Anesthetic Management of Dextrocardia with Situs Inversus for Caesarean Section- A Case Report

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ABSTRACT

Dextrocardia is a congenital cardiac malrotation in which the heart is situated on the right side of the body (dextroversion) with the apex pointing to the right. It may be associated with many cardiac and non-cardiac anomalies. It can also occur as a part of Kartagener's Syndrome (KS). Presence of congenital and cardiac anomalies make the anesthetic management challenging and knowledge of the same is essential for better patient outcome.

Keywords: Dextrocardia, caesarean section, situs inversus

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INTRODUCTION

Dextrocardia with situs inversus is also know as mirror image. Dextrocardia is a rare condition characterized by abnormal positioning of thoraco-abdominal viscera including heart, spleen and stomach on the right side and liver on the left side of the body. It occurs in 1 in 10,000 live births and is transmitted by autosomal recessive genes. It affects both genders equally. It may be associated with congenital cardiac structural and functional defects and many non-cardiac anomalies of spleen, spine and skeletal system. It can also occur as a part of Kartagener's Syndrome (KS). Presence of congenital and cardiac anomalies make the anesthetic management challenging and their knowledge is essential for better patient outcome.

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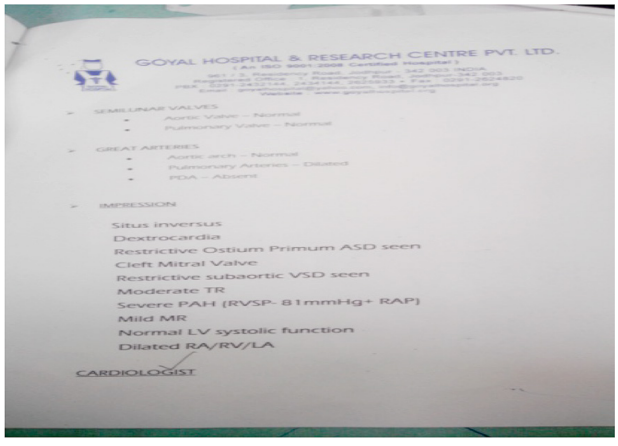
CASE REPORT

A 30 year old female weighing 38 kg was admitted to our institute at 34 weeks of gestation. She didn't have any ante-natal checkup (ANC) before this visit. She was a diagnosed case of dextrocardia with situs inversus (Fig 1). The ultrasonography (USG) of abdomen revealed breech presentation, severe oligohydramnios and intrauterine growth retardation (Fig 2). The patient was scheduled for elective caesarean section (CS). The patient had history of palpitation since childhood for which she did not consult any hospital. There was no history of dyspnea, recurrent sinusitis or bronchiectasis. She had good exercise tolerance with no deterioration of cardiorespiratory functions during pregnancy. General physical examination did not reveal any obvious findings. Airway examination revealed restricted neck extension (30 degrees), with mouth opening of 3 cm (inter-incisor distance), thyromental distance of 6 cm and Mallampatti grade III. On auscultation of chest, grade 4/6 pansystolic murmur in 5th intercostal space was heard associated with thrill. Her haematological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SEMILUNAR VALVES
- Aortic Valve – Normal
- Pulmonary Valve – Normal

GREAT ARTERIES
- Aortic arch – Normal
- Pulmonary Arteries – Dilated
- PDA – Absent

IMPRESSION
Situs inversus
Dextrocardia
Restrictive Ostium Primum ASD seen
Cleft Mitral Valve
Restrictive subaortic VSD seen
Moderate TR
Severe PAH (RVSP- 81mmHg+ RAP)
Mild MR
Normal LV systolic function
Dilated RA/RV/LA

CARDIOLOGIST

Fig 1. Echocardiography report.


MARWAR DIAGNOSTIC & IMAGING CENTRE
Dr. Mohd. Hasan

Both Uterine Arteries reveal Normal Low Resistance Continuous Diastolic Flow – +/o No Uteroplacental Insufficiency.

Fetal MCA Reveals normal High Resistance Blood Flow.

Umbilical artery at its origin, Midpart and insertion shows normal Low Resistance Continuous Diastolic Flow, no Diastolic notch noted.

Fetal Ductus Venosus also reveals normal Flow velocities with normal Waveform Pattern.

Cerebro Placental Ratio – 1.32/1.10 =1.2 (Normal).

No loop of umbilical cord seen around fetal neck during examination.

IMPRESSION: Single, Live Intrauterine Fetus with Average Maturity 33W 1D.
Cephalic Presentation. EDD:21/01/2015
Placenta Posterior wall upper segment.
Normal Obstetric Doppler Study (umbilical, uterine artery, fetal MCA and aorta). No e/o fetal hypoxia at present.
Severe oligohydromnios , Amniotic fluid index-1.47cm.
Breech Presentation.
Maternal USG: Malrotation of organs, liver on left side, spleen on right side, heart is slso
right side.

Dr.Mohammed Hasan
M.D.
Consultant Radiologist

This is a radiological opinion ,not the final diagnosis .It only helps in arriving at the final diagnosis, which is to be taken in correlation with detail of clinical and lab data. In case of any discrepancy with imaging opinion, review of the same may be done with clinical correlation.

*सोनोग्राफी के मरीज अपने साथ डॉक्टर की पर्ची व पहचान पत्र अवश्य लाएं।

Fig 2. USG report.

investigations and bleeding profile were within normal limits (hemoglobin was 11.5 g/dl, hematocrit- 35%, TLC- 9,000/mm^3, platelets- 2.5 lacs/mm^3). Chest X-Ray was not done as the patient was pregnant. Electrocardiograph showed right axis deviation, positive QRS complexes (with upright P and T waves) in leads aVR and I, inversion of all complexes (inverted P wave, negative QRS, inverted T wave), and absent R wave progression in the chest leads (dominant S waves throughout). Echocardiography revealed dextrocardia, atrial and ventricular septal defects, moderate mitral and tricuspid regurgitation, severe pulmonary artery hypertension (PAH) (mean pulmonary artery pressure was 81 mmHg), dilated right atrium and ventricle with normal left ventricular systolic function. USG showed malposition of abdominal organs. The patient was counseled preoperatively about her existing cardiac conditions and the possible intraoperative and postoperative complications. Phenylephrine (50 ug/ml) was especially loaded and kept ready for BP control during surgery. We secured a peripheral venous access and radial arterial line for continuous blood pressure monitoring.

The patient was pre-medicated with metoclopramide 10 mg and ranitidine 50 mg intravenously (IV) one hour before surgery. Standard monitorings were attached. Patient's pulse rate was 70/min, blood pressure was 124/76 mmHg and arterial oxygen saturation was 98% on room air. Instead of the usual left uterine displacement, patient was placed in right uterine displacement for vena caval decompression. ECG electrodes were placed in opposite way. Combined spinal epidural (CSE) anesthesia was chosen and bupivacaine heavy 0.5%, 5 mg with fentanyl 25 mcg were given in subarachnoid space and ropivacaine 0.25%, 12 ml administered into the epidural space. After achieving a level of sensory blockade up to T4 level, surgery was started. There was no episode of hypotension and bradycardia. Oxytocin 2 units intravenous bolus was given after the delivery of baby. Patient remained hemodynamically stable throughout the surgery and received 700 ml of IV Ringer Lactate. Surgery was completed uneventfully in 40 minutes with a blood loss of around 600 ml. The urine output during surgery was 300 ml. She was shifted to post anesthesia care unit. Postoperative pain was managed with epidural 0.2% Ropivacaine at 4 ml/hr infusion. Patient remained hemodynamically stable in the postoperative period while she was being followed up for subsequent 48 hrs. The epidural catheter was removed after 2 days.

DISCUSSION

Situs inversus is a Latin phrase used to describe the inverted position of thoracic and abdominal organs. It is called situs inversus totalis when there is a total transposition of abdominal and thoracic viscera.[1,2] Situs inversus is usually associated with congenital heart disease, most commonly transposition of the great vessels. Eighty percent of these patients have a right-sided aortic arch. Up to 20% of patients with situs inversus can have KS.[3] As our patient did not have history of recurrent sinusitis, chest infection or primary infertility, she did not appear to be a case of KS.

Patients with situs inversus require certain intraoperative considerations. The ECG and pacing or defibrillator pads should be placed in reverse. If not, the polarity change for the ECG can erroneously display a picture of perioperative ischemia. Successful cardiopulmonary resuscitation and defibrillation requires proper knowledge of cardiac anatomy. Transesophageal echocardiography imaging and interpretation must take into account the possibility of uncovering abnormalities missed in previous work-ups. Central venous cannulation should occur in the left internal jugular vein, which provides a direct route to the right atrium and lessens the incidence of thoracic duct injury. Lastly, in the obstetric patients, uterine displacement should be to the right. In our patient also, we maintained the right uterine displacement.

Regional as well as general anesthesia (GA) has been safely used to anesthetise patients with situs inversus. The associated spinal deformities like split cord, spina bifida, meningomyelocele, scoliosis etcetera either contraindicate or make the administration of neuraxial blockade difficult.[4] Anesthetic management depends on the presence of shunt, PAH, ventricular dysfunction, and arrhythmias.

In patients with situs inversus, caesarean section can be done under GA or central neuraxial block depending upon the patient's presentation and her response to physiological changes of pregnancy with cardiovascular stability being the goal. The advantage of regional anaesthesia, especially epidural anesthesia and analgesia appears to be superior because of the lower risk of high motor block. CSE has the advantage of providing rapid onset and further enhancement can easily be done by repeated small boluses of epidural local anesthetic, thus minimizing the chances of significant hemodynamic instability.[5] In patients with complex congenital anomalies, 50-100 μg of phenylephrine boluses are used to treat hypotension as this would improve systemic vascular resistance without any effect on heart rate and uterine blood flow.[6] While oxytocin can induce vasodilatation and arterial hypotension, ergometrine can cause arterial hypertension. These adverse cardiovascular effects may be catastrophic in patients with complex congenital heart disease when administered rapidly or given in high doses. Hence, titrated dose of oxytocin should be used and ergometrine should be avoided.[7,8]

Anesthetic induction in patients with pulmonary hypertension can be challenging due to the high resting sympathetic tone and resultant deficiency of catecholamine levels. This can result in exaggerated hemodynamic compromise following induction. Slow titrated doses of local anesthetic to obtain a surgical block can be safely used. Tight hemodynamic monitoring and control is of utmost importance.

Anesthetic goals for pulmonary hypertension include avoiding elevations in PVR. Supplemental oxygen should be used and pre-operative sedation minimized to avoid hypoxia and hypercarbia. SVR should be maintained as reductions in SVR will increase right to left shunting. Myocardial depressants should be avoided and maintain myocardial contractility. Preload and sinus rhythm should be maintained. The post-operative period represents a high risk time for PH patients. They should be monitored in an intensive care setting in the first few days following surgery. Patients may benefit from epidural anesthesia for post-operative analgesia.

In patients with VSD, desirable haemodynamic goals are to maintain slightly higher preload and pulmonary vascular resistance while keeping the SVR on the lower side and at the same time maintaining heart rate and contractility.

To conclude, management of patients with situs inversus totalis revolves around maintaining hemodynamic stability depending on the type and severity of the defect with concomitant attention to other organ systems which may be affected.

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