Anesthetic considerations in a parturient with Oral-Facial-Digital syndrome and repaired tetralogy of Fallot with left ventricular dysfunction

J.E. McKinnie, A.N. Abdullah, J. Ramanathan
Department of Anesthesiology, University of Tennessee Health Sciences Center, Memphis, TN, USA

ABSTRACT
Oral-Facial-Digital syndrome or Mohr syndrome is a rare congenital disorder characterized by malformations of face, oral cavity, laryngeal structures, trachea, and digits, muscular-skeletal abnormalities, and congenital cardiac defects. In this case report, we describe the anesthetic management of a parturient with Oral-Facial-Digital syndrome type II and repaired tetralogy of Fallot with left ventricular dysfunction.
© 2014 Elsevier Ltd. All rights reserved.

Keywords: Oral-Facial-Digital syndrome type II; Mohr syndrome; Obstetric anesthesia

Introduction

Oral-Facial-Digital syndrome (OFDS) or Mohr syndrome is an autosomal recessive inherited developmental disorder characterized by facial dysmorphism, absence of medial incisors, cleft lip, cleft palate, high-arched palate, poly-lobed tongue, hamartomas of the tongue, abnormalities of the epiglottis and larynx and polydactyly of hands and feet.[1–3] Other comorbidities include severe scoliosis and pectus excavatum. In addition, patients with OFDS frequently have various cardiac anomalies such as an atrioventricular canal, atrial and ventricular septal defects, tetralogy of Fallot (TOF) and valvular stenosis.[4–7] In this report, we present the anesthetic management of a pregnant patient with OFDS type II and repaired TOF with deteriorating left ventricular function.

Case report

A 25-year-old G2P1 woman at 39 weeks of gestation with a medical history of OFDS type II and repaired TOF was admitted for induction of labor. After the TOF repair in early childhood, she had required pulmonary valve replacement with bioprosthetic valves four years before the current pregnancy. A year later she became pregnant with her first child and before delivery an echocardiogram showed normal left ventricular function with an ejection fraction (EF) of 60–65%. Epidural analgesia was administered successfully for labor and spontaneous vaginal delivery. One year before the current pregnancy, her cardiac status had deteriorated once again, and she underwent mitral valve replacement with bioprosthetic valves and right ventricle to pulmonary artery conduit replacement.

During this pregnancy, she was hospitalized at 17 and 28 weeks with congestive heart failure (CHF). She was treated with bed rest, digoxin and diuretics, and was discharged home two days after each admission. Her condition was classified as New York Heart Association Functional Class 2 in stable CHF. A week before admission for induction of labor, an echocardiogram revealed an EF of 45–55%, moderate mitral valve regurgitation, mild-moderate tricuspid regurgitation, a thickened right ventricular wall, mild pulmonary hypertension and normal chambers. Given her deteriorating cardiac status, a multidisciplinary meeting of obstetricians, anesthesiologist and neonatologist advised induction of labor, epidural analgesia and vaginal delivery with forceps assistance in the second stage.

On admission, her vital signs were blood pressure (BP) 123/72 mmHg, heart rate 71 beats/min, respiratory rate 17 breaths/min and temperature 36.1°C. She was 162 cm tall, weighed 63 kg, with a body mass index (BMI) of 23.9 kg/m^2. Her medications included daily digoxin 0.125 mg and furosemide 10 mg as prescribed by her cardiologist, and antenatal vitamins: she was not prescribed anticoagulants. Physical findings related to OFDS type II were frontal bossing, malar flattening of her left cheek, a broad nasal bridge, protrusion of the left side of the jaw, indentation on the left side of the tongue following hamartoma removal, and abnormalities involving fingers and toes. Airway examination showed a Mallampati class II airway and a thyromental distance of 6 cm. Cardiovascular examination revealed a midline scar, no jugular venous distension, a pansystolic murmur over the mitral and tricuspid area, and an ejection systolic murmur over the pulmonic area. Her lung

Accepted December 2013
Correspondence to: J. Ramanathan, Department of Anesthesiology, University of Tennessee Health Sciences Center, Memphis, TN, USA.
E-mail address: jramanathan@uthsc.edu

fields were clear on auscultation. Extremities showed no cyanosis or clubbing. Her electrocardiogram (ECG) showed sinus rhythm, right-bundle branch block, and T wave abnormalities. Her hemoglobin was 9 g/dL with a normal coagulation profile. Fetal heart rate was 135 beats/min with a normal pattern.

She received an intravenous infusion of lactated Ringer's solution at 100 mL/h. A urinary catheter was inserted. Monitoring included continuous ECG, continuous BP monitoring with a 20-gauge right radial arterial line and finger pulse oximetry. She received a co-load bolus of lactated Ringer's solution in two 250 mL increments during epidural placement. An epidural catheter was placed in the L3-4 interspace and a 3 mL test dose of 1.5% lidocaine with epinephrine 1:200 000 was given. A 10 mL loading dose of 0.125% bupivacaine with fentanyl 5 μg/mL, given in 3–4 mL increments, produced excellent analgesia, and was followed by a continuous infusion at 12 mL/h. This was higher than our institutional routine of 6–8 mL/h but was used to obtain a dense sensory block for the anticipated forceps-assisted delivery.

Labor progressed normally and, 5 h later, assisted forceps delivery without maternal pushing occurred. Apgar scores were 8 and 9 at 1 and 5 min, respectively. Following delivery of the placenta, oxytocin 20 U diluted in saline 500 mL was given as slow intravenous infusion over 2 h. Estimated blood loss was 250 mL. After delivery, prophylactic intravenous furosemide 20 mg was given to avoid postpartum pulmonary edema. Her digoxin dose was increased to 0.25 mg daily which was started 12 h after delivery with daily furosemide 10 mg. Her postpartum ECG and echocardiogram remained unchanged and she had no pulmonary edema. She was monitored for 48 h in the recovery room, and the remainder of her hospital stay was uneventful. She declined genetic testing for OFDS for her neonate. At her most recent cardiology appointment, her cardiologist recommended that she should not undergo another pregnancy.

Discussion

To our knowledge, this is the first case report of anesthetic management of a pregnant patient with OFDS type II or Mohr syndrome with a repaired TOF and worsening left ventricular function. The syndrome was described originally in 1941 by Mohr and later redefined by Rimoin and Engerton as Oral-Facial-Digital syndrome or OFDS.[1,2] This is a group of rare heterogenous congenital disorders, characterized by abnormalities of oral cavity, face, larynx, trachea, chest cavity, spine and digits.[3–6] Thirteen different forms of OFDS have so far been identified based on genetic transmission and clinical manifestation; OFDS types I and II are the most common.[3–6] Our patient has OFDS type II, inherited as an autosomal recessive condition that is usually associated with congenital cardiac defects.[3,4] Cardiac anomalies include atrioventricular canal, single atrium, atrial and ventricular septal defects, coarctation of aorta, valvular stenosis and TOF.[4–7] It has been suggested that OFDS type II with cardiac defects should be considered a distinct subtype called orocardiodigital syndrome.[4] The classification of OFDS continues to evolve.[6]

Other anesthetic considerations in various types of OFDS include the presence of congenital malformations that may pose difficulties with tracheal intubation and ventilation. Large hamartomas of the tongue, micrognanthia, retrognanthia, cleft palate and high arched palate may be present and lead to difficulties with laryngoscopy and visualization of vocal cords.[6] Epiglottic hypoplasia with recurrent pulmonary aspiration and increased susceptibility to respiratory infections has been reported,[6] as has severe congenital tracheal stenosis.[8] The presence of skeletal abnormalities such as pectus excavatum and scoliosis can lead to difficult epidural placement, as well as problems with ventilation and oxygenation. Large intracranial cerebellar hamartomas may be present and cause alterations in the intracranial pressure.[9] A recent case report described severe reversible pulmonary hypertension in a morbidly obese patient with Mohr syndrome and a BMI of 44 kg/m^2; this patient had obesity hypoventilation syndrome with hypoxemia. Her echocardiogram revealed a massively dilated right atrium and ventricle and severe pulmonary hypertension. Treatment with nocturnal nasal continuous positive non-invasive ventilation resolved symptoms of obesity hypoventilation and reversible pulmonary hypertension.[10]

Our patient had typical features of OFDS involving the face, oral cavity, upper and lower limbs, but no scoliosis, pectus excavatum or anomalies of upper airway or trachea. However, she had previously undergone surgical repair of a severe form of TOF. In the four years before this pregnancy, she had undergone three major cardiac surgical procedures, and during the present pregnancy had two episodes of heart failure. The second episode of CHF, at 28 weeks of gestation, was likely to have been caused by the normal physiologic increase in cardiac output seen in the second trimester.[11] Early epidural analgesia to avoid tachycardia and increases in BP, cardiac output and systemic vascular resistance (SVR) associated with painful contractions was chosen. Dilute local anesthetics with opioids were used to minimize the decrease in SVR, hypotension and tachycardia.

A forceps delivery was achieved easily under epidural anesthesia with minimal hemodynamic fluctuation. Current recommendations encourage vaginal delivery for most patients with repaired TOF, and cesarean delivery is reserved for obstetric indications only.[12] Indeed, some experts recommend this 'cardiac delivery' for such

Download English Version:

https://daneshyari.com/en/article/2757507

Download Persian Version:

https://daneshyari.com/article/2757507

Daneshyari.com