

Case Report

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An efficient multidisciplinary approach in a pregnant patient with hereditary spastic paraparesis treated by intrathecal baclofen therapy: A case report

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Abstract

Literature regarding cases of pregnant patients with hereditary spastic paraparesis (Strumpell-Lorrain disease) and those treated by intrathecal baclofen therapy is sparse. There are currently no specific guidelines to manage pregnancy and delivery in these patients. A 42-year-old woman presented to our clinic with hereditary spastic paraparesis and was treated with intrathecal baclofen for spasticity. She conceived twice and delivered successfully by caesarean section under subarachnoid anaesthesia. The two pregnancies had favourable outcomes with no increase in spasticity and no problems encountered with the intrathecal device. In this report, we discuss the evolution of both pregnancies and the anaesthetic management for deliveries in the presence of an intrathecal catheter.

Keywords: Caesarean section; case report; hereditary spastic paraparesis; intrathecal baclofen; pregnancy.

Introduction

Hereditary Spastic Paraparesis (HSP), originally described in 1880 by Strumpell-Lorrain and also known as Strumpell-Lorrain disease, is a rare, genetic neurodegenerative disorder (3–10 cases per 100,000 population in Europe) characterised clinically by progressive spasticity and weakness of the lower limbs due to axonal degeneration in the corticospinal tracts and dorsal columns of the spinal cord [1]. The disease has been divided into uncomplicated and complicated forms. Complicated forms are less frequent and associated with other neurological disorders. There is a dearth in the literature regarding cases of pregnant patients with HSP and those treated by intrathecal baclofen therapy. There are currently no specific guidelines to manage pregnancy and delivery in these patients. We describe a successful multidisciplinary approach to manage pregnancy and delivery in a patient with HSP who underwent two caesarean sections.

Case report

The patient provided written informed consent for this report.

A 42-year-old woman presented to our centre in 1998 with uncomplicated HSP that had been diagnosed in 1994. Due to increasing spasticity in both lower limbs limiting her gait, she was treated with Intrathecal Baclofen (ITB) using a pump (Medtronic Synchronised II) that was implanted in her left hypochondrium in 2004. The catheter was replaced in 2005 and 2007 (in the L2–L3 space) due to a rupture in the interspinous space, and the pump was replaced in 2011. Since 2011, the daily baclofen dosage has been 150 µg/day, which allows adequate spasm control and facilitates routine activities. The patient has used a wheelchair since 2013 due to disease progression.

Based on favourable genetic advice, she underwent *in vitro* fertilisation in 2013. Unfortunately, she had a miscarriage due

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to amniotic sac puncture during a prenatal diagnosis.

In 2015, she became pregnant with a favourable clinical evolution; no change in spasticity, and no problems or complications were observed with the ITB pump, which had shifted slightly to the left without discomfort and remained easy to refill. In December 2015, she gave birth to a boy weighing 3.8 kg at 38 weeks of amenorrhea by caesarean section. Caesarean section was planned due to gestational hypertension, breech presentation, and HSP. After a pre-anaesthetic assessment, spinal anaesthesia was administered in the L5–S1 space using a 25-gauge Whitacre needle to inject 10 mg hyperbaric Marcaine plus 2.5 µg sufentanil to obtain a T4-level block. The patient developed significant hypotension, which was treated with fluid administration, ephedrine, and neosynephrine. The Apgar score was 5 at 1 min and 8 at 5 min, due to which the newborn was admitted to the neonatal unit for 24 h; both mother and child were discharged 5 days later. The patient opted to breastfeed. Two weeks later, she developed postpartum suppurative ovarian vein thrombophlebitis, which was treated successfully with antibiotics.

In 2017, she conceived again with a favourable clinical evolution. She gave birth to a girl weighing 3.2 kg in February 2018 via a planned caesarean section due to HSP and a scarred uterus under spinal anaesthesia in the L4–L5 space with a T4-level block obtained with 10 mg hyperbaric Marcaine and 2.5 µg sufentanil. The Apgar score was 8 at 1 min and 9 at 5 min. The patient was discharged after 3 days. One month later, she developed an appendicitis-associated abscess that was treated successfully with surgery and antibiotics.

The ITB pump was replaced 3 months after delivery due to a drained battery. Due to disease progression, long after the pregnancies, the baclofen dosage has been progressively increased to 300 µg/day currently.

Discussion

Genetic transmission of HSP may be characterised as either autosomal dominant (70%) or recessive. Family history is frequently observed; our patient's father developed spasticity between the ages of 45 and 50.

The treatment of spasticity includes administration of baclofen, a gamma-aminobutyric acid agonist that inhibits mono- and polysynaptic spinal reflexes. For patients who are unresponsive to or experience side-effects with oral anti-spastic medication, ITB is a highly effective treatment [2,3]. The treatment goal is to reduce spasticity while preserving and/or improving mobility functions.

Little is known about the effects of baclofen during pregnancy. Oral baclofen has been shown to increase the incidence of omphalocele in rat foetuses; however, there have been no studies on its effects in pregnant women. One case in the literature reports withdrawal symptoms in a newborn whose mother had been treated with oral baclofen [4]. The intrathecal dose is 100- to 1000-times lower than the oral dose and systemic absorption is <1% of the intrathecal dose; thus, ITB is considered safe for pregnancy and breastfeeding. To date, there have been no reports of adverse effects in the development of children born to patients who have been administered ITB [5,6]. However,

studies with larger sample sizes are recommended to further confirm the efficacy and safety of ITB.

The literature on pregnancies in patients with HSP and those treated by ITB is sparse [6]. No case reports, until now, have described the use of spinal anaesthesia for caesarean section in patients with HSP with an ITB pump.

We were unaware of the effects of a gravid uterus on the ITB pump and catheter, and those of pregnancy-associated physiological changes on spasticity.

The ITB pump was placed in the left hypochondrium and had moved slightly laterally during the pregnancy without causing any discomfort to the patient; there was no erosion or difficulty in medication refilling either. Moreover, it was located away from the region of the usual Pfannenstiel incision performed for caesarean sections, leading to no interference.

The catheter, which was replaced twice due to breakage in the interspinous space, was long enough to avoid stretching or disconnection from the pump. No reports have described such a complication, and it did not occur during our patient's pregnancies [7,8].

Spasticity has been noted to increase with pregnancy in 10–20% of patients with spinal cord injury [9]. It may result from noxious stimuli from conditions such as urinary tract infections, constipation, pressure injuries, and deep vein thrombosis, which has an increased incidence in women with spinal cord injury. Our patient did not experience any increase in spasticity, and the intrathecal dosage was maintained at 150 µg/day despite postpartum infectious complications.

Although vaginal delivery was possible [10], a caesarean section was planned by the consulting obstetrician due to breech presentation, gestational hypertension, and HSP. Anaesthesia management for delivery was a challenge and required a multidisciplinary approach [11].

In the presence of ITB, general anaesthesia is considered safe for the integrity of the catheter; however, this technique is known to be associated with a high risk of morbidity and mortality due to anaphylactic reactions and Mendelson syndrome [12], and is rarely used in elective cases. Regional anaesthesia was planned after an anaesthetic assessment with a multidisciplinary approach. McTiernan has suggested that an epidural has theoretical advantages over spinal anaesthesia [13], as it allows slow titration of the local anaesthetics, affording a greater control of the anaesthesia level to prevent development of hypotension and respiratory difficulties. This has been confirmed by Ali Sakr Esa *et al.*, who emphasised the need for a multidisciplinary approach [14]. Knowing the position of the intrathecal catheter during needle insertion is important to prevent damage to the catheter; therefore, anaesthesia administration should be performed on levels below the catheter [14,15]. The procedure should be performed under sterile conditions with the administration of prophylactic antibiotics to account for the increased risk of infection due to the presence of foreign material.

Finally, the anaesthesiologist, who was the same for both caesarean sections and was aware of the intrathecal catheter

position, opted for spinal anaesthesia, which is the most used technique for caesarean sections and has superiority over epidural in terms of speed and efficiency [12]. To the best of our knowledge, only two such cases have previously been described in patients with HSP [16,17]. Our patient did not encounter any complications except hypotension, which is a frequent occurrence (50–90% of all cases) during spinal anaesthesia for caesarean sections [12]. The injection sites in both surgeries based on anatomical landmarks were L5–S1 and L4–L5, respectively; i.e., three and two levels below the intrathecal catheter, respectively. However, identifying lumbar interspaces by palpation may lead to mistakes in 50–71% of all cases [18]. Thus, the use of ultrasound is recommended to improve the efficacy and safety of neuraxial anaesthesia, especially if an intrathecal catheter is already in place [19].

Our patient chose to breastfeed her two babies due to its nutritional, immunological, and relationship-based benefits. It has been previously demonstrated that during ITB, a very low concentration of baclofen is secreted in breast milk, leading to its approval for breastfeeding by the American Academy of Pediatrics [5,6]. Unfortunately, our patient had to interrupt breastfeeding due to postpartum infections requiring hospitalisation.

Conclusions

Repeated pregnancies in a patient with HSP treated with ITB may be safe and uncomplicated, provided a multidisciplinary approach is opted for, combined with regular follow-ups. Spinal anaesthesia, which is the most used procedure for caesarean sections, is safe and easy to perform, on the condition that ultrasound is used to avoid damage to the intrathecal catheter. Breastfeeding may be encouraged in such cases.

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