OUTCOME OF MITOCHONDRIAL DISEASES

Researchers at Children's Hospital, University Medical Center Ljubljana, Slovenia; Molecular Neurogenetics, Milan, Italy; and Paracelsus Medical University, Salzburg, Austria, evaluated the functional outcome of 22 children with mitochondrial diseases. Twelve children (55%) had pyruvate dehydrogenase complex deficiency, 4 had Leigh syndrome, and 10 had respiratory chain enzyme deficiencies. Complex IV was the most frequent enzyme deficiency. The morbidity was high, but the mortality was low (18%). Total Mitochondrial Disease Criteria score was above 7, 82% above 8. Using the Pediatric Evaluation of Disability Inventory and scaled scores, a large variability in functional skills was seen in self-care, mobility, and social function. Also, a large variability in caregiver assistance, from independence to total dependence on assistance was observed. Few patients used assistive devices. Children with mitochondrial disease have severe limitations in cognition and communication and are dependent on their parents. Functional status is a necessary part of the evaluation of children with mitochondrial disease. (Rogac M, Meznaric M, Zeviani M, Sperl W, Neubauer D. Functional outcome of children with mitochondrial diseases. **Pediatr Neurol** April 2011;44:340-346). (Respond: Dr Rogac. E-mail: mihael.rogac@mf.uni-lj.si).

COMMENT. Evaluation of skills in self-care, mobility, and social function is important in the management of children with mitochondrial disease. The Pediatric Evaluation of Disability Inventory is an effective bedside measure of estimating a child's need for caregiver assistance and assistive devices.

INFECTIOUS DISORDERS

POTT'S PARAPLEGIA IN AN INFANT

Neurosurgeons at the Barrow Neurological Institute, St Joseph's Hospital, Phoenix, AZ, report a 13-month-old Hispanic boy who presented with paraplegia caused by tuberculosis affecting the mid-thoracic spine. The infant walked at 9 months, and at 11 months, he and his family were diagnosed with pulmonary TB. Despite a full course of anti-TB medications, the boy developed a progressive paraplegia. He was spastic and hyperreflexic, and ankle clonus and Babinski signs were elicited. A kyphotic gibbus deformity was revealed on MRI, and with contrast, a paraspinal and epidural abscess was seen from T2 to T6, ventral, extending into the spinal canal, and compressing the spinal cord with displacement to the right. CT spine showed complete destruction of T4 vertebral body and collapse of T3 onto the body of T5. Surgery involved laminectomy, corpectomies from T2 to T4 via posterior approach, drainage of abscess, Steinmann pins, and grafts from T1 to T6. He was continued on antibiotics for 4 months and at 26 months after surgery, he could walk 5 steps and had no progression of the kyphotic deformity. (Consigilieri G, Kakarla UK, Theodore N. Pott disease in a 13-month-old: Case-report. **Neurosurgery** May 2011;68(5):E1485-E1490). (Respond: Nicholas Theodore MD, FACS, Barrow Neurological Institute, 350 W Thomas Rd, Phoenix, AZ 85013. E-mail: neuropub@chw.edu).

COMMENT. Pott's disease is a leading cause of paraplegia in developing countries, but occurs rarely in the US. This case may represent one of the youngest reported, according to the authors' review of published cases, totaling 243, and one, an 8-month-old Turkish infant. Greenfield JG, in his textbook **Neuropathology** (Baltimore, Williams & Wilkins, 1963, p 662-3), refers to associated myelin degeneration in the lateral columns, and compression and TB arteritis of the intervertebral radicular artery, accounting for some acute onset cases. Percival Pott, surgeon at St Bartholomew's Hospital, London, first described Pott's disease (1779, 1782) as paraplegia associated with tuberculous spinal caries. David JP (1779) also described the condition in France.

LIMBIC ENCEPHALITIS IN CHILDHOOD

Researchers at the University of Innsbruck, Austria, and 11 additional centers in Europe and Oxford, UK, report 10 patients <18 years of age with symptoms of limbic encephalitis (LE) of <5 years' duration and MRI evidence of mediotemporal lobe inflammation (hyperintense T2/FLAIR signal). The characteristic symptoms of LE were impairment of recent memory, temporal lobe seizures, and affective disturbances. Only 1 patient had a tumor, a neuroblastoma. Median age at disease onset was 14 years (range 3-17). Eight patients had defined autoantibodies (Hu, GAD, VGKC) known to be associated with adult-onset limbic encephalitis. Two patients identified with anti-NMDAR encephalitis without limbic dysfunction were not included. After a median follow-up of 15 months and corticosteroid or IV immunoglobulin treatment, 2 patients recovered, 8 remained impaired and one died.

Diagnosis of limbic encephalitis requires signs and symptoms predominantly (but not exclusively) of limbic involvement for < 5 years, MRI evidence of mediotemporal inflammatory disorder (hyperintense T2/FLAIR signal), and specific autoantibodies. A tumor and paraneoplastic disorder must be excluded. Other differential diagnoses include cortical dysplasia, infectious encephalitis (HSV, HHV6, VZV), anti-NMDAR encephalitis with no mediotemporal involvement, and chorea-acanthocytosis. (Haberlandt E, Bast T, Ebner A, et al. Limbic encephalitis in children and adolescents. **Arch Dis Child** Feb 2011;96:186-191). (Respond: Dr Christian G Bien, Dept of Epileptology, University of Bonn Medical Centre, Sigmund-Freud-Str. 25, 53105 Bonn, Germany.E-mail: christian.bien@ukb.uni-bonn.de).

COMMENT. Limbic encephalitis, an inflammatory disorder of paraneoplastic or non-paraneoplastic origin, and characterized by memory deficits, temporal lobe seizures or affective disorders, is recognized in adults but until recently, rarely diagnosed in children <18 years of age. Reporting the case of a 16-year-old boy who presented with subacute neuropsychiatric symptoms following a gastrointestinal illness, McCoy B et al, **(J Child Neurol** 2011;26(2):218-222) describe the disorder as an emerging pediatric condition. The MRI in this child revealed progressive hippocampal signal abnormality and swelling, and NMDAR antibody was detected in the serum. A series of 14 cases of limbic encephalitis in childhood reported from Japan revealed a predominance of seizures, disturbed consciousness, and frequent extralimbic signs as presenting symptoms. The majority had antecedent febrile illness, and a child-specific phenotype of