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Short communication

Transitioning pediatric patients receiving ketogenic diets for epilepsy into adulthood

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ABSTRACT

Purpose: To examine the process for transitioning adolescents on dietary management for intractable epilepsy to adult neurologic care.
Methods: Ten patients, ages 6–16 years when the ketogenic or modified Atkins diet was initiated, were identified; age at transition was 18–43 years. All patients were seen at Johns Hopkins Hospital, Baltimore, Maryland.
Results: Seven remain on diets (4 Ketogenic, 3 modified Atkins diet) at this time with mean diet duration of 15.5 years (range 4–32 years). Most patients had excellent but not complete seizure control and several had recurrence or worsening of seizures with attempts to wean dietary therapy, hence the need to continue. Three main transition options were utilized: (1) continue being followed in pediatric clinic ($n = 2$), (2) establish care with an adult neurologist ($n = 2$), or 3) receive care from the Adult Epilepsy Diet Center ($n = 6$). Those patients followed in the Adult Epilepsy Diet Center were slightly more likely to remain on dietary therapy following transition (5/6 vs. 1/4, $p = 0.12$).
Conclusion: It is important for adolescents with epilepsy receiving ketogenic diets to have transition plans in place for when they become adults. Adult epilepsy diet centers are the ideal option when possible.

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1. Introduction

Adolescents with chronic diseases require detailed transition plans from pediatric to adult medical care, typically around 18 years of age.[1,2] For patients with epilepsy, the complex issues regarding driving, pregnancy, employment, and independence are significant and current recommendations are for pediatricians to begin discussions regarding transitioning to adult clinics as early as age 12 years.[3–5] The American Academy of Pediatrics has recently published formal recommendations for transitioning patients with chronic conditions into adult clinics.[1]

The ketogenic diet (KD) is a high fat, low carbohydrate dietary therapy for children with medication-refractory epilepsy.[6] Most children remain on the KD for up to 2 years, at which point it is weaned to discontinuation. However, some will have recurrence or worsening of seizures with weaning of the KD, requiring long-term dietary management.[7] To the best of our knowledge, there have not been any recommendations for how best to manage patients who continue to use the KD as they reach adulthood.

The modified Atkins diet (MAD) was first described in 2003 as a less restrictive version of the KD.[8] Patients do not calculate protein, calories, or fluids and restrict carbohydrate intake to 10 g per day while eating significant amounts of fat. The MAD has been investigated for use in adults since its creation, primarily due to its inherent flexibility as compared to the KD.[8–10] In August 2010, Johns Hopkins Hospital opened an Adult Epilepsy Diet Center (AEDC) with an adult epileptologist (MCC) and dietitian (BJH) primarily responsible for dietary management of patients and mostly using the MAD de novo with support, guidance, and training from a pediatric epileptologist with expertise in dietary therapies (EK).

This case series was designed to investigate epilepsy treatment outcomes in patients who started either the KD or MAD as children at Johns Hopkins and remained on diet therapy while transitioning to adulthood. We hypothesized that our AEDC provided a useful option for those patients requiring long-term, adult dietary management.

Abbreviations: KD, ketogenic diet; MAD, modified Atkins diet; AEDC, adult epilepsy diet center.

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Table 1
Patients who reached adulthood still receiving dietary therapy.

Patient	Gender	Etiology	Age at diet (years)	Age at transition (years)	Current age (years)	Current diet (ratio)	Transition option	Seizure control over baseline at transition (%)
1	Female	Mitochondrial complex 1, 3	6	18	20	KD (4:1)	AEDC	90
2	Female	JME	16	18	20	MAD	AEDC	100
3	Male	Mitochondrial complex 3	7	20	21	MAD	AEDC	50
4	Male	LGS	10	20	25	Off	Adult neurologist	50
5	Male	HIE	14	21	23	Off	AEDC	50
6	Male	HIE	6	23	25	Off	Adult neurologist	50
7	Male	Tuberous sclerosis	6	26	33	KD (3:1)	AEDC	99
8	Male	LGS	12	43	44	KD (4:1)	AEDC	100
9	Female	HIE	11	N/A (not transitioned)	26	KD (4:1)	Continue with pediatrics	90
10	Female	None (idiopathic partial)	15	N/A (not transitioned)	30	Off	Continue with pediatrics	50

2. Methods

Medical records since 1994 were reviewed for all adults with epilepsy treated with the KD or MAD at Johns Hopkins Hospital who were actively receiving dietary therapy at the time they turned 18 years of age. All adult patients were seen in either the pediatric ketogenic diet center, AEDC, or with a different adult neurologist for their clinic visits. Patients treated with the KD as children were part of an ongoing study of the effectiveness of the ketogenic diet, one patient participated in a study of the MAD,[11] and all adults seen in the AEDC consented to be part of an ongoing study of dietary therapy for adults. Categorical data were analyzed using a Fisher's exact test ($P = 0.05$). All studies were approved by the Johns Hopkins Committee on Clinical Investigation.

3. Results

Ten patients were identified, current ages 20–44 years (Table 1). All started dietary therapy as children or adolescents (mean age 10.3 years, range 6–16 years), with all but one (Patient 2) initially starting with the classic KD. Etiologies varied and included hypoxic ischemic encephalopathy (3), Lennox Gastaut Syndrome (2), mitochondrial disorders (2), juvenile myoclonic epilepsy (1), tuberous sclerosis complex (1), and idiopathic partial epilepsy (1).

Patients in this study had somewhat prolonged use of dietary management; mean 15.5 years (range, 4–32 years). There were multiple reasons why these adults continued dietary treatment of their epilepsy. Eight achieved good seizure control with the KD (50–99% seizure reduction compared to before the KD was started). Two patients (Patients 2 and 8) were seizure-free for years after several anticonvulsants failed to control their seizures. At one time during treatment, four had either attempted to reduce the KD ratio, completely wean the KD, or exceed the recommended carbohydrate intake on the MAD with resultant immediate worsening of seizures.

Eight patients had moderate to severe developmental delay and were dependent on parent or sibling caregivers for food preparation. All of these care providers reported no concerns about compliance or difficulty administering the KD or MAD. Two patients were of normal intelligence or had mild developmental delay (Patients 2 and 10). Two patients (Patients 1 and 5) received liquid ketogenic diet formulas through gastrostomy tubes and therefore compliance was not an issue. This is similar to the 20% of children in our pediatric ketogenic diet center on formula only at this time.

Eight adolescents were transitioned to adult clinics. The mean age at transfer to adult clinic was 23.6 years, with the two oldest presenting to AEDC at ages 26 and 43 years following several years of self-management.[12] Four adults were switched from the KD to the MAD for convenience, without any resultant seizure increase. All patients remain on anticonvulsants (mean 2.4 anticonvulsants; range, 1–4), most commonly levetiracetam (7) and clonazepam (3).

Three major categories of transition options were identified as used by young adults with epilepsy on dietary therapy. First, the most common, was a transition to the Johns Hopkins AEDC. Six adults were seen in this clinic; three had sporadic care by private dietitians or self-management then chose to be followed in the AEDC for more formal management, two had been followed for 7 and 8 years in the pediatric ketogenic diet center at Johns Hopkins and were then referred, and one was an 18 year old with juvenile myoclonic epilepsy enrolled in a MAD study since age 16 years.[11]

The second option identified was to transfer care to an adult neurologist who was not a member of a ketogenic diet team. This was utilized by two patients (Patients 4 and 6), and both discontinued diets (KD and MAD respectively) within two years of the transfer of care. Both were developmentally disabled individuals with parents who continued the exact same dietary regimen without dietitian involvement, then gradually discontinued it without complications.

The third option was to continue care through a pediatric ketogenic diet center. Two adults had local adult neurologists available for emergencies and returned to Baltimore every 2–3 years for care. One patient's mother (Patient 10) emailed one year ago indicating she had discontinued her daughter's KD after 14 years total duration, without worsening seizures. Another (Patient 9) continues to return for care for now.

At this time, six remain on dietary treatment (4 on KD, 2 on MAD). A greater percentage of patients who transitioned to the AEDC instead of selecting another alternative remain on dietary therapy at the current time, although this difference did not reach statistical significance because of the relatively small sample size (5 of 6 (83%) vs. 1 of 4 (25%), $P = 0.12$). The five adults still followed in our AEDC are all local to Maryland and surrounding states.

4. Discussion

As adolescents with intractable epilepsy approach adulthood, it is critical to have a thorough plan in place for transition of their medical care.[3–5] For the majority of adults with epilepsy, anticonvulsant management is not vastly different between pediatric and adult providers. However, dietary therapy is potentially more complicated to continue as an adult because it requires a trained adult neurologist familiar with either the KD or MAD, and an adult dietitian as well.[9] Although pediatric ketogenic diet teams can continue to provide care, intermittent hospitalizations to adult units and issues such as pregnancy, living independently, and different nutritional requirements make this potentially problematic for adults. At Johns Hopkins Hospital, although no upper age limit requirement for transition exists; most patients are transitioned to adult providers by age 21 years.

In this limited case series, we identified several possible options for continuing to provide appropriate care of pediatric patients receiving dietary epilepsy management when they reach adulthood. Individual hospitals and pediatric neurology clinics can make their own personal decisions on how best to handle transitioning these patients, however, we believe that having an AEDC is ideal for several reasons. The AEDC team was interested in seeing new patients, motivated to keep them on dietary therapy, and in frequent communication with both the referring internists and pediatric ketogenic diet teams. We have found that having the AEDC in the same location as the pediatric ketogenic diet center allows for close communication between neurologists and a smoother transition process. Additionally, for the adults who were being "self-managed", the AEDC became a medical "home" for managing potential side effects, providing menu variety, and communicating with local primary care providers that had not previously existed.

Future directions include finding ways to improve the transition process even further. Several studies have proposed the concept of a "transition clinic" with both pediatric and adult neurologists present during a clinic visit.[4,5] This strategy was successfully utilized during several clinics at the AEDC, and could be implemented as well as having both pediatric and adult dietitians together to discuss the diet transition plan. Most of these 10 patients were local to Maryland, so there may be some value in providing dietary management services via email or video teleconferencing as another transition plan, with a local adult neurologist in charge of the majority of care.

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