associated features
- Present social and economic position

1.3 Type of seizure:
1. Generalised: Grand mal
   - Absence
   - Myoclonic
2. Focal:
   - Simple
   - Complex
   - Secondarily generalised
3. Unclassified

1.4 Differential diagnosis:
- Syncope
- Cardiac dysrhythmia, eg heart block
- Transient ischaemic attack
- Hyperventilation
- Breath holding
- Reflex anoxic seizures
- Migraine
- Simulated fits

1.5 Establish a cause:
- History from patient and witness(es)
- Physical examination (usually normal)
- Consider investigations: CXR; U + E, LFT, glucose, Mg, Ca++ to rule out underlying cause, eg metabolic, neoplastic
- All new cases to be referred to neurologist, paediatrician or paediatric neurologist for accurate diagnosis and initiation of treatment if indicated

2.1 Medical management of seizure
Objective: Total abolition of seizures except where patient wishes otherwise.
Three parts:
- Counselling
- Drug therapy
- Follow up

2.2 Counselling:
(a) First aid in case of fit:
   - Semi prone (recovery) position
   - Do not force object between teeth
   - Rectal diazepam
     one to three years: 5mg; over three years: 10mg. May repeat in five minutes if still fitting

(b) Driving limitation: Licence to drive heavy goods and public service vehicles permanently revoked once epilepsy diagnosed. Licence to drive all other vehicles given for limited period of time once fit free for two years.
(c) Avoid precipitating factors/hazards.
(d) Free prescriptions.
(e) Personal identification: Epi-Alert bracelet.
(f) Contraception: Interaction of enzyme inducing anticonvulsants with oral contraceptive pill reducing its efficacy: non hormonal contraceptives preferable.
(g) Pre-conception drug review:
   - Use lowest effective dose of anticonvulsant
   - Consider withdrawal if seizure free for two to three years (preferably)
   - Periconceptual folate supplements
(h) Once or twice daily dose regime (Table 1). Optimise compliance – warn about side effects of anticonvulsants (Table 2).
(i) Employment and rehabilitation.
(j) Avoid overprotection.
(k) Irish Epilepsy Association:
   - Provide professional help and advice
   - Improve public understanding
   - Assist research
   - Promote employment, rehabilitation and education

2.3 Drug Therapy (Table 1)
Principles:
(a) Treat early after second seizure has confirmed diagnosis.
(b) Use single drug.
(c) Monitor seizures using seizure diary.
(d) Increase dose slowly until adequately controlled or toxic effects begin to appear (sedation, nystagmus ➔ ataxia, dysarthria ➔ confusion and drowsiness).
(e) When changing drug, introduce new anticonvulsant in small doses and increase gradually over several weeks while withdrawing previous drug.
(f) In women of child bearing years, monotherapy with carbamazepine drug of choice. All anticonvulsants potentially teratogenic.
(g) Serum levels if toxicity or poor compliance suspected; poor control; mental retardation;
## Epilepsy protocol

### Six commonly used anticonvulsant drugs

<table>
<thead>
<tr>
<th>Drug</th>
<th>Indications</th>
<th>Initial dose regimen</th>
<th>Usual adult maintenance dose regimen¹</th>
<th>Usual daily dose intervals</th>
<th>Usual therapeutic range</th>
</tr>
</thead>
<tbody>
<tr>
<td>Carbamazepine</td>
<td>All seizure types except generalised absence</td>
<td>Therapy should be initiated at a low dose to avoid initial side effects, 100-200 mg nocte should be given for a week and then the dose should be increased by 100 mg or 200 mg steps at weekly intervals</td>
<td>600-1,400 mg/day. Dose changes should be made in 100-200 mg steps</td>
<td>b.d.(occasionally t.d.s.)</td>
<td>15-50 mmol/litre</td>
</tr>
<tr>
<td>Ethosuximide</td>
<td>Generalised absence (petit mal) seizures</td>
<td>250 mg/day, increasing by 250 mg steps every five days</td>
<td>500-1,500 mg/day. Dose changes should be made in 250 mg steps</td>
<td>b.d.(occasionally t.d.s.)</td>
<td>200-600 mmol/litre</td>
</tr>
<tr>
<td>Phenobarbitone</td>
<td>All seizure types except generalised absence</td>
<td>30-60 mg nocte, increasing by 30 mg steps at weekly intervals</td>
<td>60-180 mg/day. Dose changes should be made in 30-60 mg steps</td>
<td>Once a day</td>
<td>60-180 mmol/litre</td>
</tr>
<tr>
<td>Phenytoin</td>
<td>All seizure types except generalised absence</td>
<td>200 mg nocte, increasing by 50-100 mg steps at weekly intervals</td>
<td>200-400 mg/day. Dose changes should be made in 25-100 mg steps, and should be monitored by serum level, as the dose/serum level relationship is not linear, and at times small dose increases may result in large rises in serum level</td>
<td>Once a day (occasionally b.d.)</td>
<td>40-80 mmol/litre</td>
</tr>
<tr>
<td>Primidone</td>
<td>All seizure types except generalised absence</td>
<td>62.5-125 mg nocte, increasing by 125-250 mg steps every five days</td>
<td>500-1,000 mg/day. Dose changes are usually made in 250 mg steps</td>
<td>b.d.</td>
<td>As for phenobarbitone²</td>
</tr>
<tr>
<td>Sodium valproate</td>
<td>All seizure types</td>
<td>400-600 mg/day increasing by 200 mg steps every week</td>
<td>600-2,000 mg/day. Dose changes should be made in 200-500 mg steps</td>
<td>b.d. (occasionally t.d.s.)</td>
<td>Not very helpful. Usual level with therapeutic dose (blood taken two hours after dose) is 250-700 μmol/litre</td>
</tr>
</tbody>
</table>

¹In some patients, higher doses may be required

²For primidone, monitor the serum levels of the derived phenobarbitone
presence of renal or hepatic disease; phenytoin or multiple drug treatment.

(h) No difference in efficacy between phenobarbitone, phenytoin, carbamazepine and valproate for the treatment of specific seizure types in newly diagnosed adults or children.

(i) Phenobarbitone not now recommended as drug of first choice.

(j) Seizure frequency may increase with high drug levels.

(k) Newer agents have fewer adverse effects.

(l) 70% remission on initiation of therapy.

(m) After two or more years seizure freedom on monotherapy, 58% remain seizure free on drug withdrawal.

2.4 Follow up

(a) Should be planned and regular.

(b) Ideally should be shared with consultant neurologist, while on long term anticonvulsants.

(c) See patient and relatives frequently initially. Ensure easy access to primary health care team.

(d) Assess problems and understanding of seizure control, compliance, side effects. (Table 2).

(e) Blood tests:
   1. Serum levels of valproate, primidone and benzodiazepines unhelpful. Measure levels if poor compliance or toxicity suspected; if patient is on phenytoin or multiple drug treatment; presence of renal or hepatic disease; mental retardation.
   2. Check FBC Ca++, Phos, Alk Phos, U+E, LFT at six monthly intervals. Blood tests should never take precedence over clinical evaluation.

(f) Plan to next review.

(g) Follow up facilitated by call/recall system, disease register, labelling of notes and flow charts.

(h) When seizure free for two or more years, consider drug withdrawal over minimum of six months under supervision. Usually done in conjunction with specialist.

Special points

3.1 Paediatrics

(a) Generally poor compliance.

(b) Check developmental assessment.

(c) Neuro-cutaneous signs, eg Sturge-Weber syndrome.

(d) Distinguish jitteriness from fits, especially in small babies.

3.2 Pregnancy

(a) Rule out eclampsia as cause of seizures in pregnancy.

(b) Higher incidence of foetal abnormality.

(c) Other problems – low folate, risk of rickets, neonatal haemorrhage, neonatal withdrawal symptoms.

(d) Mothers may breast feed while taking anticonvulsants.

(e) Use folate supplement periconceptually and during pregnancy.

(f) Consider drug withdrawal preconception if fit free for two to three years. Seizures may also pose risk to foetus however.

Febrile convulsions

4.1 Definition

An epileptic seizure occurring in a child aged between six months and five years precipitated by a fever arising from infection outside the nervous system in a child who is otherwise neurologically normal.

Distinguished from epilepsy which is characterised by recurrent non febrile fits at any age.

Characteristics

♦ Two thirds of cases single and brief
♦ Not epilepsy
♦ Affects 3% of children
♦ 90% due to viral upper respiratory tract infections
♦ Seizure tends to occur at height of pyrexia

Management

4.3 Acute febrile convulsion

(a) Telephone advice – lay child prone and keep cool.

(b) Rectal diazepam:
   One to three years 5 mg ‘Stesolid’
   Over three years 10 mg ‘Stesolid’
   Repeat these doses after five minutes if fitting continues.

(c) Consider admission to hospital:
   – After first seizure
   – Age over 18 months
   – Complicating factors, eg social

4.4 Coping with subsequent febrile illness

♦ 33% risk of recurrence of febrile convolution in subsequent febrile illness
♦ Use temperature indicator
Paracetamol and minimal clothing
Temperature control fails to prevent recurrent febrile convulsions

4.5 Prognosis
Recurrence rate of febrile convulsions is 33%. It is more likely if:
- Age less than one year
- Family history febrile convulsions in first degree relative

4.6 Risk of subsequent epilepsy
Risk is minimally increased from 0.8% to 1.04% in uncomplicated seizures (ie less than 15 minutes and no focal signs or neurological sequelae).

4.7 Prophylaxis
- Long term drugs rarely indicated
- Intermittent rectal diazepam given at onset of, or during fit, now drug of first choice

Dose: One to three years = 5 mg.
Over three years = 10 mg.
May be repeated after five minutes if fitting continues.

4.8 Immunisation
- Full immunisation important
- May induce fever; take antipyretic precautions after immunisation (regular paracetamol, minimal clothes, fluids, monitor temperature)

4.9 Pertussis vaccine
Not contraindicated in child with history of febrile seizures. Ensure antipyretic precautions taken after immunisation, especially in first 24 hours.

4.10 MMR vaccine
Not contraindicated; take antipyretic precautions following vaccination; do not give concomitant immunoglobulin.

Table 2

<table>
<thead>
<tr>
<th>Drugs</th>
<th>Idiosyncratic</th>
<th>Intoxication</th>
<th>Chronic</th>
</tr>
</thead>
<tbody>
<tr>
<td>Carbamazepine</td>
<td>Rash, blood dyscrasia</td>
<td>Diplopia, unsteadiness, dizziness, drowsiness</td>
<td>Hyponatraemia</td>
</tr>
<tr>
<td>Ethosuximide</td>
<td>Rash, blood dyscrasia</td>
<td>Unsteadiness, headache, drowsiness, ataxia,</td>
<td>Headache, behaviour change</td>
</tr>
<tr>
<td>Phenobarbitone</td>
<td>Rash, blood dyscrasia</td>
<td>Sedation, headache, unsteadiness, behaviour</td>
<td>Sedaion, behaviour change,</td>
</tr>
<tr>
<td>Phenotoin</td>
<td>Rash, blood dyscrasia, immunological</td>
<td>Unsteadiness, sedation, involuntary movements,</td>
<td>Sedaion, intellectual blunting,</td>
</tr>
<tr>
<td></td>
<td>reactions</td>
<td>dizziness, nausea, headache, behaviour</td>
<td>mood change, intellectual</td>
</tr>
<tr>
<td>Primi done</td>
<td>Acute dizziness, unsteadiness, nausea</td>
<td>Sedation, headache, unsteadiness, behaviour</td>
<td>Sedaion, behaviour change,</td>
</tr>
<tr>
<td>Sodium valproate</td>
<td>Rash, blood dyscrasia, acute hepatic</td>
<td>Sedation, unsteadiness, behaviour change,</td>
<td>Sedation, behaviour change,</td>
</tr>
<tr>
<td>valproate</td>
<td>failure, acute pancreatitis,</td>
<td>nausea</td>
<td>mood change, intellectual</td>
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<tr>
<td></td>
<td>acute thrombocytopenia</td>
<td></td>
<td>blunting, metabolic bone</td>
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<td></td>
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<td>disease, connective tissue</td>
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<td></td>
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<td>disorders</td>
</tr>
</tbody>
</table>

Common or important side effects of the major anticonvulsant drugs

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Epilepsy protocol

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Irish College of General Practitioners
Careful history

- from patient
- from witness(es)

Physical examination
including developmental assessment
and neuro-cutaneous signs

Not Epilepsy

Epilepsy?

Refer to hospital
Consider pre-hospital work-up

Epilepsy Confirmed

Counselling
(may be carried out by GP
and/or other professional)

- First aid in case of fit
- Driving limitations
- Avoid Ppt factors/hazards
- Free prescriptions
- Personal identification
- Contraception
- Pre-conception drug review
- Folate supplements
- peri-conceptually and
during pregnancy
- Mothers may breast feed
- while on anticonvulsants
- Higher incidence of foetal
abnormalities
- Avoid overprotection
- Irish Epilepsy Association
- Optimise compliance
- generally poor in children
- Employment and
rehabilitation

Drug Therapy

- Treat early after second seizure
- Use a single drug
- Use a seizure diary
- Start with low dose of drug
- Increase dose slowly until
adequately controlled or
toxic S/E appear
- Monotherapy with carbamazepine
in females of child bearing
years
- Serum levels if
  - Toxicity
  - Poor compliance
  - Poor control
  - Mental retardation
  - Renal or hepatic disease
  - Phenytoin therapy
  - Multiple drug therapy
- Seizure frequency may increase
  with high drug levels
- Newer agents have fewer adverse effects
- When changing drug:
  Introduce new drug in small doses;
  gradually increase dose while
  withdrawing previous drug over
  several weeks

Follow up

- Should be planned, regular and shared with a consultant
- See patient and relatives frequently initially;
  ensure easy access to health care team
- Assess problems, understanding, compliance, side effects
- Blood tests – serum levels if appropriate; FBC, Ca, Phos,
Alk Phos, U+E, LFT at 6/12 intervals
- Plan to next review
- When seizure-free for two or more years, consider supervised
  drug withdrawal over minimum of six months
Step by step action in case of febrile convulsion

**Telephone advice**

- Lay child prone in recovery position
- Minimal clothes
- Rectal diazepam if fitting is more than five minutes
  
  If one to three years use 5 mg
  If over three years use 10 mg
  
  Repeat after five minutes if fitting continues

**On arrival**

- Rectal diazepam only if still fitting and second dose has not been given as above
- Check for cause of fever
- Consider hospital admission
  - After first seizure
  - Age of child is less than 18 months
  - Complex convulsion
  - Complicating factors, eg social
- Explanation and support

**Subsequently**

- 33% recurrence rate especially if positive family history and aged less than a year
- Use temperature indicator, paracetamol and antipyretic measures* in subsequent febrile illness
- Minimally increased risk of epilepsy in uncomplicated febrile seizures
- Continuous prophylactic drugs rarely indicated. Intermittent PR. Diazepam at onset of fit is drug of first choice
- Full immunisation important including pertussis and MMR
- Take antipyretic measures* after immunisation

* Antipyretic measures: Minimal clothes, regular paracetamol, fluids, monitor temperature