Journal of Clinical Anesthesia (2012) **24**, 586–589

ELSEVIER

Journal of Clinical Anesthesia

Case Report

Case report of a parturient with cyanotic congenital heart disease palliated with a Glenn procedure☆

Adam L. Wendling MD (Assistant Professor of Anesthesiology)*, Aaron Seller MD (Resident in Anesthesiology), Everett Peterson MD (Resident in Anesthesiology)

Department of Anesthesiology, University of Florida College of Medicine, 1600 SW Archer Rd., P.O. Box 100254 JHMHSC, Gainesville, FL 32610-0254, USA

Received 9 August 2011; revised 3 February 2012; accepted 16 March 2012

Keywords:
Cesarean delivery;
Cyanotic congenital heart disease;
Glenn procedure, obstetrical anesthesia;
Pregnancy

Abstract As the prevalence of adults with palliated congenital heart disease continues to increase, so, too, does the number of these patients who will become pregnant. Practicing physicians need to be familiar with the impact that normal physiologic changes associated with pregnancy and delivery has on patients with palliated congenital heart disease. The physiologic impact of pregnancy on a patient with palliated cyanotic congenital heart disease and the management of her delivery are presented.

1. Introduction

While the true prevalence of severe congenital heart disease (CHD) in adults is difficult to ascertain, the number of CHD patients is likely increasing, and may be as high as two per 1,000 [1]. As these adolescents reach adulthood, it is probable that an increasing number will become pregnant. Practicing physicians need to be familiar with the impact that normal physiologic changes associated with pregnancy and delivery have on patients with CHD. A patient lost to follow-up after palliation of complex cyanotic CHD with a Glenn procedure and an aortopulmonary shunt is presented.

2. Case report

A 23 year old woman with an intrauterine pregnancy at 38 weeks' gestation and with a history of tricuspid and pulmonary atresia, with a resultant hypoplastic right ventricle that was palliated with a right-sided Glenn procedure and an aortopulmonary shunt, is presented (Fig. 1). She had emigrated from Romania, which led to a lack of cardiology follow-up. She had one previous pregnancy complicated by a high-grade perineal laceration during instrumental vaginal delivery. She did not have neuraxial analgesia for that earlier delivery. She described the experience of that delivery as "torture", and requested elective Cesarean delivery for this reason. Before the current pregnancy, her oxygen saturation (SpO_2) was reported in the 90% range, with otherwise stable cardiac symptoms of occasional chest pain, orthopnea, lower extremity edema, and infrequent episodes of paroxysmal nocturnal dyspnea.

☆ Supported solely by institutional and departmental sources.
* Correspondence: Adam L. Wendling, MD, Department of Anesthesiology, University of Florida College of Medicine, P.O. Box 100254 JHMHSC, 1600 SW Archer Rd., Gainesville, FL 32610-0254, USA. Tel: +1 352 273 6575; fax: +1 352 392 7029.
E-mail address: awendling@anest.ufl.edu (A.L. Wendling).

0952-8180/$ – see front matter
http://dx.doi.org/10.1016/j.jclinane.2012.03.008

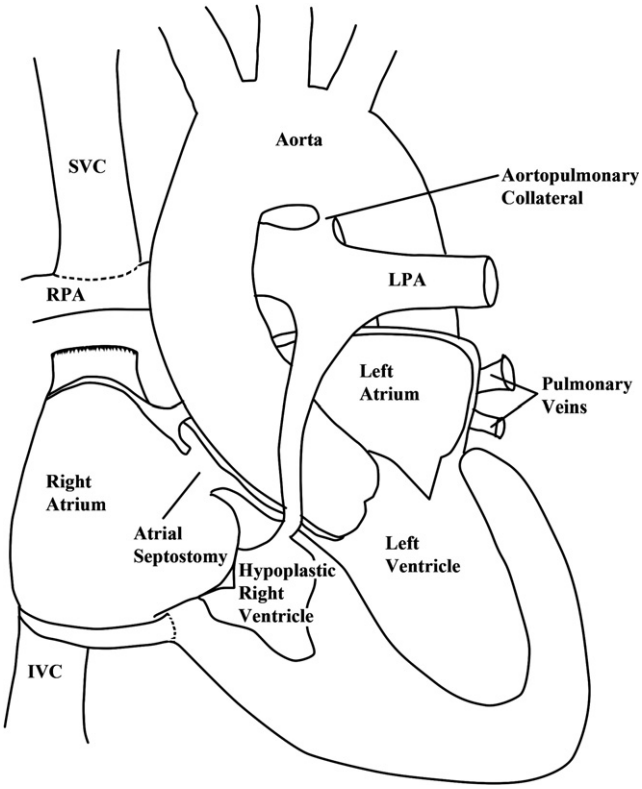

Fig. 1 Illustration of the patient's palliative anatomy. SVC = superior vena cava, IVC = inferior vena cava, RPA = right pulmonary artery, LPA = left pulmonary artery.

On presentation, she was deemed to be a New York Heart Association (NHYA) Class II heart failure, experiencing shortness of breath with activities such as climbing a flight of stairs. Since this pregnancy, her dyspnea on exertion and orthopnea had worsened slightly. Her room air SpO_2 levels were between 83% and 87%. Further physical examination showed a continuous 5/6 murmur appreciated on both sides of the sternum and definite clubbing and cyanosis of her fingertips. Aside from intrauterine growth restriction and oligohydramnios, antenatal testing was reassuring. Her hemoglobin (Hb) was 15.1 g/dL. Subsequent evaluations showed moderately suppressed ventricular function with an ejection fraction (EF) of 38%. Her electrocardiogram (ECG) showed sinus tachycardia [heart rate (HR) 110 beats per minute (bpm)] with intraventricular conduction delay and left ventricular hypertrophy. The only medications that she took prior to delivery were prenatal vitamins with iron and occasional acetaminophen.

Abdominal delivery was deemed necessary due to the history of perineal trauma, significant oligohydramnios, and patient preference. In addition to ASA standard monitors, a left radial arterial catheter was placed preoperatively. Epidural anesthesia was selected for surgical anesthesia. Twelve mL of 2% lidocaine with 1:200,000 epinephrine was administered in 3-mL increments over 20 minutes. Fentanyl 100 μg was also given through the epidural catheter to improve the density of the block. A bilateral T4 dermatome level was achieved to pin prick. The Cesarean delivery was carried out through a primary low transverse Pfannenstiel incision, followed by bilateral tubal ligation. The patient was hemodynamically stable, awake, and comfortable throughout the procedure; she delivered a baby boy with Apgar scores of 8 and 9. The patient was transfused two units of packed red blood cells (PRBCs) to increase her oxygen-carrying capacity and one L of crystalloid. She had an estimated blood loss of 800 mL and a urine output of 170 mL during the operation. Her postoperative Hb was 15 g/dL two hours postoperatively and 14.4 g/dL on postoperative day (POD) 1.

Postoperative diuresis was augmented with furosemide. Prophylactic anticoagulation was achieved with subcutaneous heparin, oral aspirin, and sequential compression devices. Continuous ECG and systemic arterial pressure monitoring was maintained for the first postoperative day in the surgical intensive care unit. The patient had an uncomplicated recovery, and on POD 3 she was discharged from the hospital. Of note, there were no congenital anomalies in either of her children.

3. Discussion

The physiologic changes of pregnancy include increased blood volume (BV), decreased hematocrit, decreased systemic vascular resistance (SVR), increased HR, and increased cardiac output (CO). These normal physiologic changes in pregnancy are variably tolerated by patients with palliated CHD, and represent risks to both the mother and, especially, the fetus [2,3]. The risk of preterm delivery and fetal death *in utero* are estimated to be as high as 16% and 57%, respectively [4].

Our patient had a number of palliative cardiac operations that resulted in her superior vena cava (SVC) flowing into her right pulmonary artery (RPA). Her pulmonary blood flow (PBF) was augmented by aortopulmonary collateral blood flow. With the available medical records and imaging, it was unclear whether her aortopulmonary collateral blood flow represented congenital aortopulmonary fistulas or an acquired, surgically created collateral conduit, such as a persistent Blalock-Taussig shunt (a surgically created anastomosis between one of the subclavian arteries and the pulmonary artery). Her inferior vena cava (IVC) flowed into a single atrium (presumably created by a surgical resection of the intra-atrial septum). Her pulmonary veins also flowed into this single atrium. In essence, our patient had a single systemic ventricle providing systemic blood flow and augmenting PBF (through the aortopulmonary collateral). Passive venous return from her upper body provided the remainder of her PBF. Preload from her IVC and pulmonary veins flowed into her single atrium, and from there to the single ventricle, and was ejected from that single ventricle partially to the pulmonary circulation, with the remainder to the systemic circulation. Fig. 1 depicts her cardiac anatomy.