neonates were contributory in 24 (41%). Metabolic tests in 45 cases contributed to the final diagnosis in 9 (20%). EMG and NCS in 23 neonates were contributory in 10, but misleading and incorrect in 3 (spinal muscular atrophy mistaken for demyelinating neuropathy, and negative reports in a case of myotonic dystrophy and one of congenital muscular dystrophy). Muscle biopsy in 14 cases was helpful in 6, and tests for congenital myasthenia in 10 were negative. (Laugel V, Cossee M, Matis J et al. Diagnostic approach to neonatal hypotonia: retrospective study on 144 neonates. **Eur J Pediatr** May 2008;167:517-523). (Respond: Dr Vincent Laugel, Sevice de Pediatrie 1, CHU Strasbourg-Hautepierre, Avenue Moliere, F-67098 Strasbourg Cedex, France. E-mail: vincent.laugel@chru-strasbourg.fr).

COMMENT. The hypotonic infant is a fairly common pediatric problem, accounting for 1 in 25 admissions to a tertiary medical center and neonatal unit. The authors provide a diagnostic algorithm based on the yield of the initial neurological examination and various tests. Central (cerebral) causes are most frequent, and the initial physical examination has a higher positive predictive value for this type of hypotonia than for peripheral (neuromuscular) types. First line tests to confirm a central cause are neuroimaging and EEG. For peripheral type hypotonia, examination of the mother (in suspected myotonic dystrophy or neonatal transient myasthenia) and DNA-based tests are most reliable. In some cases a longer follow-up may be necessary to determine the course of the disease and the correct diagnosis.

The hypotonic or limp infant syndrome has been reviewed in the literature over many years. (Walton JN. The limp child. **J Neurol Neurosurg & Psychiat** 1957;20:144-154) (Millichap JG. The hypotonic child. In **Practice of Pediatrics**. Vol IV, ed. Brennemann-Kelley, Chap 16, Hagerstown, MD; WF Prior Co. 1966). Infants with so-called benign congenital hypotonia, a term coined by Walton, are limp at birth, sitting and walking are delayed, but muscle wasting is not profound. The muscle fibers are generally small for the age. Muscle strength and motor development gradually improve, but mental retardation and other congenital abnormalities are not uncommon. In recent years, molecular genetic diagnosis of myopathies has uncovered several inherited diseases characterized by neonatal hypotonia, including nemaline myopathy and congenital fiber type disproportion (Clarke NF et al. **Ann Neurol** March 2008;63:329-337) (Rifai Z et al. **Neurology** 1993;43:2372-2377). Defining the genetic basis of these diseases will explain their heterogeneous clinical manifestations and lead to improvements in family counseling.

MOVEMENT DISORDERS

DOPA-RESPONSIVE DYSTONIA WITH DELAY IN WALKING

A 2-year, 8-month old boy with a previous diagnosis of cerebral palsy was referred to UCSF because of "awkward" gait and "walking on the toes." His gait abnormality progressed, worsening in the evening, and he was found to have a dopa-responsive dystonia caused by an autosomal-dominant GCH1 mutation. He was treated successfully with oral carbadopa-levodopa (Sinemet). Three other family members were affected, presenting with stiffness in the thighs, motor impairment, speech and swallowing difficulties, postural tremor and depressive anxiety, also responsive to carbadopa-levodopa. (Cheyette BNR et al. **Pediatr Neurol** April 2008;38:273-275).