

# A NATIONAL APPROACH TO EPILEPSY MANAGEMENT IN CHILDREN AND ADOLESCENTS

## 1. INTRODUCTION

The National Sentinel Clinical Audit of Epilepsy-Related Death concluded its review with the following statement: *“The Chief Medical Officer for England has recommended that within 3 months of completion of this audit an action plan should be in place in England to cut the level of preventable epilepsy-related deaths. The plan will need to address the problems identified in this audit, particularly: inadequate access to appropriate epilepsy care; lack of education of healthcare professionals about the principles of epilepsy management and the risks of epilepsy-related deaths; poor communication with patients and their families and other carers; and deficiencies in post-mortem investigations”*.

This paper is a response to that statement with particular reference to the situation in paediatrics. It also takes account of the findings of the expert review undertaken at Leicester into the service provided for children thought to have epilepsy.

## 2. BACKGROUND

Epilepsy is a generic term encompassing a wide range of epileptic disorders and syndromes, of variable severity and prognosis. Epilepsy is a common condition, with 0.05% (1 in 2,000) developing one of the epilepsies of childhood each year. The prevalence figure is about 0.5% (1 in 200) indicating that most large secondary schools would have six or so children with an epilepsy of childhood at any one time.

The management of the epilepsies of childhood and adolescence is made difficult by a number of factors:

- 2.1 Diagnostic difficulty. There are other conditions in childhood, some of them normal and developmental, some of them very common such as syncope (a tendency to faint), which can masquerade as epilepsy. Differential diagnosis may be a particular problem in children with pre-existing difficulties especially learning and behavioural disorders, but the problem is not confined to this group.
- 2.2 There are no diagnostic tests as such. Modern electroencephalography (EEG) techniques allow an increasing opportunity to capture a seizure, which is usually diagnostic when typical discharges appear. However, the timing of events often makes this difficult, in which case the EEG never offers more than a varying degree of corroboration of clinical assessment.
- 2.3 There are a number of epilepsies of childhood and adolescence. Knowledge of these amongst non-specialist clinicians is generally poor. Lack of awareness of epilepsy syndromes may result in inappropriate treatment with either lack of efficacy or worsening of epilepsy. Awareness of epilepsy syndromes allows more accurate advice to be given regarding potential complications and outcome.

Epilepsy may occur in isolation or in conjunction with other disabling conditions. In the latter situation, there may be particular difficulties in distinguishing between epileptic and behavioural phenomena.

- 2.3 Some children with focal epilepsy may be cured with surgery after careful assessment. The investigations are very specialised but early intervention can avoid significant disability. The system at times currently fails to identify and refer those children who might benefit from assessment with a view to the possibility of surgery.
- 2.4 In many areas of the country access to special investigation resources is limited. Neurophysiology services require someone with special expertise in reading the EEGs of neonates and children. Neuroradiological resources similarly need someone with specialist paediatric neuroradiological expertise to interpret the results of investigations. An acceptable waiting time for a non-urgent EEG recording is two weeks (six weeks for more specialised studies). An MRI scan should be performed where appropriate and be routinely available within three months.
- 2.5 The quality of written published information available for families and children is very variable. To date there has been no process to validate the age appropriateness of written information.
- 2.6 Appropriate liaison with educational, and where necessary, social services is often found to be deficient.
- 2.7 Insufficient use is made of the internet using official websites as a source of information for children, parents and professionals alike.
- 2.8 In general, services for transitional care between adolescence and adulthood could be improved.

### 3. THE EVIDENCE BASE FOR PRACTICE

Research is continuing to define the answer to many important questions in epilepsy management. In paediatrics especially there is a need to develop syndrome specific management.

#### 3.1 Which anti-epileptic drugs are most effective?

A Cochrane review comparing odds ratios on recurrence risk for modern anti-epileptic drugs indicates very little difference between drugs available, with a varying profile for each in relation to unwanted effects. The Single And New Antiepileptic Drug study promises more information to allow rational drug choice.

National Institute for Clinical Excellence and Scottish Intercollegiate Guidelines Network are currently working to establish good practice guidance in this respect and a recent consensus conference was held in Edinburgh in September 2002.

#### 3.2 The place of epilepsy surgery is currently the subject of a Cochrane review.

#### 4. THE DEFICIENCIES OF THE CURRENT SERVICE

Five government sponsored reports and some 20 patient-based surveys of (dis)satisfaction since the establishment of the NHS have documented variable and inadequate standards in epilepsy care. The Clinical Standards Advisory Group involved 2,394 people with epilepsy and found that 43.4% with severe uncontrolled epilepsy had not seen a hospital consultant in the previous year and 14% had no medical supervision at all.

Misdiagnosis at the initial stage is a common problem. Detailed reviews of diagnostic rates indicate that between 20% and 40% of people may be attributed the wrong diagnosis at an early stage.

The National Clinical Audit of Epilepsy related deficiencies in care provision although there was no control group of people who did not die in the same period. More widespread deficiencies may have been revealed if this methodology had been followed.

There are some 50,000 children with epilepsy in the United Kingdom and 63 UK Paediatric Neurologists. Of these, 12 have a specialist interest in epilepsy and only 4 practise solely within epilepsy. The challenge for our services is how to cascade specialist knowledge out from Regional Centres into local District General Hospital based paediatric services and the Primary Care Trusts they serve.

#### 5. THE PROPOSAL

The model of care we propose has the following primary configuration:

- A managed clinical network bringing together key professionals to enhance the lives of young people with epilepsy.
- A well-defined integrated care pathway.
- Appropriate clinical guidelines for each relevant step on the pathway.

##### 5.1 The Integrated Care Pathway

- 5.1.1 Child presents (usually to a primary care health professional or to A & E) with an unusual episode for assessment.

Action: In many, a benign physiological phenomenon such as syncope would be identified; advice as appropriate. Diagnostic uncertainty should lead to referral to secondary care: local district based paediatric services.

- 5.1.2 Assessment by secondary services: District Based Consultant Paediatrician one of whom should have special responsibility for epilepsy. A 'first event' protocol should be established taking referrals from general practice and A & E, with clear guidelines for referral. Ideally a child would be seen after a period of no more than 2 weeks.

In some districts that host tertiary services, the tertiary paediatric neurology service may take on responsibility for this service.

- a) The paediatrician may share clinical assessment with nurses with a special responsibility for epilepsy as appropriate.
- b) Referral for further investigations as appropriate.
- c) Diagnostic information given according to good practice guidelines.
- c) Written validated information provided to support information given verbally in clinic.
- d) Nurse with special responsibility for epilepsy provides follow up and support.
- e) Liaison with education, social services and other health professionals as appropriate to develop integrated care plan for child.

**The Role Of District Based Nurses  
With Special Responsibility For Epilepsy**

We propose one or two district based nurses with special responsibility for epilepsy working with the second level paediatrician with responsibility for epilepsy.

1. Co-ordination of District Epilepsy working group to include education, learning disability and social services, school nurses, Child and Adolescent Mental Services, parents and children.
2. Counselling.
3. Source of validated information.
4. Family support/child guidance liaison.
5. GP liaison.
6. Medication check/dosage adjustment.

For each tertiary regional centre we propose two Specialist Paediatric Epilepsy Nurses working solely in the field of epilepsy. They would have a similar role but a higher level of knowledge and the ability to support district based nurses.

### **EEG Resources**

There are many children without neurological disease who have an abnormal, even “epileptiform” EEG and a significant proportion of children with epilepsy whose EEG is persistently normal. There is a high misdiagnosis rate for childhood epilepsy and over-reliance on EEG data is a common source of this. A clinical diagnosis of epilepsy based on thorough history taking is the essential first step.

Every network should have guidelines established for requesting EEG studies but some general principles can be stated:

1. The value of the EEG is in supporting a diagnosis and particularly in identifying epilepsy syndromes.
2. The EEG may influence management and there is recognition that experience in reporting paediatric EEG is crucial to interpretation.
3. EEG should be available on an emergency basis.

- 5.1.3 Diagnostic doubt / inadequate response to treatment, or complications, leads to referral to a designated paediatric neurologist via the district based paediatrician with special responsibility for epilepsy.

Assessment by a tertiary neurologist either at outreach clinic or tertiary centre (whichever appointment appropriate or offered soonest).

Referral guidelines should be developed locally in liaison with the district based paediatrician.

- 5.1.4 Referral to quaternary paediatric neurologist with special expertise in epilepsy.
- a) This referral often for selection for surgery, continuing diagnostic doubt or further opinion in relation to treatment, associated problems or to meet parental expectation.
  - b) Assessment for surgery is often very specialised work. Neuroimaging may be normal and invasive EEG monitoring with indwelling electrodes may be required. Early intervention can avoid significant disability.
- 5.1.5 For those with intractable epilepsy (up to 30% of the total) continuing good liaison between tertiary, secondary district-based services and Primary Care Trusts is required.

- 5.1.6 The district team is responsible for providing information, support for and liaison with families, education authority, and social services to optimise opportunity and encouragement for educational progress and employment.
- 5.1.7 Transitional care arrangements for those passing from adolescence to adult services need to be established. Services should provide information on genetics where appropriate, teratogenic drugs and pregnancy, drug interactions (especially in relation to the contraceptive pill). In many areas services are particularly deficient where the epilepsy accompanies a severe learning disability.
- 5.1.8 High quality paediatric pathology services are essential for the advancement of knowledge after the death of a child with epilepsy.

Resources are required to provide an integrated care pathway: different levels of resources will be needed at each stage but an integrated pathway will have access to all components:

- Primary Care Physician
- District Paediatrician with special responsibility for epilepsy
- Nurses with specific expertise and responsibility for childhood epilepsy
  
- Neurophysiology (EEG) services
- Neuroimaging
- Psychology including neuropsychology and clinical psychology and other therapy/assessment services including speech and language services
  
- Specialist Paediatric Epilepsy Nurse
- Paediatric Neurologist with outreach to each District or equivalent service and/or Paediatric Neurologist with special interest in epilepsy able to co-ordinate assessment including specialised EEG, neuroimaging, neuropsychology and epilepsy surgery assessment
- Child Psychiatry
  
- Validated information for families:
  - on investigations
  - on epilepsy syndromes
  - on treatment including surgery

### **THE RETURN on INVESTMENT**

The aim is to improve the specialist care of children with epilepsy and to deliver a locally based service where possible.

Predicted improved outcome will be seen in these areas:

- More direct and rapid access to specialist advice, diagnosis and treatment
- Lower rates of misdiagnosis
- Fewer unwanted medication effects with appropriate drug treatment
- Increased cure rate through selection for surgery
- Earlier appropriate intervention by educational services
- Earlier appropriate intervention by social services

The number with intractable epilepsy will probably not be reduced (save for the small proportion helped by surgery) but for those who retain their epilepsy treatment will be optimal, access to validated information and support systems will improved and educational, social and employment services will be offering the best level of opportunity and encouragement.

6. ADDITIONAL RESOURCE REQUIREMENTS FOR A MANAGED CLINICAL NETWORK

- 6.1 Paediatric nurses with a special responsibility for epilepsy at a district level and tertiary specialist Paediatric Epilepsy Nurses. Expansion of numbers of district and tertiary centre based Specialist Paediatric Nurses with a specified training programme and continuing professional development. (d/w ESNA)
- 6.2 Paediatricians with special responsibility for epilepsy. One third of paediatric consultations have a neurological component. There is a wide variation in the current provision of paediatricians in each district with this designated responsibility with some districts having no such named person. Development of such posts may require additional resources or expansion of the existing incumbent's interest.
- 6.3 Epilepsy information resource to be available to all stakeholders .
- 6.4 An agreed process for commissioning the network and managerial support for the clinical network in each region.
- 6.5 Improved access to tertiary and quaternary centres will require consultant paediatric neurology expansion.
- 6.6 Improvement of access to neurophysiology (currently shortage of technicians and consultant neurophysiologists). The proposal is for at least one consultant with an expertise in paediatric EEG for each region (this role could be fulfilled by a paediatric neurologist with appropriate additional training).
- 6.7 Expansion of access to neuroimaging and appropriate number of specialists in paediatric neuroimaging.
- 6.8 Funding for the regionally based epilepsy interest groups. Funding is required for supporting secretarial activity, travel expenses, room hire, invited speaker expenses, updating a web based internet information.
- 6.9 Continuing professional development for paediatricians with special responsibility for epilepsy services (secondary being district based), secretarial support, venue hire, speaker fees, material development, etc.

### **The Role Of Paediatricians With Special Responsibility For Epilepsy**

1. To maintain standards of diagnosis and treatment and appropriate referral patterns along the clinical pathway.
2. Liaison with district-based colleagues over the selection of children requiring referral to tertiary based services.
3. To attend specialist epilepsy outreach clinics.
4. Regular attendance at the regionally based special interest groups in epilepsy.
5. To supervise the continuing care of children with intractable epilepsy with special responsibility for education/social service liaison.

To audit and improve local service delivery.

### **The Role of the Regionally Based Epilepsy Interest Groups**

- For district paediatricians with special responsibility for epilepsy, paediatric neurologists, specialist paediatric epilepsy nurses.
- 3-monthly meetings.
- Sharing difficult diagnostic cases.
- Collation audit activity on standards.
- Co-ordination of research activity.
- Education linked to continuing professional development.
- Business planning for developing the network.

### **Educational Courses Suitable for the Continuing Professional Development of Paediatricians with Special Responsibility for Epilepsy / General Paediatricians / Specialist Paediatric Epilepsy Nurses**

Proposal:

- To be held annually.
- Three or four such courses to be established in the United Kingdom.
- Course certification.
- Attendance required, at a minimum, every 4 to 5 years.