REVIEW OF INTRATHECAL BACLOFEN THERAPY FOR SPASTIC AND RIGIDITY DISORDERS

S. JOHN OBRINGER
KENNETH M. COFFEY
Mississippi State University

ABSTRACT

Intrathecal baclofen therapy, a treatment for cerebral palsy and other spastic and rigidity disorders, is showing promise as an effective intervention. This article synthesizes both the medical and rehabilitation conceptual literature to update educators and related service providers as to the efficacy of this intervention. Implications for teachers and therapists of students with physical disabilities are put forth.

Cerebral palsy refers to "a group of conditions that affect muscle movement and control or coordination" (Bowe, 2000, p. 107). Of the three major types of cerebral palsy, spasticity is by far the most common, comprising 60% of the affected population (Albright, Barron, Fasick, Polinko, & Janosky, 1993). The essential features of spastic cerebral palsy, also called hypertonia, include stiff, difficult, and uncoordinated movements resulting in possible contractures (Meythaler, Guin-Renfroe, & Hadley, 1999).

Recently, two articles in this journal have reviewed literature concerning treatment and management of individuals with spastic cerebral palsy. These articles addressed for educators and related service providers two of the more promising treatment protocols for the management of spastic cerebral palsy, Botox and Selective Dorsal Rhizotomy (SDR). Technically, there is a trilogy of management options used in most clinics today. The third option is Intrathecal Baclofen (ITB) therapy.

A sizeable number of students with spasticity are receiving ITB therapy at specific centers throughout the country, as the intervention is now in the final stages of investigation and refinement. The purpose of this article is to review the conceptual literature on ITB therapy and synthesize it for special education teachers and related service providers. Additionally, this review will compare ITB therapy to other current treatments for spasticity.

Baclofen, with a trade name of Lioresal, was developed by Novartis Pharmaceuticals and is supplied nationally by Ciba-Geigy (Meythaler, Steers, Tuel, Cross, & Haworth, 1992). It is a highly effective antispasmodic drug which works by blocking the release of neurotransmitters from nerve endings within the spinal cord (Albright, Barry, Painter, & Shulz, 1998; Coffey et al., 1993). It was originally developed solely for oral use and is currently used to treat slight spasticity and other conditions involving muscle spasms. Oral baclofen is produced in 10 mg and 20 mg tablets (Medical Economics Company, 1998). Because oral baclofen crosses the blood-brain barrier poorly, it reaches relatively low concentrations in the spinal fluid. Therefore, the dosages needed for moderate and severe spasticity make oral use an unacceptable option for many individuals (Gormley, 1999; Meythaler, 1997).

An alternative to oral use, the continuous infusion pump-intraspinal catheter system (ITB therapy), is a device for delivering baclofen directly into the cerebral spinal fluid. The tubing or catheter is placed into the lumbar subarachnoid space, allowing the baclofen to be delivered directly to the spinal nerves (Meythaler, McCary, & Hadley, 1997). The pump itself is an inch thick and three inches in diameter (Albright, 1996). It is implanted in the lower right abdomen. Two incisions are made, one for the pump and one for the catheter. The procedure is performed under general anesthesia (Meythaler et al., 1999).

The pump has been approved by the US Food and Drug Administration since June, 1996 (Albright, 1996). There are currently two major manufacturers of implantable pumps suitable for ITB therapy. "At the present time, the SynchroMed pump (Medtronic, Minneapolis, MN) is the only commercially available implantable pump whose rate of infusion can be externally adjusted" (Albright, 1996, p. S32). The pump has a number of advantages over surgical procedures (e.g. SDR). The adjustability of muscle tone reduction is a primary benefit. As the half-life of baclofen is 4 hours, the pump can be adjusted to as many as 10 intervals per day for peak functioning during regularly scheduled activities of daily living (Gilmartin et al., 2000). As noted by Meythaler (1997), "a programmable delivery system for intrathecal baclofen allows the physician to customize dosage for the individual needs of the patient without irreversible consequences" (p. 90). The reservoir supply-

ing the pump must be refilled at a maximum of 90 days (Meythaler et al., 1999). The pump is battery operated with a longevity of 4 to 5 years (Albright, 1996). The reduction of spasticity is typically observable within two hours after administration of baclofen (Albright et al., 1998). The dosage for long-term continuous infusion ranges from 300 mcg. to 800 mcg. per day (Medical Economics Company, 1998). The lowest dose with optimal response is the treatment goal.

ADVERSE EFFECTS

The complication rate for ITB therapy is substantially higher than a surgical alternative. For SDR the complication rate is less than 1% compared to 20% for ITB therapy (Gormley, 1999). Of the 51 patients enrolled in a recent clinical trial, 41 reported one or more adverse events (Gilmartin et al., 2000). The most common adverse events included: hypertonia, seizures, somnolence, headache, nausea, vomiting, dizziness, increased salivation, and constipation. More serious complications included: meningitis, cellulitis, and catheter malfunction (Gormley, 1999). Sudden withdrawal of baclofen can lead to hallucinations and seizures (Meythaler, 1997). Individuals with obsessive compulsive disorder have been reported to experience difficulty with ITB therapy by constantly manipulating their ITB pump soon after placement. This phenomenon has been labeled twiddler's syndrome (Meythaler et al., 1997). In almost all cases, individuals who have experienced adverse effects have continued with the therapy.

RESEARCH/LITERATURE RESULTS

The following are recent findings for the use of ITB therapy with individuals who have spastic and rigidity disorders:

- Meythaler et al.(1992) examined the impact of ITB therapy on individuals with spinal cord spasticity. While the investigators reported an average drop of 2.3 points (from a score of 4.3 to 2.0) on the Modified Ashworth scale (a numeric 1–5 scale that is a clinical measure of spasticity), all individuals required in-patient physical therapy and a 10-day hospital stay. It was further noted that the dosages required to control spasticity significantly increased, almost doubling, within the first year. The researchers speculated that the nervous system builds up a tolerance to baclofen over time.

- Coffey et al. (1993) in a randomized double-blind study found a reduction of spasticity on the Ashworth scale (a numeric 1–4 scale that is a clinical measure of spasticity) of 2.2 points (from a score of 3.9 to 1.7) and a reduction in muscle spasms of 2.1 points (from a score of 3.1 to 1.0). The researchers further found that the dosage of baclofen required to maintain a therapeutic effect increased over time. However, by using a drug holiday of two to four weeks for selected patients, drug tolerance was a manageable phenomenon.
- Albright et al. (1993) treated 37 patients with cerebral palsy with ITB therapy. The experimental intervention indicated the following: (1) muscle tone was significantly decreased in upper and lower extremities, (2) range of motion was appreciably increased in knee extensions, (3) overall upper extremity function was significantly increased, and (4) activities of daily living were substantially improved. The study further indicated no significant improvement in ankle dorsiflexion, hip abduction, and/or position transitions.
- Albright, Barry, Fasick, and Janosky (1995) reported a clinical study comparing 38 pairs of children who had spastic cerebral palsy. The study involved a comparison of upper extremity spasticity at 6 and 12 months after treatment with either ITB therapy or SDR. ITB Therapy 5 Results indicated the following: (1) both treatments were effective in addressing upper extremity spasticity, (2) ITB therapy was probably the treatment of choice for those ambulatory patients who did not have enough strength to walk without support from spasticity (sufficient and necessary muscle tension), and (3) SDR was probably the treatment of choice for those ambulatory patients who had sufficient strength in their lower extremities.
- Steinbok, Daneshvar, Evans, and Kestle (1995) performed a cost analysis on both ITB therapy and SDR in the treatment of two matched groups of children ($n = 9$, $n = 10$) with spastic cerebral palsy. The researchers noted that both ITB therapy and SDR were effective in treating the spasticity associated with cerebral palsy. The analysis conducted with the Canadian dollar indicated the following after one year of treatment: (1) ITB therapy averaged $64,163 per patient, and (2) SDR averaged $16,913 per patient. The researchers further noted that the drug required for ITB therapy would necessitate an ongoing expenditure of approximately $2,000 per year. In addition, a relatively high rate of complications were found in the group that received ITB therapy while no complications were found in the group who received SDR.

- In a counterpoint to the Steinbok et al. (1995) study, Albright (1996) questioned the relevance of a cost comparison between ITB therapy and SDR as these two treatments are indicated for children with different types and etiologies of spasticity. He reported a study which indicated that the ITB therapy was cost effective when considering the high cost of later orthopedic surgery (i.e. to deal with contractures) and long-term physical therapy.
- Albright (1996) also reported that ITB therapy is most appropriate for four distinct groups of individuals with spasticity: (1) children who are ambulatory with poor underlying strength, (2) children who are 16 years of age or older, (3) children who are nonambulatory due to quadriplegia and whose spasticity limits their comfort and endurance, and (4) children who have such severe spasticity that it is difficult for a single caregiver to perform personal hygiene and other basic care.
- In a double blind, randomized, multicenter study of 22 patients over a one year period, Middel et al. (1997) examined the use of ITB therapy for persons who did not respond to oral medication including oral baclofen. The researchers reported that the use of ITB therapy resulted in significant improvement in sleep, mobility, body care, recreation, and generalized movement. However, no change was seen in two measures of psychosocial behavior.
- Meythaler et al. (1997) reported that ITB therapy often resulted in an increase in functional strength due to the reduction of abnormal motor tone. In addition, the investigators strongly suggested avoiding irreversible procedures in the treatment of spasticity.
- In a longitudinal study, Rawicki (1999) found that 17/18 patients benefitted from ITB therapy. Results included a reduction in tone which reduced the need for nursing care and/or improved overall function. On the Modified Ashworth scale, the mean reduction in all patients was two full points (from a score of 5.0 to 3.0).
- Gilmartin et al. (2000) described a protocol to predict the efficacy of ITB therapy by a fairly simple procedure. This procedure identifies appropriate candidates for ITB therapy prior to a much more invasive intervention, surgical implantation of a pump. In a 12-center clinical trial using randomized, double-blind procedures, the researchers identified 51 possible candidates for ITB therapy by injecting 50 mcg., 75 mcg., or, in rare cases, 100 mcg. of baclofen using a standard lumbar puncture. The Ashworth scale was administered before and after the trial injections to document an appropriate reduction in spasticity.

- Stempien and Tsai (2000) surveyed 115 centers who utilized ITB therapy in their treatment of persons with spasticity. They stated that over 90% of center directors reported significant improvements in orthotic wear, sitting tolerance, ambulation endurance, and limb contractures. Other major findings were noted: (1) pump accuracy is decreased as the reservoir approaches the empty point and (2) ITB therapy is not risk free as approximately 10% of treatments required re-operation. Baclofen infusion team members generally include a diverse group of specialists including neurosurgeon, physiatrist, physical therapist, occupational therapist, and speech therapist,
- Van Schaeybroeck et al. (2000) in a double-blind study of 11 individuals with spastic cerebral palsy reached two major conclusions about ITB therapy: (1) swallowing and speech were somewhat improved and (2) individuals with hemiplegia may respond positively without unwanted side effects in their unaffected limbs.

IMPLICATIONS

Special education teachers and related service providers need to be apprised when a student begins ITB therapy. The following modifications or changes must be considered:

Orthotic wear—Because students very well may have reduced spasticity, they may need to be fitted with different, less restrictive orthotic wear;

Physical and occupational therapy—Because physical, and possibly occupational, therapy are needed following ITB therapy, a change in the IEP may well be needed;

Seating and posturing—Some students may be able to sit in a better position with better posture. Classroom seating may need to be modified to accommodate the changes in muscle tone:

Adaptive devices—Some students returning to the classroom may be able to better use accommodative switches, adaptive keyboards, and more sophisticated communication systems due to less spasticity. A new trial of adaptive devices is recommended;

Medication changes—Because some students develop a tolerance for baclofen, the programmable dose may well need to be increased. The special education teacher is in an excellent position to observe changes in muscle tone and alert the parents and physician; and

Consultation—ITB therapy incorporates a programmable pump which may be customized to individual needs. The pump has the ability to change doses of medication up to ten times per day., teachers and therapists may need to establish a firm schedule when specific activities are performed. The pump then can be programmed in anticipation of these daily events (i.e. physical therapy, eating lunch, writing, toileting).

SUMMARY

Returning to the trilogy theme previously mentioned, Albright's (1996) examination of the 100 most recently seen children in the Spasticity Clinic at the Children's Hospital of Pittsburgh, revealed a hierarchy of treatment procedures. The three most common interventions were (1) SDR (n = 30), (2) ITB therapy (n = 21), and (3) Botox (n = 12). This clearly illustrates the importance of ITB therapy and its relative relationship to the interventions examined in this journal previously.

In conclusion, students with cerebral palsy and other disorders causing spasticity now have treatment choices not readily available even twenty-five years ago. Medical practitioners now have options to improve physical function, relieve constant pain or discomfort resulting from spasticity, prevent secondary disorders such as contractures, and add to the quality of life for individuals with spasticity and their caregivers. Like other treatment options previously discussed in this journal, ITB therapy is not a panacea nor does it change the underlying disorder. It is, however, an important treatment tool.

REFERENCES

Albright, A.L. (1996). Intrathecal baclofen in cerebral palsy movement disorders. *Journal of Child Neurology, 11 (Suppl.* 1) S29–S35.

Albright, A.L., Barron, W.B., Fasick, M.P., Polinko, P., & Janosky, J. (1993). Continuous intrathecal baclofen infusion for spasticity of cerebral origin. *Journal of the American Medical Association, 270*(20), 2475–2477.

Albright, A.L., Barry, M..J., Fasick, M.P., & Janosky (1995). Effects of continuous intrathecal Baclofen infusion and selective posterior rhizotomy on upper extremity spasticity. *Pediatric Neurosurgery, 23*, 82–85.

Albright, A.L., Barry, M.J., Painter, M.J., & Shultz, B. (1998). Infusion of intrathecal baclofen for generalized dystonia in cerebral palsy. *Journal of Neurosurgery*, 88, 73–76.

Bowe, F.G. (2000). *Physical, sensory. and health disabilities: An introduction.* Upper Saddle River, NJ: Merrill.

Coffey, R.J., Cahill, D., Steers, W., Park, T.S., Ordia, J., Meythaler, J., Herman, R., Shetter, A.G., Levy, R., Gill, B., Smith, R., Wilberger, J., Loeser, J.D., Chabal, C., Feler, C., Robertson, J.T., Penn, R.D., Clarke, A., Burchiel, K.J., & Leibrock, L.G. (1993). Intrathecal baclofen for intractable spasticity of spinal origin: Results of a long-term multicenter study. *Journal of Neurosurgery, 78,* 226–232.

Gilmartin, R., Bruce, D., Storrs, B.B., Abbott, R., Krach, L., Ward, J., Bloom, K., Brooks, W.H., Johnson, D.L., Madsen, J.R., McLaughlin, J.F., and Nadell, J. (2000). Intrathecal baclofen for management of spastic cerebral palsy: Multicenter trial. *Journal of Child Neurology, 15*(2), 71–77.

Gormley, M.E., Jr. (1999). Management of spasticity in children: Part 2: Oral medications and intrathecal baclofen. *Journal of Head Trauma Rehabilitation, 14(2),* 207–209.

Medical Economics Company, Inc. (1998). *Physicians' desk reference* (52nd ed.). Oradell, NJ: Author.

Meythaler, J.M. (1997). Intrathecal baclofen for spastic hypertonia in brain injury. *Journal of Head Trauma Rehabilitation, 12*(1), 87–90.

Meythaler, J.M., Guin-Renfroe, S., & Hadley, M.N. (1999). Continuously infused intrathecal baclofen for spastic/dystonic hemiplegia. *American Journal of Physical Medicine and Rehabilitation, 78(3),* 247–254.

Meythaler, J.M., McCary, A., & Hadley, M.N. (1997). Prospective assessment of continuous intrathecal infusion of baclofen for spasticity caused by acquired brain injury: A preliminary report. *Journal of Neurosurgery, 87,* 415–419.

Meythaler, J.M., Steers, W.D., Tuel, S.M., Cross, L.L., & Haworth, C.S. (1992). Continuous intrathecal baclofen in spinal cord spasticity: A prospective study. *American Journal of Physical Medicine and Rehabilitation, 71(6),* 321–327.

Middel, B., Kuipers-Upmeijer, H., Bouma, J., Staal, M., Oenema, D., Postma, T., Terpstra, S., & Stewart, R. (1997). Effect of intrathecal baclofen delivered by an implanted programmable pump on health related quality of life in patients with severe spasticity. *Journal of Neurology, Neurosurgery, and Psychiatry, 63,* 204–209.

Rawicki, B. (1999). Treatment of cerebral origin spasticity with continuous intrathecal baclofen delivered via an implantable pump: Long-term follow-up review of 18 patients. *Journal of Neurosurgery, 91,* 733–736.

Steinbok, P., Daneshvar, H., Evans, D., & Kestle, J.R.W. (1995). Cost analysis of continuous intrathecal baclofen versus selective functional poste-

rior rhizotomy in the treatment of spastic quadriplegia associated with cerebral palsy. *Pediatric Neurosurgery, 22,* 255–265.

Stempien, L., & Tsai, T. (2000). Intrathecal baclofen pump use for spasticity: A clinical study. *American Journal of Physical Medicine and Rehabilitation, 79*(6), 536–541.

Van Schaeybroeck, P., Nuttin, B., Lagae, L., Schrijvers, E., Borghgraef, C., & Feys, P. (2000). Intrathecal baclofen for intractable cerebral spasticity: A prospective placebo-controlled, double-blind study. *Neurosurgery, 46*(3), 603–612.

Address correspondence to S. John Obringer, Department of Curriculum and Instruction, Box 9705, Mississippi State University, Mississippi State, MS 39762. 662-325-7114; email sjol@ra.msstate.edu