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Failure of neuraxial anaesthesia in a patient with Velocardiofacial syndrome

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ABSTRACT

Velocardiofacial or 22q11 deletion syndrome is a genetic condition caused by deletion 22q11, the deletion of a small segment of the long arm of chromosome 22. To our knowledge this is the first case report of a woman with Velocardiofacial syndrome presenting in late pregnancy for caesarean delivery. She had undergone a Tetralogy of Fallot repair as an infant and had residual pulmonary regurgitation. In addition examination revealed micrognathia and scoliosis. Neuraxial anaesthesia was unsuccessful and subsequent conversion to general anaesthesia was necessary despite concerns regarding her facial abnormalities, pulmonary regurgitation and mild intellectual impairment.

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Keywords: Velocardiofacial syndrome; Tetralogy of Fallot; Scoliosis; Caesarean section; Anaesthesia: Spinal, Epidural, General

Introduction

Deletion of a small segment of the long arm of chromosome 22, deletion 22q11 causes several syndromes such

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as Velocardiofacial syndrome (VCFS) and DiGeorge syndrome. The estimated prevalence is 1:2000, making it the second most common genetic syndrome. Common presenting features include facial dysmorphism, cleft palate, cardiac defects, learning impairment and thymic hypoplasia. Over 180 possible features are associated, therefore clinical presentation is variable. Previously, affected children rarely survived to adulthood, but with advances in the treatment of cardiac and palatal abnor-

V. Cohen et al.

malities, more women with the condition are surviving to childbearing age.¹

Case report

An 18-year-old woman with VCFS was diagnosed as pregnant at 34 weeks of gestation following a plain spinal radiograph for scoliosis surveillance. She attended the obstetric high-risk clinic and, following multidisciplinary discussion, caesarean delivery was planned at 38 weeks. Her medical problems included thoracolumbar scoliosis (Fig. 1) under conservative management, psoriatic arthropathy, rheumatoid arthritis and Hashimoto's thyroiditis. She also had severe pulmonary regurgitation resulting from tetralogy of Fallot, which had previously been treated by Blalock shunt and intracardiac repair as an infant, and impaired right ventricular systolic function (right ventricular ejection fraction 48%). Left ventricular size and function were normal (left ventricular ejection fraction 76%). The patient was on the waiting list for pulmonary valve replacement. An exercise stress test according to the Bruce protocol had deteriorated over the previous four years from 9 to 6 min.

Other features of VCFS included mild intellectual impairment, submucous cleft palate and micrognathia (Fig. 2). Airway assessment revealed a Mallampati



Fig. 1 Photograph demonstrating thoracolumbar scoliosis (reproduced with patient's consent).

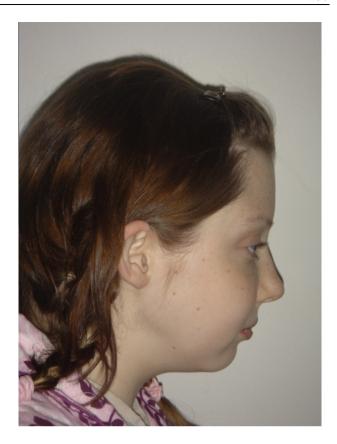


Fig. 2 Photograph demonstrating micrognathia (reproduced with patient's consent).

grade 3 view, and a thyromental distance of 4 cm. However, her mouth opening, neck movement and weight were normal, and tracheal intubation was thought likely to be straightforward. She was 156 cm tall and weighed 57 kg (body mass index 23 kg/m²). Although mild intellectual impairment had been diagnosed, she appeared to understand the implications of anaesthesia. A cardiac anaesthetic consultation indicated that general anaesthesia, if required, was likely to be uneventful.

On the day of surgery, baseline blood pressure was 130/95 mmHg, heart rate 95 beats/min and oxygen saturation (SpO₂) 98% on air. Following routine non-invasive monitoring and intravenous cannulae placement, an arterial cannula was inserted. The lumbar spine appeared unaffected by scoliosis, and combined spinalepidural (CSE) anaesthesia was attempted using a needle-through-needle technique. The epidural and intrathecal spaces were identified readily and 7.5 mg of 0.5% hyperbaric bupivacaine 7.5 mg, morphine 100 μg and fentanyl 15 µg fentanyl were injected intrathecally, after which the epidural catheter was inserted. Intravenous ephedrine 9 mg maintained cardiovascular stability. Using both loss of cold and light touch, sensory block was present to T4 bilaterally but with missed segments on the right from T12-L3, which was unchanged after 10 min. Incremental epidural boluses of 2% lidocaine with 1 in adrenaline 200 000 to a total of 20 mL,

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