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SPECIAL ANNUAL ISSUE

Intrathecal baclofen for childhood hypertonia

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Abstract
Introduction Intrathecal baclofen was first introduced in 1985 to manage childhood hypertonia. There has been an evolution in thought as to how candidates should be identified and what forms of hypertonia respond to this treatment.
Purpose This manuscript reviews the pharmacology of the drug, the assessment of candidates, the implantation of the infusion pump, and the usual doses delivered. Side effects and complications are reviewed as are outcomes.

Keywords Intrathecal baclofen · CSF infusion pump · Childhood hypertonia · Spasticity · Dystonia · Mixed hypertonia

Introduction

Hypertonia may be defined as increased muscle tone, either at rest or during movement, and does not connote either a hyperkinetic or hypokinetic movement disorder. The most common causes of hypertonia in children are spasticity (a velocity-dependent, increased resistance to passive muscle stretch) and dystonia (a hyperkinetic movement disorder characterized by sustained muscle contractions that cause twisting and abnormal postures). Twenty years ago, pediatric neurosurgeons had few options to treat either spasticity or dystonia but now use rhizotomies, botulinum toxin injections, and intrathecal baclofen (ITB) for hundreds of children that previously would have been untreated. Although ITB was used initially to treat spasticity of spinal origin and then spasticity of cerebral origin, it is now used to treat spasticity of virtually any etiology, dystonia, and hypertonia caused by such varied disorders as near-drowning, tetanus, and Rett's syndrome.

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Pharmacology

Baclofen, 4-amino-3-(4-chlorophenyl)-butanoic acid, was synthesized as an agonist of the neuro-inhibitory transmitter, γ-aminobutyric acid (GABA), adding a chloro-phenyl group to GABA to make it more lipophilic. After oral administration, baclofen is absorbed rapidly from the gastrointestinal tract, but it crosses the blood–brain barrier poorly. Oral administration of 30–90 mg/day is associated with plasma baclofen levels of 68–650 ng/ml and corresponding cerebrospinal fluid (CSF) levels of 12–95 ng/ml [28]. After intrathecal infusion of 400 μg of baclofen/day, CSF levels are typically ~380 ng/ml [34]. The half-life of ITB is 4–5 h; thus, it takes approximately 24 h to reach steady state after a dose adjustment, and ITB doses should not be adjusted more than once a day. Baclofen is cleared with CSF, at CSF clearance rates. After intrathecal infusions, plasma baclofen levels are virtually undectable [3].

Baclofen's site of action in treating spasticity is thought to be in the superficial layers of the spinal cord, Rexed layers I–II, where it inhibits the release of glutamate. Its site of action when treating dystonia is unknown but may be at

a cortical level, suppressing the excessively stimulated supplementary motor and premotor cortex.

Indications for ITB

When treatment with ITB is considered, it is essential that the treatment goal(s) be carefully considered because "reducing spasticity" per se is not a treatment goal. Some common goals include (a) to facilitate care of the child, (b) to diminish discomfort (and improve sleep), (c) to retard or prevent the progression of musculoskeletal contractures, and (d) to improve function, such as increasing upper extremity range of motion with less wrist/finger flexion and improving use of a communication device. The treatment goals need to be understood by the physician, patient, and parent. It is not uncommon for the physician to recommend ITB to diminish the progression of contractures but for the patient to infer that it is to improve function. Even when goals are carefully enumerated and agreed to by the patient, there are often unspoken hopes of unrealistic improvements that lead to postoperative disappointment and, at times, to depression, particularly in young adults.

When treating pediatric hypertonia, ITB is almost always administered via a programmable pump—rather than a fixed-rate infusion pump—because of the multiple dose adjustments that are needed. Currently, available pumps are sufficiently small that there are no longer any size or weight restrictions; I have inserted a pump into a 9-kg, 9-month-old child, but the need for ITB in children of that age and size is obviously rare.

Spasticity

ITB is used most frequently to treat spastic quadriparesis, whether secondary to cerebral palsy (CP), traumatic brain injury, or other causes. That spasticity is moderate to severe, with mean Ashworth scores of 3–4 of 5 in the lower extremities and 2–3 of 5 in the upper extremities, which has not responded sufficiently to oral medications (e.g., baclofen, dantrolene, tizanidine) and/or botulinum toxins and is increasing the burden of care or causing musculoskeletal deformity, disability, or discomfort. Post-traumatic spasticity may develop within a few weeks after serious head injury. In such children, if oral medications and botulinum toxin injections do not provide adequate benefit, ITB is generally highly effective; it can be used within a few weeks after injury, far earlier than the 1-year guideline that was recommended several years ago.

ITB is probably the treatment of choice to treat the spastic diplegia associated with familial spastic paraparesis, and it is used occasionally in an older adolescent or young adult with spastic diplegia associated with CP for whom a lumbar rhizotomy is thought to have an unacceptable risk of causing permanent hypotonia.

Children with spastic hemiparesis may be treated with ITB if their spasticity is impeding gait or causing progressive contractures. In such cases, ITB appears to diminish spasticity in the affected side without altering tone on the normal side [38]. Focal spasticity can be effectively treated with peripheral selective fasiculotomies and is almost never treated with ITB.

Dystonia

Only moderately severe or severe dystonia are treated with ITB. Before considering ITB, most dystonic children are treated (unsuccessfully) with oral medications such as baclofen, trihexyphenidyl, or levo-dopa but not always. Because severe generalized secondary dystonia responds relatively poorly to oral medications, some children with severe, disabling dystonia are recommended to have ITB without prior oral medications.

Treatment goals when using ITB for dystonia are somewhat different than when treating spasticity. In dystonia, the most common treatment goals are increased comfort and increased ease of care; dystonic arching of the trunk and neck is often painful, makes care-giving more difficult, and results in broken head or foot rests on wheelchairs. ITB is often given to treat the dystonia causing such posturing. Improved function occurs at times after ITB for dystonia, but improving function is an uncommon indication for treatment. Musculoskeletal contractures occur far less often with dystonia than with spasticity and—by themselves—are rarely indications for ITB treatment.

ITB has been used to treat secondary dystonia (that secondary to a structural brain anomaly, e.g. CP, traumatic brain injury) or heredo-degenerative dystonia (that associated with a heredo-degerative disorder, e.g., Wilson's disease, pantothenate kinase deficiency, glutatic aciduria), more often than to treat primary dystonia (that unassociated with any known abnormality other than perhaps genetic mutations). Secondary dystonia comprises 80–90% of pediatric dystonia cases. Secondary dystonia may be graded with the Barry–Albright Dystonia (BAD) scale, a validated scale that grades dystonia in eight body regions on a 0-to-4 scale; most candidates for ITB have scores of 12 or greater [9]. Primary dystonia is graded with the Burke–Fahn–Marsden scale, a 0-to-120 scale that grades dystonia in nine body regions; too few patients with primary dystonia have been treated with ITB to comment on their usual preoperative scores [11]. Primary generalized dystonia is probably treated more effectively with deep-brain stimulation of the internal globus pallidus than by ITB.

ITB is used most often to treat generalized dystonia (affecting the face, neck, trunk, and upper and lower

extremities). It is occasionally used to treat hemidystonia but rarely to treat segmental or focal dystonia. Generalized dystonia occasionally worsens abruptly and severely as "dystonic storms"; they are typically improved by ITB.

Mixed spasticity–dystonia

ITB usually improves both spasticity and secondary dystonia, and the children—most of whom have CP—with mixed spasticity and dystonia are candidates for ITB, whether the proportion of the movement disorders is 30/70 or 70/30.

Miscellaneous forms of hypertonia

ITB has been used to treat generalized athetosis and chorea, but the numbers treated are too small to know how much of an indication those forms of hypertonia are. There are multiple reports of ITB being used to treat tetanus and case reports of its use for spasticity associated with adrenoleukodystrophy in a child and for dystonia associated with reflex sympathetic dystrophy and with Rett's syndrome in adults [27].

Screening

For spasticity

When ITB therapy in children began around 1988, its effectiveness in decreasing their spasticity was unknown, and screening lumbar bolus doses of baclofen were always given to determine the effects before a pump was inserted. Doses of 50 μg were given most often, although 25 μg was used in some small children and 100 μg in large ones and in those who did not respond to 50 μg. After the screening boluses, spasticity in the lower extremities was shown to diminish at 2 h, to hit its nadir at 4 h, and to return to baseline within 8–12 h [1]. Response to the bolus dose was evaluated by grading Ashworth scores in the lower extremities. A one-point decrease in the mean Ashworth score was considered to be clinically significant when evaluating cerebral spasticity and a two-point decrease or greater when evaluating spinal spasticity.

The purpose of a screening trial was considered to be only this: to determine if ITB decreased spasticity in the lower extremities. It was not done to see if gait improved; it often worsened because legs became hypotonic. Since then, hundreds of children have received screening doses, and it has become apparent that spasticity almost always responds, so frequently, in fact, that we and several other large centers in the USA no longer do a screening trial for spasticity. Some clinicians continue to perform screening trials, however, and evaluate effects with the modified Tardieu scale or changes in range of motion. A few clinicians insert a percutaneous lumbar catheter and perform one or more screening injections through it.

If screening trials are done, it is best to use topical anesthetics rather than sedatives for the lumbar puncture, to avoid the residual sedation that confounds interpretation in some children. The lumbar punctures will occasionally cause headache, nausea, and vomiting, which also confound interpretation of the results.

For dystonia

Screening dystonia responses to dystonia is more difficult than screening spasticity responses. The first patient we treated with ITB had double-blinded injections of placebo or baclofen, 25, 50, and 100 μg each day for 6 days. We could observe no difference in her dystonia, but she was able to correctly determine every day whether she had received baclofen or placebo. At the encouraging of her neurologist, we inserted a pump, and after 2 days of continuous ITB infusion, her dystonia improved dramatically.

After screening more than 200 dystonic patients with ITB, it seems that younger, smaller patients, e.g., less than 8 years and less than 25 kg, may be evaluated by a lumbar bolus dose of baclofen, either 50, 75, or 100 μg, but for older and larger children, a catheter continuous-infusion trial is a better way to screen. For that, an intrathecal catheter is inserted in the operating room and connected to a subcutaneous infusion port, e.g., a Bard port or Mediport (Fig. 1). A Huber needle is inserted percutaneously into the port and connected to a microinfusion catheter on the bedside for increasing baclofen doses. We begin with 200 μg/day and increase by either 50 μg every 8 h or 75 μg every 12 h until (a) a clinically significant response (25% decrease in BAD dystonia scores), (b) an unacceptable adverse side effect (e.g., lethargy), or (c) no significant response at 900 μg/day. Approximately 90% of children respond to the infusion screening [4].

We no longer screen children with severe generalized secondary dystonia because of the high response rate and because they have no other good therapeutic option if they have failed oral medications (deep-brain stimulation is an option, but its effectiveness for secondary dystonia is significantly less than for primary dystonia).

For other forms of hypertonia

For other forms of dystonia, such as severe, generalized chorea or athetosis, where the response to ITB is unclear, we evaluate response with an infusion screening trial as described above.

Some children have headache, vomiting, and postural hypotension for 2–3 days after insertion of the screening catheter. The screening infusion should be delayed until

those symptoms subside. Aseptic meningitis has been reported in two patients receiving a screening baclofen bolus, but that is exceedingly rare [10].

Pump implantation

Operative details were recently described [7]. Pumps are inserted under general anesthesia, usually with the patient in the left lateral decubitus position. The presence of a gastrostomy in the left upper quadrant does not appear to increase the infection rate. After intravenous prophylactic antibiotics and meticulous skin preparation, an oblique incision is made about a finger breadth below the costal margin, and a pocket is created for the pump. For thin or small patients, a subfascial pocket provides better pump positioning, wound healing, and cosmesis (Fig. 2). Although fixed rate and adjustable rate pumps are commercially available, when treating a child with cerebral spasticity or dystonia, adjustable rate pumps are used in nearly every case. In the USA, the Synchromed II pump (Medtronic, Minneapolis, MN) with a 40-ml reservoir is used in approximately 80% of the cases and the 20-ml pump in the remainder.

Posteriorly, a 1-in. paramedian incision is made at about the L3–L4 level, and a Touhy needle is inserted obliquely cephalad into the thecal space under fluoroscopic guidance; then, the intrathecal catheter is inserted through the needle and advanced cephalad. Grabb et al. [25] reported that a catheter tip at T6–T7 was associated with better control of upper extremity spasticity than catheter tips at the traditional T11–T12 level, without associated symptoms. McCall and MacDonald [33] placed catheter tips in cervical locations in 23 patients and observed no increase in complications. We currently position catheter tips at C5–T2 for spastic quadriparesis and at C1–C4 for generalized dystonia. I have inserted intraventricular catheters in two children, with no adverse effects [8].

If the child has had—or is going to have—a spine fusion, the catheter is tunneled paraspinally cephalad and inserted into the thecal sac via a small laminectomy (Fig. 3). Liu and Walker [32] used a similar technique to insert catheters in three patients with previous fusions and directed the catheters caudal to the dural insertion site. A purse-string suture can be placed around the catheter where it enters the dura to decrease the risk of a CSF leak. The rate of CSF leaks with that technique is exceedingly small. The alternative technique of drilling through the spine fusion mass is distinctly more difficult and seems to have a higher rate of CSF leaks.

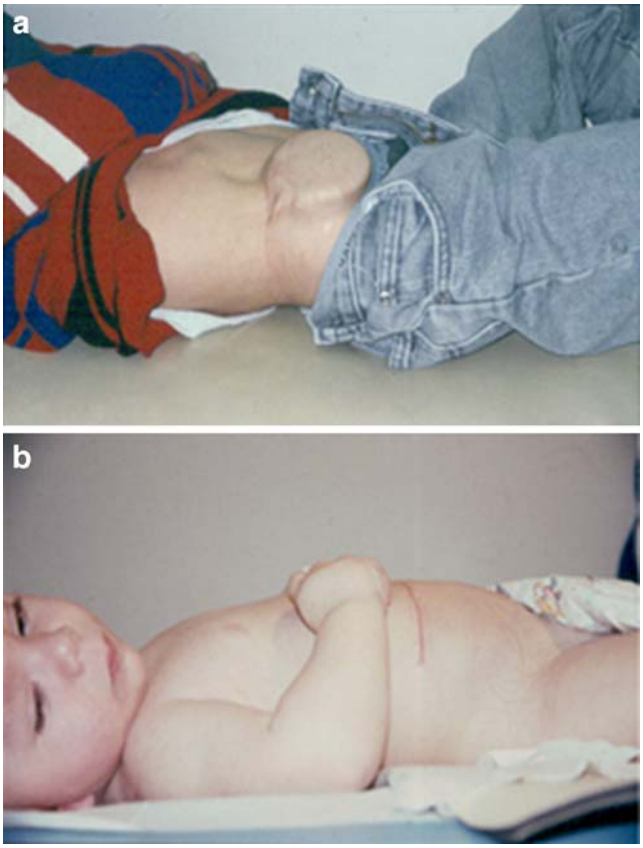

Fig. 2 Photographs comparing the appearance of subcutaneous (**a**) and subfascial (**b**) pump placements

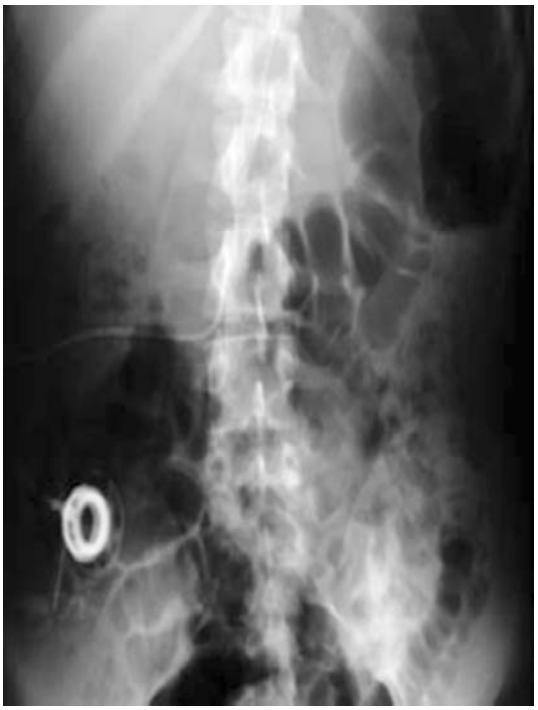

Fig. 1 Radiograph demonstrating an intrathecal catheter and subcutaneous infusion port for a continuous baclofen infusion trial

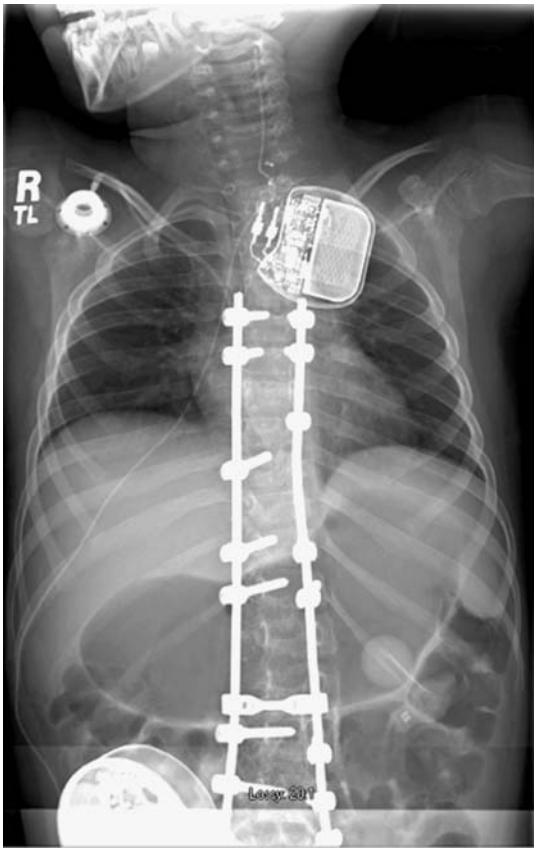

Fig. 3 Radiograph of an intrathecal catheter inserted in a child with a spine fusion. The catheter can be seen entering the thecal sac at the base of the neck with the tip resting in the upper cervical region. The child also has an implanted nerve stimulator

Single-piece catheters appear to be associated with fewer complications, particularly disconnection, than two-piece catheters.

ITB dosing

In the postoperative period, ITB dosing begins in a continuous infusion mode, customarily at 100 μg/day for spasticity and 200 μg/day for dystonia. Spasticity doses are typically increased by 5–20%/day for the first few days until spasticity is reduced to approximately the desired extent; then, the fine tuning is done at 2, 4, and 8 weeks and whenever children return for a pump refill. Doses are titrated to optimize tone and function. At 1 year after pump implantation, the mean ITB dose when treating spasticity is ~300 μg/day [5].

ITB doses to treat dystonia are increased more rapidly postoperatively, often 50–100 μg/day until some effect is noted, and then the rate of dose increase is slowed. At 1 year postimplant, the mean ITB dose is ~600 μg/day, but doses of 600–1,000 are not unusual. If the desired results are not evident by 1,000 μg/day, a trial of intermittent bolus dosing is appropriate, e.g., giving 200 μg bolus doses every 4 h with the pump running at a basal rate between boluses. When treating either spasticity or dystonia, doses are increased at times to 1,500 μg/day, but above that level, significant additional benefit is unlikely to be seen.

Sometimes, an increase ITB dose after a period of clinical stability may be associated with less response. Cooper and Ridley [16] reported two cases in which dose reductions were associated with better clinical response. Most often, however, the lack of response after a dose increase is a reflection of a catheter malfunction (disconnection, leak, obstruction, etc.).

Clinicians should beware of ITB "compounding," the practice whereby local pharmacists make up baclofen solutions in varying concentrations. There is a reason why ITB is not commercially available in concentrations greater than 2,000 μg/ml: It cannot be reliably mixed in high concentrations, particularly those greater than 3,000 μg/ml. Such high mixtures may result in precipitation of baclofen so that the amount infused is less than expected, and the pump warranty is invalidated.

Trouble-shooting

If a child has an excellent response to a screening bolus and then "looses" the response or becomes "refractory" to ITB, a system malfunction should be suspected. The site of the problem is almost never the pump. In troubleshooting, the first maneuver is usually an X-ray to determine if there is a catheter disconnection or catheter migration out of the thecal sac. If the X-ray is normal, the child is taken to the radiology department, and a rotor study is performed to confirm rotation of the roller mechanism within the pump. If that is normal, a 24-ga Huber needle is inserted into the side port of the pump, and CSF is aspirated through the intrathecal catheter. If no CSF can be aspirated, the catheter needs to be surgically explored. If CSF can be withdrawn, contrast material such as Iohexal is injected through the catheter under fluoroscopic guidance. Sites of leakage or disconnection may be identified. If there is no flow from the catheter, it is important to remember that injection through the catheter may infuse the baclofen within the catheter into the CSF and cause a minor overdose; for example, if the catheter volume is 1.5 ml and the baclofen concentration is 2,000 μg/ml, approximately 300 μg will be injected as a bolus, which may cause lethargy and hypotonia for a few hours. Unless the contrast injection shows excellent distribution of dye within the subarrachnoid space, the child should be taken to the CT scanner and a scan obtained around the catheter tip. Catheter tips in the subdural space can only be detected by this technique (Fig. 4). If no malfunction can be found but there is still a serious suspicion that there is a malfunction, a lumbar puncture and bolus baclofen injection

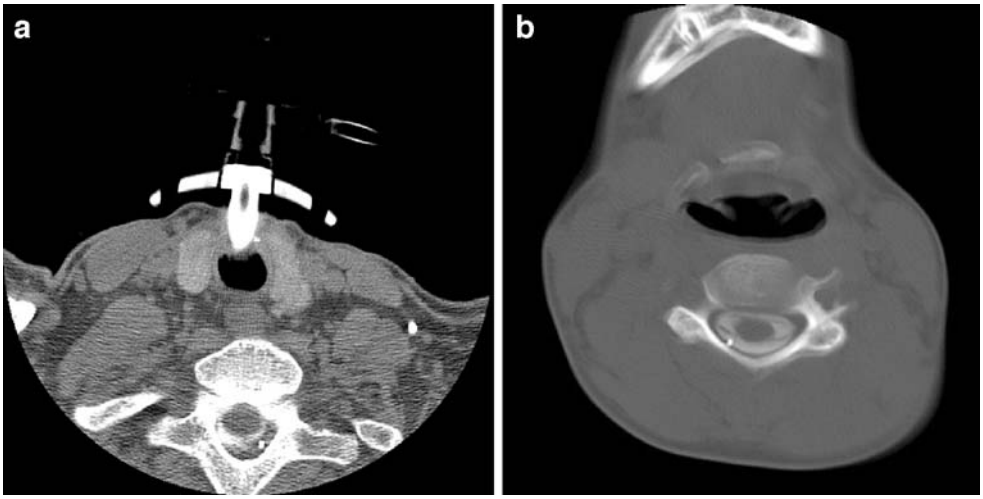

Fig. 4 CT myelograms demonstrating subdural (**a**) and subarrachnoid (**b**) catheters

usually answers the question; if the child responds to the bolus dose, the presence of a problem somewhere in the system is confirmed.

Side effects and complications

The most common acute side effect associated with ITB therapy is urinary retention, which usually may require intermittent catheterization for 2–3 days but always resolves. The most common chronic side effect is probably constipation, which is present preoperatively in many children with CP and may worsen postoperatively. Reflux appears to worsen in some children. If the ITB dose is too high, alertness will be dulled. Allergic reactions to ITB probably do not occur.

The relationship between ITB and seizures is unclear. ITB was synthesized as an anticonvulsant, and in its initial trials in adults with seizures, it apparently had no significant anticonvulsant activity. There are a few case reports of seizures worsening or developing after ITB, but in the largest series addressing the question, there was no significant correlation. Buonaguro et al. [15] evaluated 150 children treated with ITB, 40% of whom had seizures pre-ITB. In that 40%, seizures decreased after ITB in 13.3% and worsened in two. Of the 60% who did not have seizures pre-ITB, one child developed seizures afterward.

Complications occur more commonly in children with ITB than in adults [43]. Gooch et al. [23] reported complications in 100 consecutive children with 117 pumps. Twenty-four patients had 48 complications; the most common was catheter disconnection at the pump (9%), which rarely occurs with current catheters.

The three most common complications after ITB are infections, CSF leaks, and catheter problems. Infections develop 2–6 weeks postoperatively, most often because of *Staphylococcus aureus*, and may develop in the pump pocket, the CSF, or both [45]. If both are infected, the entire system is usually removed, intravenous antibiotics are given for 2 weeks, and the pump can be reinserted 6–8 weeks later. If only the pump site is infected, the pump can be removed in the operating room, the wound debrided and irrigated (e.g., 100 ppm iodine solution), and the pump replaced, with postoperative intravenous antibiotics. The success rate of that treatment is perhaps 50%. Boviatsis et al. [14] reported successfully treating pump infections with intrapocket antibiotics, in three patients, without any operative intervention. Because *S. aureus* is the most common infecting organism, consideration can be given to preoperative cultures to identify *Staphylococcus* carriers and to administering intranasal antistaphylococcal gels.

CSF leaks occur more commonly in children with ITB than in adults, ~12 vs 3% [7]. The increased leak rate may be related to several factors, including malnutrition, thin tissues, and occult hydrocephalus [6]. Subcutaneous CSF accumulations in either the lumbar region or around the pump are treated with bed rest, blood patches, lumbar drains, and operative closures.

Common catheter problems include disconnection, migration, and leaks. Microfracture of a catheter can cause intermittent under- and overdosage and cannot be seen on dye studies, only detected at operation [18]. Intermittent under- and overdosing can also apparently occur if the catheter tip perforates the arachnoid and lies in the subdural space. Changes in position may allow accumulated subdural baclofen to re-enter the subarrachnoid space and result in a transient overdose. A granuloma at the catheter tip has been reported in only one adult with ITB and never in a child [36].

Changes in personality occur after chronic ITB but are rare, less than 1% in my experience.

The relationship between ITB and scoliosis has also been debated, partly because candidates for ITB usually

have either spastic quadriparesis or dystonic CP, conditions in which scoliosis often develops. Sansone et al. [40] reported rapid progression of scoliosis in four children after pump insertion.

Overdose

Most overdoses are iatrogenic and occur during transitioning either to a higher baclofen concentration or to a catheter dye study. Mild overdoses cause only lethargy and hypotonia, generally do not need to be treated, and resolve within 8–12 h as the baclofen is metabolized. Moderate overdoses cause obtundation and bradypnea; they can be treated by intravenous scopolamine (usually with only transient improvement), but the mainstay is assisted ventilation if necessary to maintain adequate oxygenation. Severe overdoses cause coma and respiratory arrest and require intubation and assisted ventilation until the baclofen is metabolized. Consideration can be given to doing a lumbar puncture, withdrawing 20 ml CSF, and then barbitaging 20 ml normal saline two to three times to speed the recovery. ITB is not neuro-toxic, and there are no neurological consequences of overdoses, only the psychological ones.

Withdrawal

The hallmark clinical symptoms of ITB withdrawal are generalized itching, increased spasticity, and agitation. Symptoms can occur abruptly and be severe or gradually and go undetected. Mild overdoses are treated with oral baclofen, e.g., 20 mg every 6 h. Moderate withdrawal is treated by larger oral baclofen doses, e.g., 20 mg every 2–4 h plus oral benzodiazepine (e.g., clonazepam or diazepam). Severe withdrawal can be associated with delirium, hyperthermia, psychosis, seizures, and multisystem organ failure and is a medical emergency requiring immediate intervention. The mainstay of treatment is infusion of ITB, either by emergency surgery to restore infusion via the implanted pump or insertion of a lumber catheter through which a bolus of baclofen can be given, or baclofen can be infused via an external microinfusion pump [46]. At least one death has been reported worldwide from ITB withdrawal.

Tolerance

Tolerance has been reported in patients receiving ITB for both spasticity and dystonia; however, I am not sure tolerance ever occurs in spasticity and think it is probably rare (<5%) in dystonia. We now know that many children who were thought to have tolerance in fact have system malfunctions, primarily subdural catheter tip positions, and their responsiveness resumes once the catheter is repositioned.

Outcomes

Historically, the first reported use of ITB in children was by Dralle et al. [19] in 1985, which used it to successfully treat hypertonicity after near drowning in a 4-year-old child. Muller [35] used ITB to treat 20 children in a larger study and reported significant improvements in spasticity. We reported a double-blind study of bolus baclofen doses in children with cerebral spasticity in 1991 and found that spasticity in the lower extremities was significantly decreased 4 h after the injection [1]. In 1993, we reported a prospective nonrandomized trial in 27 children with cerebral spasticity and found that their spasticity was continuously improved by chronic ITB infusion [2]. More recently, we reported long-term effects of ITB in 68 children and young adult with a mean follow-up of 70 months [5]. Spasticity in the upper and lower extremities was significantly decreased up to 10 years. The dosage of ITB increased to a mean of 300 μg/day at 2 years and then remained relatively stable thereafter.

In 2000, Butler and Campbell [12] reviewed the published literature about the use of ITB for spasticity and dystonia. They concluded that spasticity was significantly improved and function was probably improved. Krach et al. [30] measured GMFMs in 31 subjects 4–29 years before and 1 year after ITB and found significant improvements in children less than 8 years old (mean change 4.1) and in those 8–18 (mean change 3.7) and also in subjects with CP classes 2 and 5. In another study, Krach et al. [31] interviewed 100 subjects—88 with CP—about their changes in function and caregiver assistance after ITB. Subjects reported improved positioning (69%), transfers (58%), dressing (69%), toileting/hygiene (51%), decreased startle (54%), improved sleep (43%), and improved comfort (53%). Twenty-two subjects had 32 therapy related events; importantly, 88% of subjects said they would undergo the procedure again.

Bjornson et al. [13] studied oral motor, communication, and nutritional status in 30 children receiving ITB. Speech improved in ten (5 in Gross Motor Function Classification System [GMFCS] level V) and worsened in two. Use of assistive technology improved in six (5 level V) and appetite improved in ten and worsened in four. Self-feeding improved in nine and worsened in two, saliva control improved in ten and worsened in eight, and stool frequency improved in eight and worsened in 14.

Care provider outcomes were evaluated by Gooch et al. [24] in 80 patients, mean age 11 years, who were treated with ITB for 1 year; all were at GMFCS levels IV or V. All care providers reported improvement in scores on the Caregiver Questionnaire. Goals chosen preoperatively were achieved as follows: decreased pain (91%), prevention of worsening deformity (91%), and improved ease of care

(88%). Range of motion was maintained in the lower extremities in 43 of 51 and lost in eight (16%). None had rapid progression of scoliosis. Ninety-five percent of the providers would have the operation again.

We reported effects of ITB on gait in 24 patients who were ambulatory to some extent pre-ITB [21]. Ambulation was graded on four functional levels: community, household, nonfunctional, and nonambulatory. The mean follow-up was 52 months. The level of ambulation improved by one level in 9 patients, did not change in 12, and was worse in three patients. Gait was thought to be improved by patients or families in 20 of 24 cases, to be unchanged in two, and to be worse in two. Dan et al. [17] reported improvement in gait control in patients with hereditary spastic paraparesis after ITB.

Post-traumatic spasticity

Turner [42] treated six children 1–14 years of age with ITB at less than 1 year after their brain injury. Preoperatively, all also had autonomic dysfunction and severe autonomic storms. Postoperatively, all had decreased spasticity of at least two points on the Ashworth scale, and their storms ceased. Francois [20] used ITB to treat three patients within 25 days after TBI and observed improvement in tone and autonomic disorders.

Orthopedic outcomes

Krach et al. [29] studied hip subluxation in 33 subjects 4–31 years old. Postoperatively, one of three had an increase migration percentage of 5% or more, and 12% had a decrease of 5% or more. No deterioration or an improvement of their migration percentage class during the year of ITB therapy was observed in 90.9%. Migration changes did not correlate with the subjects' age or severity of CP. We observed that children needed less orthopedic surgery after ITB [22].

Dystonia effects

Narayan et al. [37] were the first to report the use of ITB in pediatric dystonia; they used it to successfully treat an 18-year-old adolescent with dystonic CP. In 2001, we reported the use of ITB in 89 children with severe generalized dystonia; approximately 70% had CP, and the remainder had a variety of disorders, particularly heredo-degenerative disorders [4]. In that study, dystonia improved in 90% of cases, ease of care and quality of life improved in 85%, and speech/function improved in one third.

Walker et al. [44] used ITB to treat 14 patients with primary or secondary dystonia, grading their responses with a blinded rater. Five patients had improved symptoms, although only two had a clear clinical benefit. Etiology of dystonia did not correlate with response. Grande et al. [26] used ITB to treat a 26-year-old patient whose primary dystonia had began at age 11 and had been refractory to oral medications and a thalamotomy. They observed substantial improvement at a dose of 450 μg/day.

Tetanus

Santos et al. [41] used ITB via an external catheter to treat 22 patients with generalized tetanus and achieved control of symptoms in 21.

Cost–benefit

No high quality cost–benefit studies have been reported for ITB in children. In adults, Sampson et al. [39] evaluated the effects of ITB on function and quality-of-life measures in patients with severe spasticity. Outcomes were related to quality-of-life scores to estimate potential gains in quality-adjusted life years. They concluded that ITB has an acceptable cost–benefit ratio compared with other interventions.

Conclusions

ITB is an effective treatment for children with hypertonicity secondary to spasticity and dystonia. It is used most commonly for those with spastic quadriparesis or generalized dystonia and, for those children, often results in dramatic improvements in quality of life and in easier care. Complications occur frequently. In spite of the complications, most families and patients ask to continue ITB therapy at the end of pump battery life. Advantages of ITB include the improvement in upper and lower extremities, the ability to vary the amount of tone reduction, and the fact that the therapy is reversible. Disadvantages of ITB include the pump costs, the complications, and the fact that the therapy is not definitive (continued refills/pump replacements are needed over time).

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