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Transitioning Ambulatory Medicine from Pediatrics to Adult Care for Patients with Epilepsy and Intellectual Disability

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Abstract

Consensus statements and clinical reports exist to guide the transition of youth from pediatric to adult healthcare services. Across the range of youth with no chronic health conditions to those with the most complex disabilities, the standards of practice continue to vary broadly across the country and internationally. Youth and young adults with combined conditions of epilepsy with intellectual disability are a small subset of the total population of young adults who share common needs. These include a system of supports that supplement each person's limitations in autonomy and self-management. Caregivers play significant roles in their lives, whether they are family members or paid direct service providers. Medical decision making and treatment adherence require specific adaptations for patients whose independence due to disability is unlikely. Key issues related to tuberous sclerosis complex, neurofibromatosis, and Rett and Sturge–Weber syndromes will be highlighted.

Keywords
- epilepsy
- intellectual disability
- transition to adult care

Introduction

The study of healthcare transitions that accompany the transfer into adulthood reveals persistent gaps in care and variability between providers, even for patients with minimal medical complexity.[1] Policy statements and clinical reports addressing the transition of youth into adult health care exist in the literature for almost 20 years, and this review will highlight many examples. Yet, across the nation, we continue to seek effective, sustainable interventions that will facilitate meeting the needs of this population including well health visits and longitudinal care.[2,3] Young adults overutilize emergency departments, are more likely to participate in risk behaviors with associated morbidity and mortality, and lack appropriate primary care and scheduled follow-ups for chronic conditions.[2,4] Six core elements to foster the process have been delineated through the Got Transition/Center for Health Care Transition Improvement.[5] These include a formal policy to govern the process, tracking and monitoring of the patients from one system to the other, progressive assessments that inform planning, transfer with well-summarized information and ultimate affirmation of successful integration into adult care.[5] A consensus statement with similar recommendations was endorsed in 2016 by the American Academy of Neurology and Child Neurology Society.[6] Despite these recommendations, adult providers continue to note discomfort with providing care for conditions arising during childhood and, in particular, comorbidities that arise with intellectual disability.[7] While youth with epilepsy and intellectual disability are a small subset of the total population of transitioning youth, their care requires additional targeted actions to facilitate safe, quality care. This review highlights special considerations for

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transitioning patients with intellectual disability with comorbid medically intractable epilepsy and genetic syndromes such as tuberous sclerosis, neurofibromatosis, and Rett syndrome and Sturge–Weber syndrome (SWS).

Young Adults with Epilepsy and Intellectual Disability

Diverse models of transition for pediatric epilepsy patients have been reported that collectively affirm the necessity of directed attention to a standardized process.[3] International programs address the well-summarized past history, coordination between the adult and pediatric providers, and focus on specific themes of young adults, such as psychosocial and sexual health with variable success.[3] Beyond the need for neurology specialty services in youth with epilepsy, there is also a need to secure patient centered medical homes to deliver the longitudinal and preventative measures that oversee the total care which the individual patient needs.[8–10] In patients with intellectual disability, one commonality in the transition process is the necessary role of caregivers in adult life for activities of daily living (ADLs), instrumental activities of daily living (IADLs), and assisting the longitudinal medical team.[11–13] Caregivers may have a role in supporting medical adherence, health habits, and medical decision making.[11,13] The disability advocacy movement encourages clinicians to use guardianship as the last resort, while encouraging the supported voice of the individual through less restrictive alternatives, such as decision-making support contracts and power of attorney where appropriate for the patient's level of functioning.[11,13] An early discussion is necessary to promote the development of decision-making skills in each child and youth, so that the appropriate level of decision-making supports can be instituted at the age of consent.[13] Clinicians must engage in fostering each family's attention developmentally to the patient's ability to progressively make decisions and develop his or her own self-advocacy.[13]

Parental and caregiver stress should be acknowledged when working with this population, as parents report significant responsibility relating to the vulnerability of their child and making informed medical decisions for their child.[14] Recommendations for mental health services may be just as important for caregivers as the patients themselves, to address parental anxiety and fatigue due to their added responsibilities.[14] Clinical teams should assure that governmental and community services are made available to families to lessen the burden through both natural and paid respite and supports.[11,13]

Other considerations in transition should include close attention to antiepileptic drug choice. Antiepileptic choice should be guided by a balance of efficacy, feasibility of administration, and avoiding undesirable long-term effects and teratogenic risk in female patients.[15] Teratogenic risks and anticipatory guidance for maternity care should ideally be addressed by perinatal specialists due to some patients with medically refractory epilepsy requiring use of higher efficacy antiepileptic medications with poorer pregnancy safety profile due to frequent generalized tonic clonic seizures.[15]

Other barriers exist for young adults with epilepsy and intellectual disability. Sexual health and childbearing decisions in these patients warrant access to education services, clinicians with specialized skills, and willingness to explore the medical ethics and consequences of choices.[16] Mental illness occurs at increased frequency in those with intellectual disability and access to mental health services is significantly more challenging when compared with the general population.[17–21] Families must often travel to specialized clinics to receive needed mental health and behavioral therapy services, and transportation for patients with refractory epilepsy also can be a challenge.[22,23]

Tuberous Sclerosis Complex

Tuberous sclerosis complex (TSC) is an autosomal dominantly inherited syndrome from mutations in hamartin on chromosome 9q34 resulting in TSC1 or tuberin on chromosome 16p13.3 resulting in TSC2.[17,24] Activating growth factors for cell proliferation in many organ systems occur in both cases.[17,24] These patients require multidisciplinary care due to widespread disease of the skin, eyes, kidneys, heart, liver, and brain.[17,21,24] Within the brain, cortical dysplasias, white matter abnormalities, subependymal nodules, and subependymal giant cell astrocytomas are commonly found.[21,24] Accompanying comorbidities include intellectual disability, focal with secondarily generalized epilepsy, and neuropsychiatric diagnoses.[17,21,24]

Of TSC patients who develop epilepsy, the majority begin under age 3, and those with infantile spasms are likely to remain medically intractable or develop Lennox–Gastaut syndrome, requiring longitudinal medical, dietary, and surgical therapeutics.[19,24] It is particularly critical to assure continuous care from pediatric to adult services to reduce risk of sudden unexplained death in epilepsy or other injuries related to atonic seizures or status epilepticus.[24–27] Seizure safety should be discussed early with families of children with epilepsy, including the International League Against Epilepsy guidelines for injury prevention through restriction of higher risk activities.[23,27] Patients with tuberous sclerosis with infantile spasms often have worse developmental outcomes, even with remission of their spasms with vigabatrin, adrenocorticotropic hormone, prednisolone, or other therapies.[24] Despite worse neurocognitive outcomes and refractory epilepsy, they often survive to adulthood, necessitating transition to adult providers.[6,26,28,29] If medically intractable focal epilepsy is present in TSC patients, many patients have a dominant cortical tuber amenable to resection or ablation, and, depending on the Engle surgical result, cognitive outcomes and long-term prognosis may be improved.[30] Neuromodulatory management with vagus nerve stimulation, deep brain stimulation, or reactive neurostimulation may be pursued in patients who may not receive resection or ablation, although the comfort of adult neurologists may limit feasible access to this type of care.[7] Various pharmacotherapies for the reduction in epileptic seizures are available as well, though longitudinal care must be taken to minimize deleterious side effects.[15] Special consideration must be taken to avoid risks of neural tube defects in

female patients requiring teratogenic antiepileptics with initiation of folate and counseling against unplanned pregnancies on these medications.[15]

Non-neurologic multiorgan manifestations of TSC also must be identified and appropriate referrals made for cardiac rhabdomyoma, renal angiomyolipoma and chronic kidney disease, pulmonary lymphangioleiomyomatosis, retinal disease, and dental enamel pits.[24,31] Coordinating the redistribution of multidisciplinary pediatric care efficiently to adult subspecialty services may be challenging.[24,31] At our center, a model for transition for children with special healthcare needs was implemented using a transdisciplinary team to create medical summaries, and facilitate initiation of needed services and direct communication to new primary care and subspecialty clinicians.[32]

Neurofibromatosis

Neurofibromatosis 1 (NF1) is an autosomal dominantly inherited syndrome associated with mutations in neurofibrin 1 on chromosome 17q11.2.[18,33] About half of these mutations are de novo, while the other half present with the added challenge to the pediatric clinician of providing guidance for parents who may also need care for their own condition as well.[34] NF1 becomes evident in childhood with multiple neurocutaneous phenomena including café au lait spots, freckling in the axilla and inguinal regions, Lisch nodules of the eyes, and neurofibromas.[18,33] More importantly, patients with NF1 also need surveillance for optic gliomas, breast cancers, pheochromocytomas, endocrine tumors, rhabdomyosarcomas, glioblastoma multiforme, and malignant peripheral sheath tumors that develop from plexiform neuromas.[18,33] Moyamoya vasculopathy, aneurysms, cardiac abnormalities, or growing malignant tumors may present clinically as transient ischemic attacks or recurrent strokes.[18] While screening and treatment of malignancies are an emphasis in this condition, patients may also develop refractory hypertension due to renal artery abnormalities, bone dysplasia and scoliosis, and neuropsychiatric illnesses such as depression, attention deficit hyperactivity disorder, and cognitive impairment.[18] Focus groups that analyzed the needs and concerns of parents and patients with NF1 highlighted the lack of access and challenges in identifying providers and coordinating transition, issues of mental health care, public visibility of the neurocutaneous condition, and limited independence and socioeconomic concerns.[33]

Neurofibromatosis 2 (NF2) is an autosomal dominantly inherited syndrome associated with mutations in the merlin/schwannomin gene on chromosome 22q12.[35] The severe childhood onset form known as Wishart phenotype presents commonly with vision changes, peripheral neuropathy, myelopathy, headaches, epilepsy, and rapid hearing loss with a larger tumor burden.[35,36] The adult onset type in the 20s known as Gardner phenotype demonstrates slower hearing loss and fewer tumors due to the presence of more functional merlin/schwannomin gene product.[35] Central nervous system tumors classically include acoustic neuromas, multiple meningiomas, and spinal cord glial tumors.[35–37] These patients have fewer cutaneous signs of disease than NF1 patients.[35–37] Surveillance should include imaging studies, audiology testing, ophthalmologic exams, and neurosurgical as well as oncologic co-management. Early intervention along with support from a multidisciplinary medical home may be helpful for maintaining quality of life, though studies generally demonstrate decline of quality of life overall due to the evolution and progression of disease from childhood to adulthood.[38]

Patients with NF1 and patients with NF2 with the Wishart form both have early onset multiorgan system disease requiring multiple providers for adequate care that require complex support in the transition to adulthood.[32]

Rett Syndrome

Rett syndrome is a common cause of X-linked dominant intellectual disability in females and is lethal in males without mosaicism, most often due to mutations in the methyl CpG binding protein 2 (MECP2) gene on Xq28.[39,40] Rett syndrome is classically associated with normal development until early regression of motor and speech skills, followed by the emergence of hand wringing movements.[39] Secondary microcephaly is observed within 10 years of life.[39–41] While there are several causes of atypical Rett syndrome, there is a specific mutation in the cyclin-dependent kinase-like 5 gene that has a similar phenotype to MECP2 patients but with increased incidence of infantile spasms.[42] In general, after a period of regression, patients with Rett syndrome stabilize though often have intellectual disability and persistent motor disability.[43]

In children with Rett syndrome who do walk, gait is characterized by ataxia and spasticity, resulting in slower initiation and kinetics of walking when compared with age matched controls.[44] During the regression stage of Rett syndrome, the majority of patients lose the ability to walk as well as verbal and nonverbal communication loss.[44,45] More longitudinally, decline in mood and interest is seen in cross-sectional studies of a wide age range of patients.[43] Issues with weight loss, physical disability, poor bone health due to immobility, and respiratory complications add to the need for multidisciplinary care teams in adult life.[39,41]

Breath holding spells and stereotypies are both nonepileptic events associated with classic and atypical Rett syndrome that can make the diagnosis and management of comorbid epilepsy challenging.[41] Additional seizure mimics include mechanical falls leading to fractures, dislocations, or head injuries that may occur due to impaired mobility. Trauma from altered consciousness and loss of motor control are an important cause of morbidity in patients with epilepsy in general.[27] A high suspicion for seizure should be considered in the evaluation of unwitnessed falls in patients with Rett syndrome. Once the first unwitnessed fall is reported, precautions to prevent injury even prior to a definitive diagnosis of epilepsy are indicated.[23]

The regressive nature of this disease can add a notable additive factor in the transition to adult care, though many patients stabilize around the time of transition requiring

assistance with ambulation and communication.[39,43] Family and hired caregivers often are required through adult life.[11,12]

Sturge–Weber Syndrome

SWS is a neurocutaneous disorder associated with capillary malformations of the skin, typically in areas innervated by the superior branch of the trigeminal nerve, and leptomeningeal capillary malformations caused by de novo somatic mutations in the guanine nucleotide-binding protein alpha-Q (GNAQ) on chromosome 9q21.[46] There is often focal epilepsy with atrophy and calcification of the ipsilateral cortex most commonly underlying the cutaneous lesion, headaches, stroke-like episodes, and focal weakness that may progress over time.[29,46] The location of lesions leads to heterogeneity of symptoms at presentation and variability of progression and outcomes depending on the extent and location of the intracranial disease.[47] Over half of children with SWS will develop epilepsy before 2 years old.[48] If early epilepsy occurs, intellectual disability, behavioral problems, and learning disabilities are more likely to co-occur.[48,49] Intellectual or learning disabilities, as well as attention deficit disorder, occur commonly. Glaucoma can originate in childhood or adulthood and choroidal hemangioma may also occur, necessitating lifelong ophthalmology surveillance.[29,47]

Due to the de novo somatic mutation of SWS, neither screening of parents nor anticipation of recurrence in the offspring is expected.[46] Patients with SWS typically live into adulthood, necessitating transition services.[29] In a cross-sectional survey, about one-third of patients with SWS are financially independent in adulthood and about half will marry, though the majority will likely require some educational supports due to learning disabilities as well as assistance with adaptive limitations and supported decision making from caregivers.[11,13,47]

Conclusion

Patients with syndromes in which epilepsy is accompanied by intellectual disability often have multiorgan system comorbidities, physical disability requiring assistance with ADLs, and mental health concerns. They typically require assessment of their decision-making abilities to determine capacity for self-advocacy and/or necessary supports from caregivers. The identification of a multidisciplinary care team to meet the specific needs of each patient requires attention to the core elements of transition. A policy governing the process should be created. Patients should be tracked through their transition process and attention should be given to effective transfer of information between the sending and receiving clinicians. Effective transitions of care involve identification of a team of adult providers who are comfortable with three aspects of caring for a medically complex patient: providing surveillance for anticipated comorbidities, exploring both patient and caregiver concerns, and addressing supported decision making while maximizing participation and consent or assent from the patients themselves.

Conflict of Interest
None declared.

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