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CASE REPORT

Anaesthesia for caesarean section in a patient with dopa-responsive dystonia or Segawa's syndrome

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ABSTRACT
Dopa-responsive dystonia, also known as hereditary progressive dystonia with diurnal fluctuation, or Segawa's syndrome, is a rare hereditary progressive dystonia with two striking clinical features: a marked diurnal fluctuation of symptoms with symptoms worsening throughout the day and improving after sleep, and a dramatic response to levodopa therapy. Whilst rare, it is treatable, with function being normal or near normal after levodopa therapy. We present our experience of providing anaesthesia for caesarean section in a patient with dopa-responsive dystonia and discuss the safety of levodopa therapy during pregnancy and the anaesthetic management of these patients.

Keywords: Dopa-responsive dystonia; Pregnancy; Caesarean section; Anaesthesia; Combined spinal-epidural

Introduction

Dopa-responsive dystonia (DRD), also known as hereditary progressive dystonia with diurnal fluctuation or Segawa's syndrome, was first described in Japan in 1971.[1,2] Cases have since been reported worldwide.[3–8] The main pathological lesion is a functional defect of dopaminergic tracts in the basal ganglia, which is due to mutation in the guanosine triphosphate cyclohydrolase 1 (GCH-1) gene in 80–90% of cases.[9–12] DRD typically presents in the first decade of life and is a rare hereditary progressive dystonia with two striking clinical features: a marked diurnal fluctuation of symptoms with symptoms worsening throughout the day and markedly improving after sleep, and a dramatic response to levodopa therapy.[2,9,13] It is important to make the diagnosis of DRD, as whilst rare, it is treatable, with function being normal or near normal with levodopa therapy.[3,8]

We present our experience of providing anaesthesia for caesarean section in a patient with DRD. The case is important for a number of reasons including the rarity of the condition and the limited information in the literature regarding anaesthesia in DRD. A literature search revealed only two reported cases of anaesthesia in DRD, both being for caesarean section, one under general the other under epidural anaesthesia, in the same patient who continued levodopa therapy during pregnancy.[8] Our case differs in that our patient discontinued levodopa therapy before conception due to a lack of consensus regarding its safety in pregnancy. Whether levodopa therapy has continued is of relevance to the anaesthetist and we discuss its safety during pregnancy and the anaesthetic management of these patients.

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Case report

A 29-year-old primiparous woman with DRD presented for elective caesarean section as she was experiencing intermittent, uncontrollable leg spasms that would have significantly interfered with labour and vaginal delivery. The spasms were a result of discontinuation of her levodopa therapy. She had previously been taking co-careldopa, which consists of levodopa and the peripheral dopa-decarboxylase inhibitor (pDDI) carbidopa. After discussion with her neurologist and obstetrician she and her partner had been adamant that they would discontinue her co-careldopa during conception and pregnancy because of concerns regarding its safety in pregnancy.

The woman had been born prematurely at 34 weeks of gestation by caesarean section. After delivery, she spent approximately six weeks on the special care baby unit suffering from respiratory distress and severe jaundice. As a child she had sufficient strength and co-ordination to be able to run but had always been clumsy and experienced frequent falls. She began to develop further

problems in her early teenage years with pain and muscle spasms around her right knee, these problems being initially ascribed to knee injuries. However, her condition gradually deteriorated with dystonia, significant abnormal muscle spasms and postures, which necessitated the use of crutches and eventually a wheelchair. The spasms initially only affected the right leg but gradually the problem ascended to involve her axial muscles. Despite numerous investigations and trials of a variety of medical therapies her situation did not improve and she underwent a number of orthopaedic procedures including a right ankle fusion. The diagnosis of DRD was made while she was at university and her condition improved dramatically after starting co-careldopa. This enabled her to have an almost normal gait and way of life. At around this time she also had a tendon transfer performed on her ankle, which further improved her mobility. She was maintained on a total daily dose of levodopa 100 mg and carbidopa 400 mg. Although she had no family history of dystonia, she was well informed about the hereditary nature of DRD and received genetic counselling both pre-pregnancy and in the first trimester.

Her past medical history also included mild asthma and bronchiectasis for which she took regular corticosteroid and selective β_2-agonist inhalers. She suffered from a cough and underwent daily postural drainage. She had undergone several procedures under general, spinal and epidural anaesthesia. After general anaesthesia she had suffered from postoperative nausea and vomiting. On one occasion she had been admitted to intensive care because of her asthma and bronchiectasis; she had required continuous positive airway pressure (CPAP) for 24 h but was not ventilated.

The patient discontinued her levodopa therapy during attempts to conceive and became pregnant after three months. No levodopa was taken throughout the pregnancy. She experienced clumsiness and lack of co-ordination in her lower limbs by six weeks, needed crutches by the end of the first trimester and nearing the end of the third trimester required a wheelchair. She experienced uncontrollable, intermittent leg spasms, rigidity, pain and some falls; her symptoms were worse towards the end of the day and when she was tired. These symptoms were similar to those she had suffered before starting levodopa treatment. The pregnancy was otherwise relatively uneventful with morning sickness and one attendance to hospital with possible spontaneous rupture of membranes for which she was not admitted. As her blood group was O rhesus negative, she was given prophylactic anti-D immunoglobulin after the falls.

Pregnancy did not exacerbate her asthma or bronchiectasis, although postural drainage had become impossible later in pregnancy due to severe reflux. As she had not experienced any respiratory compromise, lung function tests were not performed.

A decision to deliver by elective caesarean section was made at 22 weeks of gestation because of maternal and obstetric concerns that dystonic symptoms would interfere with labour and vaginal delivery. She was pre-assessed by a consultant anaesthetist at 28 weeks when it was noted that she was 162 cm tall and had weighed 48 kg at booking (BMI 18 kg/m^2). Airway assessment revealed a Mallampati class 1 airway with no limitations of mouth opening, jaw protrusion or neck extension. In view of her asthma and bronchiectasis it was felt that a regional technique would be beneficial and it was decided to perform the elective caesarean section under combined spinal-epidural (CSE) anaesthesia. A physiotherapist was involved to assist with alternative methods of postural drainage in order to reduce mucus retention.

Pre-operatively she received ranitidine, metoclopramide and sodium citrate. Intravenous access was secured with a 16-gauge cannula and Hartmann's solution started. As is our practice, regional anaesthesia was established with a two space CSE technique with the patient sitting. Using an aseptic technique and 1% lidocaine to the skin, an epidural catheter was sited first in the L2/3 space via a 16-gauge Tuohy needle following loss of resistance to saline. The spinal was then performed at the L3/4 level with a 25-gauge Whitacre needle and 2.3 mL of 0.5% heavy bupivacaine plus fentanyl 25 μg. Both procedures were uneventful. The patient was then positioned in right lateral and then supine with a left lateral tilt. A block to cold between T4 and S5 bilaterally was achieved. Anaesthesia to pinprick was also tested over the incision site before surgery. At the time the caesarean section took place it was not routine practice to test the block with light touch. Immediately after delivery of the baby, Syntocinon was administered by bolus followed by a 4-h infusion, and a prophylactic dose of co-amoxiclav was given. A further litre of Hartmann's solution was given during the caesarean section. Surgery was uneventful and the patient remained haemodynamically stable. A healthy female infant was delivered weighing 3400 g, with Apgar scores of 9 at 1 and 5 min, and venous and arterial cord pH of 7.243 and 7.326 respectively. Postoperatively oral analgesics, prophylactic low-molecular-weight heparin and intravenous fluid were prescribed. Her respiratory function was not compromised by regional anaesthesia or caesarean section and she was able to recommence her normal method of postural drainage immediately post partum. An epidural infusion of 0.1% bupivacaine with fentanyl 2 μg/mL was continued at 7 mL/h for two days to provide analgesia and to help reduce uncomfortable muscle spasms. The epidural and urinary catheters were removed on day 3 and mother and baby were discharged home on the fourth postoperative day.

On the day of delivery the patient re-started her co-careldopa. Initially, her symptoms prevented her from providing full care for her baby and additional support