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## Title page

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Behavioural disorder in people with an intellectual disability and epilepsy: a report of the Intellectual Disability Task Force of the Neuropsychiatric Commission of ILAE

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## Summary

The management and needs of people with intellectual disability (ID) and epilepsy are well evidenced; less so, the comorbidity of behavioural disorder in this population.

'Behavioural disorder' is defined as behaviours that are difficult or disruptive, including stereotypes, difficult or disruptive behaviour, aggressive behaviour toward other people, behaviours that lead to injury to self or others, and destruction of property. These have an important link to emotional disturbance. This report, produced by the Intellectual Disability Task Force of the Neuropsychiatric Commission of the ILAE, aims to provide a brief review of some key areas of concern regarding behavioural disorder among this population, and proposes a range of research and clinical practice recommendations generated by Task Force members. The areas covered in this report were identified by experts in the field as being of specific relevance to the broad epilepsy community when considering behavioural disorder in persons with epilepsy and ID; they are not intended to be exhaustive. The practice recommendations are based on the authors' review of the limited research in this field combined with their experience supporting this population. These points are not graded but can be seen as expert opinion guiding future research and clinical practice.

### Keywords:

Comorbidities

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Disability

## *Epidemiology*

ID is typically diagnosed when individuals are assessed as having limitations in both intellectual functioning (an IQ of 70 or under) and adaptive behaviours (conceptual, practical and social skills) with onset occurring during the developmental period<sup>1</sup>. Approximately 1.04% of the population are estimated to have intellectual disability<sup>2</sup>. Challenges arise however in determining prevalence estimates for this population as they typically rely on persons known to specialist service providers and therefore may exclude a 'hidden' population who do not engage with services, likely those with mild ID.

The prevalence of epilepsy among individuals with ID has been found to be well in excess of that reported in the general population; at 22% (CI95% 19.6-25.1) in a pooled estimate determined from a recent meta-analysis of 48 studies published since 1990<sup>3</sup>.

Prevalence estimates for the general population, in contrast, are considerably lower (0.80%)<sup>4</sup>.

Early studies examining the prevalence of behavioural disorder among this population reported elevated levels among children with ID and epilepsy<sup>5</sup>. Molteno et al (2001)<sup>6</sup>, in a study of 355 children attending special schools in South Africa, reported higher levels of psychopathology among those who had epilepsy, notably in the domains of self-absorbed and autistic related behaviours. Similarly, McGrother et al (2006)<sup>7</sup>, in a large sample of 2,393 adults with ID, found that those with epilepsy were more likely than those without epilepsy to engage in disturbing others at night, seeking attention (16.1% vs. 10.6%), and being uncooperative (20.0% vs. 12.6%). In contrast to these findings, Robertson et al (2015)<sup>3</sup> identify a number of studies which report no difference in rates of behavioural problems between individuals with intellectual disability who have epilepsy and those who do not.

A small number of studies have reported a decreased rate of behavioural disorder among persons with epilepsy and ID. A population-based prevalence study of 416 individuals with severe and profound ID in Finland reported a higher percentage of individuals without

epilepsy presenting with behavioural disturbance (27.9% vs 17.6%)<sup>8</sup>. Similarly, Arshad et al (2011)<sup>9</sup> found that rates of mental health problems were significantly lower among participants with epilepsy (9.4%) than participants without epilepsy (48.2%) in a sample of 752 individuals with ID who were consecutive referrals for assessment to a specialist mental health service. Specifically, rates for schizophrenia (7.1% vs. 20.3%), personality disorder (3.8% vs. 8.6%), and anxiety (4.5% vs. 7.9%) were significantly lower among those with epilepsy.

While the evidence base is limited, these studies suggest a complex relationship among ID, epilepsy and behavioural disorder. It is likely that methodological differences in samples, case definitions, and diagnostic criteria impact the variation in findings. A reduction in these differences towards more standardised methodologies may contribute to a greater understanding of the combined impact of these complex conditions.

#### ***Practice points for epidemiology***

1. Population-based epidemiological studies are recommended to ensure that individuals at all levels of intellectual disability are represented.
2. Standardised definitions of intellectual disability, epilepsy and behavioural disorder should be agreed upon to ensure consistency among epidemiological studies.

#### ***Aetiology***

Current evidence suggests a number of key variables may influence the likelihood of behavioural disorder for individuals with ID and epilepsy: genetic causation, severity of ID, and the presence of autistic spectrum characteristics. Powis and Oliver<sup>10</sup> for example,

reviewing the prevalence of aggression in a range of genetic disorders, noted that certain conditions appear to have greater prevalence of aggression as compared with others.

The environment has also been identified as impacting the development and maintenance of behavioural problems. Environmental factors may have specific impact for known genetic syndromes and often the impact leads to the phenotypic behaviours<sup>11</sup>.

Exposure to life events, for example, increases the risk of psychopathology while environmental responses, such as reinforcement and punishment, may act as maintaining factors of these behaviours<sup>12</sup>.

When considering epilepsy variables in the aetiology of behavioural disorder, the role of factors such as seizure frequency, peri-ictal events and antiepileptic drugs (AEDs) have been examined. The impact of AEDs is covered later in this report. Psychiatric phenomena in the peri-ictal period, in particular post-ictal psychosis, are well described (see for example Clancy et al, 2014<sup>13</sup>) and an association with violent acts has been observed. In the general epilepsy population, both post-ictal confusional states and rage have been documented<sup>14</sup>. Such behaviours may reflect alterations in cortical excitability<sup>15</sup>. Considerable challenges remain for clinicians to determine the source of behavioural disorder, whether due to epilepsy, use of medication, a combination of both or other factors<sup>16</sup>.

#### ***Practice points for aetiology***

1. Assessment of the aetiology of behavioural disorder should include identification of the cause of an individual's ID.
2. The presence of autistic traits should be identified in any individual presenting with behavioural disorder.
3. Assessment of challenging behaviour should include an assessment for comorbid psychopathology such as ADHD.

4. In all individuals the association between seizures and peri-ictal behavioural changes should be identified.

5. In all individuals the association between seizure worsening or improvement and behaviour change should be assessed.

### ***Behavioural side effects of antiepileptic drugs***

There is a dearth of high quality information on behavioural changes associated with AED use among individuals with epilepsy and ID. A Cochrane Review found that the majority of studies in this field typically used no or non-reliable measures of behavioural exacerbation, were uncontrolled, and were mostly retrospective in nature<sup>17</sup>.

The impact of AEDs on behaviour and cognition ranks second within 11 areas prioritized in consensus guidelines developed by the Health Special Interest Group of IASSIDD (International Association for the Scientific Study of Intellectual and Developmental Disability)<sup>17</sup>. Whilst newer AEDs in general offer some advantages as compared to older AEDs, mainly a lack of enzyme-inducing properties, they may also result in behavioural side effects.

Access to AEDs will be restricted in many countries globally; the restriction being a lack of more modern AEDs. It is important to recognise that some medications that are rarely used because of behaviour concern (an example of which could be phenobarbitone) may be used frequently in countries with low income and limited availability of AEDs. This may lead to an increase in behavioural effects.

Research indicates that some AEDs may have both positive and negative behavioural side effects. Helmstaedter and colleagues, for example, reported that 59% of 228 consecutive

out-patients prescribed levetiracetam reported behavioural changes compared to 9% of controls; of these 37% reported negative changes in behaviour and 22% positive changes<sup>19</sup>.

These side effects may also occur in persons with ID, in some cases to a higher frequency than that observed for the general epilepsy population specifically, for those prescribed levetiracetam, aggressive behaviour has been reported to occur more frequently in persons with ID<sup>19</sup>.

Research assessing lamotrigine, though generally believed to have a positive impact on behaviour, has reported negative changes. Seven out of 20 individuals with ID (five with Lennox Gastaut syndrome) who were prescribed lamotrigine as part of different AED regimens spontaneously reported behavioural changes using the Aberrant Behaviour Checklist (ABC)<sup>20</sup>. Behavioural improvement was observed for four individuals, whilst adverse behavioural effects were noted for three; findings which the authors state reflect the varied influence of lamotrigine on behaviour. The authors also note that serum concentrations of lamotrigine were not predictive of behavioural change.

A randomized, double-blind, placebo-controlled trial of topiramate reported improved behaviour in both active and control groups with no statistical difference between the groups<sup>21</sup>. In contrast, an open prospective study showed significant adverse cognitive and behavioural side effects of topiramate in children and adolescents with ID<sup>22</sup>. More recently, an evaluation of routine clinical data reported that individuals with epilepsy and ID may experience the same cognitive side effects under topiramate as individuals who do not have ID<sup>23</sup>.

Behavioural improvement for those prescribed vigabatrin was associated with seizure freedom in seven children with epileptic, formerly infantile, spasms<sup>24</sup>. The positive impact of seizure improvement is likely to be true across AEDs but has not been researched. Other

AEDs that are associated with such behavioural side effects in persons without ID, for instance zonisamide or perampanel, can cause these effects in persons with ID, although no clinical trial data specific to people with ID and epilepsy exist. High drug load may also be associated with negative behavioural changes and is frequent in persons with epilepsy and ID<sup>25</sup>. Older AEDs (e.g. primidone and phenobarbital) may also have behavioural side effects, including when drug withdrawal is tried.

In summary, the impact of AEDs on behavioural change is, in many cases, unpredictable due to a lack of trial information. To address this issue, clinicians are encouraged to monitor behavioural change closely, employing established instruments for the assessment of adverse events such as the Adverse Event Profile (AEP)<sup>26</sup> or the FENAT<sup>27</sup>. Similarly, the ABC is a widely-used instrument assessing behaviour profiles in persons with ID<sup>28</sup> and could be considered in clinical practice.

#### ***Practice points for behavioural side-effects of anti-epileptic drugs***

1. AEDs may have positive or negative behavioural side effects in persons with ID.
2. Behavioural effects should be monitored closely.
3. Validated assessment scales are needed for patients with epilepsy and ID.
4. There is a major need for research into behavioural safety of AEDs in persons with ID.

#### ***Pharmacological treatments of behavioural disorder***

Multiple difficulties hinder efforts to include people with ID in randomised controlled trials (RCTs). These difficulties include, amongst others: consent, difficulty in applying strictly controlled protocols, heterogeneity of aetiology, and blurred boundaries between

target symptoms. Consequently, data reliably proving or disproving the effectiveness of specific psychotropic agents are lacking, especially among those with co-morbid epilepsy.

Table one summarises current clinical recommendations for the use of psychotropic medication for aggression.

**[Insert Table 1 here]**

The lack of clinical trial information, compounded by evidence of the impact of antipsychotics on seizure frequency leads to considerable clinical equipoise<sup>39</sup>. For the clinician in epilepsy it should be considered that no drug is often the best drug for this group of patients. When medication is used, it should be originated from services competent in ongoing assessment of the behaviour, the environment and other associated psychopathology.

#### ***Practice points for pharmacological treatments of behavioural disorder***

1. Behavioural disorder is multifactorial and a thorough assessment, including a functional analysis of behavior, is needed before medication is started.
2. Use of medications to manage behaviour is not recommended for non-experienced epilepsy services; shared care with psychiatric services is needed.
3. When used, courses should be short and monitored for efficacy.
4. Psychotropic medication can be used to treat mental illness contributing to behaviours that are challenging.

#### ***Epilepsy surgery and the risk of cognitive or behavioural change***

The *European Federation of Neurological Sciences (EFNS) and the Epilepsy Surgery Guidelines and Proposed ILAE Criteria for Epilepsy Surgery in Children* do not consider ID

as a contraindication to proceed with a surgical procedure<sup>40-41</sup>. Moreover in the UK, National Institute for Health and Care Excellence (NICE) Guidelines state that children, young people and adults with ID must not be discriminated; therapies and investigations for the general epilepsy population should be offered<sup>42</sup>. The number of patients with a low IQ that are offered surgery, however, remains less than those functioning within the normal intelligence range<sup>43</sup>.

Limited information on the behavioural outcome of surgery exists to help in decision making. An analysis of 664 patients found no association between seizure outcome, postoperative cognitive development and behavioural outcome (using the Child Behaviour Checklist) and IQ level ( $\leq 70$ , 70-85,  $>85$ ) when patients were matched according to surgical variables such as age or surgical procedure<sup>44</sup>. Behavioural outcome showed lower scores one year after surgery in all groups, indicating fewer behavioural difficulties after surgery than before. Other factors, such as duration of epilepsy, were also associated with seizure outcome in patients with a low IQ<sup>45</sup>.

No deterioration of cognitive functioning was observed in patients with a low IQ compared to those functioning within the normal intelligence range<sup>44</sup>. A further study of a series of 31 patients evaluated with the Washington Psychosocial Seizure Inventory showed an improvement in psychosocial functioning for those who became seizure free<sup>45</sup>.

Liang et al, described low IQ as a factor associated with cognitive improvement in a small series of 25 patients with tuberous sclerosis complex (TSC) who underwent epilepsy surgery<sup>46</sup>. Another review paper identified the benefits of epilepsy surgery in 177 patients with tuberous sclerosis, where IQ was reported in 62 cases (50 patients with ID); the authors concluded that deterioration of cognitive functioning may be prevented with epilepsy surgery<sup>47</sup>.

A predominance of large surgical resections (multilobar, hemispherectomy) was observed in patients with lower IQ<sup>48</sup>. Other palliative procedures like corpus callosotomy (CC) and vagal nerve stimulators, have also been extensively used in this population.

Although postsurgical cognitive complications like disconnection syndrome have been reported more frequently in patients after CC, a recent paper found that after this procedure, half of patients showed attention enhancement (related to improvement in drop attacks) and behavioral outcome was better at earlier age of surgery<sup>49</sup>.

### ***Practice points for epilepsy surgery and ID***

1. ID is not a contraindication for epilepsy surgery and a presurgical evaluation should be offered in refractory cases as in other patients with epilepsy.
2. Epilepsy surgery can benefit cognitive and behavioural outcomes especially in patients who remain seizure free.
3. Patients and families should be advised that although current data is positive, the precise impact of surgery on an individual's epilepsy, and indeed their intellectual functioning, cannot be fully predicted.

### ***Psychological management of behavioural & emotional issues***

Psychological therapies and behavioural management techniques are effective in improving the quality of life of people with ID<sup>50-51</sup>. Despite this, there is limited research investigating cognitive or behavioural treatments, and currently, there are no randomised controlled trials assessing the efficacy of psychological therapies in persons with comorbid ID and epilepsy<sup>52</sup>. This is concerning given the range of psychosocial challenges facing individuals with ID, including unemployment and poverty, a lack of meaningful friendships

or intimate relationships, stressful family circumstances, trauma and abuse, and elevated rates of mental health difficulties<sup>51</sup>. Moreover, for persons with ID and epilepsy, there is limited information on the long-term effects of seizures on their cognitive and behavioural functioning, and they face the added psychosocial challenges of living with often ‘hard to treat’ epilepsy, including reduced daily living skills, self-care and adaptive social behaviours, social stigma, lack of independence, exploitation by others, and increased carer burden and burnout<sup>53</sup>.

There appears to be a barrier to referral for treatment; adults with ID are less likely to be referred for psychological therapy than adults without ID<sup>50</sup>. Also relevant is poor detection of mental health problems associated with: (i) ‘diagnostic overshadowing’, where a mental health problem is not recognised due to difficulties differentiating it from challenging behaviours associated with ID<sup>51</sup>, (ii) the lack of diagnostic assessment tools with robust evidence of reliability and validity for detecting mental health problems, and (iii) the treatment gap between mental health and ID services, which have distinct cultures, and can be detrimental to identifying problems and providing continuity of care<sup>51</sup>.

In considering the cognitive abilities necessary for psychological therapy in people with ID, the success of cognitive behaviour therapy (CBT) in children without ID highlights that fully developed adult abilities are not needed to gain treatment benefits. Despite this, the cognitive content of treatment (i.e., what a person thinks) has generally been overlooked in favour of the cognitive process (i.e., how a person thinks), even though the former can underpin an individual’s psychosocial difficulties and can be targeted in treatment<sup>51</sup>.

A cognitive deficit model has been commonly used with people with ID, which promotes increased self-monitoring through instructional training, often in the form of a behaviour modification program, to ameliorate cognitive and behavioural difficulties. This contrasts

with the cognitive distortion model of CBT traditionally employed with adults without ID, where the therapist elicits negative automatic thoughts, identifies the relevant cognitive distortions, and helps the individual to modify or reframe thinking to improve mood and wellbeing. Since the latter has been shown to have better generalisability across behaviours and environmental settings, ideally both models should be considered when referring people with ID and epilepsy for treatment<sup>51,54</sup>.

A 'step-wise' approach to psychological treatment in patients with ID is recommended, whereby patients should first undergo formal neuropsychological assessment of their cognitive abilities and skills prior to commencing treatment to identify cognitive strengths and weaknesses<sup>54</sup>. Since over 80% of people with ID have mild ID, neuropsychological testing can profile the patient's general intellect (IQ), memory, attention, information processing speed, verbal communication and comprehension, and executive functions (i.e., planning, abstract reasoning, mental flexibility, working memory), with higher verbal IQ linked to better treatment outcomes in some studies<sup>51-54</sup>. Also beneficial is an assessment of the patient's metacognitive profile, particularly relating to emotional recognition, self-awareness, insight, and an ability to understand the links between cognition and emotion, as these skills are directly relevant to the success of CBT<sup>54</sup>.

Following the initial assessment, the therapist can then build on existing patient skills and work to develop new skills where required to maximise the effectiveness of the intervention. Where abilities or skills are unable to be developed, the intervention should be adapted<sup>51, 54</sup> with the goal of enhancing communication and understanding of the patient experience so that a shared understanding between the patient, carer and therapist can be achieved<sup>55</sup>. Examples of ways in which psychological therapy can be tailored to the cognitive abilities and skills of the patient are provided in Table 2<sup>56-57</sup>.

[Insert Table 2 here]

Table 3 contains a summary of psychological treatment studies of mental health problems in individuals with ID using a cognitive behaviour approach. Across studies, the strongest evidence supports the efficacy of CBT in treating aggression and anger in people with ID<sup>58</sup>. Anger is a challenging emotion to treat in adults both with and without ID, and has clinical salience in the ID population as aggression can lead to institutionalisation and over-prescription of medications for behavioural control. Moreover, resolving anger can remove attentional biases and cognitive distortions associated with threat perception, as well as memory biases for distressing experiences that are challenging to process<sup>58</sup>. Thus, the use of CBT for the effective treatment of anger and aggression in people with ID speaks to the viability of cognitively-based psychological treatments for improving patient quality of life.

[Insert Table 3 here]

***Practice points for Psychological management of behavioural & emotional issues***

1. People with ID should not be excluded from psychological therapies but should be able to access psychological therapies when needed.
2. A 'step-wise' approach is recommended, including neuropsychological assessment of cognitive strengths and weaknesses and subsequent tailoring of the therapy to build on existing patient skills.
3. Both behaviour modification and CBT should be considered, and for persons with mild ID who present with anger, psychological therapies such as CBT may be beneficial.
4. Clinicians should also consider treatment of comorbid mental illness in people with ID with behavioural challenges.

***Challenges in adolescence: autism, behaviour and epilepsy.***

Up to 33% of people with epilepsy of childhood-onset have persistence of seizures into adulthood, and 19-35% never achieve remission<sup>61,62</sup> Whilst this non-remission group often have associated neurodeficits, unless severely affected, most will survive into adulthood<sup>63</sup>. With age, complications are common affecting cognitive, behavioural and psychosocial functioning.

Tuberous Sclerosis Complex offers a useful model for the transition of children with epilepsy, behaviour and developmental disability. People with TSC have ID in 50% of cases, which may be severe, and 40% have Autism Spectrum Disorder (ASD). As the child grows into adulthood the emphasis of care shifts from seizure control and developmental issues to renal, psychiatric disease and other issues<sup>64</sup>. Remission of epilepsy occurs for many patients permitting AEDs to be tapered off<sup>65</sup>. In patients with TSC the severity and phenotype of the autistic features are inextricably linked with intelligence and epilepsy outcomes. Mental health issues occur in 66% of individuals with TSC, with anxiety and obsessive-compulsive tendencies common and handicapping.

Transition to adult services is complex as there are many medical issues which change with age and fall outside the neurological system<sup>66</sup>. Hence a “medical team” is needed. Effective transition programs from paediatric to adult care need both services to work together. Support for family and carers must be in place including identification of guardianship, establishment of trust funds, exploration of residential living options, preparing for changes in the family such as parental aging, carers having their own medical issues, and addressing the parental concerns of what will happen to the child who outlives them<sup>64</sup>.

Adult epilepsy or neurology waiting rooms and clinics are ill-equipped for younger patients who present with behavioural difficulties, and adult neurologists can be uncomfortable with such patients, especially those with aggressive behaviour, sexuality expression, and sleep disorders<sup>67</sup>.

Input from a pharmacologist is needed since there is often polypharmacy not just related to AEDs<sup>67</sup>. Involvement of other medical and non-physician services should include primary care for basic health maintenance such as nutrition, influenza immunization, and dentistry<sup>67,68</sup>. Also input from speciality physicians and rehabilitation therapists are needed<sup>68,69</sup>.

It is important to plan early for transition, to identify on-going caregivers and decide which is better, an actual isolated once off “hand-over” clinic or a chronic combined service. The clinician must be aware that other health issues may dominate such as behavioural disorders, and psychiatric manifestations. The social challenges can be huge and it may be better for a non-neurologist to lead the chronic on-going care plan. It is important to work as a flexible team.

In order for the above to be effective it is essential for the child neurologist to prepare and plan ahead, and to identify and involve key role players.

***Practice points for challenges in adolescence: autism, behaviour and epilepsy***

1. Transition of patients with complex disability should be planned.
2. The adult service will need to reproduce an often comprehensive paediatric model of care including physical, cognitive, psychiatric and behavioural needs.
3. The presence of ASD should be noted.

4. Individual aetiology of the ID is crucial and allows prospective care planning.
5. Adult services should be identified prior to transition.

### ***Social Policy - supporting family caregivers of those with complex disability***

Families are distinguished as both recipients and contributors to the care and support of individuals who have ID and epilepsy. The evidence-base, while limited, suggests a level of dissatisfaction by families in both roles.

As recipients of services, families report challenges sourcing specialist expertise, notably at primary care level; consultations characterised by poor communication and insufficient time; professionals lowering their expectation of treatment options for this population; and reluctance by some professionals to provide support regarding behavioural problems<sup>70</sup>. Moreover, interagency collaboration amongst these professionals is poor, resulting in families navigating a complex and fragmented pathway to care<sup>71</sup>.

While professional services are challenged in supporting individuals with such complex needs<sup>72</sup>, it is families that carry most of the care burden<sup>73</sup>. The evidence base on caregiver burden in epilepsy is sparse in comparison with research in other, less prevalent, neurological conditions and even more so for caregivers supporting those with both epilepsy and ID<sup>74</sup>. Albeit limited, the evidence indicates a substantial caregiver burden across multiple areas including physical, social and psychological domains<sup>75</sup>.

Physically, parents report chronic fatigue and sleep deprivation as a consequence of their caring duties<sup>76</sup>. The responsibility of providing care for individuals with such complex needs is substantial, and even the most vigilant of families seem unable to prevent injuries

occurring; consequently, the pool of friends and extended family willing to provide respite care is diminished<sup>77</sup>. The social burden of care is evident in strained marital relationships, marginalisation of other siblings whose needs become overshadowed, and a restriction in social activities as families perceive their presence is unwanted or feared by others enjoying social occasions<sup>78,70</sup>. Psychologically, caregiving for those with complex needs is associated with impaired psychological health, emotional health, quality of life and well-being<sup>79</sup>. Behavioural difficulties provide an additional source of stress<sup>80</sup>.

The ILAE has published a White Paper on the medical and social needs of people with epilepsy and ID<sup>71</sup>. The White Paper identifies the pivotal role of family caregivers and calls for recognition of their expertise and the promotion of shared care through greater knowledge transfer and communication with professionals. The White Paper also highlighted the need for more person centred approaches to consultations and the need for informed choice to be fostered among those with limited capacity. The ILAE, as the leading professional association within the epilepsy field, is charged with highlighting the needs of both individuals and families, and with providing guidance to both epilepsy and ID services on how to optimally support families using practical solutions such as respite.

#### ***Practice points for Social Policy standards***

1. Clinical services should recognize family burden and stress especially in families of people with behavioural disorder.
2. Interagency collaboration is required between disability and epilepsy services – family members should be a key stakeholder in this collaboration.

3. Practical supports for families, such as respite and access to information, are urgently required.

### ***Conclusion***

Behaviour and its manifestations have a pervasive impact on people with an ID and epilepsy. Many areas of need are not provided by epilepsy services. Moreover, both epilepsy and intellectual disability vary by degree of severity, with few research papers exploring the impact of severity. Any child or adult with an ID, epilepsy and behavioural disorder should be provided with multi-disciplinary care to ensure quality of professional input and improved quality of life for the individual. A dearth of sound scientific evidence has been shown in several sections of this report. This is surprising and concerning after so many years of research.

### **3-5 Key Bullet points:**

- Behaviour and its manifestations have a pervasive impact on people with intellectual disability and epilepsy.
- A thorough assessment is required prior to pharmacological treatment as is close monitoring of side effects.
- Clinical trial data for pharmacological and psychological interventions is limited for people with intellectual disability and epilepsy.
- Intellectual disability is not a contraindication for epilepsy surgery, but the precise impact cannot be fully predicted.
- Specific consideration is needed at times of transition (e.g. from child to adult services) and for family members.

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We confirm that we have read the Journal's position on issues involved in ethical publication and affirm that this report is consistent with those guidelines.

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**Table 1:** Clinical recommendations for the use of psychotropic medication for aggression

<b>Recommendation:</b>	<b>Source:</b>
Except for acute aggressive emergency interventions, antipsychotics may be more harmful than helpful.	(Tyrrer et al. 2008 <sup>29</sup> ; albeit contrasting results were reported by Gagiano <sup>30</sup> et al 2005).
If necessary, atypical antipsychotics are recommended rather than traditional ones because of lower toxicity during long-term use.	(Simon et al. 1996 <sup>31</sup> ; Aman et al. 2004 <sup>32</sup> ).
Clozapine should be the last antipsychotic to be chosen because of its potential pro-convulsive nature, as well as unpredictable interactions with Carbamazepine and Valproate.	(Alldredge 1999 <sup>33</sup> ; Mula & Monach 2002 <sup>34</sup> ; Mula et al. 2004 <sup>35</sup> ).
While methylphenidate is considered to effectively control some behavioural problems arising from hyperactivity in paediatric patients with ID and/or epilepsy, relevant data on safety and efficacy are lacking in regard to adults with ID and epilepsy.	(Simonoff et al. 2013 <sup>36</sup> ; Baptista-Neto et al. 2008 <sup>37</sup> ).
Only consider antipsychotics when:  Psychological or other interventions alone do not produce change within an agreed time or treatment for any  coexisting mental or physical health problem has not led to a reduction in the behaviour or the risk to the person or others is very severe (for example, because of violence, aggression or self-injury).	(NICE 2015 <sup>38</sup> )

Only offer antipsychotic medication in combination with psychological or other interventions.

**Table 2:** Recommendations for adapting psychological therapy in people with ID<sup>56-57</sup>.

<b>Therapeutic element</b>	<b>Definition</b>
Simplification	Less complex/technical; smaller chunks, shorter sessions
Language	Reduce vocabulary/sentence structure and length of thought
Activities	Augment typical activities; use of art, homework to make concepts concrete
Developmental level	Integrate developmental level into presentation; use of games, relevant social contexts
Directive methods	Explicit outline of goals and progress
Flexible methods	Adjust usual methods to suit cognitive level and progress rate
Involve caregivers	Use family and support staff; help with homework
Transference/countertransference	Clear therapeutic boundaries; attachments can be stronger and take a parental role
Sensitive interview methods	Avoid response biases; agreeableness, suggestibility, confabulation
Disability/rehabilitation approaches	Address the disability; reflect issues relating to self-identity and support positive self-review, mastery

**Table 3:** Psychological treatment studies of mental health problems in people with ID <sup>57, 59</sup>.

<b>Population</b>	<b>Intervention</b>	<b>Effect</b>
<b>Depression &amp; ID</b> 3 studies 1 treatment vs WL control <sup>60</sup>	CBT CBT group therapy	Reduced depression (behavior ratings and self-ratings) Decreased depression, negative thoughts, increased positive self-perceptions
<b>Anxiety &amp; ID</b> 8 studies	CBT, relaxation	Reduced anxiety, improved cognitive performance
<b>Anger &amp; ID</b> 6 studies	CBT (anger management)	Reduced anger and aggressive behaviours
<b>Psychosis &amp; ID</b> 3 studies	Behavioural treatments	Reduced displays of psychotic speech
<b>Offending &amp; ID</b> 10 studies	CBT	Changes in attitudes towards offensive behaviour, reduced offending-related cognitions and offending
<b>ID</b> 92 studies <sup>50</sup>	Psychotherapy	Moderately beneficial effect across a range of outcome measures, primarily behaviour (79%)