Intrathecal Baclofen pump dosing for Dystonia: Two case studies

Carolyn Chowne RN, MScN, Sunny Hill Health Centre for Children, BC, Canada

Background

Intrathecal Baclofen (ITB) pumps are an established treatment for spasticity in children with Cerebral Palsy (CP). Dystonia is often present in conjunction with spasticity but can also be present on its own. Dystonia can create more functional barriers to activities and daily life. Episodes of extreme tone or ‘bostonic storms’ can be completely debilitating.

There are few medications for effective management of dystonia. Other treatments, such as Deep Brain stimulation, are invasive and not readily accessible to the wider population.

There is good support and knowledge around ITB pumps for treatment of spasticity but management of dystonia with ITB pumps requires significantly different dosing and support. There are no guidelines or supports to inform practice. Experienced multidisciplinary practitioners rely on their clinical judgement to make decisions.

Intrathecal baclofen can provide good control of dystonia and enhanced sharing of experiences and treatment methods will deepen our knowledge base and support best practice.

Case 1 “SG”

SG - a ten year old boy with mixed spastic/dystonic CP, GMFCS V, W 23 week prem with cognitive impairment and complex medical history. He is nonverbal, anti-seizure medication (AED) are required. He has recurrent Pneumonia, neurogenic bladder with frequent UTIs, osteopenia with spontaneous fracture of his right femur and frequent long lasting episodes of dystonic storms. Medications included: Baclofen, Trihexyphenidyl, Lansoprazole, Clonazepam, Bromocriptine, Gabapentin, Nitrofurantoin, Lorazepam, Ondansetron, Budesonide and Pulmicort, Gabapentin, Midazolam and Cannabinoid tetrathydrocaps

Baclofen therapy was started in 2008 with a 40 ml pump and a 6 hour period.

Intrathecal Baclofen pump was placed by neurosurgeon with catheter tip at level C-1.

Dosing:

Step 1: Initial dosing at 100mcg/24 hours simple continuous.

Step 2: Increase by 50 mcg/hr ultimately to 13 mcg/hr basal rate.

Step 4: Titrate increases to effect by up to 25% increases every 24 hours initially and eventually weekly to final dose of 220 mcg every 4 hours and basal rate of 20 mcg/hour total daily dose of 1789.4 mcg/day and discharged.

Other medications were decreased and discontinued including baclofen upon initial start of ITB therapy followed by fentanyl, morphine, Bromocriptine and Trihexyphenidyl.

In the 16 months since pump implantation he has had 4 hospitalizations for a total of 35 days. 7 days average length of stay.

Dosing has remained relatively steady with current dosing at 200 mcg q 4 hours and basal rate of 25 mcg/hour total daily dose of 1773.8 mcg/day.

Dosing might have caused.

Case 2 “MD”

MD - a 12 year old boy with mixed spastic/dystonic CP, GMFCS V, Ex 24 week prem with IVH. He is Gastrostomy tube and orally fed with significant scoliosis, previous bilateral hip surgeries and osteopenia. He is verbal and very social with some cognitive impairment. He is generally healthy with few hospital admissions until dystonic storm episode the winter prior to pump implant. Medications included: Baclofen, Clonazepam, Lansoprazole, Trihexyphenidyl, carbidopa/levodopa 25mg/100mg, dantrolene, gabapentin, and ondansetron, midazolam and cannabidoil tetrathydrocaps spray. In the 12 months prior to pump implant MD was hospitalised 3 times for 20-28 days each time with ICU stay to manage dystonic storms.

Intrathecal Baclofen pump was placed with catheter tip at T1 level after multiple attempts to advance further.

Dosing:

Step 1: Initial dosing at 100 mcg/24 hours simple continuous.

Step 2: Increased by 25-50 mcg q 4 hours for 4 days.

Step 3: Initiated complex/flex bolus dosing of 50 mcg q 4 hours with basal rate of 1 mcg/hr.

Step 4: Titrate dosing to effect weekly then monthly to family living quite far from centre. Final daily dose is 150 mcg bolus q 4 hours with 13mcg/hour basal rate for total daily dose of 1205 mcg/day.

Medication weaning started with baclofen upon initiation of ITB therapy followed by levodopa/carbidopa. Then in order, dantrolene, gabapentin, midazolam and cannabidoil tetrathydrocaps were reduced but not weaned completely. After a few months Trihexyphenidyl was also reintroduced at a reduced dose from previous levels. Pre-medication of cannabidoil remains but also remains by orally spastic.

MD has successfully undergone scoliosis surgery with a remarkable recovery and has not been hospitalised for any other reasons since ITB therapy.

Dose remains at 1205 mcg/day using 150 mcg bolus q 4 hours with 13mcg/hour basal rate.

Discussion/Significance

Dosing: Treatment of dystonia differs significantly from that of the treatment of spasticity. Typical dosing for spasticity involves titrating simple continuous doses to effect, often a total daily dose of 250 to 500 mcg/day.

Dosing for dystonia involves significantly higher daily doses and complex, aggressive dosing with regular boluses and continuous basal rates. The step wise approach of increasing the total daily dose and then initiating the complex/bolus dosing provided an underlying medication base to work from. This hopefully reduced the potential of droveness, drop in BP, respiratory rate or other potential negative side effects that sudden introduction of bolus dosing might have caused.

Catheter placement: Optimal catheter placement is in the C-1-3 level, with some centers placing them intraventricularly based on better effect in the basal ganglia. However, due to physiologic challenges this is not always an option. Fluoroscopic guidance in placement can be of benefit.

Pump size: Due to the higher levels of drug needed for dosing the pump size plays a considerable role in optimizing care and reducing travel and appointment burden on patients and families. Both patients were quite small and maintaining and gaining weight was an ongoing issue. It was eventually decided to use the 40mc pumps and carefully monitor skin condition and weight. A benefit of the significant dystonia reduction for both these patients has been weight gain so concerns about pump size have been mitigated.

Understanding how to meet the needs of the Dystonic CP population will lead to better overall care and improved quality of life for these patients and their families, reduced hospital stays, lower incidence of polypharmacy and reduced risk of negative medication effects. Sharing our experiences will build a body of knowledge and evidence to help in the development of practice guidelines and promote best practice.

References


Acknowledgements

Thanks to Dr. Esias Van Rensburg of Sunny Hill Health Centre for Children, Dr. Ash Singhal and Dr. Raj Heran of British Columbia Children’s Hospital.

Thanks to Dr. Esias Van Rensburg of Sunny Hill Health Centre for Children, Vancouver, BC, Canada.