COMMENT. Acute facial nerve palsy is a common symptom of LNB, and a significant number (21%) will persist at follow-up. In a previous study of long-term outcome (3-5 years) of facial palsy in LNB, one-half of patients with subjective symptoms of residual facial palsy had signs of mild to moderate dysfunction on clinical examination, III-IV on the House-Brackman grading scale (I normal-VI no movement). (Bagger-Sjoback D et al. **Otol Neurotol** 2005 Jul;26(4):790-5). Subjective symptoms, objective signs, and neurophysiological test results show no clear correlation.

Since LNB is amenable to antibiotic treatment, a high index of suspicion and early diagnosis of acute neurologic complications is important. Examples of more common neurologic manifestations include, in addition to facial palsy, lymphocytic meningitis, mononeuropathy multiplex, and painful radiculoneuritis. (Halperin JJ. **Curr Infect Dis Rep** 2011 Aug;13(4):360-6).

SPINAL TUBERCULOSIS (POTT'S DISEASE)

Researchers at Great Ormond Street Hospital for Children and Institute of Child Health, London, UK reviewed their experience of childhood spinal tuberculosis (TB) over a 15-year period (1995-2010). Of 21 patients identified (median age 9.7 years, range 3.4-15.9 years) 11 were Black African, 7 Asian, 2 Middle Eastern and 1 Caucasian. Nine were born in the UK, 1 in the Netherlands and the remainder outside Europe. Ten had traveled to a country endemic for TB within the year before diagnosis. Four (19%) had a previous diagnosis of TB, 11 (52%) a known contact, 10 (48%) had received BCG vaccine and none was HIV-positive. Clinical presentations included systemic symptoms in 18 (night sweats, weight loss, fever and anorexia), back pain in 16, neurological symptoms in 12 (weakness and limp in 7, sensory change in 5), and spinal deformity in 5. *Mycobacterium tuberculosis* was isolated in 14 patients (67%) by vertebral biopsy or from paraspinal abscess. Spinal cord compression or stenosis occurred in 8 (38%), vertebral collapse in 13 (62%), and paraspinal abscess in 15 (71%). Chest x-ray showed TB lung disease in 8 patients (38%). Extra-spinal disease was co-existent in 12 (57%) patients, including psoas abscess in 5 (24%). All patients received TB treatment for at least 12 months, 7 underwent surgery, and 75% resolved fully. All patients were alive and without neurologic deficit at a median follow-up of 24 months. (Eisen S, Honywood L, Shingadia D, Novelli V. Spinal tuberculosis in children. **Arch Dis Child** 2012 Aug;97(8):724-9). (Respond: Dr Sarah Eisen, Department of Infectious Diseases and Microbiology, Institute of Child Health, 30 Guildford St, London WC1N 1EH, UK. E-mail: saraheisen@hotmail.com).

COMMENT. The authors list key features that should alert the clinician to a diagnosis of spinal TB: TB contact or travel to endemic area, history of previous TB, systemic symptoms, back pain and long duration of symptoms. Treatment should be supervised closely and prolonged. Late onset paraplegia, a feature of Pott's disease, was not a complication in the authors' cases. In a series of 8 patients with late onset Pott's paraplegia due to kyphosis, this complication was treated successfully with decompression and grafting. A mean period of 24 years (range, 9-46 years) had elapsed from the onset of active disease and the age at neurological deterioration. (Bilsel N, et al. **Spinal Cord** 2000 Nov;38(11):669-74). This report re-emphasizes the need for long-

term antibacterial therapy, careful follow-up and monitoring with spinal x-ray and neurologic evaluation, as indicated in the Great Ormond Street experience of spinal TB.

METABOLIC DISORDERS

CLINICAL CHARACTERISTICS OF 5 PHENOTYPES OF COENZYME Q10 DEFICIENCY

Researchers at Columbia University Medical Center, New York; University of Genoa, Italy; and University of Granada, Spain reviewed 149 cases of coenzyme Q10 (ubiquinone) deficiency, including their own cohort of 76 patients diagnosed from 1997-2010. Cerebellar ataxia was the principal phenotype and the presenting symptom in 94 children (63%). Less frequent phenotypes included encephalomyopathy in 4 patients, isolated myopathy in 14, infantile-onset multisystemic disease in 17, nephropathy (with or without sensorineural hearing loss) in 11, and atypical presentations in 9. Other manifestations include neuropathy, seizures, congenital hypotonia, dystonia, ophthalmoplegia, retinitis pigmentosa, optic atrophy, agenesis of corpus callosum, and hypogonadism. Onset was primarily in childhood; 82% were aged < 13 years including 23% in infancy (<12 months). Mortality rate was 8%.

Direct measurement of CoQ10 in skeletal muscle by liquid chromatography is the most reliable test for diagnosis of CoQ10 deficiency. Morphological and biochemical findings differ in the various clinical forms. Family history suggests autosomal recessive inheritance. Pathogenic mutations are described in patients with the infantile multisystemic syndrome and some juvenile-onset cerebellar ataxia cases. Response to oral supplementation with CoQ10 is frequent but variable; one patient with infantile spasms failed to respond. (Emmanuele V, Lopez LC, Berardo A, et al. Heterogeneity of coenzyme Q10 deficiency. Patient study and literature review. **Arch Neurol** 2012 Aug;69(8):978-83). (Respond: Michio Hirano MD, H Houston Merritt Clinical Research Center, Department of Neurology, Columbia University Medical Center, 630 W 168th St, P&S 4-423, New York, NY 10032. E-mail: mh29@columbia.edu).

COMMENT. The occurrence of primary and secondary CoQ10 deficiencies adds to the difficulty in study of the molecular classification of this heterogeneous disorder. (Quinzii CM, Hirano M. **Biofactors** 2011 Sep;37(5):361-5). Pathogenic mutations are identified in genes involved in the biosynthesis of CoQ10 (primary CoQ10 deficiencies) or in genes not directly related to CoQ10 biosynthesis (secondary CoQ10 deficiencies). Respiratory chain defects may contribute to the pathogenesis of primary CoQ10 deficiencies.

HEADACHE DISORDERS

MANIFESTATIONS OF FAMILIAL HEMIPLEGIC MIGRAINE

Researchers at University of Arkansas, Little Rock, AR report 3 cases of familial hemiplegic migraine complicated by reversible cerebral edema and followed by