

still paralyzed. The phrenic nerve has a longer anatomic course than the recurrent laryngeal nerve and may subsequently show a slower rate of recovery. Following stretch trauma of the phrenic nerve, the normal diaphragmatic function gradually returns over the succeeding six to twelve months.¹⁶

In conclusion, the present case report describes an interesting complication of postoperative unilateral paralysis of both the phrenic nerve and the recurrent laryngeal nerve, following surgery at a site far from their anatomical pathways. The possible etiologic factors predisposing to this complication are discussed. The occurrence of unilateral paralysis of both nerves in the same patient, on the same side and at the same time suggests a single etiology. However, the possibility still remains that separate etiologies were involved.

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Anesthesia for a Child with Leigh's Syndrome

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Leigh's syndrome¹ or subacute necrotizing encephalomyelopathy (SNE) is a chronic neurologic disease, usually discovered during infancy. Feeding problems, weakness, external ophthalmoplegia, swallowing difficulties, ataxia, and convulsions are frequent clinical features; with respiratory difficulties occurring late in the course of the disease. The neuropathologic findings, both histologically and topologi-

cally, are distinctive,² with several similarities to Wernicke's encephalopathy. Increased blood lactate and pyruvate concentrations are also frequently present. The most likely enzyme abnormality is a defect in activation of pyruvate dehydrogenase.³

These patients have a chronic relapsing course, and acute exacerbation has been reported following surgery.⁴ The anesthetic course in a fourteen-month-old child who required an open liver biopsy for diagnosis is described below.

REPORT OF A CASE

The patient was a 14.5-month, 7-kg girl who was born prematurely at 28-weeks gestation with Apgar scores of 6 and 9. She was treated with continuous positive airway pressure and oxygen, and was discharged at age two months. The child was mildly developmentally retarded, but was pulling to stand and imitating sounds by

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10 months. At 13 months, she underwent surgical correction of strabismus and myringotomy under halothane anesthesia. Following this surgery, her developmental milestones regressed to the point where she could not sit. No biochemical abnormality was found and a CT scan of the brain was reported as "essentially normal." She improved over two weeks and surpassed her previous developmental skills. However, during the next 1.5–2 months, her neurologic function gradually declined and she was admitted for evaluation. A CT scan at this time was consistent with cerebral degenerative disease. Biochemical tests were normal except for consistently elevated blood lactate and pyruvate levels. Mean blood lactate concentration was 52.4 ± 36 SD mg/dl (normal 3–13 mg/dl), and mean pyruvate concentration was 1.01 ± 0.39 SD mg/dl (normal 0.3–0.7 mg/dl). She was scheduled for an open liver biopsy. Pre-medication was achieved with atropine, 0.07 mg, im. Anesthesia was induced with thiopental, 25 mg, and maintained with nitrous oxide (67 per cent), meperidine (12.5 mg), and *d*-tubocurarine (6 mg). Immediately following intubation of the trachea, pH_a was 7.31, P_{aCO_2} 45 torr, P_{aO_2} 110 torr, and base excess -3.5 mEq/l. Hematocrit was 29 per cent; serum sodium 136 mEq/l, serum potassium 3.9 mEq/l, serum chloride 106 mEq/l, blood lactate 34.5 mg/dl, and blood pyruvate 1.55 mg/dl. The surgery proceeded uneventfully. Normal saline, 120 ml, packed erythrocytes, 10 ml, and fresh frozen plasma, 10 ml, were infused. At the end of surgery, reversal of muscle paralysis was accomplished with atropine and neostigmine. The trachea was extubated in the recovery room. While breathing room air spontaneously, pH_a was 7.34, P_{aCO_2} 38 torr, P_{aO_2} 89 torr and base excess -5.0 mEq/l. Blood lactate and pyruvate levels were 39.5 mg/dl and 1.95 mg/dl, respectively. Her postoperative course was satisfactory and neurologic function remained the same as preoperatively. Microscopic examination of the liver biopsy revealed markedly abnormal-appearing mitochondria. Special tissue studies suggested a defect in the activation of pyruvate dehydrogenase.

DISCUSSION

Chronic lactic acidosis has been described in association with many childhood syndromes.⁵ Several enzyme defects could prevent the entry of pyruvate into the citric acid cycle, which would necessitate the conversion of pyruvate to lactate and alanine. With such an enzyme defect (provided tissue oxygenation is adequate), the lactate-to-pyruvate ratio will be normal (approximately 10:1).

The diagnosis of Leigh's syndrome rests on the characteristics of the neuropathologic findings. Symmetrical lesions are usually found in the brain stem and in the lateral walls of the third ventricle. The location of these lesions helps explain the clinical features, which are variable but include respiratory difficulties, hypotonia, ataxia, convulsions, and other autonomic nervous system symptoms. The syndrome is usually diagnosed before four years of age and has a progressive course marked by remissions and acute exacerbations. Approximately 15 per cent of the reported cases live past the age of 4 years, and several cases have been diagnosed after the second decade.²

Although the clinical and pathological features are adequately described, the precise biochemical abnormalities are less well understood. A decrease in hepatic

pyruvate carboxylase,^{6,7} inhibition of thiamine pyrophosphate-ATP phosphoryl transferase,⁴ and defective activation of pyruvate dehydrogenase³ have all been proposed. The latter defect is most likely. Patients with SNE may lack the ability to control the activity of this enzyme complex appropriately under various metabolic conditions, which might explain the fluctuations in their clinical course.

I have reported a neurologically and biochemically uneventful anesthetic in a child with Leigh's syndrome. Because of possible brainstem lesions, close attention was paid to airway management and to monitoring, including body temperature and arterial *pH*. Hyperventilation was avoided, since a low P_{aCO_2} might inhibit pyruvate carboxylase and worsen the lactic acidemia.⁸ Normal saline was used for crystalloid with colloid and blood components given as required. No exogenous lactate was administered. Selection of appropriate anesthetic agents was difficult, since both barbiturates and inhalational agents are known to interfere with mitochondrial respiration.^{9,10}

Elevated spinal fluid endorphin levels were recently reported in a patient with SNE.¹¹ A transient response of his clinical symptoms to naloxone administration was found. If this finding is confirmed, use of narcotics in patients with SNE will have to be carefully evaluated.

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