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Transition and transfer from pediatric to adult health care in epilepsy: A families' survey on Dravet syndrome

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ABSTRACT

We used a questionnaire to ascertain the perception of transition and transfer from pediatric to adult health-care system in patients with Dravet syndrome and their families. Sixty families received the questionnaire. We had a response rate of 85%. Sixty-one percent of patients experienced a transfer. Factors that positively impacted transfer were the quality of transition preparation ($p < .000001$), a longer duration of follow-up by the same child neurologist ($p < .001$), the availability of the child neurology staff ($p < .01$), a transfer into the adult health-care system after the age of 18 ($p < .01$), and a stable medical condition before transfer ($p < .05$). All families reported a positive experience in the pediatric health-care system. Child neurologists were considered as welcoming, available, and helpful. Their experience in the adult health-care system was similar to pediatric care. Almost all patients who experienced "transfer" reported no gap in this process.

1. Introduction

In 2001, van Dyck et al. estimated that 12.8% of children in the US had specific health-care needs resulting from chronic illness [1], and more than 95% of children born with chronic illness survive beyond the age of 20 [2,3]. For instance, in 1974, the median age at death for patients with cystic fibrosis was 8 years [3], whereas in 2010, the median predicted age of survival had risen to 38.3 years [4]. "Many illnesses once considered to be confined to childhood must now be thought of as diseases that begin in childhood but continue into adult life" [5]. These advances have encouraged many national pediatric societies including those from Canada [6], the USA [7], and the UK [8] to make recommendations to optimize the transition to the adult health-care system for children with chronic diseases.

By transition, we mean a process beginning in childhood and aiming to prepare older youth with chronic illness and their families for "adult" health care. Transfer is the formal handing over of care from the pediatric to the adult health-care system. To better understand this transition and transfer process, three perspectives have to be considered: those of the providers (physicians), the patient, and the family. Pediatric providers have identified many barriers [9,10] to optimal transfer into the adult health-care system: their own lack of time, the small number of available adult providers (family physicians, internal medicine physicians, specialists) who can handle adolescents with specific health-care needs, the lack of multidisciplinary care in the adult health-care system, and the financial impact on the transfer for the patient and his family. The personal experience of the pediatric providers may also affect the transition and the transfer as well as their potential prejudices. Perhaps one of the most important obstacles is the reciprocal attachment between pediatricians and their patients and their families. For instance, a study questioning child neurologists and pediatric caregivers about transition in Lennox–Gastaut syndrome, a severe form of epilepsy, emphasized that "an adult practitioner would take less time with the patient and family" or that "the adult providers would not be familiar with the patients' medical condition" [9]. The same provider's concerns have been reported in the literature in other childhood chronic diseases [10,11], with two medical conditions playing a major role: cognitive ability and progressive disability with specific health needs [11].

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The patients' and families' points of view have been evaluated in some chronic diseases such as cystic fibrosis, diabetes mellitus, and inflammatory and sickle cell diseases. However, few data are available regarding the patients who suffer from epilepsy especially when cognition is impaired, which is frequent in severe childhood epilepsies persisting into adulthood. The patients' and families' views are important to emphasize. The single study reporting concerns of the families of children with chronic diseases associated with developmental disabilities showed a discrepancy between the pediatric providers' and the families' priorities [10].

In order to better explore patients' and families' opinions about transition and transfer in epilepsies with cognitive impairment, we proposed a structured questionnaire for a cohort of patients with Dravet syndrome and their families. Dravet syndrome (DS) is a severe epileptic encephalopathy with both pharmacoresistant epilepsy and cognitive delay. It is characterized by the onset of seizures before the first year of life, often triggered by fever. After the second year, various types of seizures such as atypical absences, focal seizures, generalized seizures, and segmental or massive myoclonus also occur. Epilepsy might remain active in adulthood with fever sensitivity and often necessitate AED polytherapy. Patients are cognitively impaired with more than half having an IQ under 50 [12].

We aimed first to describe patients' and families' opinions on pediatric care in comparison with care in the adult health-care system and their opinion on the transfer and then identify the factors that improved the transfer in this cohort.

2. Methods

2.1. Survey development

The data analyzed in this report are part of a larger questionnaire developed by a group of experts (reported in the Acknowledgments) under the aegis of the Wyeth Foundation based on the available literature, with input from the IPSOS Institute, France (an institute that specializes in survey management and data collection). The overall questionnaire was originally developed to study the long-term outcome of various chronic pediatric diseases such as diabetes mellitus, renal transplantation, epilepsy, type 1 neurofibromatosis, juvenile arthritis, and the effects of radiotherapy. Specific questions were adapted to each disease in addition to a common structure. The epilepsy questionnaire consisted of 198 close-ended or semi-open-ended questions which evaluated medical, educational, socioprofessional, and sexual outcomes. Part of the questionnaire evaluated transition and was divided into three categories: pediatric follow-up, transition/transfer to the adult health-care system, and the first two years of adult health-care follow-up. The transition/transfer part of the survey consists of 95 questions. Many questions used a Likert method with responses in 4 or 5 categories with 1 meaning “very good” and 4 or 5 meaning “very bad”.

2.2. Participants

We sent the questionnaire to the parents of all patients with a diagnosis of Dravet syndrome who had had at least one outpatient visit at our institution during childhood and were aged over 18 years on January 1st 2011. Patients were identified from our department database.

2.3. System of care for patients with rare and severe epilepsies

Pediatric health care and adult health care in France are free for all patients with severe epilepsies and cognitive disabilities. These patients are usually followed at pediatric neurology and epilepsy units into adulthood in university hospitals. Families and patients have, in addition to health care, easy access to psychologists, neuropsychologists, and social workers. The improvement of transition and transfer was one of the major aims of the referral center for rare epilepsies. Since its creation in 2005, this center allowed the identification of adult neurologists' partners and contacts.

2.4. Statistical analysis

After verification of validity conditions, especially Levene's quality test of variances, we performed statistical comparisons using Student *t*-test and Fisher's exact test to evaluate binary questions.

We compared the following:

- factors related to good versus unsatisfactory transfer;
- responses to a series of questions concerning the perception of pediatric versus adult follow-up;
- characteristics of patients who achieved a transfer to adult care versus those who remained in pediatric follow-up.

This study was approved by the ethics committee of our institution, Necker Hospital, APHP. All participants or their legal guardians signed an informed consent to be included in the data analysis. For statistical analysis, $p < 0.05$ was considered as statistically significant, but in the case of multiple comparisons, the Bonferroni method was used, and a new level of significance was established according to $p = 0.05 / n$, where n is the number of repeated comparisons. Results are expressed as the average $\pm$ standard deviation. In the event of missing data, percentages were calculated per number of responses obtained, item by item.

3. Results

3.1. Global characteristics of the responses

Responses were obtained from 53 of 60 mailed questionnaires to patients with Dravet syndrome (Fig. 1). Two responses were excluded because of the absence of signed consent, giving a total response rate of 85% (51 patients). The Cronbach's alpha coefficient [13], which is commonly used as a measure of reliability by studying the stability of the responses to the same question asked in two instances in the questionnaire, was 0.99 for the age of the first consultation with the neurologist, 0.93 for the knowledge of the antecedents by the neurologist, and 0.73 for the estimation of the availability of the adulthood care team ($\alpha \geq 0.9$: excellent internal consistency, $0.8 > \alpha \geq 0.7$: acceptable consistency).

All families reported that patients received help in completing the questionnaire because they were not able to read, write, or, in many instances, understand the questions.

The age of the patients ($n = 51$) ranged from 22 to 59 years with an average of 29 ± 6.5 years. All patients were residents of France. Ninety percent stopped their classical school education before the end of kindergarten (at the age of 6 years) and were followed in special education centers since the age of 6 years. Ten percent followed specialized primary school classes and were oriented to the special education system by the age of 10 years at most. Thirty-six patients (71%) still had active epilepsy (at least one seizure in the last 12 months preceding the survey) with an average of 73 ± 33 seizures per year. Almost all (98%) were still treated with polytherapy, receiving a mean of 3.3 ± 0.93 antiepileptic drugs. Ninety percent (46/51) of questionnaires had the transfer part completed and could be further analyzed. Thirty-one (61%) experienced transfer to the adult health-care system and 20 (39%) did not (Fig. 1).

3.2. Patients without any transfer experience ($n = 20$)

In this group, four were treated since the diagnosis of DS by adult neurologists, eight were still followed by a child neurologist after the age of 18, three had a double follow-up with an adult and a child