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ORIGINAL PAPER

Understanding the reasons for delayed referral for intrathecal baclofen therapy in pediatric patients with severe spasticity

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Abstract
Objective Intrathecal baclofen therapy (ITB) has been used in the treatment of spasticity and dystonia. In our pediatric movement disorder clinic, we noted a delay in referral of patients for consideration of ITB. Often, only after years of failed medical therapy, a baclofen pump is considered. This study attempts to investigate the prevalence, length and causes of the delay.
Methods A retrospective, outcome analysis was performed. We conducted a survey of 30 pediatric patients who received baclofen pumps between the ages of 5 and 23. Patients were divided into two groups (before and after ITB approval by the US Food and Drug Administration in 1996) (FDA/Center for Drug Evaluation and Research, 2014; Ridley and Rawlins, J Neurosci Nurs 38:72–82, 2006; Medical Advisory Secretariat, Ont Health Technol Assess Ser 5:1–93, 2005). Information was collected regarding their onset of spasticity, attempted treatments, pump referral, satisfaction, and resulting change in the quality of life.
Results There was a delay in referral in most cases investigated. Average time to baclofen pump implantation, after initial onset of spasticity, was 5.14 years (group A) and 11.7 years (group B). Out of the subjects who reported diminished effects or no effect of pharmacological treatment, 93 % of these respondents reported that ITB had a dramatic long-lasting effect on their spasticity. Of 30 patients, 28 reported effectiveness of ITB, and 26 of 30 subjects reported an improved quality and ease of life.
Conclusion Despite the limitations of this subjective retrospective analysis of outcomes and delay in referral, the opinions of the parents and caregivers should be considered. Earlier referral for ITB therapy may better treat severe spasticity in pediatric patients.

Keywords Anoxic brain injury · Botulinum toxin · Cerebral palsy · Contractures · Delay in treatment · Hypertonia · Hypoxic-ischemic encephalopathy · Intrathecal baclofen · ITB · Periventricular leukomalacia · Spasticity

Introduction

Patients who have experienced impairment to the central nervous system including the motor and premotor cortex, the basal ganglia, or the spinal cord can develop spasticity and dystonia. Most of our movement disorder clinic patients are children who have suffered hypoxic-ischemic injury and have permanent static encephalopathy and periventricular leukomalacia (PVL). Damage to the upper motor neuron function results in the absence of inhibitory influence on alpha motor neurons [1]. This disrupts the normal communication between alpha motor neurons and afferent muscle nerve fibers [1]. The imbalance of excitatory and inhibitory inputs leads to increased muscle activity resulting in spasticity. Hypoxic-ischemic encephalopathy (HIE) often results in cerebral palsy with spastic diplegia or spastic quadriparesis. Spasticity is seen in patients who have experienced brain or spinal cord injury from other causes as well, such as stroke, trauma, infection, toxic-metabolic disorders, and anoxic brain injury.

Intrathecal baclofen therapy has been accepted as an effective means of treating spasticity in these patients. ITB requires the surgical implantation of a baclofen pump subcutaneously or below the fascia. Baclofen is a gamma amino butyric acid (GABA) agonist, which may act at the level of the spinal cord to reduce spasticity [2]. Baclofen inhibits afferent nerve fibers by binding to $GABA_B$ receptors in the dorsal horn of the

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spinal cord, and causes presynaptic inhibition of alpha motor neurons [3]. This restores proper function between the alpha motor neurons and muscle afferents, reducing abnormal muscle activity [4]. Baclofen in the cerebrospinal fluid may also affect periventricular areas in the brain if the baclofen concentration is high enough in the ventricles. Intra-ventricular baclofen therapy (IVB) has been used by Albright and others for the treatment of spasticity and dystonia [5].

Pharmacological, non-invasive treatment of spastic, dystonic, and mixed-movement types of movement disorder has been reported to be rather ineffective in most cases [6]. This includes botulinum toxin injections, alcohol blocks, oral baclofen as well as other oral medications. Oral baclofen is not absorbed well by the cerebrospinal fluid, so treatment requires high doses with sedative side effects [1]. When given intrathecally, baclofen can be given at just 1 % of an equivalent oral dose, and without systemic side effects [1]. Although proven to be so effective, there appears to be delayed referral to neurosurgeons for intrathecal or intra-ventricular baclofen. This study attempts to identify the presence and nature of this delay and explore the reasons for it in an effort to improve future treatment and quality of life of patients with severe spasticity and dystonia.

Methods

As part of a quality improvement initiative for the Pediatric Gait and Spasticity Clinic, a retrospective analysis of the pediatric neurosurgery General Electric (GE) Centricity database from 2005–2014 was accomplished. A query was done in our electronic medical record (EMR) database to select patients who underwent an implantation of a baclofen pump. In a retrospective study of 164 consecutive patients who have received baclofen pumps since 2005, 57 of them were under the age of 23 at the time of implantation. Five of the 57 pediatric patients were lost to follow up after pump implantation. We were able to contact 52 patients and we were able to obtain consent and participation of 30 of these pediatric patients or caregivers (57.7 % response rate) who had received intrathecal baclofen pumps in order to complete an outcome survey about the management of severe spasticity or dystonia. Children were between the ages of 5 and 23 at the time of implantation. For patients who were not able to communicate at the time of the survey due to their condition, parents or primary caregivers were consented and surveyed on their child's behalf. We acquired information regarding the patient's age of onset of their spasticity and tone. The nature of initial treatment of spasticity was investigated including the type of physician they initially saw, the type of treatment they first received, the age of this intervention, and the effectiveness of the treatment. The remainder of the questions evaluated patients' experience with their ITB pump, including the parent's opinion of the pump's effectiveness on their child's spasticity and quality of life. The questionnaire that was utilized is depicted in Fig. 1.

By looking at the age of the patient when they developed spasticity, as well as the age they received a pump, we were able to determine whether there was a delay for baclofen pump referral. It is important to consider that ITB therapy for treatment of spasticity of cerebral origin was not approved by the US Food and Drug Administration (FDA) until 1996 [7–9]. Although ITB was first approved by the FDA in 1992, at that time it was approved only for treatment of spasticity of spinal origin [7, 8]. Our study analyzed the age of individuals when ITB therapy was authorized by the FDA in 1996 for treatment of spasticity of cerebral origin [7–9]. Since some of our subjects were born before 1996 when ITB was not yet a treatment option, we had to account for this when determining their delay in referral. The subjects were divided into two groups: group A and group B. Group A included 22 out of the 30 subjects who were born after ITB therapy approval. Group B consisted of 8 out of the 30 subjects who were born before the pump was approved and made available. For these 8 patients, we were able to calculate the time between

1. At what age did your child first have problems with tone and spasticity?

2. To what type of doctor were you referred to for care of the spasticity?
   a) Physiatrist
   b) Neurologist
   c) Neurosurgeon
   d) Orthopedist

3. At what age did your child start treatment?

4. What medications or treatments did your child use to treat the spasticity?
   a) Botox
   b) Oral Baclofen
   c) Alcohol Blocks
   d) Other

5. Were any of the treatments effective at all?

6. At what age did your child receive the baclofen pump?

7. Did your physician ever mention the idea of a baclofen pump prior to the start of the other treatments?

8. Did you or your physician ever seem hesitant about placement of a pump?

   -if yes: for what reasons?

9. Has there been any complications with the pump?

   -if yes: what were the complications?

10. Regarding your child's spasticity, would you say the pump has had:
    a) No effect
    b) Small effect
    c) Dramatic effect

11. Regarding your child's quality and ease of life, would you say the pump has had:
    a) No effect
    b) Small effect
    c) Dramatic effect

Fig. 1 The questions that were used to obtain data in our study are displayed. The survey was executed by phone call

implantation of the pump and the age of each subject when ITB treatments were authorized. For both group A and group B, by additionally calculating the time between the implantation of the pump and the start of the initial less-invasive treatments, we were able to assess the amount of time patients experimented with other treatments before ultimately receiving the pump.

Results

Out of the 30 pediatric patients who received an intrathecal baclofen pump between the ages of 5 and 23, 15 subjects had spasticity due to "cerebral palsy" (CP) or hypoxic-ischemic encephalopathy (HIE) due to perinatal causes, 11 patients had documented anoxic brain injury at an age greater than 4, 1 patient had a spinal cord injury, 1 patient had viral encephalitis, and 2 patients that were related had hereditary spastic paraparesis. The outcome of ITB treatment for these patients seemed to be independent of the cause of spasticity. The characteristics of the 30 subjects in this study are shown in Fig. 2.

As we hypothesized, there was an evident delay in the referral of ITB. It was found that there was at least 1 year delay in 96.7 % of the cases recorded. In group A, there was an average of 5.14 years between the time of onset of spasticity, and the time the patient received a pump. The longest delay recorded was 11 years, while the shortest was less than a year. This delay in ITB referral for subjects born after 1996 is represented in Table 1. This information was determined by taking the difference between the respondent's age at pump implantation and age at the onset of spasticity.

Group B subjects have a larger average delay than those subjects in group A. This may be accounted for by the fact that group B participants were born between the years of 1984 and 1994, and ITB therapy was not approved by the FDA until 1996 [7–9]. However, even after ITB was approved it was years before these subjects were finally referred. These individuals all received pumps sometime after 2004, which is at least 8 years after ITB approval. In group B, there was an average of 11.7 years between the time of ITB therapy approval and the time the patient received a pump. These eight patients each struggled with spasticity for more than 7 years before ITB therapy was instigated. The longest delay in ITB referral was 16 years, while the shortest period of delay was 7 years. It is likely that lack of general awareness and acceptance of ITB therapy in its early years of approval contributed to this large delay. This latency in ITB referral for group B subjects is represented in Table 2.

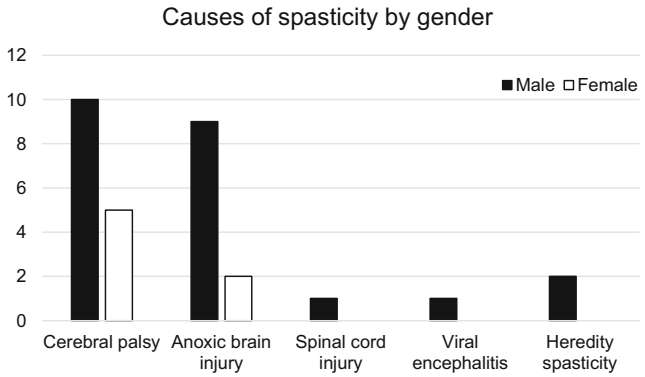

Fig. 2 Bar graph showing the distribution of causes of spasticity among the respondents. Gender of the subjects is also displayed

Looking at both groups A and B subjects together, our results show a much greater delay in patients who have cerebral palsy due to perinatal causes than in patients who had brain or spine injuries later in life. Interestingly, most subjects who had HIE or anoxic brain injury later in life had a significantly shorter postponement of ITB therapy.

During the years of delay, all but 4 subjects in groups A and B tried other treatments and medications. Nineteen tried botulinum toxin injections, 15 took oral baclofen, eight reported that they had tried alcohol injections, and many tried physical therapy. Many of the subjects tried more than one therapy. Because their treating physician attempted to control spasticity through various modalities and medications, there was a delay in referral to ITB therapy consistent with almost all respondents. Although some subjects report that these therapies were minimally effective, all participants stated that the treatments ultimately were insufficient. Six patients reported that the medications stopped working after some time. Two patients said that if they had continued with medications, the dose would have been too high to withstand. One patient reported that the amount of medication they were receiving caused intolerable side effects and was then discontinued. Twelve patients stated that their previous treatments had no effect at all. Only one of the 30 subjects was initially referred directly to a neurosurgeon. About 30 % of patients saw a physiatrist, 57 % saw a neurologist, and 27 % saw an orthopedist for initial consult. Several patients saw more than one of these specialists. It was these physicians who prescribed the initial treatments to care for the patients' spasticity. Group A patients spent an average of 4.16 years (range 0–10.5 years), and group B spent an average of 14.1 years (range 6–19 years) trying multiple types of treatments before they were ever informed about ITB therapy. Of the 30 patients, 24 reported that their physician had not mentioned the idea of a baclofen pump when the other treatment options were proposed. It was not until after the other treatments proved to be ineffective that a referral to a neurosurgeon was made. In the caregivers' opinions, it was the development of recurrent muscle contractures that prompted the referral for neurosurgical consultation. Although the parents felt that the oral medications and injection therapies were ineffective long before the referral was made, is was not until multiple recurrent contractures

Table 1 Length of delay of ITB referral after approval (group A)

Subject	Cause of spasticity	Age of onset of spasticity (years)	Age at ITB implantation (years)	Delay of ITB referral (years)
1	ABI	13	13	0
2	ABI	12	13	1
3	ABI	17	18	1
4	ABI	18	19	1
5	ABI	22	23	1
6	VE	13	15	2
7	ABI	16	18	2
8	ABI	18	20	2
9	ABI	17	19.5	2.5
10	ABI	18	21	3
11	CP	0	5	5
12	SCI	5	10	5
13	CP	9	14	5
14	ABI	0	7	7
15	CP	0	7	7
16	HS	2	10	8
17	CP	0	9	9
18	CP	2.5	12	9.5
19	CP	1	11	10
20	HS	2	12	10
21	CP	0	11	11
22	ABI	4	15	11

CP cerebral palsy, *ABI* anoxic brain injury, *SCI* spinal cord injury, *HS* hereditary spasticity, *VE* viral encephalitis

developed that their child was referred for ITB evaluation. Unfortunately, the development of contractures found in many of our patients was treated with repeat tendon lengthening several times before the referral for more aggressive treatment of spasticity was made.

About 50 % of caretakers had hesitations regarding pump implantation. Eleven parents were most concerned about their child undergoing an invasive surgical procedure and did not like the idea of their child having a foreign device in their body. Two parents were concerned about potential complications, such as infection. One parent was skeptical that their child might have been too young for this procedure. However, after prolonged consideration, all parents recognized that their child's spasticity was too poorly controlled and decided to go ahead with surgery despite their fear and hesitations. It is possible that this hesitation and prolonged consideration may have added to the delay of pump implantation. Parents may have subjective recall or poor memory, and ITB may have been discussed with them prior to their initial visit with a neurosurgeon.

Parents were also asked of their opinions about the effectiveness of ITB on their child's spasticity. Two patients

Table 2 Delay of ITB referral before ITB approval (group B)

Subject	Cause of spasticity	Age when ITB was approved (years)	Age at ITB implantation (years)	Delay of ITB referral (years)
23	CP	12	19	7
24	CP	8	17	9
25	CP	12	21	9
26	CP	7	16.5	9.5
27	CP	5	19	14
28	CP	5	19	14
29	CP	6	20	14
30	CP	2	18	16

CP cerebral palsy

reportedly had no effect from the pump. Fifteen respondents reported a small to moderate effect, and 13 reported a dramatic effect from the intrathecal baclofen. This totals to 28 out of 30 respondents who reported a beneficial response to ITB. The parents were also questioned about ITB's effect on their child's quality and ease of life. Four parents reported no change, 16 parents reported a small to moderate effect, and ten respondents experienced a dramatic effect. This totals to 26 out of 30 patients that experienced an improvement in their quality of life. It is important to note that it was not only the patient's quality of life that was improved but also their caretaker's. Parents reported that caretaking was less problematic due to easier mobilization during daily activities.

Of the 30 patients included in the study, three (10 %) reported infections at the abdominal incision site and three (10 %) were found to have a spinal catheter disconnection. In all three patients with catheter malfunctions, scoliosis and dystonia were noted. No pump malfunctions were observed. Although some patients did have these complications, all but two subjects decided to continue ITB therapy due to its perceived effectiveness.

A total of five patients had the baclofen pump removed. Two patients felt that their spasticity had not improved after ITB therapy. One patient had the pump removed because their spasticity had resolved after 3 years of rehabilitation following an anoxic brain injury. Two subjects had the pump removed secondary to an infection at the abdominal incision site, as described above.

However, 93.3 % of respondents reported a significant reduction of spasticity after the initiation of ITB therapy. This may suggest that the benefits of ITB outweigh the risks of this procedure. Many studies have shown that when patients with spasticity are not satisfactorily treated, other complications may arise [10].

Discussion

Despite latency in the referral of patients for intrathecal baclofen, there is much data supporting the effectiveness of ITB. In a study of 20 pediatric patients with severe spastic CP, overall tone decreased over the 18 months following implantation of an intrathecal baclofen pump [1]. Many patients report that ITB therapy causes their spasms to become less frequent or severe and patients and caregivers report an increased range of movement with ITB. With improved range of motion, patients were less likely to develop contractures from the inability to mobilize their joints [1]. Over a period of 18 months following pump implantation researchers investigated the quality of life through a caregiver questionnaire. Researchers reported that patient's comfort, mobility, and daily care had positively advanced [1]. A similar study used the Child Health Questionnaire-PF50 to measure quality of life and also found large improvements in children's pain and mental health [11]. Several reported clinical studies support the use of intrathecal baclofen, including a study of 35 children with cerebral palsy who reports reduced pain and improved sleep within 6 months of treatment, social function improvement within 6 months, and mobility improvement [4]. Statistically significant reduction of spasticity has often been reported with the implementation of ITB [4]. Another study of 30 patients with an average age of 51 years old reported good clinical response to treatment of spasticity and rigidity, improved quality of life, pain reduction, and patient satisfaction with short length of admission [12]. In a publication of 20 adult patients with "severe progressive spinal spasticity that was not responsive to medical therapy," all subjects demonstrated a reduction in muscle tone, spasms, and pain secondary to receiving ITB [13]. On average, patient Ashworth scores decreased from 4.4 to 1.8 [13].

In order to produce an optimal outcome, a time line has been suggested for the management of spasticity in children. Increased muscular rigidity or decreased range of motion may first be noted by the parents or caregivers of the child. In pediatric patients with cerebral palsy, a primary care physician may be consulted in reference to their child's increased tightness. At that point, a referral to a neurologist or physiatrist can be made to assess the child's signs and symptoms of spasticity. These physicians typically evaluate the child and start oral anti-spasmodic medications if spasticity is severe. If a medical diagnosis of spasticity is made, various treatment options are recommended depending upon the condition of the child. A thorough neurologic examination may be performed followed by neuroimaging, if necessary. X-rays of the hips and spine may be recommended to assess the child's gait. In addition, a referral to an ophthalmologist can be made to check the presence or absence of papilledema. A medical algorithm for the most ideal time for treatment of spasticity is represented in Fig. 3. Treatment for these patients can range from conservative to aggressive. Potential treatment options may include therapeutic referrals such as occupational therapy, physical therapy, and speech therapy (if necessary); pharmacological treatments such as oral baclofen, botulinum toxin injections, and alcohol blocks; and surgical referrals including orthopedic and neurosurgical procedures [14]. The timing of medical invention for spasticity is important to prevent the development of muscle contractures. When spasticity is first noted, the muscles and joints of the child may be mobilized and flexible [14]. However, as time progresses or if treatment interventions are not pursued, the mobility of the joints and muscles decrease and contractures may develop [14]. At this point, but before the development of contractures, a referral to a neurosurgeon for evaluation of ITB therapy can be made. An assessment for a selective dorsal rhizotomy (SPR) for patients with spasticity may be recommended as another treatment option. A patient is considered to be a good candidate for

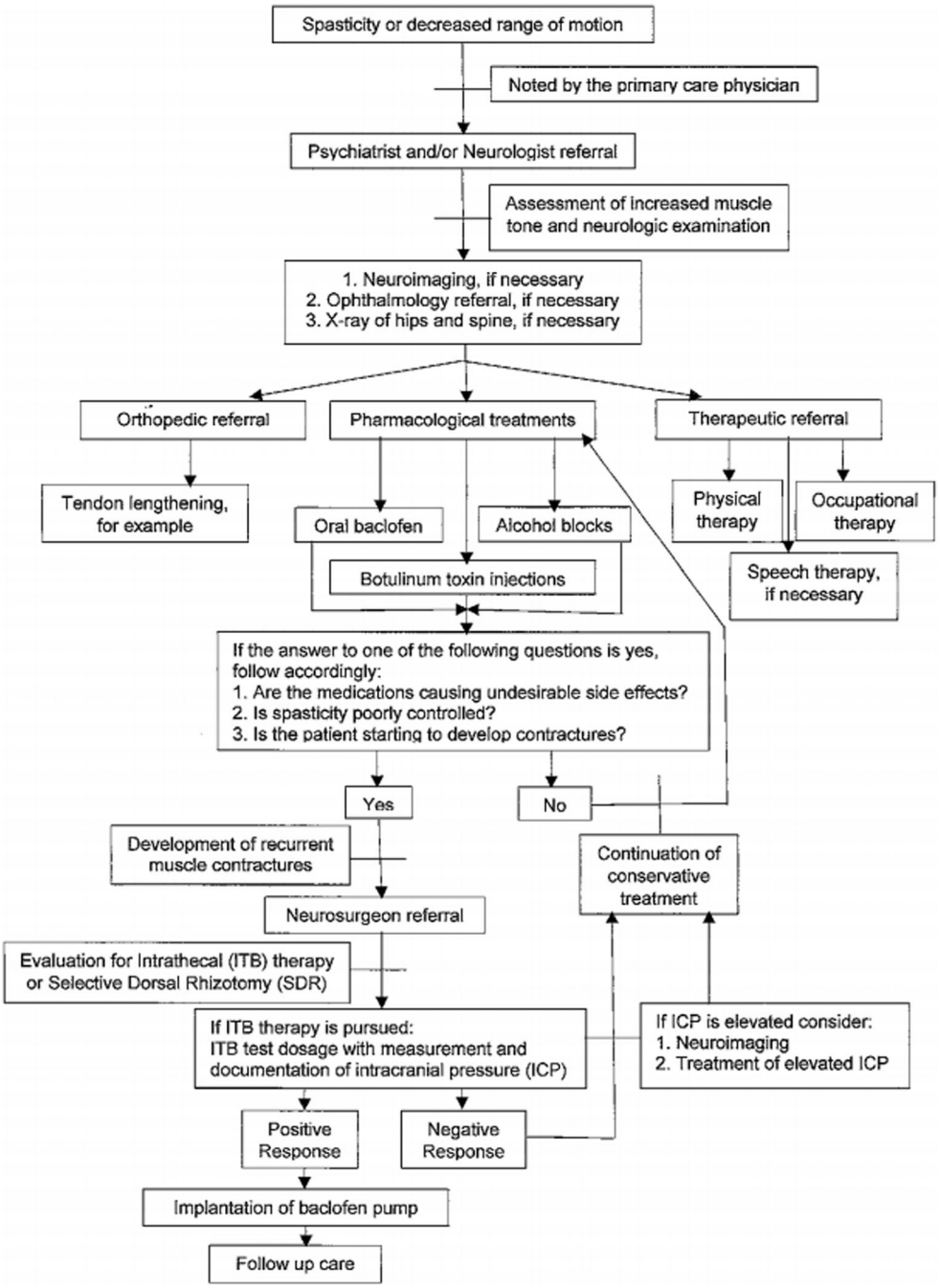

Fig. 3 Medical algorithm for recommended timing of spasticity treatment [14, 15]

ITB therapy if conservative treatments such as the oral baclofen, botulinum toxin injections, or alcohol blocks are causing intolerable side effects or an undesirable response [14, 15]. In addition, a patient must be of a certain age and body size to "accept" the implanted pump [9]. If the decision is made to proceed with baclofen pump implantation, a test dosage of intrathecal baclofen with measurement and documentation of intracranial pressure (ICP) is required. If the patient's ICP is elevated, alternative recommendations such as neuroimaging or treatment of the elevated ICP may be decided. A reduction of the patient's spasticity as per the patient Ashworth score indicates a positive response to the ITB test dosage [15]. Contrarily, lack of functional improvement in the patient's spasticity indicates a negative response, and conservative treatments should be continued [15]. After the pump is implanted, the patient will require a refill of the intrathecal baclofen medication every few months with their designated physician. Follow-up care for these patients is important to assess

the intrathecal baclofen dose administered, as well as the benefit the patient is receiving from ITB therapy.

Medical Advisory Secretariat of Ontario has advised that children must be of a certain age and body size to tolerate the implanted pump [9]. Literature states that intrathecal baclofen therapy can be considered in individuals at least 3 years of age at the time of pump implantation [4, 16, 17]. Although all participants in group B with the exception of one subject were greater than 3 years old when ITB treatment was approved in 1996, none of them received a baclofen pump for at least another 8 years post-approval. Despite being old enough to have the procedure, there was still an average of 11.7 years between the time of ITB therapy approval and the time the patient received a pump. It is possible that this delay may correlate to the general lack of acceptance and knowledge of baclofen pump placement by the medical community and caretakers. Patients with spasticity are commonly first treated by neurologists and physiatrists, and these physicians often defer ITB referral for several years while trying conservative treatments such as oral medications, botulinum toxin injections, and alcohol blocks for 2 plus years first. Some physicians may never refer to a neurosurgeon for ITB or still may consider the pump as new technology. Hesitations may also be due to fear of associated medical complications such as seizures; however, seizures only occur rarely in cases of overdose or withdrawal [18]. We realize the limitations of our study and acknowledge that parents' opinions may be subjective to poor recall or selective memory. It is possible that some parents or patients may not have followed up with their neurologists or physiatrists, delaying their treatment process. Latency in referral may have resulted from parents' own apprehension about surgical intervention.

Despite multiple reports about the efficacy of ITB in the treatment of spasticity and dystonia, we have noted a large delay of referral for consideration of baclofen pump therapy in our study. It has been our experience that many of our patients have suffered for years and have incurred many of the complications of spasticity before they are referred to see a physiatrist or a neurosurgeon for treatment. Many of our patients have had multiple orthopedic surgeries for contractures that have developed from poorly controlled spasticity. Pharmacological therapies that are initially tried are reported to have limited effect [6, 19]. Interestingly, our data shows a much greater delay in patients who have cerebral palsy due to perinatal causes than in patients who had brain or spine injuries later in life. It is possible that this is due to an earlier referral to a neurosurgeon in the event of brain trauma. This earlier referral may also be occurring because the spasticity or dystonia is more profound, severe, or apparent in an older child with new HIE. It is also possible that patients with HIE from perinatal causes were referred to neurologists as infants, when their spasticity was not so evident. Neurologists would typically evaluate the infant and only start oral anti-spasmodic medications if spasticity were severe. Symptoms and signs of spasticity may have developed in a more insidious fashion after perinatal HIE, so it is possible that when the infant first saw the neurologist they did not have severe spasticity. Some parents or patients may not have followed up regularly with their neurologists or there may have been years of treatment with oral medication, botulinum toxin, and/or alcohol blocks before referral for ITB therapy or neurosurgical referral.

Clinical experience and many authors have shown that ITB is effective in reducing spasticity and tone. A study of 13 adult patients evaluated the effects of intrathecal baclofen placement at a very early stage in acquired brain injury (ABI) to determine if there are any negative effects of early implantation [3]. The study concluded, "ITB therapy in ABI should be considered as early as possible. The implants are safe and effective in reducing spasticity." [3] Another publication of 14 patients who underwent ITB pump implantation less than 12 months after a traumatic brain injury (TBI) had a similar conclusion [10]. Patients were able to achieve functional improvements sooner including progress made in their gait and motor function, as well as the reduction of pain [10]. It has been the experience of various clinicians that when these patients are not satisfactorily treated within a duration of time that complications may arise [10]. Unlike other pharmacological treatments that are often tried, the effectiveness of the ITB does not appear to decrease over time [4]. A significant number of subjects in our study reported diminished effect of pharmacological treatment, and 93 % of these respondents reported that ITB has had a beneficial long-lasting effect on their spasticity. These effects correspond to the continuation of ITB therapy, leading to increased independence and decreased spasticity as indicated by the patient Ashworth score.

In an article reporting 86 patients ranging in age from 3 to 42 years, 92 % of subjects who had received ITB therapy had diminished spasticity as of their 24-month follow-up [16]. In a study of 44 patients with upper motor neuron disease, none of the patients who received ITB requested discontinuation of therapy. This suggests that "the benefits of ITB on symptom relief are meaningful, even when complete mobility cannot be maintained, and that the therapy is well tolerated." [20] Although many parents in our study initially had hesitations towards pump implantation, there is a significant satisfaction rate post-operatively. About 93 % of patients reported that the baclofen had a small to dramatic effect on their spasticity, and 86.6 % reported a small to dramatic effect on the patient's quality of life. These findings are consistent with the reviewed primary literature. In Albright's study of 86 patients with generalized dystonia, 86 % of respondents reported an improvement in the quality and ease of life for the patients in addition to their caretakers [16].

Treatment of spasticity is not only beneficial for the patient, but also for their caretakers. Many caretakers must transfer,

dress, and mobilize the patient, and reduction in muscle tone significantly improves the ease at which they can do so. Despite the large hesitance towards this more invasive procedure, 95 % of subjects in a study of 80 caretakers of pediatric patients reported that they would have this procedure performed again [17]. In Dario et al.'s study of patients with "severe progressive spinal spasticity," the subjects' physical disability was measured according to the Functional Independence Measure (FIM) [13]. The FIM scale evaluates an individual's disabilities by determining how much aid is needed on a daily basis [21]. Dario et al. investigated patient function in terms of bathing, dressing, and transferring the individual. After receiving ITB, the mean FIM score increased from 33.8 to 58.7 [13]. This indicates a decreased burden on caretakers and increased patient quality of life as a result of ITB therapy. Our study coincides with this previous research, as several parents that we spoke to reported that care of their child was made easier post-surgery.

Invasive procedures inevitably warrant hesitation, but in weighing the benefits against the risks of pump implantation, studies consistently highlight the benefits of effective treatment of spasticity with ITB. Much research confirms both the efficacy and safety of ITB therapy. In a study of 68 subjects with spasticity of cerebral origin, overall upper and lower extremity spasticity decreased over the 70 months following implantation of an intrathecal baclofen pump [22]. The long-term effectiveness of ITB therapy was contributed to the continuous relief, stable dosage, and decreased spasticity as indicated by the patient Ashworth score [22]. The long-term effects of ITB therapy was evaluated in another study of 24 patients 10 years following pump implantation [23]. These individuals described that after a decade of receiving ITB treatments, they still had a moderate-to-high level of satisfaction with the pump [23]. Low amounts of pain and less frequent muscle spasms correlates to their decision to continue ITB therapy [23]. In another article of 37 patients with spasticity of cerebral origin, a substantial reduction of overall tone of the upper and lower extremities was noted at the 6- and 12-month follow-up appointments [24]. A connection between the intrathecal baclofen dose prescribed and overall muscle tone reduction was found [24]. The dose of baclofen can be easily adjusted along the course of treatment, and its direct effect on the spinal cord makes it extremely effective when oral baclofen fails to provide desired results. The side effects associated with intrathecal baclofen as well as the dosage prescribed are much lower compared to that of oral baclofen [1, 25]. Various articles support the utilization of long-term ITB therapy for patients with spasticity who have not responded to other less invasive modalities and treatments [25, 26]. The benefits of ITB therapy appear to outweigh the risks and complications of this procedure. "Safe and efficacious, this mode of treatment appears to be the gold standard for treating severe spasticity." [12] The responses from most patients in our study support this statement, as 93 % found relief of their severe spasticity after years of trying other therapies.

Conclusion

With an average delay of ITB referral being 5.14 years in group A and 11.7 years in group B, we conclude that our patients with severe spasticity did experience a delay in referral for neurosurgical evaluation and consideration of ITB therapy. In our patients, other treatments that were ultimately ineffective were utilized for many years before referral. About 80 % of subjects reported that their physician never informed them about an ITB pump until after all other treatment options had been exhausted and multiple tendon lengthening procedures had been performed for recurrent contractures. A limitation of our study is that parents' opinions or recall may be subjective, and it is possible that they were informed of surgical alternatives years before they consented to a neurosurgical referral. Another limitation of this study is that the parents' opinions of ITB's clinical results are subjective and obviously not blinded. After analyzing the nature of our patients' delayed ITB referral, we conclude that there must be an increased awareness of ITB therapy. The main underlying issue seems to be parental lack of knowledge of this treatment due to physician's delay in its consideration. This study serves to inform the public of ITB's significant effectiveness in controlling severe spasticity and show that it should be considered as one of the first treatment modalities in managing this chronic disorder.

Conflict of interest The authors report no conflict of interest concerning the materials or methods used in this study or the finding specified in this paper.

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