children: case report and analysis of liver transplantation outcomes in the United States. **J Pediatr** May 2011;158:802-807). (Response and reprints: Ayse L Mindikoglu MD, U Maryland Sch of Med, Baltimore, MD. E-mail: amindiko@medicine.umaryland.edu).

COMMENT. Children who undergo liver transplantation for VPA-associated acute liver failure have a significantly worse survival rate compared with children with LF caused by other drugs. The authors conclude that in a child with a progressive seizure disorder and acute liver failure after VPA therapy, the VPA should be discontinued and carnitine initiated while investigating a presumed mitochondrial disorder. Liver transplant should be contraindicated for VPA-associated liver failure, even when a mitochondrial disease is not confirmed.

NEURODEGENERATIVE DISORDERS

CARDIAC INVOLVEMENT IN BATTEN DISEASE

The onset and progression of cardiac involvement in juvenile neuronal ceroid lipofuscinosis (Batten disease) are studied in 29 children and adolescents with genetically verified disease at Aarhus University Hospital, Skejby, Denmark. One third of initial EKGs had abnormal deeply inverted T waves. Repolarization disturbance of ventricular myocardium at initial recording correlated with risk of death during the 7-year observation period. Heart rate and variability were significantly reduced with increasing age, suggesting a decreased parasympathetic activity on the heart or negative influence on sinus node automaticity. Bradycardia, arrhythmia, sinus arrests and atrial flutter indicated an age-dependent decrease in sinus node activity. In their early 20s, ventricular hypertrophy was a frequent finding. (Ostergaard JR, Rasmussen TB, Molgaard H. Cardiac involvement in juvenile neuronal ceroid lipofuscinosis (Batten disease). **Neurology** April 8, 2011;76:1245-1251). (Response and reprints: Dr John R Ostergaard, Department of Pediatrics, Aarthus University Hospital, Skejby, Denmark, E-mail: john.ostergaard@skejby.rm.dk).

COMMENT. Juvenile neuronal ceroid lipofuscinosis (JNCL, Batten disease), caused by mutations in the CLN3 gene and failure to respond to oxidative stress (Tuxworth RJ. **Hum Mol Genet** 2011;Mar 15[Epub ahead of print]), is the most common type of inherited lysosomal storage and neurodegenerative disease. The clinical course is characterized by progressive visual failure, dementia, and seizures. In JNCL, visual failure occurs at age 4-7 years, and blindness within a few years. Psychomotor deterioration becomes evident in the early school years, and seizures start at a mean age of 10 years. Extrapyramidal symptoms develop at age 12-15, and death usually occurs by the 3rd decade. Cardiac complications have not previously received much attention. In Menkes Textbook of Child Neurology 3rd ed, 1985, cardiac involvement is not mentioned among the various forms of NCL, classified according to age at onset: Infantile (Santavuori) 9-19 months; Late-infantile (Batten-Bielschowsky) 2-4 years; Variant (Batten) 5-7 years; Juvenile (Spielmeyer-Vogt) 4 years-puberty. To-date, 10 types of NCL are described caused by mutations in recessively inherited genes, 8 of which are characterized.