Epilepsy and intellectual disability
a workshop

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Aim and structure

• To understand diagnostic skills at the borderlands
• To be able to assess and ameliorate epilepsy related risk
• To formulate a treatment plan
We are in the borderlands of epilepsy

Sir William Gowers. 20.03.1845-04.05.1915

His original book consisted of those whose presentation challenged the diagnostic scheme he had developed and raised difficult questions by demonstrating the limits of his understanding.
An assessment model

Lessons from NEAD
Simple pathway
Limited tools
An event of diagnostic uncertainty

Define the event

Identify comorbidity especially intellectual disability

Verbal history
Association with external factors
Video
Further investigation
What tools do we have?

- Investigation results
- Knowledge of differential diagnosis
- Your ability to communicate in the setting of clinical equipoise
- Ability to be objective
Minimum requirements for the diagnosis of psychogenic nonepileptic seizures: A staged approach
A report from the International League Against Epilepsy Nonepileptic Seizures Task Force

*†W. Curt LaFrance Jr., ‡Gus A. Baker, §Rod Duncan, ¶Laura H. Goldstein, and #Markus Reuber

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SUMMARY

An international consensus group of clinician-researchers in epilepsy, neurology, neuropsychology, and neuropsychiatry collaborated with the aim of developing clear guidance on standards for the diagnosis of psychogenic nonepileptic seizures (PNES). Because the gold standard of video electroencephalography (vEEG) is not available worldwide, or for every patient, the group delineated a staged approach to PNES diagnosis. Using a consensus review of the literature, this group evaluated key diagnostic approaches. These included: history, EEG, ambulatory EEG, vEEG/monitoring, neurophysiologic, neurohumoral, neuroimaging, neuropsychological testing, hypnosis, and conversation analysis. Levels of diagnostic certainty were developed including possible, probable, clinically established, and documented diagnosis, based on the availability of history, witnessed event, and investigations, including vEEG. The aim and hope of this report is to provide greater clarity about the process and certainty of the diagnosis of PNES, with the intent to improve the care for people with epilepsy and nonepileptic seizures.

LaFrance et al, ILAE NES TF.
**Minimum requirements for the diagnosis of psychogenic nonepileptic seizures: A staged approach**

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<table>
<thead>
<tr>
<th>Dx level of Certainty</th>
<th>History</th>
<th>Witnessed Event (semiology)</th>
<th>EEG</th>
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<tbody>
<tr>
<td><strong>Possible</strong></td>
<td>+</td>
<td>By witness or self-report / description</td>
<td>No epileptiform activity <em>interictal</em> routine or sd-EEG</td>
</tr>
<tr>
<td><strong>Probable</strong></td>
<td>+</td>
<td>By clinician on video or in person (semiology c/w NES)</td>
<td>No epileptiform activity <em>interictal</em> routine or sd-EEG</td>
</tr>
<tr>
<td><strong>Clinically Established</strong></td>
<td>+</td>
<td>By clinician on video or in person (semiology c/w NES)</td>
<td>No epileptiform activity on ictal ambulatory or rout. EEG</td>
</tr>
<tr>
<td><strong>Documented</strong></td>
<td>+</td>
<td>By clinician, (semiology c/w NES), while on video EEG</td>
<td>No epileptiform activity captured on ictal video EEG</td>
</tr>
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</table>

*(LaFrance et al, ILAE NES TF.*
Where are we now – the burden of care

- Mortality
- Hospitalisation
- Emotional, Cognitive, and behavioural
- Social & Cultural
Why a risk assessment?

Epilepsy is associated with a range of risks, drowning, injury, scalding, Sudden Unexpected Death in Epilepsy. Some of these are preventable (e.g. drowning) and in some appropriate intervention can reduce the risk (SUDEP).

NICE Guidance (NICE clinical guideline 137; guidance.nice.org.uk/cg137) states: Children, young people and adults with epilepsy and their families and/or carers should be given, and have access to sources of, information about…Risk Management.

It further define this as:

All children, young people and adults with epilepsy and learning disabilities should have a risk assessment including:

Bathing and showering

Preparing food

Using electrical equipment

Managing prolonged or serial seizures

The impact of epilepsy in social settings
N.I.C.E

• The suitability of independent living, where the rights of the child, young person or adult are balanced with the role of the carer.
• It is an expectation that all patients on the epilepsy pathway are NICE compliant.

• Competencies
• All professionals completing risk assessments should be competent in, or if not recognise the need for advice on:
  • 1. Identifying and classifying seizure types.
  • 2. Knowing key drug side effects
  • 3. Identifying individual risk of SUDEP
• These competencies should be supported by competency in communication with people with a learning disability, their carers and other health professionals including the assessment of capacity.
The 3 I’s
Identify the risk

• The precise risk should be defined. This for example would be for drowning in the case of bathing or death in the case of SUDEP.
Individualise the seizures

• Identify each seizure type the person has and its relative likelihood to impact on the risk issue.

• For example a drop attack may not have much risk of electrocution but a complex partial seizure occurring with automatisms in a kitchen may.

•

•
**Implement measures to reduce risk**

- This will usually be in two parts.

- The first practical measures specific to the risk such as night time monitoring for SUDEP or showering for drowning whilst bathing.

- The second is epilepsy management assessment. This will be to look at ensuring epilepsy management is appropriate. This could be identifying that further treatment is necessary for convulsive seizures or use of rescue medication to improve community access.

- It is strongly recommended that this element is undertaken in consultation with the medical team.
<table>
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<th>Identify</th>
<th>Individualise</th>
<th>Implement</th>
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<tbody>
<tr>
<td><strong>The risks from bathing are:</strong>&lt;br&gt;1. Drowning&lt;br&gt;2. Scalding (in bath)</td>
<td>All seizure types can lead to drowning with exception of auras when there is clear consciousness and no motor involvement.&lt;br&gt;An individual who has been seizure free for over a year and who is not on programme of medication is at low risk. However even in such cases it is worth recognising that the occasional recurrent seizure can occur.</td>
<td>Ensure every effort has been made to ensure seizure freedom&lt;br&gt;Discuss option of shower rather than bath&lt;br&gt;The only safe bathing plan is when an individual is directly observed at all times. A carer outside the door listening is not safe</td>
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## Managing prolonged or serial seizures

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<th>Individualise</th>
<th>Implement</th>
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<tr>
<td><strong>The risks are:</strong> Progression into status, death</td>
<td>Take clear history of all seizure types and identify episodes of prolonged events, especially those of greater than 5 minutes duration. Identify if convulsive seizure. Ensure the timing does include the recovery period. For cluster seizures assess impact on individual of cluster is it dangerous</td>
<td>All patients with prolonged seizures or clustering need an assessment by an epilepsy specialist for the use of rescue medication</td>
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SUDEP

<table>
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<th>Individualise</th>
<th>Implement</th>
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<tr>
<td>The risk of SUDEP is death</td>
<td>Whilst SUDEP is associated with all types of epilepsy research suggests that generalised tonic clonic convulsions (including secondary generalised) are at highest risk. As immediate care is potentially protective it is also important to identify the presences of nocturnal seizures.</td>
<td>Ensure every effort has been made to ensure seizure freedom. SUDEP reduction is through best possible seizure control and direct care during and after a seizure. There is evidence to suggest that having someone with you after a convulsive seizure until the time you recover may reduce SUDEP thus a post seizure plan is needed for all people with epilepsy and convulsive seizures. This should include identification of the seizure, including at night, and the availability for someone to sit with the patient following the seizure.</td>
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Formulating a treatment plan:
Know the person, know the family, understand the interventions
make a plan
For all options there is a fairly simple assessment to do

Adapted to the individual’s needs and outcome

Indicated → Efficacious → Safe

Workable, measurable? Most of all it is communicated as an Action not Inaction
Treatment options - can be more than one modality at one time

- Watchful waiting
- Pharmacotherapy
- Surgical treatments
- Psychological therapy
- Diet
The impact of AEDs – responder rate

<table>
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<tr>
<th>Responder rate</th>
<th>Relative effect (95% CI)</th>
<th>No of Participants (studies)</th>
<th>Quality of the evidence (GRADE)</th>
<th>Comments</th>
</tr>
</thead>
<tbody>
<tr>
<td>Responder rate (≥50% reduction in overall seizure frequency)</td>
<td>RR 2.58 (1.60 to 4.14)</td>
<td>382 (3).</td>
<td>low</td>
<td>The included studies assessed different AEDs (lamotrigine, rufinamide, topiramate). Similar age ranges were included in the studies and they had a similar duration of treatment. Two studies were rated as having unclear risk of bias and one study had low risk of bias.</td>
</tr>
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</table>
Surgical treatments resective

A randomised, controlled trial of surgery for temporal lobe epilepsy
Wiebe et al. N Eng J Med 2001 Aug 2; 345 (%): 311-8
Results

• 80 randomised to surgery or treatment with AED for one year

• “free of seizures impairing awareness”

• At one year:
  • 58% in the surgical group & 8% medical group free of seizures impairing awareness (p<0.001).
  • Better QOL in surgery
  • 10% adverse effects surgery
  • One patient died in medical Group
Surgery – non resective- VNS


• 3 Class 1 evidence
• 2 Class 2 evidence
• Many other studies

• “An effective and relatively safe adjunctive therapy in patients with medically refractory epilepsy not amenable to resection. However it is important to recognise that complete seizure freedom is rarely achieved using VNS and that a quarter of patients do not receive any benefit from therapy
Knowing the person and family
Searching beyond the seizures

Patient related outcome measures.
With thanks to Ritty George
Patient Reported Outcome Measures (PROMs)

- PROMS are principally self-completed questionnaires, but can also be in the form of interviews, diaries or by means of a computer or any other handheld electronic devices (Devlin et al., 2010). PROMs can be used to measure a variety of constructs such as functional and heath status, quality of life, symptom and symptom burden, personal experience of care and health related behaviours (Black., 2013). These measures can be either generic or condition-specific.
PROMS in people with an intellectual disability

Who?
• Patient, carer or other

What
• Are issues same for people with an intellectual disability?

When
• How frequent, resource implications?
Usage in my service*

The Glasgow Epilepsy Outcome Scale

Out of the 360 patient records, 69.7% (n=251) had a GEOS form filled and 30.3% (n=109) did not. There was variation in the level of concerns of informal and formal carers in all four sub-sections of the GEOS scale (p=0.000): concerns about seizures (A), concerns about drugs for epilepsy (B), concerns about injury (C) and concerns about daily life (D). Informal carers also seemed to give higher scores for concerns than formal carers.

*Ritty George Cardiff University Unpublished BSc Thesis 2016
Usage in my service*
The Glasgow Epilepsy Outcome Scale

The five concerns that both informal and formal carers reported which were in the top 10 concerns in the GEOS form were concerns about ‘fits causing damage to the brain’, ‘not seeing seizures happen/not being there’, ‘having fits during the night’, ‘having fits despite drugs’ and ‘SUDEP (unexplained death in epilepsy)’

*Ritty George Cardiff University Unpublished BSc Thesis 2016
Management guidance
The recommendations on choice of treatment and the importance of regular monitoring of effectiveness and tolerability are the same for those with learning disabilities as for the general population.

Every therapeutic option should be explored in children, young people and adults with epilepsy in the presence or absence of learning disabilities.
Consensus Guidelines*

• “Selection of the drug for a given individual must be based on an understanding of the individual, each drug’s pharmacology, side effect protocol interactions and risk. For the individual with intellectual disability the impact of AEDs on their cognition, behaviour and mobility should be particularly considered. (Grade C) “

*Kerr et al JIDR 53 * 687-694 August 2009
Special Groups – syndromes/conditions?

**Down syndrome**
- Recognise late onset myoclonic seizures and use appropriate AEDs for generalised seizures

**Autism**
- Assessing the impact of treatment change in individuals with autism is particularly difficult. A baseline assessment of behaviour and physical status is regarded as essential

**Tuberose sclerosis**
- Awaiting the potential use of MTOR inhibitors
Special groups-epilepsy syndromes

- Level 1 evidence for Lamotrigine, topiramate, Rufinimide and Clobazam
A pathway for achieving efficacy and effectiveness

Individual choice and characteristics including seizure syndrome

Carer environment knowledge and skills

Agreed outcome: Seizure retention tolerability PROMS

Choose AED to match these

Measure seizures and PROMS
Conclusion

• Whatever your role with people with intellectual disability and epilepsy a good knowledge of why and how we treat epilepsy is valuable