2004;26:213-218). Most of the known genes code for ion channels, but 2 are not channelopathies (MASS1/VLGR1 coding for a G-protein coupled receptor in one family with febrile and afebrile seizures, and LG11, a tumor suppressor gene, identified in several families with autosomal dominant (AD) familial lateral temporal lobe epilepsy). Other epilepsies with identified genes are AD familial nocturnal frontal lobe epilepsy (CHRNA4), generalized epilepsy with febrile seizures plus (SCN1A, SCN2A), severe myoclonic epilepsy of infancy (SCN1A), and juvenile myoclonic epilepsy (GABRA1). A large number of genetic factors probably contribute to seizure susceptibility.

The clinical spectrum of SCN1A mutations ranges from febrile seizures, febrile seizures plus, a mild and a classical form of severe myoclonic epilepsy in infancy (Dravet syndrome). (Berten PGM et al. Clinical correlations of mutations in the SCN1A gene: from febrile seizures to severe myoclonic epilepsy in infancy. **Pediatr Neurol** April 2004;30:236-243).

## EPILEPSY RISK FACTORS FOLLOWING NEONATAL SEIZURES

Clinical and polysomnographic risk factors as early predictors for the development of postnatal epilepsy were determined in 158 infants presenting with two or more seizures, in a study at Hospital Sao Lucas, Porto Alegre, Brazil. Epilepsy rate after neonatal seizures was 22% within 12 months follow-up and 33.8% within 48 months. The types of epilepsy syndromes included: West syndrome in 12 infants; focal symptomatic epilepsy in 10; infantile epileptic encephalopathy in 3; other generalized symptomatic epilepsies in 5; and miscellaneous (13). Perinatal asphyxia, electrolyte imbalance, and bacterial meningitis were the most frequent etiologic factors in neonatal seizures. An abnormal neurologic examination, cerebral palsy, cognitive deficits, and abnormal polysomnographic recording were predictors of an unfavorable outcome and development of postnatal epilepsy. Prematurity, birth weight, and perinatal asphyxia were not significantly related to outcome, but the incidence of postnatal epilepsy was significantly correlated with neonatal bacterial meningitis. Neonates with seizures that required large initial doses and maintenance anticonvulsant drugs were at greater risk of postnatal epilepsy. (Da Silva LFG, Nunes ML, Da Costa JC. Risk factors for developing epilepsy after neonatal seizures. **Pediatr Neurol** April 2004;30:271-277). (Respond: Dr Nunes, Division of Neurology, Hospital Sao Lucas, Pontificia Universidade Catolica do Rio Grande do Sul School of Medicine, Av Ipiranga 6690, cj220, 90610-000 Porto, Alegre, RS, Brazil).

COMMENT. In this selected population that included infants in intensive care, a higher incidence of epilepsy than that seen in population-based studies could be expected. Multiple risk factors for neonatal seizures, and especially bacterial meningitis, are associated with a higher probability of developing postnatal epilepsy. An abnormal neurologic examination and polysomnographic abnormalities are predictors of an unfavorable outcome.

## HYPOTHYROIDISM AND NEONATAL SEIZURES

A term infant born to a mother with gestational diabetes and a history of hypothyroidism presented with seizures on the 6$^{th}$ day of life that responded to L-thyroxine, in a report from The Children's National Medical Center, Washington, DC. The mother had

not required thyroid replacement therapy during or for 9 years before this pregnancy. She was taking daily antidepressants for major depression. Birth complications included meconium in the amniotic fluid, cesarean section for abnormal fetal heart rate, neonatal respiratory distress, and treatment in the neonatal intensive care unit. The Apgar scores were 8 and 9 at 1 and 5 minutes. Birth weight was at the 10-25$^{th}$ %, length <10$^{th}$ %, and head circumference of 34 cm (25-50$^{th}$ %). Cranial sutures were wide and the posterior fontanel measured 4 cm. Oxygen saturation was 95%, and respiratory distress resolved in a few hours. Rhythmic jerking started in the right arm on day 6 and became generalized. Seizures were unresponsive to an initial phenobarbital bolus, and were controlled by addition of lorazepam and phenytoin. Lumbar puncture showed normal CSF, and blood electrolytes, glucose, and amino acids and urine organic acids were normal. EEG, CT scan, and MRI were normal. Thyroid function studies on day 9 were as follows: TSH 0.67 (N: 0.4-4.7 uIU/mL); free T4 0.5 (N: 0.7-1.8 ng/dL); free T3 1.41 (N: 1.5-3.5 pg/mL). On day 15 after L-thyroxine 25 ug/day, the TSH was 0.71 and free T4 1.4. Maternal TSH and T4 were normal. Seizures stopped and antiepileptic medications were discontinued, with one relapse at 2 months associated with hypothyroidism (hoarse cry, wide fontanel, delayed bone age, temperature instability). Seizures responded to an increase in thyroxine. The neurologic evaluation at 6 months follow-up was normal. (Aly H, Kanter DE, Fisher-Owens SA. **J Pediatr Neurol** 2004;2:111-113). (Respond: Hany Aly MD, 900 23$^{rd}$ Street, NW Room G-132, Washington, DC 20037).

COMMENT. Multiple factors may have contributed to the seizures in this case (hypoxic-ischemic encephalopathy, maternal diabetes and antidepressants) in addition to hypothyroidism. The authors argue against alternative causes and find the response to thyroxine and successful withdrawal of AEDs to be strongly supportive of an association. The expected rise in TSH with congenital hypothyroidism may be delayed in some newborns, and the initial low TSH and low free T4 are compatible with the diagnosis. (Nelson Pediatrics gives newborn normal values: TSH 1.0-9.1 and T4 2.0-4.9). The routine neonatal metabolic screening may be normal, as in this infant, and this finding should not discourage further testing for possible hypothyroidism as a cause of neonatal seizures refractory to therapy. Seizures are known to complicate myxedema in adults but are rare with congenital hypothyroidism.

## PHOTOSENSITIVE EPILEPSY

The evolution and disappearance of photosensitivity (PS) was studied long-term in 42 patients (17 males, 25 females; mean age at onset 6 years 9 months, range 5 to 12 years) with electroencephalographic evidence of photosensitive epilepsy at University of Chieti, San Valentino Hospital, and Brindisi Hospital, Italy. Valproate (VPA) monotherapy was begun after the second seizure in 36 patients, VPA with carbamazepine or lamotrigine in 4, and stimuli avoidance but no drugs in 2. Mean duration of follow-up was 8 years 1 month, and the mean age at end of follow-up was 15 years 2 months. At end of follow-up, photoparoxysmal responses (PPRs) were present in 25 and had disappeared in 15. Of 31 (75%) responding to antiepileptic drugs (AEDs), 19 had persistent PS and 12 were PS free. Photosensitive epilepsy has a good prognosis for seizure control, independent of persistence or disappearance of PS. Response to AEDs is not dependent on disappearance of PS. PS is