Case Report
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Primigravida with Trilogy of Fallot (Severe Pulmonary Stenosis, Right Ventricular Hypertrophy, and Huge Atrial Septal Defect) for Emergency Lower Segment Cesarean Section

Poonam Ghodki[1], Shalini Sardesai[1], Jyoti Deshpande[1], Kalyani Patil[1], Kedar Bangal[1]

Abstract

Trilogy of Fallot; a triad of pulmonary stenosis, right ventricular hypertrophy and atrial septal defect is an uncommon acyanotic congenital heart disease. We present a severe case of trilogy of Fallot diagnosed for the first time in pregnancy. The patient was otherwise in a compensated state and had become symptomatic due to physiological changes in pregnancy. The anaesthetic goals therefore included maintaining the existing physiology and preventing decompensation thereby avoiding Eisenmengerisation. The anaesthetic management of the severe and complex heart disease keeping patient safety at the core is discussed.

Keywords: Caesarean section, congenital heart disease, trilogy of Fallot.

Introduction

There are very few reports of management of pregnancy with pulmonary stenosis. There is no literature available about the incidence of pregnancy with trilogy of Fallot comprising pulmonic stenosis (PS), right ventricular hypertrophy (RVH), and atrial septal defect (ASD) [1]. Furthermore, the recommendations for anesthetic management of such patients do not exist. Typically, pulmonary valvular stenosis is characterized by a bifoliate (20%) or trifoliate valve, with varying degrees of fusion of the commissures and fibrous thickening [2]. Congenital obstruction can be at the pulmonary valve, below the pulmonary valve, or above the pulmonary valve. PS accounts for 10-12% of congenital heart disease in adults, and the probability of survival up to childbearing age are high [3]. The right ventricular outflow obstruction results in the RVH increased right-sided pressures and subsequent right heart failure (RHF). Pregnancy-induced increase in intravascular volume and heart rate can further precipitate RHF and cardiac arrest. Pulmonary stenosis is generally well tolerated in pregnancy in the absence of other hemodynamically significant lesions [4]. Isolated ASD, on the other hand, is one of the most common congenital defects seen in pregnant women [5]. The chronic left to right shunt with exaggerated intravascular volume in pregnancy along with severe PS in our patient posed unique anesthetic challenges. The case presentation with pathophysiology, hemodynamic goals, and anesthetic management is hereby discussed.

Case Report

A 24-year-old primigravida weighing 68 kg with 35 weeks plus 5 days gestation in labor was admitted to the obstetric ward. The patient had breathlessness on accustomed activity (NYHA class 3) in the 5th month and was admitted. 2D echo revealed severe PS, RVH, and ASD. The patient had been examined by cardiologist, and he had advised pulmonary balloon valvotomy. She was referred to cardiac center, but the patient was financial constraints she neither underwent a valvotomy nor did she follow-up with obstetrics unit here. The patient presented directly at 35 weeks plus 5 days gestation in labor. On examination, she was well compensated with no signs of congestive cardiac failure and preferred to lie in the lateral position (left/right). She was afebrile; pulse rate was 86/min, blood pressure 136/80 mmHg, and respiratory rate 18/min with room air saturation (SPO2) 95%. She had parasternal heave, diastolic shock, and pansystolic murmur. Her electrocardiography (ECG) was suggestive of the RVH and strain pattern in the inferior leads. The 2D Echo showed the right atrium and RVH, severe pulmonary stenosis (Peak pressure gradient/mean pressure gradient = 144/94 mmHg), large ostium secundum ASD (39 mm), left to right shunt, moderate tricuspid regurgitation, RSVP = 97 mmHg, ejection fraction 55%, and the impression was of acyanotic congenital heart disease. A decision of emergency cesarean section was taken after discussion with cardiologist.

[1]Department of anaesthesiology, SKN Medical College, Pune, India.

Address of Correspondence
Dr. Poonam Ghodki,
Department of anaesthesiology, SKN Medical College, Narhe, Pune Pin Code 411041, Maharashtra, India.
Email: drpoonamghodki@gmail.com


Dr. Poonam Ghodki


Dr. Shalini Sardesai


Dr. Jyoti Deshpande


Dr. Kalyani Patil


Dr. Kedar Bangal

Our primary aim was not to disturb the compensated physiology of the patient which meant maintaining her pre-operative systemic and pulmonary vascular resistance levels relative to each other even under anesthesia. We chose general anesthesia for this patient. The patient was kept starving in view of impending cesarean section for 8 h. Emergency cardiac drugs and vasopressors were kept ready. The necessary laboratory investigations were within normal limits. She received prophylactic injection ranitidine and injection metoclopramide intravenously (IV) in ward as a protocol. The patient was shifted to operation theater and all the standard monitors were attached – pulse oximeter, ECG, and non-invasive blood pressure monitor (NIBP). Preoperatively, SPO_2 was 95-96%. The patient was preoxygenated, premedicated with IV injection fentanyl 2 mcg/kg, injection ondansetron 4 mg, and preservative-free injection lignocaine 1.5 mg/kg was given. Priming was done with injection vecuronium and induction of anesthesia was done by injection etomidate 10 mg IV. Then, the patient was intubated and air entry was confirmed by auscultation. The patient was put on ventilator and hyperventilated to maintain end-tidal CO_2 between 30 mmHg and 34 mmHg. The anesthesia was maintained on 100% oxygen and sevoflurane. Immediately after delivery of a healthy baby; infusion of injection oxytocin 20 units IV was given over 30 min in the second IV fluid. Urine output after 1 L of IV infusion was <20 ml at the end of surgery (50 min), subsequently, injection frusemide 5 mg IV was given. Postoperatively, before reversal, the patient was given bilateral ilioinguinal and iliohypogastric nerve blocks with loss of resistance technique with injection bupivacaine 0.25% 20 ml and dexamethasone 4 mg on either side. Injection tramadol 50 mg was added to IV drip. All the hemodynamic parameters were maintained throughout the surgery. The patient was reversed with injection neostigmine and injection glycopyrrolate and extubated. She was observed for half an hour and shifted to intensive care unit for careful observation with oxygen mask with O2 flow rate at 4 L/min. She was pain-free and her post-operative recovery was uneventful. Both mother and baby were discharged on the 7th post-operative day with an advice to follow-up with the cardiologist.

Discussion

We successfully managed a patient of trilogy of Fallot with severe individual components who was diagnosed for the first time in pregnancy. She had severe PS with huge ASD and RVH, each component compensating the other. The hemodynamics had changed with physiological changes in pregnancy, and that is, the reason she had become symptomatic. Our main aim of anesthesia was to maintain the compensatory mechanism, hence not allowing an increase in PVR, decrease in SVR, preventing RVF, and reversal of shunt [6,7]. After diagnosis of pulmonary stenosis in the 5th month of pregnancy, the patient was advised to follow-up with opinion of cardiologist who had advised a balloon pulmonary valvotomy. However, she did not comply. She directly presented to us when she was in labor. A multidisciplinary (cardiologist, obstetrician, and anesthesiologist) decision for cesarean section was taken as the stress of labor and normal delivery would prove deleterious to the patient considering her cardiac status. Our patient had large ASD with severe pulmonary stenosis as noted by the pressure gradients. Perhaps the huge ASD did not let the pressure built up in the right atrium and thus was protecting against RHF. The ASD was so huge that practically there was a single atrium functionally. Subsequently, there was mixing of blood in the common atrial chamber as evident from the low baseline oxygen saturation. Due to the compensation, she never had any complaints until the compensatory mechanisms started to fail due to pregnancy. Therefore, the aim of anesthesia was not to let the patient decompensate from the any anesthetic intervention. The main challenge was to avoid a fall in the systemic blood pressure below the pulmonary blood pressure by maintaining both the cardiac output and the systemic vascular resistance to prevent the right to left shunt or Eisenmengerisation [8, 9]. The patient was adapted to the high pulmonary pressures, and as the pulmonary stenosis was organic, the use of pulmonary vasodilators (sildenafil, milrinone, etc.,) was not indicated [10, 11]. The concern was to avoid sudden hemodynamic changes, hypoxia, hypercarbia, and acidosis and further rise in the pulmonary vascular resistance. These hemodynamic goals could be best achieved with general anesthesia; although there is no contraindication to graded epidural anesthesia, one has to be extremely careful as the fall in venous return is not taken up so well and it is also difficult to prevent/treat ensuing hypotension. To the horizon of our knowledge and literature search, there are no case reports of trilogy of Fallot with such severe PS and huge ASD in a pregnant patient for programmed cesarean section. Sanikop et al. managed a case of pregnant patient with severe pulmonary valvular stenosis gradient of 68 mmHg and concluded that general anesthesia offers better advantage as compared to spinal or epidural anesthesia [12]. As neuraxial block has a risk of reduced venous return and hence reduced cardiac output, it was feared that our patient may land up in failure[9]. Use of cardiostable drugs such as fentanyl, etomidate, and sevoflurane along with 100% oxygen helped us in achieving the hemodynamic goals. There are reports of similar cases conducted under central neuraxial block [8]. Shah et al. performed the case of pulmonary stenosis (peak systolic gradient of 67 mmHg) and a history of undergoing balloon pulmonary valvotomy under epidural anesthesia [3]. In spite of adequate preloading, the patient had hypotension up to 70/50 mmHg after epidural bolus injection. She complained of visceral pain due to stretching of tissues that was tackled with IV ketamine 50 mg, which may prove detrimental by increasing the heart rate and pulmonary vascular resistance [13]. Pain is deleterious in these patients as it leads to decompensation. We routinely practice a multimodal analgesic regimen that includes a transversus abdominis plane/ilioinguinal-iliohypogastric block along with parenteral drugs for post-operative analgesia in lower segment cesarean section. We followed the same protocol in this patient, who too stayed comfortable and pain-free postoperatively. Cardiologist suggested a valvotomy before ASD closure because in our patient the ASD was functioning as a conduit between the two atria thus preventing congestive cardiac failure. We had to take special care to prevent thromboembolism which could be critical in our patient with large ASD having a high propensity of paradoxical embolism.

That was one of the reasons why a central line was not opted for. An arterial line was desirable, but we chose not to put it and monitored with NIBP only.

Conclusion

Detailed preparation and meticulous knowledge of the pathophysiology are the key for successful management of this rare case of trilogy of Fallot in pregnancy. General anesthesia with the use of appropriate cardiostable drugs and providing good pain relief is the prime components for maintaining hemodynamic stability and safe management of such cases.

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