Letters to the editor

'Effects of oral baclofen on children with cerebral palsy'

SIR–Baclofen is widely considered an appropriate medication for use in spastic cerebral palsy (CP)[1,2] but it is poorly studied.[3]

We investigated the use of baclofen through a prospective, uncontrolled, clinical trial. Participants were eleven children (nine males, two females; mean age 4y 3mo, range 3 to 5y, SD 11mo) with moderate CP, as defined by their Gross Motor Function Classification System scores of II, III, or IV. None received other spasticity interventions. Before receiving baclofen, each patient underwent a baseline assessment, including the Gross Motor Function Measure (GMFM),[4] physical examination, and parent questionnaires. The patients then went through dose escalation and continued treatment with oral baclofen for a total of six months. At that time, a follow-up assessment was performed with the same measures. This study was approved by the hospital Institutional Review Board and consent for participation was obtained from each child's parent or guardian.

Most patients reached a dose of around 2mg/kg/day of baclofen. Five patients experienced sleepiness that usually resolved without dose alterations. There were no adverse events that were directly attributable to patients' baclofen treatment. One patient had new onset seizures while on 0.5mg/kg/day of baclofen and another had an exacerbation of her seizure disorder.

Patients demonstrated smaller gains on the GMFM–66 than expected,[4] although this did not reach significance with a stratified *t*-test (Table I). Only two patients demonstrated greater than expected improvement in their GMFM–66 score. Equal numbers of patients demonstrated increases and decreases.

Range of motion and spasticity, assessed by goniometry and modified Ashworth scale, did not change. On several Child Health Questionnaire subscales, including Physical Summary, Role-Physical, and Bodily Pain, patients' scores decreased. This reflected poorer health status following baclofen treatment, but these changes were not statistically significant.

Ten parents reported that their child's functioning was improved while taking baclofen, four felt that their child was more comfortable, and six found that it was easier to care for their child. Sleepiness was noted to have worsened in four patients, improved in one, and was unchanged in the remainder. Eight of 11 patients continued to be treated with baclofen after completion of the study.

This uncontrolled study did not provide objective support for the use of oral baclofen among young children with spastic CP. The only meaningful positive changes were observed through parent reports. Baclofen may have had negative effects on motor function, as the patients did not demonstrate functional gains reported in similar children with CP. Several of the outcomes reflected worsening function or quality of life; however, these changes did not reach significance, potentially due to type 2 error.

In this group of 11 children, baclofen was found to be well tolerated, with the most common side-effect being transient sedation. Thus, this study does not support the assertion that oral baclofen is too sedating to be used in children with CP.

Baclofen is widely used to treat children with CP, but it may prove to be harmful for these children. This study suggests that baclofen may impair motor progress or reduce quality of life parameters. A controlled study of oral baclofen in a larger cohort of children with CP is warranted to better understand its effectiveness and to identify ideal target populations, if any, for its use.

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Table I: GMFM–66 change scores, standard deviations (SD) for study sample and normative cerebral palsy sample, by age and Gross Motor Function Classification System (GMFCS) level

Age (y)	GMFCS level	Norm change	Norm SD	Norm (n)	Study[a] change	Study SD	Study (n)
2–3	2	3.03	3.39	16	–4.56	6.45	2
2–3	3	2.43	2.75	14	–3.24	–	1
2–3	4	2.63	3.07	15	2.88	4.36	3
4–5	2	0.96	2.90	10	–0.68	7.45	2
4–5	3	0.02	2.99	17	–1.12	–	1
4–5	4	0.71	2.49	30	–0.53	0.75	2

Overall comparison: *z* score=–2.6, *p*=0.1. [a]Mean. GMFM–66, Gross Motor Function Measure–66; Norm, normative sample (subgroups aged 2–3 and 4–5; Russell et al. 2002.); SD, standard deviation. –, not calculated due to sample size of 1.

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'Weight and height gain after intrathecal baclofen pump implantation in children with spastic tetraparesis'

SIR–Chronic intrathecal baclofen (ITB) infusion is widely used to treat severe spasticity of cerebral and spinal origin in children. ITB therapy effectively improves spasticity, decreases muscle tone, increases the range of hamstring motion, and prevents musculoskeletal contractures both in spastic paraparesis and tetraparesis, thus improving hygiene and facilitating caregivers' duties.[1, 2]

Children with spasticity are often below the normal centile for height and weight when compared with healthy children without disability of the same age and sex. Nutritional problems in patients with moderate to severe cerebral palsy (CP) seem to arise mainly from mild or severe feeding dysfunction.[3, 4] Little is known about the role in malnutrition of other factors, such as spasticity.

In 2001 a report[5] suggested that by improving spasticity ITB therapy may also reduce caloric expenditure, thus normalizing children's growth.

We retrospectively collected weight and height data for three children who attended our service and underwent implantation of an ITB pump (Syncromed Model EL, 8627–10, Medtronic Inc, Minneapolis, USA). All children had spastic quadriplegia: patient 1, following post-anoxic damage after a cardiocirculatory arrest at age of 2 years and 6 months; patient 2, as a result of having CP; and patient 3, due to having chromosomal encephalopathy (deletion 22q11.2).

Children's age, sex, weight, height, and spasticity outcome measures are listed in Table I. The following scales were performed in order to assess spasticity: modified Ashworth scale; Penn Spasm Frequency Score; and Deep Tendon Reflexes Scale (see Table I legend for relative scores).

Firstly, these three patients show that ITB therapy markedly improves spasticity, thus helping to improve childcare and, thereby, inducing secondary benefits in general terms. Secondly, they show that ITB therapy achieves excellent benefits in terms of growth, with a conspicuous gain in weight and height.

After ITB therapy, functional outcome measures improved in all three children. Soon after ITB implantation, the modified Ashworth scale score decreased by at least 2 points, painful muscle spasms disappeared in patients 1 and 2 and decreased in frequency in patient 3. Hyperreflexia was less pronounced in all three patients. These excellent results remained unchanged throughout 18 months follow-up.

In agreement with others,[5] we believe that the caloric expenditure which is spent to sustain involuntary muscular contraction owing to spasticity could be shifted to provide the amount necessary for normal linear growth. Before ITB pump therapy, our patients showed a poor growth rate increase for their age and sex; however, after ITB pump therapy all patients achieved excellent growth, gaining rapidly and markedly in height and weight. Children who were under the normal centile values reached the normal centile (see Table I). Caregivers reported that, in all three children, food intake remained unchanged after ITB. The most sensitive measure related to improved patient spasticity was weight. Although height did increase in all children, a significant increase was only seen in patient 2. Conversely, weight increased significantly in all three children ($p < 0.05$, t-test for paired data). These changes could reflect a decreased total caloric expenditure owing to spasticity.

Our study was conducted retrospectively, and we could not give any objective data about children's caloric intake. We were only able to interview parents and caregivers about children's food intake.

In conclusion, we feel that children with CP must receive ITB therapy to reduce spasticity. The therapy reduces the number and severity of painful muscular spasms so that children sleep better and are easier to care for, thus benefiting rehabilitation and improving hygiene conditions, and gives excellent results in severe cerebral spasticity. Our results suggest that ITB therapy could also present an important secondary benefit: it may diminish caloric expenditure due to spasticity, thus, allowing the expenditure saved to be used for achieving normal linear growth. In order to investigate our hypothesis it would be necessary to carry out prospective trials to analyze nutritional

Table I: Weight, height, modified Ashworth scale score (MAS), Penn Spasm Frequency score (SFS), and Deep Tendon Reflexes Scale score (DTR) of three children with spastic quadriplegia following intrathecal baclofen pump implantation

| | Patient 1 (female) | | | Patient 2 (male) | | | Patient 3 (male) | | |
	Pre	Pump	Post	Pre	Pump	Post	Pre	Pump	Post
Age, y:m	7:0	8:0	9:6	4:6	5:6	7:0	2:5	3:5	4:11
Weight, kg	11	11	24	13	17	24	11	14	17
Centile	<3rd	<3rd	10th	<3rd	10th	50th[a]	5th	25th	75th[a]
Height, cm	102	108	119	96	103	133	82	91	96
Centile	<3rd	<3rd	3rd	10th	<3rd	90th[a]	<3rd	3rd	10th
MAS	3 bilat	3 bilat	0 bilat	4rl, 3ll	4rl, 3ll	2rl, 1ll	4 bilat	4 bilat	2 bilat
SFS	3 bilat	3 bilat	0 bilat	2 bilat	2 bilat	0 bilat	3 bilat	3 bilat	2 bilat
DTR	4 bilat	4 bilat	2 bilat	5 bilat	5 bilat	3 bilat	4 bilat	4 bilat	3 bilat

[a]$p < 0.05$. Pre, 12 months before intrathecal baclofen pump implantation; pump, at moment of implantation; post, 18 months after implantation; bilat, bilaterally; rl, right leg; ll, left leg. MAS score – 0, no increase in muscle tone; 1, slight increase in muscle tone, manifested by a catch and release or minimal resistance at end of range of motion when affected part is moved in flexion or extension; 1+, slight increase in muscle tone, manifested by a catch followed by minimal resistance throughout remainder (less than half) of range of motion; 2, more marked increase in muscle tone through most of range of motion, but affected part easily moved; 3, considerable increase in muscle tone, passive movement difficult; 4, affected parts rigid in flexion or extension. SFS score – 0, no spasms; 1, mild spasms induced by stimulation; 2, infrequent full spasms occurring <1/h; 3, spasms occurring >1/h; 4, spasms occurring >10/h. DTR score – 0, absent; 1, hyporeflexia; 2, normal; 3, mild hyperreflexia; 4, three or four beats clonus only; 5: clonus.

status and caloric intake before and after ITB therapy. We then believe that prospective trials with more dietetic, nutritional, and growth information are required to further investigate this issue.

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'A child with severe night terrors and sleep-walking responds to melatonin therapy'

SIR–Night terrors (pavor nocturnus) occur in up to 6% of children.[1] The onset of this parasomnia tends to be between 2 to 4 years of age and rarely persists into adulthood.

The clinical presentation is striking. Usually 1–2 hours after falling asleep the affected child will suddenly sit up with their eyes wide open. They are confused, and scream and struggle; it is impossible to reason with them, and their speech is unintelligible. There is rapid heart rate and respiration, profuse sweating, and extreme anxiety. When they walk during these episodes (somnambulism) they bump into objects and often sustain injuries. Generally these episodes last from 10–15 minutes and there is no subsequent recall of these events by the child.

Specific treatment is not required in milder cases. Parents are reassured and requested to protect, but not awaken, their children. In more severe cases, behavioural management techniques and low doses of benzodiazepines are used for night sedation.

A 12-year-old male with Asperger syndrome and marked chronic sleep-phase onset delay was referred for melatonin therapy. He also had severe coexistent night terrors and sleep-walking for several years. These episodes occurred 2–3 times almost every night. His sleep disorder affected his parents so much that they said they could no longer cope. The child was also regularly sustaining cuts and bruises. Oral administration of controlled release melatonin (5mg), 30 minutes before the desired bed time, corrected the sleep phase onset within two days. The night terrors and the sleep-walking episodes abruptly disappeared and have not recurred for over six months.

Melatonin therapy for sleep-phase onset delay is well described in the literature.[2] Night terrors and sleep-walking episodes are both due to a faulty transition from slow wave to REM sleep state. This process is executed by complex neuronal and hormonal mechanisms. The exact pathophysiology of these parasomnias is unclear.[3] It is known, however, that deep sleep predisposes children to these events, which is why they tend to occur most frequently 1–2 hours after falling asleep at night when the slow wave sleep cycle is deepest.

Due to our patient's severe sleep deprivation, he slept deeply. With melatonin therapy his sleep deprivation was corrected, which may explain the abrupt disappearance of his parasomnia. It is also quite likely that melatonin, in some way, corrects the faulty process of transition from slow wave to REM sleep in the brainstem, where this change occurs.

Over the years we have heard anecdotal reports of children whose night terrors subsided with melatonin therapy. We have also corresponded with paediatric sleep pathologists who have had similar observations (personal communication, Dr M Davey 2004).

We believe that well-designed melatonin trials for night terrors and sleep-walking episodes should be undertaken. As melatonin is remarkably free from adverse effects, it offers significant advantages over the use of benzodiazepines in severe cases.

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