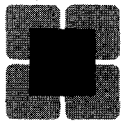

PANEL DISCUSSION

FRITZ E. DREIFUSS, MD, MODERATOR; JEAN AICARDI, MD; RICHARD HRACHOVY, MD; DIETER JANZ, MD; PIERRE LOISEAU, MD

Dr. Dreifuss: We heard this morning about two well-defined syndromes of so-called idiopathic epilepsy, which carry a very benign outlook, and about two syndromes, which are referred to as either cryptogenic or symptomatic epilepsy, where the outlook is less certain and where the response to therapy is probably less favorable.

It is extremely important, I think, to attempt to define these syndromes in order to be able to predict, when one first sees a patient, whether the outcome will be sufficiently favorable to avoid the use of antiepileptic drugs altogether or, if one has to use them, whether one can predict a termination point for such treatment, or whether the syndrome is, like the Janz syndrome, one in which continuous long-term medication is going to be necessary. This is one of the practical uses of being able to classify patients into these various syndromes. Dr. Janz has given us a further look at what I think the future holds for identification of the individual syndromes: that is, a discussion of what is ultimately going to prove to be the *fons et origo*—namely the gene itself. Juvenile myoclonic epilepsy is perhaps offering us the very first glimpse into the nirvana of being able to identify the underlying cause of some of the problems that are at the present time referred to as the epilepsies.

Question: I want to ask Dr. Aicardi one question. We all have the same experience of that particular group of children who start with what appear to be simple febrile convulsions and develop myoclonic encephalopathy. I am asking if he has any way to tell us how to predict which of these children, after beginning with perhaps two or three mild convulsions, and maybe one prolonged convulsion, go on 6 months later to develop this more or less malignant form of epilepsy?

Dr. Aicardi: I think there is a way. The fact that seizures very often start before the age of 8 or 9 months and then are very frequently unilateral and long-lasting (15 to 30 or more minutes), the fact that the seizures repeat themselves within a short time in the same way (usually within 1 or 2 months), and occasionally the finding of photosensitivity on the EEG—all these signs are not necessarily predictive of severe myoclonic epilepsy but rather they indicate the likely development of a severe epilepsy, which may or may not be myoclonic. They constitute fairly good prognostic indicators rather than definite indications of the type of epilepsy.

Question: In the treatment of infantile spasms, what are the clinical or EEG criteria of a favorable response?

Dr. Hrachovy: We use both clinical and EEG criteria. The clinical criterion is cessation of spasms. We also like to see improvement in the EEG; that is, disappearance of the hypsarrhythmic pattern. We use both of those criteria as far as the acute effects of hormonal therapy are concerned.

Question: What is the differentiation between juvenile myoclonic epilepsy and impulsive petit mal?

Dr. Dreifuss: They are the same condition, just a different terminology.

Question: What is the long-term outcome of infantile spasms treated with nitrazepam?

Dr. Dreifuss: Nitrazepam was used in one of our studies in a comparison with ACTH. The immediate outcome was almost identical. The long-term outcome has not yet been determined for the group treated with nitrazepam. It would appear that the long-term outlook is not as favorable as for ACTH, but this series of patients included a very large number of symptomatic

infantile spasm patients for whom the natural outcome is bad.

Question: Where does the myoclonic astatic epilepsy of Doose fit in the category of Aicardi?

Dr. Aicardi: It does not fit at all, for the reason that Doose and I use totally different criteria. The criteria used by Doose, as I understand them, are mainly etiologic. Among the group of patients with brief myoclonic or astatic seizures, he separates cases of Lennox-Gastaut syndrome, those who display signs of brain damage of whatever type, and the group dominated by genetic factors, which he calls myoclonic astatic epilepsy.

Basically, the division made by Doose is on etiologic grounds, whereas what I try to do is to divide the patients on clinical and electroencephalographic grounds. That, of course, leads to something that overlaps with Doose's classification, but does not coincide in any way, because the criteria used are different.

Question: At what point do you distinguish brief tonic from myoclonic seizures?

Dr. Aicardi: This is difficult to tell. Clinically, and from the electromyographic point of view, myoclonia, either isolated or repeated, is quite different from a tonic seizure, which is a continuous contraction. Myoclonus is essentially a very brief muscle contraction that may remain isolated or recur in a saccadic manner.

Question: Which drugs would you use for myoclonic epilepsy?

Dr. Aicardi: This is a vast topic and a very difficult problem. Few of the myoclonic epilepsies respond well to any form of treatment. For such patients, the two drugs which can be useful—but are not necessarily so—are sodium valproate, especially for Group I cases, and the benzodiazepines, although patients often develop tolerance for the latter after a short time. These, alone or in combination, are the two most useful drugs in my experience.

I have very little experience with the ketogenic diet, which is used by many people. Apparently in France it is quite difficult to get patients to eat that sort of thing.

In some cases, either of Lennox-Gastaut syndrome or of severe myoclonic epilepsy, I also use steroids temporarily in the hope of tiding the patient over a difficult period, but not on a permanent basis. Usually, when they work, they do so only for a brief period of time, and very frequently they do not work at all.

Question: Do you have more data about nocturnal seizures in benign childhood epilepsy with centrotemporal spikes?

Dr. Loiseau: Ten or 20 seizures have been correctly observed from an electroclinical point of view. The ictal EEG does not always demonstrate a focal onset. It may show, especially when there are very frequent seizures, bilateral asymmetrical slow spike waves. But very few seizures have been directly observed during the sleep of these children.

Question: Does one ever see independent centrotemporal spikes in association with benign rolandic epilepsy?

Dr. Loiseau: Independent centrotemporal foci are observed in only 30% of cases.

Question: What are the necessary and sufficient criteria for benign childhood epilepsy with centrotemporal spikes?

Dr. Loiseau: It is necessary to have clinical proof and EEG proof. We cannot make this diagnosis only on clinical findings *or* electroencephalographic findings. Both are necessary and sufficient.

Questions: Can clonazepam be used in JME, especially prior to recurrence of tonic-clonic seizures? What is the treatment and prognosis of nonresponders on valproate?

Dr. Janz: One investigator has reported very good results with clonazepam in patients with JME. I do not know for how long. That is always a question with benzodiazepines; they do not work very long. The drug of choice for nonresponders on valproate, however, is always primidone, which is also effective in JME.

Question: What is the difference between pyknolepsy and juvenile absence?

Dr. Janz: A very pragmatic answer is that daily occurring absences are called pyknoleptic and nondaily are called nonpyknoleptic.

Question: Can you define awakening grand mal?

Dr. Janz: In awakening grand mal, the patient's attack usually occurs between minutes and one or two hours after waking. There is no certain definition, but in practice it is not very difficult to define.

Question: Do you think that photosensitivity is a separate disorder from JME and other generalized epilepsy?

Dr. Janz: It is a very important question. Photosensitive epilepsy is not a special clinical syndrome, because if you divide patients with epilepsy into those who are photosensitive and those who are not, then you will find all types of generalized epilepsy in these cases. Photosensitivity is a genetically determined pathogenetic condition, which contributes to the manifestation of generalized epilepsy.