extremities, inappropriate laughter, and seizures. Seizures are
generalized in 90% of patients. Deafness has been reported previously,
but the abnormal auditory brain stem responses recorded in this study
appear to be unique.

Eight sporadic cases of Angelman syndrome associated with chromo-
some 15q12 deletion are reported from the Central Hospital, and Kobato
Gakuen, Aichi Prefectural Colony, Japan (Matsumoto A et al. Epilepsia
1992; 33: 1083). Angelman syndrome is included in the etiology of West,
Lennox-Gastaut and other infantile-onset epileptic syndromes.

KETOGENIC DIET FOR INTRACTABLE SEIZURES

The efficacy of the ketogenic diet in 58 children with multiple seizure
types resistant to antiepileptic drugs has been evaluated at the Johns Hopkins
University School of Medicine, Baltimore. All patients had severe neurologic
handicaps: mental retardation (84%), cerebral palsy (45%), and microcephaly
(15%). Mean age at diet initiation was 60 months (range 12-235 months). EEGs
showed Lennox-Gastaut pattern (33%), hypsarrhythmia (19%), and focal
activity (14%). Antiepileptic drugs, including valproate, showed no adverse
interaction. Seizures were completely controlled in 29%, and reduced by half
in 38%. Diet was continued for an average of 24 months in those controlled and
for 4 months in the unimproved group. Dietary benefit was not determined by
seizure type. Renal stones required withdrawal of the diet in 2 patients.
(Kinsman SL et al. Efficacy of the ketogenic diet for intractable seizure
disorders: Review of 58 cases. Epilepsia Nov/Dec 1992; 33:1132-1136). (Reprints:
Dr SL Kinsman, Kennedy Krieger Institute, 707 N Broadway, Baltimore, MD 21205).

COMMENT. Despite further demonstration of its efficacy and relative
safety, the ketogenic diet is not a popular method of therapy among
neurologists generally. Apart from the Mayo Clinic and Johns Hopkins,
the diet is not frequently promoted in the training of pediatric neurolo-
gists and dieticians. Furthermore, the bias against fat in the diet in
today's society adds to the difficulty in convincing mothers to become
enthusiastic partners in the treatment program. Millichap JG at the
Mayo Clinic (Epilepsia 1964; 5: 239) and Schwartz RM at the John
Radcliffe Hospital, Oxford (Dev Med Child Neurol 1989; 31: 145) studied
the metabolic effects of the diet and were unable to document any
significant changes in blood lipid profiles using the classical diet.
For an account of the mechanism of action of the diet, see Millichap JG.

FELBAMATE IN LENNOX-GASTAUT SYNDROME

The Felbamate Study Group report results in 73 patients ages 4 to 36
years with the Lennox-Gastaut syndrome, using a double-blind, placebo-