OPEN ACCESS

GAVIN PUBLISHERS

Annals of Case Reports

Ellison MB, et al. Ann Case Report 10: 216.

DOI: 10.29011/2574-7754/100216

Case Report

Management of a Term Parturient with Severe Pulmonic Stenosis and Congenital Heart Disease

Matthew B. Ellison[1], John Bozek[2], Daniel Sloyer[1], Heather K. Hayanga[1], Jeffrey Puette[1], Roy E. Henrickson[1], Brian W. Grose[1], Pavithra R. Ellison[1]

[1]Department of Anesthesiology, West Virginia University, USA

[2]Department of Anesthesiology, Lenox Hill Hospital, USA

[*]**Corresponding author:** Matthew B. Ellison, Department of Anesthesiology, West Virginia University School of Medicine, PO Box 8255, 1 Medical Center Drive Morgantown, WV 26506, USA. Tel: +1-3042851931; Fax: +1-3045984930; Email: mellison@wvumedicine.org

Citation: Ellison MB, Bozek J, Sloyer D, Hayanga HK, Puette J, et al. (2019) Management of a Term Parturient with Severe Pulmonic Stenosis and Congenital Heart Disease. Ann Case Report 10: 216. DOI: 10.29011/2574-7754/100216

Received Date: 12 April, 2019; **Accepted Date:** 02 May, 2019; **Published Date:** 13 May, 2019

Abstract

A 20-year-old primigravida was admitted for induction of labor at 38 weeks following rupture of membranes. The patient had a history of a ventricular septal defect that was repaired at five months of age. Upon admission, the patient was dyspneic with minimal exertion. A transthoracic echocardiogram demonstrated severe pulmonic stenosis, moderate tricuspid regurgitation, and right ventricular hypertrophy. After consultation with multiple specialists, a balloon valvuloplasty was considered but ultimately abandoned because of the concern that the pulmonic stenosis was subvalvular in origin. The multidisciplinary team planned to proceed with epidural anesthesia for the management of labor pain along with an instrumented delivery. An ultrasound-guided awake arterial line was placed and fluid status optimized. An epidural catheter was placed and gradually titrated to maintain optimal hemodynamics. The patient underwent a successful vacuum-assisted delivery and was discharged on post-partum day two. After further workup and investigation, the patient underwent resection of a right ventricular muscle bundle and tricuspid valve repair four months after delivery.

Keywords: Pulmonic stenosis; Valvular heart disease in pregnancy; Physiologic changes during labor; Heart failure in pregnancy; Tricuspid regurgitation; Ventricular septal defect

Introduction

Valvular cardiac disease has become increasingly more common in pregnant patients because of advancing maternal age, improved treatment and survival of patients with congenital heart disease (CHD), and increasingly successful reproductive technology. The causes of valvular heart disease differ greatly throughout the world. In developing countries, rheumatic heart disease accounts for approximately 90% of the cardiac valve lesions in women of child-bearing age. [1]. On the other hand, CHD is the primary cause of valvular lesions in this age group in developed nations. Regurgitant valvular disease is typically better tolerated during pregnancy than stenotic disease because of the physiologic decrease in systemic vascular resistance. Mild to moderate pulmonic valve lesions are often congenital in nature and are well tolerated during pregnancy. However, severe pulmonic stenosis may lead to right ventricular failure, dysrhythmias, pre-eclampsia, and preterm delivery of the fetus with increased mortality [2].

Case Presentation

A 20-year-old gravida 1, para 0 female was admitted to labor and delivery at a gestational age of 38 weeks and 1 day for induction of labor following premature rupture of membranes. She had notable history of ventricular septal defect (VSD) repair with Gore-Tex patch at 5 months of age. Her current symptoms of severe dyspnea and chest pain with mild exertion were consistent with New York Heart Association (NYHA) Functional Class III. A multi-disciplinary team consisting of pediatric cardiology, adult cardiology, high risk obstetrics, obstetric anesthesiology, and cardiac anesthesiology was convened in order to determine the optimal perioperative management of this patient. Transthoracic

echocardiographic (TTE) studies of the mother were obtained by both cardiology services with discrepancies in their interpretation. An electrocardiogram showed a first degree atrioventricular block with right ventricular hypertrophy.

The adult TTE demonstrated a normal left ventricular ejection fraction (LVEF), a mildly dilated left atrium and a severely thickened and flattened interventricular septum consistent with right ventricular (RV) volume and pressure overload. The TTE was also significant for severe RV hypertrophy with moderately decreased function, moderate tricuspid valve regurgitation, and severe pulmonic valve stenosis with a mean and peak gradient of 45 and 82 mm Hg, respectively. The adult cardiology service was concerned that the patient's preload dependence would not be compatible with vaginal delivery. They recommended transfer to another facility where percutaneous balloon valvuloplasty could be performed prior to delivery.

The pediatric cardiology echocardiogram was read as normal LVEF, normal sized left atrium, severe RV hypertrophy, trace tricuspid regurgitation, normal pulmonic and aortic valves, and severe right ventricular outflow tract obstruction with a mean and peak gradient of 70 and 139 mm Hg, respectively, as well as a peak velocity across the pulmonic valve of 5.9 meters/second (Figure 1). The pediatric cardiology service was confident that the severely increased gradient across the pulmonic valve was secondary to an obstructing muscle bundle in the right ventricular outflow tract rather than being due to the pulmonic valve itself. This would mean that the lesion was not amenable to valvuloplasty. After extensive discussion amongst the multi-disciplinary team, it was determined that the patient would be managed at our facility. The adult cardiology service deferred cardiac management of this patient to the pediatric cardiology team.

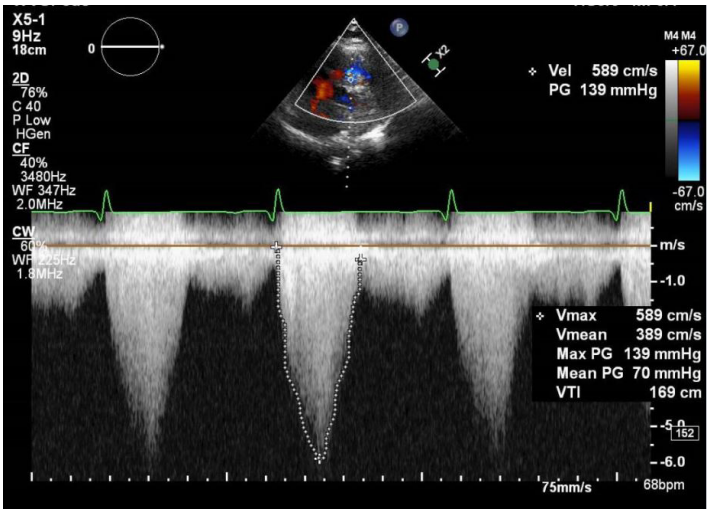

Figure 1: Pediatric cardiac echocardiogram demonstrating high velocity and gradient across the right ventricular outflow tract.

Because a second stage of labor with excessive Valsalva was worrisome in this patient due to decreased preload, an instrument-assisted delivery under epidural anesthesia was planned for the fetus. Neither the patient nor her immediate family had a history of anesthetic issues. The patient was thoroughly counseled on the risks and benefits of epidural placement as well the possible need for an emergent caesarean section. The anesthetic plan consisted of optimal fluid management guided by invasive hemodynamic monitoring and urine output followed by a "low dose" epidural infusion while avoiding aggressive epidural boluses to preserve preload and maintain systemic vascular resistance (SVR). While it was planned to place the epidural early in induction of labor given the bolus limitations, the patient elected to wait until her pain had worsened. Adjuncts for her assisted second stage of delivery, including remifentanil patient-controlled analgesia (PCA) as well as epidural opioid boluses, were discussed as options given the possible limitations of a low-dose, bolus-limited epidural.

Prior to epidural placement, an ultrasound-guided radial arterial line was placed and connected to an Edwards EV1000 FloTrac Hemodynamic Monitoring System (Edwards Lifesciences Corporation, Irvine, California). Central venous monitoring was considered but thought to be unhelpful given that the EV1000 was already in use and supplied much of the same information. Utilizing the FloTrac system, the patient's preload was carefully augmented with 250 ml of 5% albumin followed by an uncomplicated epidural placement on the first attempt. After a negative test dose, the patient received a slowly titrated bolus of 5 mL of 0.2% ropivacaine and was started on an infusion of 8 mL/hour of 0.2% ropivacaine with 2 mcg/mL fentanyl. The procedure was well tolerated without significant hypotension or changes in the FloTrac parameters. A vacuum-assisted delivery with episiotomy occurred without incident and with sufficient analgesia via the epidural without additional adjuncts. The patient was discharged on post-partum day two.

After discharge, the patient's symptoms progressively worsened over several months until she had developed NYHA Class IV symptoms of dyspnea and chest pain at rest. She was scheduled for surgery by the pediatric cardiac service at our institution secondary to her functional decline approximately four months after delivery. The patient underwent resection of an anomalous muscle bundle and a fibrous infundibulum in the right ventricle which had indeed been the cause of her severe RV outflow obstruction. She also underwent a tricuspid valve repair but the pulmonic valve was spared because it was normal in appearance and function. The postsurgical gradient across the RV outflow tract was normal and the patient was extubated in the operating room and taken to the intensive care unit. The patient was discharged home in stable condition four days after her cardiac surgery.

Discussion

The most frequent cause of valvular cardiac lesions in women of child-bearing age is rheumatic disease in developing countries, accounting for up to 90% of cases [1]. In industrialized nations, CHD accounts for up to 50% of cardiac valvular lesions in women of this age group [1]. The most commonly diagnosed lesions in the European Registry on Pregnancy and Heart Disease were mitral (63%) followed by aortic valvular disease (23%) [3]. Newly diagnosed valvular lesions in pregnancy are often first appreciated upon auscultation, which is reported to have a sensitivity of 70% and a specificity of approximately 98% for valvular disease [4]. Transthoracic echocardiography is key in determining which valve is involved as well as the severity of the lesion and the approximate cardiac chamber pressures.

When the presence of significant valvular disease has been identified in a parturient, the patient should be referred for care at a tertiary center with specialty physicians if possible. A detailed discussion between the specialist and the patient may be helpful to determine risks for the pregnancy as well as appropriate timing of delivery and cardiac intervention, if required [5]. Stenotic valvular lesions generally pose a greater risk to the parturient than regurgitant lesions because of the pregnancy associated increases in cardiac output and heart rate as well as the decrease in afterload. By the third trimester, pregnant patients experience an increase in cardiac output of approximately 30-50%, a heart rate increase of 15-20%, a 30-50% reduction in systemic vascular resistance, and a 40% increase in plasma volume [6-8].

The NYHA Functional Classification system places patients into one of four categories based on the severity of their heart failure systems. At initial presentation, our patient fell into NYHA Class III which describes marked limitation of physical activity where limited activity causes fatigue, palpitations, or dyspnea. Later, at the time of cardiac surgery, the patient had progressed to Class IV, which describes a patient who is symptomatic at rest and is unable to undergo any physical activity without discomfort [9].

In patients with severe pulmonary stenosis, as in our patient, the elevated right ventricular outflow gradient increases right ventricular work and pressure, leading to RV failure [10]. Severely stenotic lesions that reduce forward flow from the right-sided circulation may compromise global cardiac output [10]. The increased oxygen demands of labor place the patient at risk for acute hemodynamic decompensation and right-sided heart failure [11]. Additionally, Valsalva during labor may significantly compromise preload and be hazardous to these patients' fragile hemodynamic balance [7-10]. If a vaginal delivery is planned by the obstetric team, an assisted second stage labor (as in our patient) may be prudent, which may necessitate adequate pain control with options including neuraxial analgesia.

Intraoperative management of patients with severe pulmonic stenosis includes the goal of a relatively slower to normal heart rate to allow adequate timing for ventricular filling and ejection. In addition, the maintenance of adequate preload and prevention of significant increases in pulmonic vascular resistance helps to assist forward flow [7-11]. In the gravid patient, preload may be compromised by regional anesthesia, peripartum hemorrhage, aortocaval compression, and Valsalva during labor [11]. Hypervolemia, on the other hand, may overwhelm the volume and pressure-overloaded right ventricle and may precipitate right-sided heart failure and dysrhythmias [7-11].

The primary risk with epidural analgesia in the setting of pulmonary stenosis s decreased peripheral vascular resistance (a known side effect of neuraxial techniques) leading to systemic hypotension, decreased coronary perfusion pressure, decreased global cardiac output, and decreased perfusion to the fetus [12]. In addition to the potentially hazardous decline in afterload, Conway and Posner address the theoretical risk of coagulopathy should right-sided heart failure cause hepatic congestion [11]. An undiagnosed coagulopathy could pose a disastrous outcome following a neuraxial procedure and delivery [11].

Despite the dangers of neuraxial anesthesia in pulmonary stenosis, it has been pointed out that epidural-induced venodilation may actually improve right-sided heart failure [11]. This may be helpful in situations of volume overload or, in the gravid patient, with the autotranfusion that occurs following delivery [11]. Compared to general anesthesia in the parturient, the benefits of neuraxial anesthesia including superior pain control and avoidance of airway manipulation remain [7-11].

In our patient, an ultrasound guided arterial line was placed and connected to a FloTrac invasive monitoring device prior to epidural placement. This allowed for real time monitoring cardiac output, systemic vascular resistance, and Stroke Volume Variation (SVV) during the epidural and subsequent delivery.

Ideal hemodynamic management was achieved in this patient with preload augmentation (colloid and crystalloid) as well as gentle and judicious dosing of her epidural. Had she developed hypotension, she may have been best served with a bolus (or infusion) of phenylephrine to restore afterload and shift toward a low to normal heart rate. Had this patient proceeded to cesarean section, the evidence would suggest that epidural would be the most reasonable form of pain control if time permitted. Benefits of epidural analgesia include avoidance of general anesthesia with airway manipulation and risk of aspiration, intraoperative and postoperative pain control, and possible assistance of right-sided heart failure (fluid overload or auto-transfusion) with therapeutic venodilation [7]. Spinal anesthesia, unlike the slowly titratable epidural, would have posed greater risk given the possibility of a more dramatic, less controllable decrease in afterload. Continuous

spinal, however, has been described in this patient population [10]. Given the plan for an instrument assisted second stage of labor, the discussion of a remifentanil PCA and additional parenteral opioids was prudent. However, our patient had sufficient pain control for her vaginal delivery with the epidural and those adjuncts were unnecessary.

One of the most interesting factors about this case was the disagreement between the interpretations of echocardiograms by the pediatric and adult cardiology services. Both services felt that the patient had a severely increased gradient across the RV outflow tract. The difference was that the adult cardiology service was certain that the stenosis was isolated to the pulmonic valve and that the patient should undergo valvuloplasty prior to delivery. The pediatric service, on the other hand, felt very strongly that the stenosis was subvalvular in origin, likely from a right ventricular muscle bundle, which would not have been amenable to valvuloplasty. Because the patient underwent surgical repair of the lesion approximately four months after delivery, we know that the stenosis was in fact subvalvular in origin and was directly related to an anomalous muscle bundle and fibrous infundibula in the RV.

The 2008 update of the American College of Cardiology and American Heart Association Guidelines for the management of patients with valvular heart disease and the 2008 guidelines for the management of adults with congenital heart disease recommend valvuloplasty in non-pregnant patients with pulmonic stenosis and a mean transvalvular gradient greater than 40 mm Hg in asymptomatic patients and greater than 30 mm Hg in symptomatic patients [8,12]. Our patient's subvalvular lesion was not amenable to valvuloplasty. Therefore, if her symptoms had continued to worsen during the pregnancy and intervention was required, the patient may have required surgical intervention. Fortunately, the mortality rates for pregnant women undergoing cardiac valvular surgery are similar to those of non-pregnant women. However, the fetus is at significantly higher risk, as up to 1/3 of parturients undergoing cardiac surgery will suffer fetal mortality [13].

Conflicts of Interest

The authors deny any potential conflicts of interest including commercial relationships such as consultation and equity interests.

Funding Statement

The research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

References

1. Soler-Soler J, Galve E (2000) Worldwide perspective of valve disease. Heart 83: 721-725.
2. Drenthen W, Pieper PG, Roos-Hesselink JW, Schmidt AC, Mulder BJ, et al. (2006) Non-cardiac complications during pregnancy in women with isolated congenital pulmonary valvar stenosis. Heart 92: 1838-1843.
3. Roos-Hesselink JW, Ruys TP, Stein JI, Thilén U, Webb GD, et al. (2013) Outcome of pregnancy in patients with structural or ischemic heart disease: results of a registry of the European Society of Cardiology. Eur Heart J 34: 657-665.
4. Mineo K, Cummings J, Josephson R, Nanda NC (2001) Acquired left ventricular outflow obstruction during acute myocardial infarction: diagnosis of a new murmur. Am J GeriatrCardiol 10: 283-285.
5. Regitz-Zagrosek V, Blomstrom LC, Borghi C, Cifkova R, Ferreira R, et al. (2011) ESC guidelines on the management of cardiovascular diseases during pregnancy: The Task Force on the Management of Cardiovascular Diseases during Pregnancy of the European Society of Cardiology (ESC). Eur Heart J 32: 147-197.
6. Ouzounian JG, Elkayam U (2012) Physiologic changes during normal pregnancy and delivery. Cardiol Clin 30: 317-329.
7. Vidovich MI (2014) Cardiovascular Disease. In: Chestnut DH, Wong CA, Lawrence T, Ngan-Kee WD, Beilin Y, et al. (ed.). Chestnut's Obstetric Anesthesia: Principles and Practice. 6th Edition, Elsevier Saunders, Philadelphia, USA. 960-1002.
8. Warnes CA, Williams RG, Bashore TM, Child JS, Connolly HM, et al. (2008) ACC/AHA 2008 guidelines for the management of adults with congenital heart disease: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (Writing Committee to Develop Guidelines on the Management of Adults with Congenital Heart Disease). Developed in Collaboration with the American Society of Echocardiography, Heart Rhythm Society, International Society for Adult Congenital Heart Disease, Society for Cardiovascular Angiography and Interventions, and Society of Thoracic Surgeons. J Am Coll Cardiol 52: 143-263.
9. The Criteria Committee of the New York Heart Association (1994) Nomenclature and criteria for diagnosis of diseases of the heart and blood vessels. (9th Edition) Little Brown, Boston, USA.
10. Campbell N, Rosaeg OP, Chan KL (2003) Anesthetic management of a parturient with pulmonary stenosis and aortic incompetence for Caesarean section. British Journal of Anesthesia 90: 241-243.
11. Conway JB, Posner M (1994) Anaesthesia for Caesarean section in a patient with Watson's syndrome. Canadian Journal of Anesthesia 41: 1113-1116.
12. Bonow RO, Carabello BA, Chatterjee K, de Leon AC Jr, Faxon DP, et al. (2008) 2008 Focused update incorporated into the ACC/AHA 2006 guidelines for the management of patients with valvular heart disease: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines. J Am Coll Cardiol 52: 1-142.
13. Mahli A, Izdes S, Coskum D (2000) Cardiac operations during pregnancy: Review of factors influencing fetal outcome. Ann Thoracic Surg 69: 1622-1626.