Intellectual Disability (ID), Autism & Epilepsy

Management of epilepsy in adults with intellectual disability

Presented on behalf of the ID Epilepsy Strategy Working Committee
People with ID (~25% of all PWE) are more susceptible to being in the higher risk groups of treatment resistance but have less defined treatment outputs

- Mild ID – 8 -10% Moderate – profound ID - 50%
- 60% of people with ID and epilepsy will be treatment resistant
- Seizures 2\textsuperscript{nd} most common reason for premature mortality in ID
- Deaths - 43% with epilepsy - 31% had had a seizure in the previous 5 years
- SUDEP 3 -9 times her risk of SUDEP

Misdiagnosis rates

CIPOLD
ID Specific issues
Ambulatory care sensitive conditions

- English Hospital Episode Statistics (HES) data; 2005 to 2009 inclusive

- Crude rate of emergency admissions for ACSCs is 76 admissions/1000/year for adults with ID.

- This is roughly five times the rate for other people (15 per 1000)

- 5.8 days/admission for ID compared to 3.7 for others
Hospital Admissions which should not happen

Figure 2 Comparison of the causes of emergency admissions for ACSCs admissions for people with and without LD or associated conditions (cont).
Autism & it’s spectrum

- Up to 38% may develop epilepsy

- Two peaks
  - 0-5 years
  - 10 years +

- Seizures have low remission rate (16% in adulthood)

Risk of epilepsy in autism and ID

Tuchman & Rapin Lancet Neurol 2002;1:352–8
ASD subtypes & epilepsy
(Tuchman & Rapin 2002)

- Core autism (autistic disorder, AD)
  - About 30% show AR
  - About 30% develop a clinical epilepsy by adolescence
- Asperger syndrome
  - ~5-10% develop epilepsy in early childhood
- Pervasive Developmental Disorder NOS
  - Increased risk of epilepsy linked to severity of brain dysfunction
- Disintegrative Disorder
  - up to 70% develop epilepsy
- Rett syndrome
  - >90% develop epilepsy
**Table 1**

Characteristics of the individuals with or without epilepsy

<table>
<thead>
<tr>
<th></th>
<th>Epileptics (n = 33)</th>
<th>Non-epileptics (n = 97)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Gender (M/F)</td>
<td>25/8</td>
<td>81/16</td>
</tr>
<tr>
<td>Birth weight (g)</td>
<td>3227 ± 374</td>
<td>3172 ± 457</td>
</tr>
<tr>
<td>Head circumference (cm)(^a)</td>
<td>33.5 ± 1.0</td>
<td>33.5 ± 1.7</td>
</tr>
<tr>
<td>Age when walking alone (months)</td>
<td>14.4 ± 7.0</td>
<td>13.7 ± 3.6</td>
</tr>
<tr>
<td>Diagnosis of DSM-IV</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Autistic disorder</td>
<td>26</td>
<td>75</td>
</tr>
<tr>
<td>Atypical autism</td>
<td>7</td>
<td>22</td>
</tr>
<tr>
<td>Speech loss in infancy (+/-)</td>
<td>7/26</td>
<td>17/80</td>
</tr>
<tr>
<td>Cognitive level(^***)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Normal–mildly retarded</td>
<td>1 (3.1%)</td>
<td>31</td>
</tr>
<tr>
<td>Moderately retarded</td>
<td>6 (20.0%)</td>
<td>24</td>
</tr>
<tr>
<td>Severely retarded</td>
<td>8 (22.2%)</td>
<td>28</td>
</tr>
<tr>
<td>Profoundly retarded</td>
<td>18 (56.3%)</td>
<td>14</td>
</tr>
<tr>
<td>Adaptive level (SQ)(^b,)**</td>
<td>42.1 ± 16.8</td>
<td>59.4 ± 20.6</td>
</tr>
<tr>
<td>Psychotropic drugs (+/-)</td>
<td>21/12</td>
<td>39/58</td>
</tr>
</tbody>
</table>

\(^a\) Six data points were not available.

\(^b\) Two data points were not available.

\(^*\) \( p = 0.026 \) (Fisher, both side).

\(^**\) \( p < 0.05 \).

\(^***\) \( p < 0.001 \).
Current prescribing practices in ID

- Current approach – is this working?

- Can strategies that work in the general population apply directly to ID?

- Are we tailoring available AEDs using a person-centred approach?

AEDs, anti-epileptic drugs
* Side effects: behavioural, mental, and physical

* Relevant co-morbidities

* Evidence base of individual AEDs

<table>
<thead>
<tr>
<th>Drug Name</th>
<th>ID specific evidence</th>
<th>Type of evidence</th>
<th>Comments</th>
</tr>
</thead>
<tbody>
<tr>
<td>Carbamazepine</td>
<td>Kaski et al 1991</td>
<td>Improved efficacy using slow release preparation vs. standard</td>
<td>No direct evidence of tolerance or efficacy</td>
</tr>
<tr>
<td>Gabapentin</td>
<td>Crawford et al</td>
<td>Add on comparative open study with Lamotrigine – no difference</td>
<td>Power side effects of aggression?</td>
</tr>
<tr>
<td>Lamotrigine</td>
<td>Motte et al 1997</td>
<td>LGS specific, RCT using placebo</td>
<td>power, specific syndrome</td>
</tr>
<tr>
<td></td>
<td>Buchanan 1995</td>
<td>N = 34 majority showed &gt; 50% improvement</td>
<td>Power</td>
</tr>
<tr>
<td></td>
<td>Gidal et al 2000</td>
<td>N = 44, 45% &gt; 50% improvement 20% worsening</td>
<td>Power, who's who?</td>
</tr>
<tr>
<td></td>
<td>McKee et al 2006</td>
<td>N = 22 sub analysis of a larger study</td>
<td>Power</td>
</tr>
<tr>
<td>LEV</td>
<td>Kelly et al. 2004</td>
<td>N = 64 Observational study of adjunct LEV 38% seizure free</td>
<td>Improved seizure control in majority and carer satisfaction</td>
</tr>
<tr>
<td></td>
<td>Brodtorb et al 2004</td>
<td>n = 184 ID n = 56 equally effective</td>
<td>Study focus was on behaviour - worse in ID</td>
</tr>
<tr>
<td>Topiramate</td>
<td>Kerr et al 2005</td>
<td>RCT Double blind to placebo n = 57 28/29 32% reduction in seizure frequency vs. 1%</td>
<td>No negative impact on behaviour Power</td>
</tr>
<tr>
<td>Sodium V</td>
<td>SANAD?</td>
<td>Sub analysis of difficult to treat</td>
<td>Multiple issues</td>
</tr>
<tr>
<td>Lacosamide</td>
<td>Flores et al 2012</td>
<td>Real world cohort N = 403 18% ID sub analysis No differences between ID vs. non ID</td>
<td>Case selection</td>
</tr>
</tbody>
</table>
Conclusions

- Concept of ID – nebulous – all lumped into one!
- No specification of the nature or degree of ID, even when ID is mentioned
- Poor descriptions of co-morbidities, such as PDD/autism
- The newer AEDs possibly tend to be better tolerated and affect cognitive functioning to a lesser degree than older AED options. However the evidence base for safe use is extremely weak
- No Concept of what is ‘Challenging Behaviour’

Why should a psychiatrist working with people with ID have a knowledge of epilepsy?
The development of standards and initiatives

- Enhance diagnosis, pathways to investigation
- Guidelines for treatment
- Improve links among different stakeholders between primary care, MDT, social services and patient centred clinical consultations
<table>
<thead>
<tr>
<th>Recommended Actions</th>
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<tbody>
<tr>
<td><strong>Investigations and diagnosis in individuals with complex needs</strong></td>
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<tr>
<td>A Working Group to develop</td>
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<tr>
<td>• the diagnosis of epilepsy</td>
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<tr>
<td>• educational initiatives to improve clinician communication</td>
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<tr>
<td>• identify a pathway to investigation for complex needs</td>
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<tr>
<td><strong>Medication</strong></td>
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<tr>
<td>• Establish a Task Force to develop</td>
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<td>• guidelines for the treatment &amp; establishing best practice for the identification and management of AED side effects</td>
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<td>• Audit templates</td>
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<td>• Rescue medication</td>
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<tr>
<td><strong>Enhancing medical services</strong></td>
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<tr>
<td>• Guidance on</td>
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<td>• Accessible &amp; exchangeable information in clinic settings</td>
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<td>• Minimum standards on MDTs primary care, shared decisions, person centred patient care etc.</td>
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<tr>
<td>• epilepsy specialist nurse provision</td>
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<tr>
<td>• training manuals to support non-specialist services</td>
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</table>
| BRONZE                                                                 | 4. That adults with epilepsy have an agreed and comprehensive written care plan  
|                                                                      | 6. That adults with a history of prolonged or repeated seizures have an agreed written emergency care plan  
|                                                                      | 8. That adults with epilepsy who have medical or lifestyle issues that need review are referred to specialist epilepsy services  
|                                                                      | 9. That young people with epilepsy have an agreed transition period during which their continuing epilepsy care is reviewed jointly by paediatric and adult services |
| SILVER                                                               | 1. That adults presenting with a suspected seizure are seen by a specialist in the diagnosis and management of the epilepsies within 2 weeks of presentation  
|                                                                      | 2. That adults having initial investigations for epilepsy undergo the tests within 4 weeks of them being requested  
|                                                                      | 3. That adults who meet the criteria for neuroimaging for epilepsy have an MRI  
|                                                                      | 5. That adults with epilepsy are seen by an epilepsy specialist nurse who they can contact between scheduled reviews  
|                                                                      | The above are in addition to all of Bronze level indicators |
| GOLD                                                                 | 6. That adults who meet the criteria for referral to a tertiary care specialist are seen within 4 weeks of referral  
|                                                                      | The above is in addition to all of Bronze and Silver level indicators |
Adapted from Mike Kerr - – Epilepsy in People with ID: Can we reduce the Burden of Disease? Keynote presentation at Faculty Psychiatry of Intellectual Disability Annual Conference 26 – 27 September 2013
Epilepsy in People with an ID - Core Knowledge, Skills and Attitude Framework
Adapted from framework developed by Skills for Health, Skills for Care and Health Education England

Gold
• Knowledge, skills & attitudes for those who are providing **expert** epilepsy care
  • example: ID psychiatrists & neurologists who diagnose & manage complex epilepsy

Silver
• Knowledge, skills & attitudes for those who are providing epilepsy support & care
  • example ID psychiatrists, neurologists, GPs managing the epilepsy.

Bronze B
• Knowledge, skills and attitudes for roles that will have **regular contact** with epilepsy
  • example general practice, psychiatry trainees in all specialities particularly ID

Bronze A
• Knowledge, skills & attitudes for roles that require **general awareness** of epilepsy in ID
  • For example medical students, ED doctors
Implementing Strategy
Next Steps

Links with other Stakeholders

Links with training structure

Competencies & CPD

Better evidence of mortality causes - ? LeDer link

Assess current evidence base for service provision - ? Link with NASH/LD –observatory

Position paper from RCPsych of prescribing modelling STOMP
Working Group

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Advisors
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Dr Regi Alexander
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Dr Ashok Roy

“Life for people with major disabilities supported by good services will often look quite ordinary, but this ordinariness will be the product of a great deal of careful planning and management”

Mansell 2007