

# Acute sensory neuronopathy: Report of a child with remarkable clinical recovery

**Article abstract**—A 9-year-old boy presented with acute sensory neuronopathy, a condition generally associated with poor recovery. Although sensory nerve action potentials were abolished and sural nerve biopsy showed marked loss of myelinated fibers, he eventually made an almost complete clinical recovery.

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In 1980, Sterman et al<sup>1</sup> described a new clinical entity: acute sensory neuronopathy, a monophasic disorder characterized by rapid onset of generalized paresthesias, ataxia, and areflexia which "progresses rapidly to a severe, mainly proprioceptive sensory deficit."<sup>1</sup> Sensory nerve action potentials are markedly reduced or absent, but motor function remains intact. The site of the lesion is thought to be at the dorsal root ganglia and, in the similar cases reported to date, ultimate recovery has been very poor.<sup>1-5</sup>

We present a patient with the typical clinical and electrophysiologic features of this syndrome who made a good functional recovery.

**Case report.** A previously healthy 9-year-old boy presented with fever, headache, and generalized exanthema 10 days before admission. Eight days later he developed vomiting, abdominal distension, and constipation. He had not been exposed to toxic substances and had not been given pyridoxine.

On admission, arterial blood pressure was 110/60 mm Hg, heart rate 106 per minute, and respiration rate 40 per minute. Rectal temperature was 39 °C. He was fully conscious and neurologic examination was normal. Abdominal x-rays revealed a distended small intestine. There was mild leukocytosis with shift to the left. Laparotomy disclosed mesenteric adenitis. He was given clindamycin and co-trimoxazole, and the fever subsided in 2 days. On his fifth day in the hospital he acutely developed numbness of the left side of the face and tongue that rapidly extended to the entire body. He complained of dry eyes and mouth and generalized paresthesias. In addition, he developed acute bladder paresis requiring catheterization.

On examination, his mental status was normal and his pupils were mydriatic (7 mm), equal, completely unresponsive to light, and near accommodation. Light touch and pinprick sensations of the face were reduced bilaterally, and corneal reflexes were absent. Ocular movements and the rest of the cranial nerves were intact. Muscle bulk and strength were normal. There was mild loss of pain and temperature sense, severe impairment of position and vibration sense in all limbs, and astereognosis. He had profound truncal and limb sensory ataxia which prevented him from walking or standing unaided. Limb tendon reflexes were absent, but jaw jerk was normal. Plantar responses were flexor.

Blood pressure fluctuated between 120/70 and 100/60

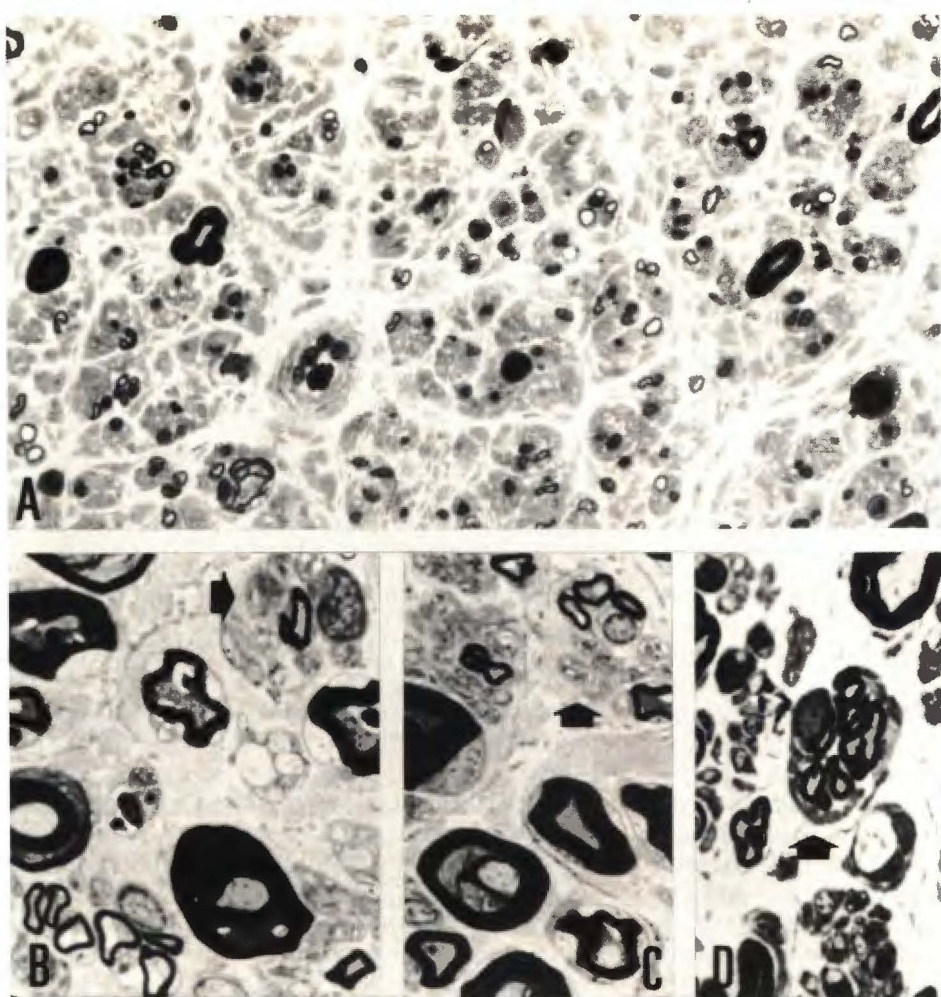
mm Hg, but no orthostatic hypotension was detected. Normal values were obtained for CBC, chemistry, serum lead and mercury, liver and thyroid functions, serology, and immunology. Urine screen for porphobilinogen and porphyrins was negative. Cultures of peritoneal fluid, blood, and stools were negative. Small-intestine biopsy revealed no pathologic changes. EEG and brain CT were normal. CSF was clear and acellular; the glucose content was 60 mg/dl and the protein was 65 mg/dl with no discrete bands on electrophoresis.

**Clinical follow-up.** Intestinal and bladder paresis improved within a week. Neurologic status remained unchanged for 2 months. At 6 months, pinprick and touch sensations and ataxia had mildly improved, but marked loss of position and vibration sensation was still present.

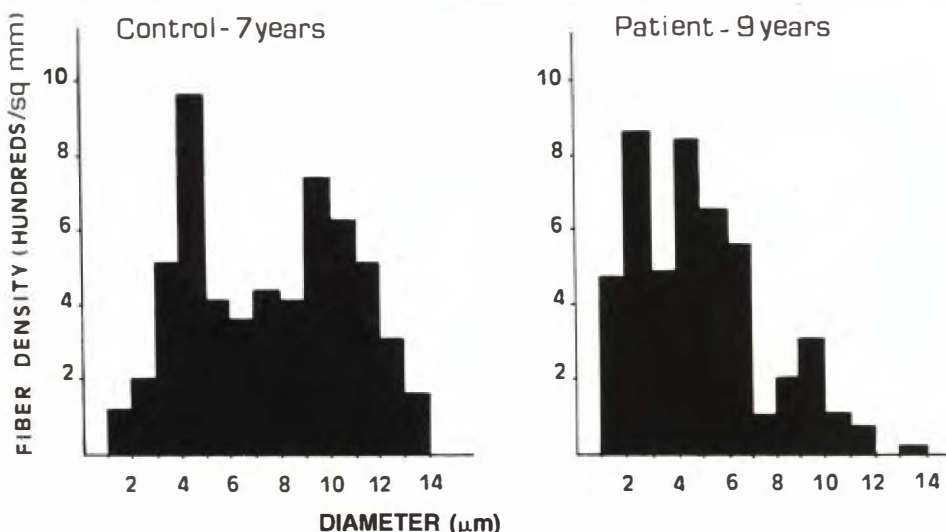
Neurologic status continued to improve gradually over time. Three years after onset he had fully reactive pupils, normal gait, and no ataxia. At the last follow-up, 6 years after onset, he had only tendon areflexia, mild facial hypesthesia, depressed corneal reflexes, and impaired stereognosis of the ulnar aspect of the right hand.

**Neurophysiologic investigations.** These were performed on several occasions, the first on the 13th day after the onset of neurologic symptoms and the last 6 years later. Concentric needle examination showed no abnormal spontaneous activity on any occasion. The voluntary pattern on maximal effort was full in proximal and distal muscles with motor unit potentials of normal configuration. In addition, turns/amplitude analysis of the interference pattern<sup>6</sup> of several muscles was always normal. Single-fiber EMG examination of the extensor digitorum communis muscle on the 13th day and again 1 month later showed normal fiber density (1.6), normal jitter values (less than 55  $\mu$ sec), and no blocking phenomena. Motor nerve conduction velocities, compound muscle action potential amplitudes (CMAPs), and F-wave latencies in the median, ulnar, peroneal, and posterior tibial nerves were normal at all evaluations. Repetitive stimulation of the ulnar nerve at 3 and 50 Hz, recorded with surface electrodes in the hypothenar eminence, was normal on the 13th day. However, sensory nerve action potentials of the median, ulnar, radial, and sural nerves were unobtainable, as were somatosensory evoked potentials and blink reflexes. H reflexes and the proprioceptive silent period of the soleus muscle were absent in all examinations and remain so. Pattern-shift visual evoked potentials and brainstem auditory evoked potentials were normal. Masseter reflex elicited by an oscilloscope-triggering hammer was normal.

**Follow-up.** Blink reflexes were abolished for 3 years. Then a single delayed response appeared, with latencies



*Figure 1. Semithin section of the sural nerve showing marked dropout of large myelinated fibers and relative predominance of small-caliber fibers, sometimes arranged in clusters (A). Axonal sproutings of variable size are also seen (arrows in B, C, and D). (Richardson stain; A,  $\times 400$  before 6% reduction; B to D,  $\times 1,000$  before 6% reduction.)*



*Figure 2. Myelinated fiber histogram demonstrating loss of large-caliber fibers and predominance of small-caliber fibers in our patient compared with a normal control.*

ranging from 70 to 85 msec. On the last examination, 6 years after onset, early and late responses were elicited with increased latencies (R1, 25 msec; R2, 55 to 65 msec). Sural sensory response was 2  $\mu$ V (sural conduction velocity [SCV], 32.4 m/sec), and right median sensory potential (third digit) was 5  $\mu$ V (SCV, 39.4 m/sec).

One year after onset of symptoms, a biopsy was obtained from the left sural nerve. The specimen was fixed in 2.5% glutaraldehyde in Na<sup>+</sup> cacodylate buffer, postfixed in 1% osmium tetroxide, and embedded in Araldite. Semithin sections were stained with Richardson stain (toluidine blue, methylene blue, and borax).



Sections of the nerve showed marked loss of both myelinated and unmyelinated fibers (figure 1A). Several sections revealed the presence of axonal sproutings (figure 1, B, C, and D). There were no cellular infiltrates in the epineurium or endoneurium, and no amyloid or metachromatic materials. In figure 2, a histogram of myelinated fibers shows loss of large-caliber fibers and predominance of small myelinated fibers in our patient compared with a normal control.

**Discussion.** This patient presented with generalized dysesthesias, autonomic dysfunction, ataxia, and profound proprioceptive deficit, but with normal motor power. Conventional and computerized EMG, motor conduction studies, repetitive nerve stimulation, and single-fiber EMG were normal, ruling out a subclinical motor disturbance or a neuromuscular transmission disorder (ie, botulism). These findings, combined with the absence of sensory potentials and of blink and H reflexes, are diagnostic of acute sensory neuronopathy.<sup>1,7,8</sup> This diagnosis implies massive loss of neurons in the dorsal root ganglia, and therefore predicts poor recovery, as actually happened in the patients described previously.<sup>1-5</sup> However, in this patient sensory functions improved considerably over subsequent years. Furthermore, there was electrophysiologic and histologic evidence of partial sensory regeneration over the following years. These findings suggest that a subpopulation of neurons underwent distal axonal degeneration—which manifested itself by a “dying back” neuropathy—but not perikaryal death. In this situation, a certain degree of functional recovery might occur by way of distal sprouting, particularly in children. More severe lesions would result in death of the dorsal ganglion cell bodies (neuronopathy), leading to irreversible degeneration of the axon.<sup>7,8</sup> Such a dose-dependent mechanism occurs in the clinical and experimental sensory neuropathy produced by pyridoxine<sup>9,10</sup> and other neurotoxic drugs that may involve selectively the dorsal root ganglion cells.<sup>8</sup> This patient's case demonstrates the clinical and pathologic overlap between the generally reversible acute sensory neuropathies and the usually irreversible acute sensory neuronopathies.

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