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Intrathecal Baclofen to Improve Functional Status in ALS: A Case Report

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Background: Intrathecal pumps are well known to benefit patients with chronic pain as well as spasticity. Intrathecal baclofen (ITB) can offer doses 100-1000 times smaller with similar efficacy, compared to oral baclofen. Only 2 previous reports detailed improvement in functional status after patients with amyotrophic lateral sclerosis (ALS) received ITB.

Case Report: Our patient presented with progressive bulbar palsy, further progressing to ALS. His lower extremity spasticity and tremors continued to progress over 3 years despite increased baclofen. At the time of implant, he expressed whole body tremors and spasticity to bilateral lower extremities, complicated by falls. Prior to the trial, the patient ambulated 50 feet. ITB was started at a rate of 100 mcg/day. After the implant, the patient's ambulation distance increased to 100 feet.

Conclusion: The patient and his wife reported resolution of his tremors and improvement in spasticity. This report details the functional improvement obtained from ITB in a patient with ALS.

Key words: Amyotrophic lateral sclerosis (ALS), intrathecal, baclofen, functional status, chronic pain, interventional pain

BACKGROUND

Intrathecal infusion systems are documented to benefit those with chronic pain (1-5) and those with spasticity (1-9). Intrathecal baclofen (ITB) is a well-studied intrathecal infusion for spasticity (1-9), offering doses 100-1000 times smaller with similar efficacy to its oral formula (9). Minimal literature discusses the use of baclofen to treat the spasticity in the setting of amyotrophic lateral sclerosis (ALS) (10,11). This case report documents a patient whose functional status benefitted from an ITB infusion system.

This case report is devoid of the patient's identifiable information; therefore, it is exempt from IRB review requirements as per the policy of our institution. Informed consent was obtained prior to publication of this report.

CASE REPORT

A 56-year-old man presented to our institution's neurology department one year after this progressive dysphagia and dysarthria symptoms began. During that year, he developed symptoms affecting his left hand and his speech. After his initial presentation, he was diagnosed with progressive bulbar palsy. The diagnosis was later changed to ALS as his disease progressed. Oral baclofen (5 mg twice a day) was started 2 years after his initial symptoms. Baclofen was initially prescribed for upper extremity spasticity, which then progressed to lower extremity spasms and ensuing balance issues a few months later. His lower extremity spasticity, tremors, clonus continued to progress over the next 3 years despite increased baclofen – 10 mg 3 times a day. He experienced significant fatigue secondary to the escalat-

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ing dose of baclofen despite also taking armodafinil. At the time of initial evaluation at the pain management center (4 years after presenting to neurology, 5 years after symptom onset), he expressed whole body tremors to bilateral lower extremities, complicated by falls from bed. He also experienced severe lower extremity spasticity.

After discussion with neurology colleagues and the patient, it was decided to move forward with a continuous ITB trial. After implantation of the trial pump, the infusion was started at 5 mcg/hour and his oral baclofen dose was reduced by 50%. During the 4-day continuous ITB trial, a 1-point decrease in his tone, via the Modified Ashworth Scale (13), was noted in his triceps, biceps (1+ to 1), quadriceps, and hamstrings (3 to 2). During the trial the physical therapy team noted an improvement of ambulation from 7 to 20 meters between postoperative day (POD) 1 and 2. The patient reported improvements in spasms and tremors and wished to move forward with permanent implant.

Permanent Implantation

The L2-L3 intralaminar space was accessed, and a Medtronic Ascenda spinal infusion catheter (Medtronic, Inc., Minneapolis, MN) was threaded to the T10 vertebral body. The catheter was tunneled underneath the flank and connected to a Medtronic Synchromed II Intrathecal Pump with 40 mL reservoir. The pump was filled with baclofen 300 mcg/mL and started at a rate of 100 mcg/day.

Follow-up

On POD 1, the baclofen infusion was increased to 120 mcg/day with continuation of oral baclofen 10 mg twice a day and 5 mg oral baclofen as needed every 4 hours. The patient described a few tremors the night of POD 0-1 and endorsed slight stiffness in his ankles. Patient rated his pain 0/10 at rest and 3/10 with activity. In the evening of POD 1, baclofen infusion was increased to 140 mcg/day due to increasing lower extremity spasticity. POD 2, intrathecal infusion rate was increased to 155 mcg/day and oral baclofen dosing decreased to 5 mg twice a day, 5 mg as needed every 4 hours. Patient continued to report improvement in spasticity in the bilateral lower extremities and was later discharged on POD 2 to inpatient rehab for further optimization of ITB dosing.

While at inpatient rehab, the ITB dose remained at 155 mcg/day. While working with physical therapy, he was able to ambulate 100 feet with a rolling walker, double the ambulation distance prior to implantation. At the first clinic visit (POD 10), the patient and his wife reported complete resolution of his tremors and nocturia. They noted improvement in his leg heaviness and spasticity. He stopped taking oral baclofen at this time. His Modified Ashworth Scale improved to one for the left lower extremities and one for the right lower extremities.

At the second clinic visit (POD 66), he noted one tremor per day for a few seconds, which was broken with manual stretching. He noted his legs feeling very heavy at times, thus the ITP infusion regimen was transitioned to patient-controlled boluses of baclofen (personal therapy manager, PTM) with a low basal rate (PTM bolus 5 mcg/1 min, 3 hr lockout with 145 mcg/day basal rate). The patient and his wife were pleased with his symptoms and functional improvement since his ITB infusion began.

DISCUSSION

Baclofen, tizanidine, memantine, benzodiazepines, dantrolene are oral myorelaxant agents mentioned in the literature (12). Because of baclofen's poor lipid solubility, high oral doses are required to reach therapeutic levels in the CSF (9). These doses lead to side effects in 25-70% of patients (9), chiefly muscle weakness, sedation, nausea, dizziness (2,3,9). Increasing doses of baclofen do not necessarily correspond with higher levels in the CSF (9). ITB is the best studied intrathecal infusion for spasticity (1-9). ITB offers several advantages. Because it is infused directly at the target site, doses 100-1000 times smaller offer similar efficacy to oral administration (9). In addition, ITB's efficacy did not diminish over a period of 5 years (9). Adverse effects of intrathecal administration are similar to oral administration (2,3,9), at rates of 4.4 – 54% (9), most commonly during the titration phase of ITB. Withdrawal is possible during titration, with risk of rhabdomyolysis and multisystem organ dysfunction (9). There are also device and procedure side effects, infection, catheter migration/disconnection, pump dysfunction, CSF leak, and spinal headache (9).

The best studied pathologies that lead to spasticity severe enough to require ITB are multiple sclerosis, spinal cord injury, and stroke (1). However, minimal literature discusses the use of ITB to treat ALS specifically. McClelland et al (4) looked into ITB to manage pain from ALS induced spasticity. This study focuses on pain control,

briefly mentioning their cohort's mean Ashworth scale (an agreed upon clinical tool to quantify spasticity) (13). Two case reports have detailed improvement in functional status after ITB was initiated in patients with ALS (10,11). The later of the 2 case reports (1) documents the functional status of the patient, noting their patient was able to walk without assistance and climb stairs. The patient described was able to tolerate 540 mcg/day as their condition progressed, maintaining his ability to complete his activities of daily living (10).

This case corresponds well to the documented functional improvement by Marquardt and Seifert (11). Our patient was able to double the distance he was able to ambulate and showed improvement in his Modified Ashworth Scale. His nocturia also resolved. In addition, he was satisfied with the improvement in his spasticity and tremors.

CONCLUSION

This case report demonstrates that ITB provided significant improvement specifically in the functional status of this patient with ALS as well as excellent relief from spasticity related pain. Further research can be done to quantify and verify these results.

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