drome of continuous muscle fiber activity and plasmacytoma with IgM paraproteinemia. *Neurology* 1994;44:560–561.
4. Newsom-Davis J, Mills KR. Immunological associations of acquired neuromyotonia (Isaacs syndrome): report of 5 cases and literature review. *Brain* 1993;116:453–469.
5. Halbach M, Hömberg V, Freund H-J. Neuromuscular, autonomic and central cholinergic hyperactivity associated with thymoma and acetylcholine receptor-binding antibody. *J Neurol* 1987;234:433–436.
6. Garcia-Merino A, Cabella A, Mora JS, Liano H. Continuous muscle fiber activity, peripheral neuropathy and thymoma. *Ann Neurol* 1991;29:215–218.
7. Rceback J, Benton S, Swash M, Schwartz MS. Penicillamine-induced neuromyotonia. *BMJ* 1979;279:1464–1465.
8. Sinha S, Newsom-Davis J, Mills K, Byrne N, Lang B, Vincent A. Autoimmune etiology for acquired neuromyotonia (Isaacs syndrome). *Lancet* 1991:338:75–77.
9. Bady B, Chauplannaz G, Vial C, Savet J-F. Autoimmune etiology for acquired neuromyotonia. *Lancet* 1991;338:1330.
10. Bobert FB. The staphylococci. In: Bobert FB, Joseph Marr J, eds. *Medical Microbiology.* Boston, MA: Little, Brown and Co, 1980: 273–285.
11. Andersson M, Alvarez-Cermeo J, Bernardi G, et al. Cerebrospinal fluid in the diagnosis of multiple sclerosis: a consensus report. *J Neurol Neurosurg Psychiatry* 1994;57:897–902.
12. Zeman A, McLean B, Keir G, Luxton R, Sharief M, Thompson E. The significance of serum oligoclonal bands in neurological diseases. *J Neurol Neurosurg Psychiatry* 1993;56:32–35.
13. Shillito P, Molenaar PC, Vincent A, et al. Acquired neuromyotonia: evidence for autoantibodies directed against K+ channels of peripheral nerves. *Ann Neurol* 1995;38:714–722.

Intrathecal Baclofen in the Treatment of Dystonic Storm

Dystonia is characterized by prolonged, involuntary muscle contractions frequently causing twisting and repetitive movements or abnormal postures.[1] When dystonia rapidly escalates from its baseline to a presentation of extreme, forceful, continuous, generalized dystonic contractions, it has been referred to as dystonic storm.[2] Dystonic storm is a neurologic emergency that requires aggressive intervention. We present a case of dystonic storm that was responsive to intrathecal baclofen (ITB).

Case Report

A 16-year-old boy with DYT1 torsion dystonia first developed right-hand clumsiness at age 8. Within 8 months, both arms were involved and he began walking with his trunk markedly flexed. By age 10, he was wheelchair-bound, unable to walk as a result of dystonic spasms involving all four limbs and the trunk. At age 12, his spasms were severe enough to cause a fracture of his right radius, requiring open reduction and internal fixation. At baseline, he was able to sit in a wheelchair and move all four extremities. He required help with feeding, dressing, and hygiene. His treatment included 60 mg/day baclofen, 40 mg/day trihexiphenidyl, and 6 mg/day clonazepam which gave him some relief from the spasms. Trials of levodopa, diazepam, carbamazepine, and phenytoin proved ineffective.

At age 16, his dystonia worsened markedly over a 5-day period caused by no clear precipitant. On admission to an intensive-care unit, his temperature was 40°C. He was awake and alert and in continuous, generalized, painful dystonic spasms at rest. His face was in a grimace with his eyes shut tightly. His trunk arched in ophisthotonic posture. His arms were flexed, and his legs scissored and flexed at the knees and hips, his heels reaching close to his face (see videotape segment 1). He did not have loss of speech, dysphagia, or respiratory impairment. His deep tendon reflexes were 2+ in his arms, 3+ at the knees, and he had non-sustained bilateral ankle clonus; his plantar responses were flexor. His serum creatinine kinase was 1032 U/mL (normal, 35–200 U/mL). His treatment included intravenous hydration and acetaminophen, and his oral medications for dystonia were increased. His dystonic spasms continued, and he was sedated with a continuous intravenous infusion of midazolam. These measures brought his dystonia under temporary control, but the spasms increased with every attempt to reduce the medication. As a last resort before generalized anesthesia, the patient underwent test doses of intrathecal baclofen (ITB).

One hour after an ITB bolus of 50 μg, he was markedly less dystonic (see videotape segment 2). The facial dystonia had resolved except for occasional twitches involving his orbicularis oculi muscles. His arms were resting comfortably by his side and he could demonstrate a functional grasp with his right hand. His legs were relaxed and fully extended, and his deep tendon reflexes were normal. Within 3 hours, his dystonia increased to its former level. A repeat test dose of 75 μg ITB produced similar benefit but caused drowsiness. An ITB pump (10-cc reservoir; Medtronic SynchroMed, Minneapolis, MN, U.S.A.) was inserted, but he had pain at the surgical site requiring 1 mg intravenous morphine every 4 hours for relief.

Over the next 2 weeks, the ITB was increased to 900 μg/day as the midazolam and morphine were concurrently tapered off. He was able to participate in physical therapy and sit in a wheelchair. On a dystonia rating scale[3] while recumbent, the dystonia severity score declined from 98 to 76 (maximum score 120); his functional capacity improved from 24 to 20 (maximum score 30) chiefly as a result of improvement in right-hand function (videotape segment 3). His oral medications were continued and dantrolene was added before discharge. A follow-up videotape segment was obtained at 3 months (videotape segment 4) showing his dystonia at its previous baseline.

Eight months after his episode of dystonic storm, the patient began to develop tolerance to chronic ITB therapy. The ITB infusion rate was not increased above 900 μg/day because of the need for more frequent refills, and the patient declined a drug holiday or other intervention related to the pump. Instead, he underwent a bilateral pallidotomy for his dystonia 11 months after undergoing ITB pump implantation, resulting in an improvement in his dystonia. At 3 months postoperatively, it is too early to know if the effect of the pallidotomy will be sustained, and the patient continues on 800 μg/day ITB.

A videotape accompanies this article.

Received September 16, 1997; revisions received December 15, 1997, and January 6, 1998. Accepted January 6, 1998.

Address correspondence and reprint requests to Blair Ford, MD, at the Neurological Institute, 710 W. 168th St., New York, NY 10032, U.S.A.

Discussion

Dystonic storm is a condition of severe, potentially life-threatening, generalized dystonia arising precipitously, often against a backdrop of chronic dystonia. Dystonic storm may be caused by trauma, surgery, infection, fever, medication withdrawal, or, as in the present case, no clear precipitant. We follow another patient who experienced dystonic storm after the abrupt cessation of ITB therapy, described as a complication in a previous report.[4] The differential diagnosis of dystonic storm includes malignant hyperthermia and neuroleptic malignant syndrome. The immediate risks to life in this setting include hyperthermia, muscular fatigue, rhabdomyolysis with acute renal failure,[5,6] dehydration, and respiratory compromise.

Dystonic storm should be treated in an intensive-care unit. Treatment consists of supportive measures, including hydration, pain control, monitoring, respiratory assistance if needed, and attempts to interrupt the intractable dystonic spasms.[2] Hyperpyrexia may respond to acetaminophen or require external cooling. Myoglobinuria requires adequate hydration but dialysis may be needed.[2] To reduce the dystonic spasms, several oral medications in high doses have been suggested.[2] Agents such as dantrolene, tetrabenazine, and neuroleptics may be tried on an empiric basis, titrating to a state of parkinsonism or weakness. Clozapine has been used for intractable tardive dystonia[7] and may also be considered in dystonic storm. Intravenous muscle relaxants and sedatives are useful as an intermediate step before skeletal muscle paralysis with pancuronium[2] or general anesthesia using thiopental or propafol.[5] However, as previous reports[2] and the current case indicate, there is a tendency for intractable dystonia to recur on withdrawal of sedating, paralyzing, or anesthetic agents.

As shown in our report, ITB may be effective in the treatment of dystonic storm and may obviate the need for general anesthesia. The technique can be effective in treating intractable dystonia[4] and in fact has been used in cases that would qualify as dystonic storm.[8,9] In an intensive-care unit, ITB titration can be more rapidly performed than is possible in the ward or ambulatory setting. ITB can interrupt the dystonic spasms and, after implantation of a pump, offers the possibility of chronic maintenance therapy,[4] an advantage over intravenous medication. In the initial phase of treating dystonic storm, adjustments in oral medication and the use of a titratable intravenous agent such as midazolam may help facilitate a smooth reduction in dystonia as the ITB dosage is adjusted. In the long-term, ITB therapy is often limited by the development of tolerance,[10] especially in dystonia.[4] Declining efficacy may necessitate an assessment of pump function, dose adjustment, drug holiday, or medication substitution[11]; in chronic refractory cases of generalized dystonia, stereotactic thalamotomy or pallidotomy[12] may be considered, as in the present case. In the acute and life-threatening setting of dystonic storm, however, ITB offers an important, safe, and rapidly acting therapeutic intervention.

Legends to the Videotape

Segment 1: Baseline examination in the intensive-care unit.

Segment 2: One hour after a bolus injection of 50 μg ITB.

Segment 3: Two weeks after ITB pump insertion (900 μg/day ITB dosage).

Segment 4: Home videotape segment 3 months after discharge from the hospital.

Arif Dalvi, MD
Department of Neurology
University of Cincinnati
Cincinnati, Ohio, U.S.A.

Stanley Fahn, MD
Blair Ford, MD
Department of Neurology
Columbia University
New York, NY, U.S.A.

References

1. Fahn S. Concept and classification of dystonia. *Adv Neurol* 1988; 50:1–8.
2. Vaamonde J, Narbona J, Weiser R, Garcia MA, Brannan T, Obeso JA. Dystonic storms: a practical management problem. *Clin Neuropharmacol* 1994;17:344–347.
3. Burke RE, Fahn S, Marsden CD, Bressman SB, Moskowitz C, Friedman J. Validity and reliability of a rating scale for the primary torsion dystonias. *Neurology* 1985;35:73–77.
4. Ford B, Greene P, Louis ED, et al. Use of intrathecal baclofen in the treatment of patients with dystonia. *Arch Neurol* 1996;53: 1241–1246.
5. Jankovic J, Penn AS. Severe dystonia and myoglobinuria. *Neurology* 1982;32:1195–1197.
6. Paret G, Tirosh R, Ben-Zeev B, Vardi A, Brandt N, Barzilay Z. Rhabdomyolysis due to hereditary torsion dystonia. *Pediatr Neurol* 1995;13:83–84.
7. Trugman JM, Leadbetter R, Zallis ME, Burgdorf RO, Wooten GF. Treatment of severe axial tardive dystonia with clozapine: case report and hypothesis. *Mov Disord* 1994;9:441–446.
8. Narayan RK, Loubser MB, Jankovic J, Donovan WH, Bontke CF. Intrathecal baclofen for intractable axial dystonia. *Neurology* 1991; 41:1141–1142.
9. Delhaas EM, Brouwers JRBJ. Intrathecal baclofen overdose: report of 7 events in 5 patients and review of the literature. *Int J Clin Pharmacol Ther* 1991;29:274–280.
10. Akman MN, Loubser PG, Donovan WH, O'Neill ME, Rossi CD. Intrathecal baclofen: does tolerance occur? *Paraplegia* 1993;31: 516–520.
11. Chabal C, Jacobson L, Terman G. Intrathecal fentanyl alleviates spasticity in the presence of tolerance to intrathecal baclofen. *Anesthesiology* 1992;76:312–314.
12. Lozano AM, Kumar R, Gross RE, et al. Globus pallidus internus pallidotomy for generalized dystonia. *Mov Disord* 1997;12:865–870.

Paraneoplastic "Rubral" Tremor: A Case Report

Paraneoplastic cerebellar degeneration is a complication of malignancies characterized by truncal and appendicular ataxia, dysarthria, and nystagmus. The pathologic hallmark of the dis-

A videotape accompanies this article.

Received July 8, 1997; revision received December 29, 1997. Accepted January 5, 1998.

Address correspondence and reprint requests to Dr. Paola Pergami at the Neurological Department, IRCCS C. Mondino, via Palestro 3, 27100 Pavia, Italy.