CASE REPORT

Graded Epidural Anaesthesia for Abdominal Hysterectomy in an Adult with Congenital Heart Disease: A Case Report

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ABSTRACT **Background:** Congenital heart anomalies pose challenges during anaesthesia with its associated morbidities; Cor-triatriatum dexter may be associated with other heart anomalies which subject the patient to increased risks.

Case Presentation: We here-in report the case of a 30 yr old woman that had cor-triatriatum dexter with associated multiple atrial septal defects, rheumatic valvular heart disease, pulmonary hypertension and associated ventricular dysfunction that had total abdominal hysterectomy under graded epidural anaesthesia.

Conclusion: High risk patients with multiple congenital anomalies may undergo anaesthesia with careful choice of anaesthesia technique.

Keywords: Cor-triatriatum dexter, Congenital heart anomaly, Graded epidural anaesthesia.

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Access this Article Online	
Quick Response Code:	Website: https://njan.org.ng
	DOI: https://doi.org/10.82223/nja.vol2no2.49

How to cite this article:
Atiku M, Salahu D, Abdullahi M.M.M, Alhassan D.M, Alhassan Z.A, Aminu A,B. Graded epidural anaesthesia for abdominal hysterectomy in an adult with congenital heart disease: A case report. Nigerian Journal of Anaesthesia. 2025;2:114-116.

INTRODUCTION

Cor-triatriatum dexter (CTD) is a rare condition where a fibromuscular band separates the right atrium into two chambers due to persistence of the right valve of the embryonic sinus venosus making the heart triatrial.[1] It has a prevalence of 0.025% among all congenital heart diseases.[1]

CTD rarely exists in isolation as it is often associated with other cardiac abnormalities such as ventricular septal defect, atrial septal defect, mitral regurgitation, tricuspid valve abnormalities and pulmonary artery stenosis or atresia.[2] Its clinical presentation depends on the size of communication between the subdivided compartments and the presence of associated anomalies. The condition is associated with right sided heart abnormalities such as hypoplasia or atresia of the tricuspid valve and/or pulmonary artery orifice. When it coexists with an ASD, features of right to left shunting could be so subtle that its diagnosis could be missed.[1]

Congenital heart diseases may range from serious conditions that are diagnosed at birth, to milder forms which could be missed at birth and later diagnosed in adulthood. Among congenital heart conditions, lesions with high pulmonary vascular resistance have been characterized as having the highest risks during anaesthesia.[3] Before the advancement of echocardiography, this condition was either diagnosed during cardiac surgery, or post-mortem.[4] Knowing and anticipating the cardiovascular effects of anaesthetic agents on these patients' cardiovascular parameters is of utmost importance.

We report the case of a patient with cor-triatrium dexter with associated multiple atrial septal defects, rheumatic heart disease (severe mitral incompetence), pulmonary hypertension, left ventricular dysfunction and pericardial effusion that underwent anaesthesia for total abdominal hysterectomy.

CASE PRESENTATION

The patient is a 30 year old woman who presented to the gynaecology clinic of Aminu Kano Teaching Hospital with complaints of cyclical lower abdominal pain and primary amenorrhea. On evaluation, a diagnosis of transverse vaginal septum with coexisting uterine

fibroids was made and she was planned for exploratory laparotomy with examination under anaesthesia.

Preanaesthesia review revealed a young woman who was small for age, she had no known comorbid conditions, examination of the cardiovascular and respiratory systems were all normal. The available investigations, full blood count and differentials, urea, electrolyte and creatinine were all within acceptable ranges. She was then scheduled for surgery.

Following induction of general anaesthesia with 150mg of propofol, and endotracheal intubation, she was noticed to develop an arrhythmias; tachycardia (250b/min) with an irregular rhythm, she then suddenly began to desaturate despite being mechanically ventilated with 100% oxygen, and immediately thereafter suffered a witnessed cardiac arrest. The inhalational agent was swithed off and high quality CPR was commenced, and 1mg adrenaline administered intravenously. Return of spontaneous circulation (ROSC) was achieved within 3 minutes of cardiac arrest. The surgery was abandoned, and patient transferred to the intensive care unit for continued post cardiac arrest care. She was reviewed by the cardiologist while in the ICU, a bedside echocardiography revealed a left ventricular ejection fraction of 45%, dilated left atrium and left ventricle, thickened mitral and aortic valves with moderate mitral regurgitation and cor-triatriatum dexter. She was placed on carvedilol, digoxin, spironolactone, torsemide, and monthly penicillin and was discharged from the ICU following recovery after 24hrs.

Six months thereafter, she was rebooked for surgery (myomectomy/total abdominal hysterectomy and vaginoplasty). On preanaesthesia review, a thorough cardiovascular examination revealed a displaced apex beat and pansystolic murmur. Repeat echocardiography revealed a decline in her cardiovascular function with severe mitral incompetence, multiple atrial septal defects with cor-triatriatum dexter, moderate pulmonary hypertension, mild pericardial effusion, and a further decline in ejection fraction to 31%. A high-risk consent was obtained.

Upon her arrival at the theatre, a multiparameter monitor showed baseline vitals to be a pulse rate of 90b/min, blood pressure of 132/90mmHg, and a saturation of 96% on room air. A 16G intravenous cannula was secured, and a size 7.5 Fr central line was inserted, baseline central venous pressure (CVP) was 15mmHg. An epidural catheter was placed at the L3/L4 space, with a test dose of 2mls of 2% lidocaine with adrenaline administered. Graded epidural with low dose plain bupivacaine with fentanyl as adjuvant to enhance anaesthesia, alongside some sedation was the anaesthetic technique of choice; 5mls of 0.1% bupivacaine with fentanyl 4mcg/ml was administered, this was followed by another 5mls of the same mixture after 5 minutes, 5mls of 0.125% bupivacaine with fentanyl after another 5 mins was again given, following which surgical anaesthesia was achieved, the dermatomal level of block was T6. She was also sedated with boluses of midazolam 2mg and ketamine 10mg Her haemodynamics remained stable with pulse rate ranging from 90–120b/min, mean arterial pressure (MAP) ranging from 94-106mmHg and oxygen saturation ranging from 97-100% throughout the surgery period, with surgery lasting for about 2 hours. She received a total of 1.5L of 0.9% saline, and urine output was 600mls. There was an estimated blood loss of 1liter, and 1 pint of blood was transfused. Continuous central venous pressure monitoring continued throughout the intraoperative period with CVP ranging from 10mmHg to 16mmHg (Appendix I).

After surgery, the patient was transferred to the Intensive Care Unit where she was observed for 24 hours and subsequently discharged to the ward.

DISCUSSION

Cor-triatriatum, a rare condition typically produces symptoms by causing pulmonary venous obstruction and pressure overloading on the right side of the heart. The most frequent initial symptoms in infants are respiratory distress, cyanosis, recurrent respiratory tract infections, and feeding difficulties; while older patients present with syncope, dyspnea, and hemoptysis.[5,6] Pulmonary hypertension worsens the workload on the right chamber; this patient had pulmonary artery pressure of 40mmHg. The most dangerous intraoperative complication from a pulmonary hypertension exacerbation is right ventricular failure causing persistent systemic hypotension.[7]

Hypercarbia should be avoided to prevent a pulmonary hypertensive crisis. In patients with pulmonary hypertension, regional anaesthesia is preferred where applicable. General anaesthesia is challenging as mechanical ventilation may reduce venous return and worsen right sided heart function, and direct laryngoscopy may result in increased sympathetic outflow leading to increased pulmonary vascular resistance.[8] Furthermore, hypoxia, acidosis, sudden haemodynamic changes, tachycardia should be avoided. The use of graded epidural further ensured stability in hemodynamics, alongside CVP monitoring which ensured adequacy of fluid therapy.

As seen on echocardiography, this patient had multiple ASDs: this signifies significant blood shunting between the atria and hence increased work load on the right heart, changes in systemic vascular resistance (SVR) should be avoided, a high SVR worsens the left-to-right shunt, increasing the volume of blood going to the lungs, while low SVR can cause the shunt to reverse, leading to right-to-left shunting and hypoxia. A reversed shunt can lead to hypoxia and increase the risk of stroke from paradoxical emboli intraoperatively.[9]

The patient was also diagnosed with rheumatic heart disease with echocardiography showing severe mitral incompetence, this implies that increased pressure is required to pump blood from the chambers most especially the left resulting in progressive chamber enlargement with a consequent risk of heart failure. Sudden changes in heart rate and blood pressure should be avoided as this may worsen the mitral regurgitation

and consequently reduce the cardiac output. Intra-arterial blood pressure monitoring would be ideal for this patient in order to monitor the beat to beat changes in blood pressure, this was however not done due to logistic reasons. Graded epidural method of anaesthesia with low concentration of bupivacaine however ensured blood pressure stability, as opposed to subarachnoid block.

Our patient had left ventricular dysfunction with an ejection fraction of 31%, global chamber hypokinesia and associated chamber enlargement suggesting a dilated cardiomyopathy. Ventricular dysfunction and chamber dilatation makes the myocardium very sensitive to negative inotropic agents such as inhalational anaesthetic agents hence increasing the risk of arrythmias and heart failure.[10]

Lee et al[6] similarly reported the successful anaesthesia for laparoscopic resection of ovarian tumour in a 45 yr old patient with cortriatrium. General anaesthesia was employed with cardiostable drugs utilised, such as etomidate, remifentanyl, sevoflurane. Their patient however had no additional cardiac anomalies, this was unlike our patient that had cor-triatriatum dexter, multiple atrial septal defects and valvular disease.

Our choice of graded epidural anaesthesia using low dose bupivacaine with fentanyl as adjuvant enabled avoidance of myocardial depression.

CONCLUSION

Patients with multiple heart anomalies undergoing surgery may be an anaesthetic challenge. Graded epidural with low dose bupivacaine offers appropriate conditions for a successful hysterectomy procedure.

Financial support and sponsorship: None

Conflicts of interest: There are no conflicts of interest.

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Appendix I: Multiparameter monitor showing intraoperative hemodynamics and CVP at 12mmHg.