A Team Approach to Caring for Individuals With Intellectual and Developmental Disabilities and Epilepsy

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Introduction:

Caring for patients with intellectual development disorders and epilepsy (IDD-E), like those with Lennox-Gastaut Syndrome (LGS), is uniquely challenging as patients with IDD-E are more likely to experience treatment-refractory seizures and often suffer from a host of comorbid conditions, including but not limited to attention deficit hyperactivity disorder (ADHD), autism, aggression, self-injurious behavior, learning disabilities, sleep disorders, anxiety, and mood disorders.[1,2] Patients with IDD-E have an elevated risk for status epilepticus, seizure-related injuries, and sudden unexplained death in epilepsy (SUDEP).[3-5] While these difficulties may seem insurmountable, there is indeed hope that these patients' overall condition and quality of life can be improved. In fact, clinical experience suggests that effective epilepsy treatment may alleviate the symptoms of other comorbid conditions, including IDD.[3] Therefore, improving epilepsy management is a critical component to the effective care of patients with IDD-E in order to achieve the best possible outcome.

The unique challenges of caring for patients with IDD-E must be met by caregivers (parents/family members), and a team of medical professionals, which may include an epileptologist, a general neurologist, a neurology nurse practitioner (NP), social workers, psychologist, genetics counselors, therapists, school professionals, and primary care providers (including pediatricians, family physicians, NPs, physician assistants, and nurses). Here, we describe the many challenges to the effective management of epilepsy in this special subgroup of patients, but offer suggestions for a comprehensive approach to medical care and solutions for both specialists and non-specialists alike.

What Defines IDD and IDD-E?

The prevalence of IDD-E increases with the severity of intellectual developmental disorders (IDD).[6] Intellectual disability (previously termed "mental retardation") involves impairments of general mental abilities that impact adaptive functioning in three domains (conceptual, social, and practical) diagnosed before the age of 18 years. The term "developmental disabilities" encompasses a variety of childhood conditions that present before 22 years of age and may persist throughout a person's lifespan.[7] About 1% to 3% of the general population are diagnosed with IDD,[8] and approximately 20% of patients with epilepsy also have IDD.[9]

Seizure types, etiology, and chronicity differ between general epilepsy and IDD-E populations.[1] Some seizure types are easily identified, such as atonic and generalized tonic-clonic seizures.[10] Other seizure types, including atypical absence, tonic, myoclonic, and partial focal seizures, are often subtle and may be more difficult to identify in a patient with IDD-E.[11-13] Tonic seizures are the type that typically persists into adulthood, but is the type most likely to be missed, as they often occur during sleep and can be confused with restlessness, nightmares or missed entirely.[14] Many patients with IDD-E exhibit behavioral disorders, such as ADHD, aggression, anxiety, and mood disorder.[15-17] Furthermore, many patients are non-verbal or have communication impairments.[14,18] Such impediments may complicate accurate seizure identification, which is central to achieving seizure control.

Treatment Challenges of IDD-E

Along with comorbid conditions that persist and often worsen with age,[2] patients with IDD-E are faced with many treatment challenges, including transition from childhood to adulthood medical services, continuity of care by both specialists and non-specialists, and access to managed care over the course of a lifetime. Increased awareness and the creation of management guidelines are helping, but progress is slow and more work is needed.[19]

Comorbidities and Epilepsy Care

Comorbid conditions such as ADHD, autism, and self-injurious behaviors are often reported to be more troublesome than seizures in patients with IDD-E.[1,20] As such, they may dominate the treatment plan, but their extensive care contributes to polypharmacy, adverse events, and increases the risk for drug-drug interactions in patients. Clinical experience suggests that better seizure control may improve comorbid conditions and even decrease the need for concomitant medications.[3]

Many patients with IDD-E have a specific type of severe, intractable epilepsy called Lennox-Gastaut syndrome (LGS). LGS is characterized by multiple intractable seizure types, abnormal electroencephalogram (EEG) with slow spike-and-wave patterns, and cognitive impairment or regression. Peak onset of LGS is between 3 and 5 years of age, but usually the syndrome persists into adulthood. Even if early development was normal, up to half of all patients with LGS will exhibit a delay in intellectual abilities within months following onset. Developmental delays and even regression in skills are cumulative, and within 5 years from onset, up to 96% of all patients with LGS will have significant intellectual or development disorders.[21-23] As patients with LGS age, the profile of the disorder evolves, with seizure types and EEG patterns changing over time.[14] Thus, misdiagnosis early in childhood not only complicates diagnosis in adulthood, but presents a major barrier to proper treatment at each stage of life.

Transition of Care

Because IDD-E and LGS are life-long syndromes, the patient's medical care must transition at least once in their lifetime. As patients age, they often move from a longtime trusted pediatric neurologist to an adult neurologist, from health insurance under their parents' plan to public insurance, and from home

care to assisted residential care. The transition process frequently occurs without much warning and without the necessary preparation, which may be extensive and take a significant amount of time. Under these conditions, obtaining complete medical histories of patients with LGS from one stage of life to another is often difficult because the patient's parents may no longer be available and/or the patient's records are scattered across multiple facilities and providers. Not surprisingly, such transitions are difficult for patients and caregivers and thus may lead to undue distress and gaps in patient care.

Medical Care From Specialists

Pediatric neurologists often know their patients from a young age, are familiar with their medical history, understand their family dynamics, and have seen them through the challenges of various developmental stages. Once the patient transitions to adult care, a new clinical team must learn about the individual, (who often has complex medical issues and an extensive patient medical history,) frequently with inadequate medical documentation and limited support from the patient. At the same time, the adult clinician must work to build rapport with the patient and family, operating at a perceived deficit as compared to the rapport already established with the pediatric clinician.

Medical Care From Non-specialists

In group care settings, primary care providers (PCPs), including family physicians and nurse practitioners, are primarily responsible for managing the comprehensive care of many patients with disparate conditions. Physical therapists, nursing aides, and other daily care providers spend the most time engaged in the day-to-day care of patients and may know the patients best, yet they are not authorized to determine treatment courses for patients and may lack the knowledge to properly identify and communicate seizure types to treating physicians.

Specialists such as an epileptologist or a neurologist are often not on staff in group care settings and are sometimes not readily available for patient treatment or direct, regular communication with primary healthcare staff and caregivers. In urgent but non-emergency situations, expert consultations may be delayed until the patient can be transported to another facility or epilepsy office or until a visiting neurologist will be on-site. Further, it may be difficult to establish doctor-patient relationships or follow-up care under these circumstances.

Funding

With each passing year, funding for patient facilities becomes more restricted, limiting appropriate services for patients with IDD-E. Staff is often overworked with limited or no formal medical training. Treatment options are often limited to those older drugs deemed most affordable, even if these fail to adequately control seizures, resulting in a higher overall economic healthcare burden. Finally, government funds are strictly earmarked for specific use (eg, transportation, on-site care), handicapping administrators from meeting dynamic patient needs.

Treating the "E" in IDD-E

Pharmacological Treatments

Identifying and utilizing the most effective antiepileptic drugs (AEDs) may not only improve outcomes for epilepsy, but also improve the IDD-E patient's overall condition. Clobazam,[24] rufinamide,[25] lamotrigine,[26] and topiramate[27] are AEDs approved for the treatment of seizures associated with LGS. These AEDs have demonstrated efficacy in reducing seizures associated with LGS and should be considered first-line treatment for patients with LGS. Other AEDs including valproate[28] and felbamate[29] may be helpful as add-on therapy to reduce overall seizure frequency; however, they rarely improve drop seizures, which are associated with serious injuries and additional risks. The use of AEDs with mood-elevating properties (eg, lamotrigine) may be particularly beneficial for patients with LGS, independent of seizure control.[26]

Non-pharmacological Treatments

In addition to AEDs, other non-pharmacological treatment options are available including the ketogenic diet, which may reduce seizures in patients with IDD-E.[30,31] However, dietary restrictions can be difficult to implement in the IDD-E and LGS patient populations as many patients have limited food preferences, eat meals in a group home setting where tightly controlled individualized diets may be unrealistic, and have difficulty complying with frequent laboratory testing. The ketogenic diet is possible to implement in patients with gastrostomy tubes,[32] but the need for frequent laboratory testing remains a significant challenge.

Surgical Options

Surgical treatment options include vagus nerve stimulation (VNS,) corpus callosotomy, responsive neurostimulation, and resective surgery. VNS may improve seizure control and elevate mood in patients with IDD-E, including those with LGS.[33] Other surgeries may also be an option in this patient population. Since IDD-E implies a diffuse encephalopathy, surgery may not provide seizure freedom even in patients with a single active seizure focus. More realistic goals with both VNS and other surgeries may be a meaningful reduction in seizure frequency, decreased drug burden, and improved quality of life.[34]

Solutions for a Lifetime of Patient Care

While there is no "one-size fits all" approach to care for this special group of patients, there are many measures that can be taken by healthcare providers and caregivers to reduce the overall burden of disease.

Solutions for the Transition of Care

1. *Start early.* It is recommended that annual discussions to prepare children and families for adulthood begin as early as age 12. This is important both for specialty care[19] and primary care.[35]

2. *Schedule overlapping visits*. Patients should have visits with their adult clinicians before ceasing care from their pediatric team to ease the transition process. Conversations must be ongoing and reevaluated every year to ease the transition and ensure that expectations are met.
3. *Educate parents and caregivers about IDD-E*. Reinforce best practices for seizure identification and documentation, current treatment options, and effective patient advocacy.
4. *Create a transition packet*. A transition packet should include all diagnoses, current medications and ancillary treatments, neuro-imaging (including actual CD-ROM), EEG reports, pertinent lab data, as well as a complete list of previous medication trials and why they failed. Accurate documentation, both before and after the transition, can aid medical professionals in providing the best treatment. Advise caregivers to keep a copy of the transition packet in case of emergencies or change in providers.
5. *Initiate insurance transitions early.* It is important to ensure seamless coverage, without gaps, to optimize patient care.

<u>Solutions for the Epileptologist and Neurologist</u>

1. *Advise patients and caregivers about the possible advantages of newer AEDs*. For some patients, newer AEDs may be equally (if not more) effective, with a better safety profile. The possibility of decreased total drug load and fewer AEs may warrant treatment changes.
2. *Make treatment changes discretely and slowly*. Implementing adjustments in treatment one at a time may allow patients, caregivers, and physicians to more rapidly identify the positive and negative effects of any new approach.
3. *Understand the patient perspective*. While seizure freedom is the ultimate goal for both patients and physicians, other factors play an important role for patients and their caregivers. Be sure to identify how the patient and caregiver view treatment effects.

Any decrease in seizure frequency can lead to meaningful patient improvements, while adverse events like sedation may be too troublesome even in light of fewer seizures.

4. *Take time to set goals*. For long-standing treatment-refractory patients, seizure freedom may not be an attainable goal. An overall focus on improved quality of life can be achieved if there are fewer side effects, even without a reduction in seizure frequency.

<u>Solutions for Primary Care Providers, Physicians, and Nurse Practitioners</u>

1. *Treat epilepsy aggressively*. Effective epilepsy management which minimizes seizures, even if the goal of seizure freedom is not met, may improve overall patient outcomes if the patient is more alert with fewer side effects of medication.

2. *Consider newer AEDs*. Providers need to examine the risks of long term adverse effects, specifically osteopenia and osteoporosis in non-ambulatory patients with older, first generation drugs. Some patients may benefit from newer drugs with better safety profiles.
3. *Reduce polypharmacy*. The goal for all patients should be seizure control, with the fewest medications and at the lowest dosages necessary. This will improve safety and possibly overall quality of life.
4. *Refer to a qualified epileptologist, especially if seizures remain refractory*. Ideally, the epileptologist, they should be working in a comprehensive epilepsy program.

Solutions for Physical Therapists and Daily Care Providers

1. *Become empowered therapists and daily care providers*. If daily care providers can aid in the identification of seizure types, epilepsy management can be tailored to fit the individual patient. This requires teaching on the part of the epilepsy team and provision of seizure diaries.
2. *Document seizures using video on mobile devices*. Although there is concern for patient privacy, the ready availability of recording devices is worth exploring to improve patient care in a cost-effective manner.

Solutions for Payors and Administrators

1. *Embrace telemedicine*. Advances in internet and mobile technology may improve care and patient quality of life, while having the potential to reduce costs and burdens associated with patient care, eg, reaching patients in rural areas, connecting with specialists, and virtual conferencing instead of transporting patient from a group home to a neurology office.[36-38]
2. *Lobby for interchangeable use of funds*. Flexible spending allows facilities to use overages in one area to supplement deficient budgets and provide better overall care, without more money.
3. *Evaluate cost savings holistically.* Newer AEDs may cost more initially, but patients with improved seizure control may see a reduction in overall illness and healthcare costs along with improved quality of life, leading to a lower overall healthcare burden.

Summary

Patients with IDD-E, including those with LGS, are more likely to experience treatment-refractory seizures and comorbid conditions. Improved seizure control via newer AEDs may reduce the burden of comorbid diseases, while allowing for a reduction in overall medications and an improved safety profile. Many suggestions for medical professionals to improve patient care were identified in this paper.

Optimal patient treatment includes: managing epilepsy with the fewest drugs at the lowest dosages and establishing a clear history of diagnosis and past medical history with prior and current treatment plans. Following these suggestions will foster open and clear communication between the transition team of providers and caregivers to create a better environment for patient care for these medically complicated patients.

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