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Improvements in Children With Cerebral Palsy Following Intrathecal Baclofen: Use of the Rehabilitation Institute of Chicago Care and Comfort Caregiver Questionnaire (RIC CareQ)

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Abstract

Implantation of an intrathecal baclofen pump is recommended for children with cerebral palsy as a means to improve care and comfort when other options fail to control severe hypertonia. Making an assessment of a child's spasticity-related limitations in both routine care and activity is a necessary component of selection of intrathecal baclofen candidates. The Rehabilitation Institute of Chicago Care and Comfort Caregiver Questionnaire (RIC CareQ) is a validated, easy-to-use questionnaire that elicits information about the ease of daily activity and caregiving in patients with severe spasticity. Questionnaires completed by caregivers and patients at a pediatric physiatry spasticity clinic over an 11-year period were reviewed to evaluate whether the Rehabilitation Institute of Chicago Care and Comfort Caregiver Questionnaire captured improved caregiving and comfort of children with cerebral palsy and severe spasticity following intrathecal baclofen pump implantation. The Questionnaire scores showed improvement after intrathecal baclofen pump implantation, consistent with subjective reports of patient and caregiver satisfaction.

Keywords

cerebral palsy, spasticity, intrathecal baclofen

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Spasticity, a velocity-dependent increased resistance to muscle stretch, is present in patients who have experienced a central nervous system injury and occurs in patients with cerebral palsy, traumatic brain injury, and spinal cord injury. Spasticity may result in pain, orthopedic deformities, and difficulty with movement, activities of daily living, and mobility. Although physical and occupational therapies are first implemented to improve function, spasticity is often severe enough to warrant medical management.

Medical management of spasticity can include use of oral medications and focal injections; however, these options may be limited by total dosage or undesirable side effects such as truncal weakness, somnolence, toxicity, or incomplete spasticity control. In patients in whom spastic tone or side effects exceed successful treatment, medication administered directly to the central nervous system can reduce negative systemic side effects by delivering the medication past the blood-brain barrier. In 1985, Penn and Kroin[1] were the first to demonstrate that baclofen, a commonly used spasticity medication, administered to the cerebrospinal fluid could reduce spinal cord injury–mediated spasticity. In 1991, Albright et al[2] found a similar decrease in spasticity of cerebral origin with intrathecal delivery of baclofen. In 1996, the intrathecal baclofen pump was approved by the Food and Drug Administration for the management of spasticity caused by central nervous system injury.

The primary goal for tone management in intrathecal baclofen pump candidates is improvement of activities of daily living, caregiving, and comfort. A secondary consideration is improvement in mobility. Without a means to objectively assess response to treatment, the practitioner will generally use the subjective response of the patient or caregiver to intrathecal baclofen pump treatment to guide management even when functional changes are not observable. One of the largest populations using

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intrathecal baclofen pump treatment is children with cerebral palsy. In those cases, primary caregivers, rather than the patient, are often the main source of feedback.

The Caregiver Questionnaire (CQ) was developed at the Rehabilitation Institute of Chicago in 1990 for children with spastic quadriplegic cerebral palsy undergoing selective dorsal rhizotomy surgery for spasticity management.[3] The Caregiver Questionnaire was developed as a health-related quality of life tool; however, conceptually it is a perceived burden of effort measurement because it captures the caregiver's role. Schneider et al[3] demonstrated that the overall Caregiver Questionnaire score correlated with the Child Health Questionnaire Parental Impact-Time subscale, which describes a caregiver's personal time limitations based on the child's health-related status. This finding was consistent with the overall goal of determining caregiver burden by use of the Caregiver Questionnaire.

Nemer McCoy et al[4] later used the Caregiver Questionnaire to develop the Care and Comfort Hypertonicity Questionnaire to identify the severity of spasticity, where higher scores correlate with patients who would be appropriate candidates for intrathecal baclofen pump implantation. The questionnaire has undergone initial validation testing.[4]

The Caregiver Questionnaire was also one of the questionnaires used to develop the Caregiver Priorities and Child Health Index of Life with Disabilities (CPCHILD), an instrument developed to measure the ease of daily activity and caregiving of children with severe cerebral palsy. The Caregiver Priorities and Child Health Index of Life with Disabilities measures the effectiveness of interventions intended to improve or preserve outcomes for children with severe disabilities.[5]

As use of intrathecal baclofen evolved, the Caregiver Questionnaire was written and revised periodically for use in the spasticity clinic. Hwang et al[6] evaluated the questionnaire in 2011 and determined that it was a reliable measure of a caregiver's perception of burden of care for the studied group of children. The questionnaire was called the CareQ at the time of publication; however, to avoid confusion with another similarly named questionnaire, it has been renamed the Rehabilitation Institute of Chicago Care and Comfort Caregiver Questionnaire (copyright 2011, Rehabilitation Institute of Chicago, Chicago, IL; all rights reserved).[6] The Rehabilitation Institute of Chicago Care and Comfort Caregiver Questionnaire focuses on daily care activities, use of equipment, positioning, and pain. Although the Questionnaire is not a health-related quality of life assessment, it does address participation, especially as impacted or limited by pain. It also provides an insight on caregiver burden by focusing on routine daily care, including the ease of using equipment. Further, it is not specifically focused to the pediatric population; both adults and caregivers have completed this questionnaire (Table 1). Finally, it allows respondents to indicate the amount that a patient is able to contribute to performance of daily care activities.

Patients or caregivers of patients with significant hypertonia are asked to complete the Rehabilitation Institute of Chicago Care and Comfort Caregiver Questionnaire in the clinic. The Rehabilitation Institute of Chicago Care and Comfort Caregiver Questionnaire can be completed before intrathecal baclofen pump implantation by or for patients who later may be candidates for intrathecal baclofen. The Questionnaire is used both before and after implantation because it is designed to educate as well as evaluate performance of the intrathecal baclofen pump. It is first used as an educational tool to assist patients, their caregivers, and the medical team in establishing realistic goals for spasticity treatment by concentrating on daily activities that could be improved with better spasticity control. This is an important function of the Rehabilitation Institute of Chicago

Table 1. Demographic Characteristics of 114 Patients Who Received ITB Pumps.

Ethnicity	
White	79
African American	21
Hispanic	12
Asian	1
Other	1
Diagnosis	
Cerebral palsy	90
Traumatic brain injury	12
Transverse myelitis	1
Spinal cord injury	6
Other	5
GMFCS	
I	0
II	3
III	2
IV	32
V	46
Tone	
Spastic	84
Dystonic	1
Spastic and dystonic	26
Missing data	3
Involved extremities	
Diplegia	10
Triplegia/quadriplegia	102
Missing data	2
Age at pump implant (y)	
3-5.9	5
6-8.9	23
9-11.9	14
12+	64
Missing data	8
Complications[a]	
Infection	16
Catheter disconnection	25
Pump malfunction	6
Other	2
None	74
Outcome	
Currently followed	67
Moved/lost to follow-up	37
Death	10

Abbreviations: GMFCS, Gross Motor Function Classification System; ITB, intrathecal baclofen.
[a]Total complications greater than 114 as some patients had more than 1 complication.

Care and Comfort Caregiver Questionnaire because patients who had received intrathecal baclofen pumps indicated a need for greater education before pump implantation.[7] The Questionnaire is also administered intermittently in continuing follow-up appointments for patients after receiving an intrathecal baclofen pump to review how goals are being met. This information guides ongoing treatment.

The Rehabilitation Institute of Chicago Care and Comfort Caregiver Questionnaire consists of 19 questions and takes only a few minutes to complete, making it ideal for a busy clinic setting. In our review of questionnaires completed in 11 years of use, all questionnaires were fully completed without assistance from clinic staff. Answers are in the form of a Likert scale, where 1 denotes an easy task and 5 indicates the greatest level of difficulty.

We undertook this study to determine whether the Rehabilitation Institute of Chicago Care and Comfort Caregiver Questionnaire captured patient and caregiver perceptions of improved caregiving and comfort following management with intrathecal baclofen for the child with severe spasticity. Our hypothesis was that caregivers of children undergoing intrathecal baclofen pump implantation would demonstrate changes in the perceived burden of care and that the Rehabilitation Institute of Chicago Care and Comfort Caregiver Questionnaire would be a sensitive measure to detect such changes.

Methods

A chart review was done to evaluate responses on the Rehabilitation Institute of Chicago Care and Comfort Caregiver Questionnaire in 11 years of clinical practice from 1996 to 2007. Basic demographic characteristics including date of birth, gender, ethnicity, diagnosis, tone type and distribution, and outcome were collected for all patients who have undergone intrathecal baclofen pump implantation at our medical center. Information including date of implantation, known complications, and other surgeries was also abstracted from the medical charts.

In order to characterize the intrathecal baclofen experience at our facility, data were abstracted on all patients receiving this medication within the interval of study and were analyzed descriptively. Differences between Rehabilitation Institute of Chicago Care and Comfort Caregiver Questionnaire scores before and after implantation of the intrathecal baclofen pump were evaluated by paired *t* test for each domain. Moreover, linear regression models that included covariates such as time since pump implantation and length of intrathecal baclofen treatment were constructed to examine the influence of these variables on post–intrathecal baclofen care and comfort questionnaire scores. Values of $P < .05$ were considered statistically significant when testing differences (paired *t* test) and when examining partial correlation coefficients (linear regression). Continuous variables are presented as mean $\pm$ standard deviation and discrete variables as n (%). Data were evaluated using the Statistical Package for Social Sciences version 15.0 for Windows (SPSS Inc, Chicago, IL).

Results

At the end date for chart review and abstraction in June 2007, 117 patients who received care in the spasticity clinic had undergone intrathecal baclofen pump implantation. Three patients were excluded because of missing diagnosis or date of birth. More males (n = 86) than females (n = 28) received intrathecal baclofen pumps in the sample of 114 patients. The mean age was 14 years (range, 4-47 years). Sixty-eight percent of patients (n = 78) had a diagnosis of moderate to severe (Gross Motor Function Classification System[8] [GMFCS] IV or V) cerebral palsy (Table 1). Seventy-four percent of patients (n = 84) presented with spasticity as the primary tonal abnormality, and an additional 23% (n = 26) demonstrated a combination of spasticity and dystonia. Nearly all patients had triplegic or quadriplegic distribution of their hypertonicity.

Table 2. Age and GMFCS Classification of Cerebral Palsy Patients.

GMFCS	N	Age at time of ITB (y)
II	3	21 $\pm$ 6.7
III	2	18 $\pm$ 3.9
IV	32	14 $\pm$ 6.0
V	43	12 $\pm$ 5.0

Abbreviations: GMFCS, Gross Motor Function Classification System; ITB, intrathecal baclofen.

Of the original 114 patients in this study, 67 continued to receive care at our medical center. Thirty-seven patients moved or received care closer to their homes, which is thought to be consistent with transition of care from a major medical center to the local community, especially as intrathecal baclofen pump availability has grown. Ten patients had died from medical comorbidities.

Seventy-four patients did not have any known complication events. Of the recorded known complications, catheter disconnection from the pump was the most frequent complication (25 occurrences), followed by infection (16 occurrences). Six patients were believed to have some type of malfunction of their pumps (Table 1). Seventy-five patients were classified with moderate to severe cerebral palsy (Gross Motor Function Classification System IV or V). Comparisons by severity of cerebral palsy were not conducted as only 5 patients were classified with less severe involvement (Gross Motor Function Classification System II or III). Notably, patients with greater severity were younger at the time of intrathecal baclofen pump implantation. There was a mean age of 14 years and 12 years at implantation in patients with Gross Motor Function Classification System IV and V, respectively, as compared with a mean age of 21 and 18 years in patients with Gross Motor Function Classification System II and III, respectively (Table 2).

By conclusion of the chart review period, 33 patients had undergone implantation of a second pump, with most undergoing replacement 4 to 5 years after receiving the first pump. Three of the 33 patients who received a second pump required replacement because of infection and again had difficulty with infection, necessitating a second explantation.

Twenty-five patients (22%) underwent orthopedic surgery prior to pump placement and 33 patients (29%) underwent orthopedic surgery after the implantation. Of the patients who had orthopedic surgery prior to pump implantation, 2 had bony

Table 3. Care and Comfort Scores Before and After ITB and *P* Value for Change.

CareQ domain	Before ITB	After ITB	*P* value
Upper extremities	3.5	2.3	<.001
Lower extremities	3.2	1.9	<.001
Positioning	3.4	2.3	<.001
Pain	3.5	3.7	NS
Orthotic devices	3.2	1.8	<.001

Abbreviations: ITB, intrathecal baclofen; NS, not significant.

procedures, 9 had soft tissue surgery, 6 had combined bony and soft tissue surgery, and 8 had multiple orthopedic surgeries. Of the patients who had orthopedic surgery after pump implantation, 14 had bony surgery, 9 had soft tissue surgery, 6 had a combination of bony and soft tissue surgery, and 4 had multiple orthopedic surgeries.

The Rehabilitation Institute of Chicago Care and Comfort Caregiver Questionnaire was completed by 84 patients or caregivers in the clinic. Sixty-three (75%) had completed questionnaires both before and after intrathecal baclofen pump implantation. A paired *t* test was used to evaluate the care and comfort questionnaire scores for each domain before and after implantation. The event of orthopedic surgeries could not be examined for influence on questionnaire domains because of the small sample size. Statistically significant improvements were seen in each domain of the care and comfort questionnaire except for pain (Table 3).

Patients or their caregivers completed the post–intrathecal baclofen questionnaire an average of 37 months after pump implantation. A linear regression model was used to examine the influence of time since pump implantation on Rehabilitation Institute of Chicago Care and Comfort Caregiver Questionnaire scores. The findings indicated that the length of time with intrathecal baclofen spasticity management did not affect Rehabilitation Institute of Chicago Care and Comfort Caregiver Questionnaire scores for upper extremity, lower extremity, and pain. There was an unclear relationship with the positioning and orthotic devices domains (Table 4).

The influence of increased age in the intrathecal baclofen recipient was examined using a linear regression model. The data indicated that age did not affect upper extremity, lower extremity, and pain scores. Pain scores were significantly lower as patient's age increased. Age at time of intrathecal baclofen pump implantation did not have a clear effect on either the positioning or orthotic devices domains (Table 4).

There were 49 documented complication events reported in 40 patients who received intrathecal baclofen pumps. Using a linear regression model, the presence of complications was used to examine influence on Rehabilitation Institute of Chicago Care and Comfort Caregiver Questionnaire scores. There was not a clear impact on the orthotic devices or positioning domains. Complications did not appear to affect improvement seen in the remaining questionnaire domains (Table 4).

Table 4. *P* Values for Post-ITB Care and Comfort Domain Scores in Linear Regression Models Controlling for Interval Since ITB Implantation, Age at ITB, and Presence of Complications.[a]

CareQ domain	Interval since ITB implantation	Age at ITB implantation	Presence of complications
Upper extremities	**.021**	**.026**	**.019**
Lower extremities	**.018**	**.010**	**.014**
Positioning	.147	.158	.179
Pain	**.003**	**.018**	**.002**
Orthotic devices	.537	.634	.449

Abbreviation: ITB, intrathecal baclofen.
[a]Values in bold indicate significance.

Discussion

Patients and caregivers generally report overall satisfaction with the intrathecal baclofen pump in spasticity management. This correlates with the statistically significant improvements seen in the Rehabilitation Institute of Chicago Care and Comfort Caregiver Questionnaire scores for upper extremity, lower extremity, orthotic devices, and positioning. All questionnaires were fully completed without supervision from clinic staff, indicating that patients and caregivers found the survey easy to finish in a busy clinic setting. The overall educational goal of the questionnaire appears to be satisfied from patient and caregiver communication of achieved expectations.

Literature suggests variable pain response to intrathecal baclofen pump management. Earlier publications have reported subjective improvements in pain.[7,9,10] Small numbers of patients reported worsening pain in one prior study,[7] and Parke et al[11] noted that patients with neuropathic pain complaints did not have any reported improvement in pain. In our patients, the small change in pain scores was insignificant.

The impact of time from pump implantation on Rehabilitation Institute of Chicago Care and Comfort Caregiver Questionnaire scores was assessed because questionnaires completed shortly after intrathecal baclofen pump placement might not reflect optimal treatment dosage. In a multicenter study, Albright et al[12] reported that most intrathecal baclofen dosage changes were made in the first 2 years. On average, our population completed a follow-up questionnaire at 37 months after pump implantation, which suggests that patients were typically at a stable dose. Krach et al[7] also found little correlation between time from implantation to any outcome measures. This is consistent with our analysis, which suggested that time did not affect most Rehabilitation Institute of Chicago Care and Comfort Caregiver Questionnaire scores.

A linear regression model that controlled for age at time of intrathecal baclofen pump implantation was used to evaluate whether proxy reports from caregivers affected Rehabilitation Institute of Chicago Care and Comfort Caregiver Questionnaire answers. Our data did show that scores in the Rehabilitation Institute of Chicago Care and Comfort Caregiver Questionnaire pain domain decreased as patients aged, and presumably were able to self-report pain. This finding

would seem to suggest that caregivers do have some difficulty with approximating their child's pain levels. Most questions on the Rehabilitation Institute of Chicago Care and Comfort Caregiver Questionnaire allow caregivers to give a direct report of their difficulty or ease with activities of daily living. This is consistent with our finding that age at time of intrathecal baclofen pump implantation did not appear to influence reported outcomes in most of the Rehabilitation Institute of Chicago Care and Comfort Caregiver Questionnaire domains.

The most common complication, catheter disconnection from the intrathecal baclofen pump, has decreased in frequency over time with revised surgical techniques and modifications made to the pump and catheter hardware. The 11-year period encompasses the period during which these evolutions occurred and accounts for the relatively large number of catheter complications in our sample.

The presence of complications did not affect scores on the Rehabilitation Institute of Chicago Care and Comfort Caregiver Questionnaire. Part of this effect could be attributed to overall satisfaction with the intrathecal baclofen pump. In the absence of a controlled study, we were not able to examine the timing with complications.

The frequency of orthopedic surgery both before and after intrathecal baclofen pump implantation is consistent with the severity of hypertonicity and the resultant deforming musculoskeletal effects in this population. A contributing factor in the larger numbers of surgeries after implantation of the pump is the frequency of planned surgeries when overall spastic tone control is improved.

Analysis performed was based on available data. In the absence of a controlled setting, conclusions about the short-term impact of complications or other interventions, such as surgery, could not be evaluated and would be of interest for future study.

Conclusion

Spasticity treatment is initiated on the premise that attempted control of spasticity may make a small but powerful difference in the life of the patient and the caregiver. Our findings indicate that the Rehabilitation Institute of Chicago Care and Comfort Caregiver Questionnaire is a measurement scale that adequately captures the impact of severe spasticity on care and comfort while helping to set realistic goals for medical management of spasticity.

The Rehabilitation Institute of Chicago Care and Comfort Caregiver Questionnaire allows a non-labor–intensive means for ongoing evaluation of spasticity treatment goals. It can be administered to both pediatric and adult populations with or without intrathecal baclofen pump management for hypertonia. The Questionnaire allows the reporting of caregiver and patient contribution to performance of activities of daily living. Although it is not a health-related quality-of-life measure, findings on the Rehabilitation Institute of Chicago Care and Comfort Caregiver Questionnaire give an approximation of activity limitations and caregiver burden.

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Authors' Note

Rehabilitation Institute of Chicago, RIC, and RIC CareQ are trademarks of the Rehabilitation Institute of Chicago.

Author Contributions

KB and DGS (author of the Rehabilitation Institute of Chicago Care and Comfort Caregiver Questionnaire) wrote the manuscript and analyzed the data. BT reviewed the manuscript. All authors contributed to data collection.

Declaration of Conflicting Interests

The authors declared the following potential conflicts of interest with respect to the research, authorship, and/or publication of this article: Gaebler-Spira has served as an advisor or consultant for Merz Pharmaceuticals and has received grants for clinical research from Cerebral Palsy Research Registry, National Institute on Disability and Rehabilitation Research, National Institutes of Health and National Science Foundation, Ultraflex, Accorda, and CNS Therapeutics.

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Ethical Approval

The research conducted in this study was under the approval of the Northwestern University Institutional Review Board.

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