Epilepsia Open

SPECIAL REPORT

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

*Mike Kerr, †,‡Christine Linehan, §Christian Brandt, ¶Kousuke Kanemoto, **Jun Kawasaki, ††Kenji Sugai, ¶Yukari Tadokoro, ‡‡Vicente Villanueva, §§Jo Wilmshurst, and ¶¶,***Sarah Wilson

Epilepsia Open, 1(3-4):102–111, 2016
doi: 10.1002/epi4.12018

Professor Mike Kerr is a professor at Cardiff University, Wales, United Kingdom.

Summary

The management and needs of people with intellectual disability (ID) and epilepsy are well evidenced; less so, the comorbidity of behavioral disorder in this population. "Behavioral disorder" is defined as behaviors that are difficult or disruptive, including stereotypes, difficult or disruptive behavior, aggressive behavior toward other people, behaviors 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 behavioral 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 behavioral 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.

KEY WORDS: **Comorbidities, Behavior, Disability.**

Accepted August 16, 2016.

*Institute of Psychological Medicine and Clinical Neuroscience, Cardiff University, Cardiff, United Kingdom; †UCD Centre for Disability Studies, University College Dublin, Dublin, Ireland; ‡Tizard Centre, University of Kent, Canterbury, United Kingdom; §Department of General Epileptology, Bethel Epilepsy Centre, Mara Hospital, Bielefeld, Germany; ¶Department of Neuropsychiatry, Aichi Medical University, Aichi, Japan; **Kawasaki Clinic, Kyoto, Japan; ††Department of Child Neurology, National Center of Neurology and Psychiatry, Kodaira, Japan; ‡‡Multidisciplinary Epilepsy Unit, Neurology Service, University Hospital and Polytechnic La Fe, Valencia, Spain; §§Department of Paediatric Neurology, Paediatrics and Child Health, Red Cross War Memorial Children's Hospital, University of Cape Town, Cape Town, South Africa; ¶¶Melbourne School of Psychological Sciences, the University of Melbourne, Melbourne, Victoria, Australia; and ***Comprehensive Epilepsy Program, Austin Health, Melbourne, Victoria, Australia

Address correspondence to Christine Linehan, UCD Centre for Disability Studies, School of Psychology, University College Dublin, F208 Newman Building, Belfield, Dublin 4, Ireland. E-mail: christine.linehan@ucd.ie

Epidemiology

Intellectual disability (ID) is typically diagnosed when individuals are assessed as having limitations in both intellectual functioning (an IQ of 70 or under) and adaptive behaviors (conceptual, practical, and social skills) with onset occurring during the developmental period.[1] Approximately 1.04% of the population is estimated to have intellectual disability.[2] Challenges arise, however, in determining prevalence estimates for this population because they typically rely on persons known to specialist service providers and therefore may exclude a "hidden" population that does 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% (95% confidence interval [CI] 19.6–25.1) in a pooled estimate determined from a recent

Key Points

- Behavior 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 are 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

meta-analysis of 48 studies published since 1990.[3] Prevalence estimates for the general population, in contrast, are considerably lower (0.80%).[4]

Early studies examining the prevalence of behavioral disorder among this population reported elevated levels among children with ID and epilepsy.[5] Molteno et al.,[6] 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 behaviors. Similarly, McGrother et al.,[7] 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.[3] identify a number of studies that report no difference in rates of behavioral 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 behavioral 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 behavioral disturbance (27.9% vs. 17.6%).[8] Similarly, Arshad et al.[9] 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.

Although the evidence base is limited, these studies suggest a complex relationship among ID, epilepsy, and behavioral disorder. It is likely that methodological differences in samples, case definitions, and diagnostic criteria affect the variation in findings. A reduction in these differences toward more standardized 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 Standardized definitions of intellectual disability, epilepsy, and behavioral disorder should be agreed upon to ensure consistency among epidemiological studies.

Etiology

Current evidence suggests a number of key variables may influence the likelihood of behavioral disorder for individuals with ID and epilepsy: genetic causation, severity of ID, and the presence of autistic spectrum characteristics. Powis and Oliver,[10] 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 influencing the development and maintenance of behavioral problems. Environmental factors may have specific impact for known genetic syndromes, and often the impact leads to the phenotypic behaviors.[11] Exposure to life events, for example, increases the risk of psychopathology; environmental responses, such as reinforcement and punishment, may act as maintaining factors of these behaviors.[12]

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

Practice Points for Etiology

1 Assessment of the etiology of behavioral 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 behavioral disorder.

3 Assessment of challenging behavior should include an assessment for comorbid psychopathology such as attention-deficit/hyperactivity disorder (ADHD).
4 In all individuals the association between seizures and peri-ictal behavioral changes should be identified.
5 In all individuals the association between seizure worsening or improvement and behavior change should be assessed.

Behavioral Side Effects of Antiepileptic Drugs

There is a dearth of high-quality information on behavioral 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 nonreliable measures of behavioral exacerbation, were uncontrolled, and were mostly retrospective in nature.[17]

The impact of AEDs on behavior 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).[18] Although newer AEDs in general offer some advantages as compared to older AEDs, mainly a lack of enzyme-inducing properties, they may also result in behavioral side effects.

Globally, access to AEDs is restricted in many countries because of a lack of more modern AEDs. It is important to recognize that some medications that are rarely used because of behavior 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 behavioral effects.

Research indicates that some AEDs may have both positive and negative behavioral side effects. Helmstaedter and colleagues, for example, reported that 59% of 228 consecutive outpatients prescribed levetiracetam reported behavioral changes compared to 9% of controls; of these, 37% reported negative changes in behavior and 22% positive changes.[19] 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 behavior has been reported to occur more frequently in persons with ID.[19]

Research assessing lamotrigine, generally believed to have a positive impact on behavior, has reported negative changes. Seven out of 20 individuals with ID (5 with Lennox-Gastaut syndrome) who were prescribed lamotrigine as part of different AED regimens spontaneously reported behavioral changes as assessed by the Aberrant Behavior Checklist (ABC).[20] Behavioral improvement was observed for 4 individuals, whereas adverse behavioral effects were noted for 3, findings that the authors state reflect the varied influence of lamotrigine on behavior. The authors also note that serum concentrations of lamotrigine were not predictive of behavioral change.

A randomized, double-blind, placebo-controlled trial of topiramate reported improved behavior in both active and control groups with no statistical difference between the groups.[21] In contrast, an open prospective study showed significant adverse cognitive and behavioral side effects of topiramate in children and adolescents with ID.[22] 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.[23]

Behavioral improvement for those prescribed vigabatrin was associated with seizure freedom in 7 children with epileptic, formerly infantile, spasms.[24] 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 behavioral 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 behavioral changes and is frequent in persons with epilepsy and ID.[25] Older AEDs (e.g., primidone and phenobarbital) may also have behavioral side effects, including when drug withdrawal is tried.

In summary, the impact of AEDs on behavioral change is, in many cases, unpredictable owing to a lack of trial information. To address this issue, clinicians are encouraged to monitor behavioral change closely, employing established instruments for the assessment of adverse events such as the Adverse Event Profile (AEP)[26] or the Fragebogen zur Erfassung von Nebenwirkungen unter Antiepileptika-Therapie (FENAT).[27] Similarly, the ABC is a widely used instrument that assesses behavior profiles in persons with ID[28] and could be considered in clinical practice.

Practice Points for Behavioral Side Effects of AEDs

1 AEDs may have positive or negative behavioral side effects in persons with ID.
2 Behavioral 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 behavioral safety of AEDs in persons with ID.

Pharmacological Treatments of Behavioral Disorder

Multiple difficulties hinder efforts to include people with ID in randomized controlled trials (RCTs). These difficulties include, among others: consent, difficulty in applying strictly controlled protocols, heterogeneity of

etiology, and blurred boundaries between target symptoms. Consequently, data reliably proving or disproving the effectiveness of specific psychotropic agents are lacking, especially among those with comorbid epilepsy. Table 1 summarizes current clinical recommendations for the use of psychotropic medication for aggression.

The lack of clinical trial information, compounded by evidence of the impact of antipsychotics on seizure frequency, leads to considerable clinical equipoise.[39] The clinician in epilepsy should consider 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 behavior, the environment, and other associated psychopathology.

Practice Points for Pharmacological Treatments of Behavioral Disorder

1 Behavioral 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 behavior is not recommended for inexperienced 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 behaviors that are challenging.

Epilepsy Surgery and the Risk of Cognitive or Behavioral Change

The European Federation of Neurological Sciences (EFNS), the Epilepsy Surgery Guidelines, and the ILAE "Proposed Criteria for Referral and Evaluation of Children for Epilepsy Surgery" do not consider ID a contraindication to surgical procedures.[40,41] Moreover, in the United Kingdom, National Institute for Health and Care Excellence (NICE) guidelines state that children, young people, and adults with ID must not be discriminated against; therapies and investigations for the general epilepsy population should be offered.[42] The number of patients with a low IQ who are offered surgery, however, remains fewer than the number of those functioning within the normal intelligence range.[43]

Limited information on the behavioral outcome of surgery exists to help in decision making. An analysis of 664 patients found no association among seizure outcome, postoperative cognitive development, behavioral outcome (using the Child Behavior Checklist), and IQ level (≤70, 70–85, >85) when patients were matched according to surgical variables such as age and surgical procedure.[44] Behavioral outcome showed lower scores 1 year after surgery in all groups, indicating fewer behavioral difficulties after surgery than before. Other factors, such as duration of epilepsy, were also associated with seizure outcome in patients with a low IQ.[45]

No deterioration of cognitive functioning was observed in patients with a low IQ compared to those functioning within the normal intelligence range.[44] 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.[45]

Liang et al.[46] 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. 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.[47]

A predominance of large surgical resections (multilobar, hemispherectomy) was observed in patients with lower

Table 1. Clinical recommendations for the use of psychotropic medication for aggression

Recommendation	Source
Except for acute aggressive emergency interventions, antipsychotics may be more harmful than helpful.	Tyrer et al. (2008);[29] albeit contrasting results were reported by Gagiano et al. (2005)[30]
If necessary, atypical antipsychotics are recommended rather than traditional ones because of lower toxicity during long-term use.	Simon et al. (1996);[31] Aman et al. (2004)[32]
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);[33] Mula & Monach (2002);[34] Mula et al. (2004)[35]
Although methylphenidate is considered to effectively control some behavioral problems arising from hyperactivity in pediatric 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);[36] Baptista-Neto et al. (2008)[37]
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 behavior or the risk to the person or others is very severe (for example, because of violence, aggression, or self-injury). Only offer antipsychotic medication in combination with psychological or other interventions.	NICE (2015)[38]

IQ.[48] Other palliative procedures such as corpus callosotomy (CC) and vagal nerve stimulators have also been extensively used in this population. Although postsurgical cognitive complications such as 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.[49]

Practice Points for Epilepsy Surgery and ID

1 Intellectual disability 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 behavioral outcomes, especially in patients who remain seizure free.
3 Patients and families should be advised that although current data are positive, the precise impact of surgery on an individual's epilepsy, and indeed their intellectual functioning, cannot be fully predicted.

Psychological Management of Behavioral and Emotional Issues

Psychological therapies and behavioral management techniques are effective in improving the quality of life of people with ID.[50,51] Despite this, limited research investigates cognitive or behavioral treatments, and, currently, no randomized controlled trials assess the efficacy of psychological therapies in persons with comorbid ID and epilepsy.[52] 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.[51] Moreover, for persons with ID and epilepsy, there is limited information on the long-term effects of seizures on their cognitive and behavioral 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 behaviors, social stigma, lack of independence, exploitation by others, and increased carer burden and burnout.[53]

There appears to be a barrier to referral for treatment; adults with ID are less likely to be referred for psychological therapy than are adults without ID.[50] Also relevant is poor detection of mental health problems associated with: (1) "diagnostic overshadowing," where a mental health problem is not recognized owing to difficulties differentiating it from challenging behaviors associated with ID,[51] (2) the lack of diagnostic assessment tools with robust evidence of reliability and validity for detecting mental health problems, and (3) 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.[51]

In considering the cognitive abilities necessary for psychological therapy in people with ID, the success of cognitive behavioral 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 favor 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.[51]

A cognitive deficit model has commonly been used with people with ID, which promotes increased self-monitoring through instructional training, often in the form of a behavior modification program, to ameliorate cognitive and behavioral 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 well-being. Because the latter has been shown to have better generalizability across behaviors and environmental settings, ideally both models should be considered when referring people with ID and epilepsy for treatment.[51,54]

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.[54] Because 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.[51–54] 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, because these skills are directly relevant to the success of CBT.[54]

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

Table 3 contains a summary of psychological treatment studies of mental health problems in individuals with ID using a cognitive behavioral approach. Across studies, the strongest evidence supports the efficacy of CBT in treating aggression and anger in people with ID.[58] Anger is a challenging emotion to treat in adults both with and without ID and has clinical salience in the ID population because aggression can lead to institutionalization and overprescription of medications for behavioral 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.[58] 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.

Practice Points for Psychological Management of Behavioral and 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 behavior 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 behavioral challenges.

Challenges in Adolescence: Autism, Behavior, and Epilepsy

Up to 33% of people with epilepsy of childhood onset have persistence of seizures into adulthood, and 19–35% never achieve remission.[61,62] Although individuals in this nonremission group often have associated neurodeficits, unless severely affected, most will survive into adulthood.[63] With age, complications that affect cognitive, behavioral, and psychosocial functioning are common.

Tuberous sclerosis complex offers a useful model for the transition of children with epilepsy, behavior disorder, and developmental disability. Fifty percent of people with TSC have ID, 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.[64] Remission of epilepsy occurs for many patients, permitting AEDs to be tapered off.[65] 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 because many medical issues change with age and fall outside the neurological system.[66] Hence, a "medical team" is needed. Effective transition programs from pediatric to adult care need both services to work together. Support for family and carers must be in place, including identifying guardianship, establishing trust funds, exploring 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.[64]

Adult epilepsy or neurology waiting rooms and clinics are ill-equipped for younger patients who present with behavioral difficulties, and adult neurologists can be uncomfortable with such patients, especially those with aggressive behavior, sexuality expression, and sleep disorders.[67]

Input from a pharmacologist is needed because there is often polypharmacy not just related to AEDs.[67] Involvement of other medical and nonphysician services should include primary care for basic health maintenance such as nutrition, influenza immunization, and dentistry.[67,68] Also,

Table 2. Recommendations for adapting psychological therapy in people with ID[56,57]

Therapeutic element	Definition
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[57,59]

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

CBT, cognitive behavioral therapy; ID, intellectual disability; WL, waitlist control.

input from specialty physicians and rehabilitation therapists is needed.[68,69]

It is important to plan early for transition, identify ongoing caregivers, and decide which is better, an actual isolated once-off "handover" clinic or a chronic combined service. The clinician must be aware that other health issues may dominate such as behavioral disorders and psychiatric manifestations. The social challenges can be huge, and it may be better for a nonneurologist to lead the chronic ongoing care plan. It is important to work as a flexible team.

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, Behavior, and Epilepsy

1 Transition of patients with complex disability should be planned.
2 The adult service will need to reproduce an often comprehensive pediatric model of care, including physical, cognitive, psychiatric, and behavioral needs.
3 The presence of ASD should be noted.
4 Individual etiology 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, though limited, suggests a level of dissatisfaction by families in both roles.

As recipients of services, families report challenges sourcing specialist expertise, notably at the primary care level; consultations characterized 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 behavioral problems.[70] Moreover, interagency collaboration among these professionals is poor, resulting in families navigating a complex and fragmented pathway to care.[71]

Though professional services are challenged in supporting individuals with such complex needs,[72] it is families that carry most of the care burden.[73] 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.[74] Albeit limited, the evidence indicates a substantial caregiver burden across multiple areas, including physical, social, and psychological domains.[75]

Physically, parents report chronic fatigue and sleep deprivation as a consequence of their caring duties.[76] 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 from occurring; consequently, the pool of friends and extended family willing to provide respite care is diminished.[77] The social burden of care is evident in strained marital relationships, marginalization 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.[70,78] Psychologically, caregiving for those with complex needs is associated with impaired psychological health, emotional health, quality of life, and well-being.[79] Behavioral difficulties provide an additional source of stress.[80]

The ILAE has published a white paper on the medical and social needs of people with epilepsy and ID.[71] 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 highlights the need for more person-centered 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 behavioral disorder.
2 Interagency collaboration is required between disability and epilepsy services—family members should be key stakeholders in this collaboration.
3 Practical supports for families, such as respite and access to information, are urgently required.

Conclusion

Behavior and its manifestations have a pervasive impact on people with 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 behavioral disorder should be provided with multidisciplinary 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.

Acknowledgments

The authors would like to acknowledge the support of Mr. Hirano and his daughter Nanase, of the Japanese Epilepsy Association, who advocate on behalf of people with epilepsy in Japan.

Disclosure of Conflicts of Interest

Dr. Brandt has received personal compensation from Otsuka, Eisai, Desitin, Pfizer, and UCB Pharma for serving on scientific advisory boards, speaking activities, and congress travel and financial support for research activities from UCB Pharma and Otsuka. Dr. Villanueva has participated in advisory boards and pharmaceutical industry-sponsored symposia for Eisai, UCB, Merck Sharp & Dohme, Bial, Pfizer, GlaxoSmithKline (GSK), Esteve, Medtronic, and Cyberonics. All remaining authors have no conflict of interest to disclose. 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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