Anaesthesia Management of a Patient with Congenital Kyphoscoliosis and Large Atrial Septal Defect Posted for Kyphoscoliosis Corrective Surgery

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ABSTRACT

The authors present the anaesthesia management of a 12-year female patient who underwent corrective surgery for kyphoscoliosis under general anaesthesia. The patient had congenital idiopathic kyphoscoliosis with left lateral curvature (left convex), C7-hemivertebrae, large atrial septal defect with left to right shunt (ostium secundum defect of 24 mm), mild pulmonary hypertension with mean pulmonary artery systolic pressure (PASP) of 39 mmHg, and dilated right heart. The authors gave general anaesthesia with vigilant monitoring of haemodynamic parameters (heart rate, ECG, blood pressure, spO_2), $EtCO_2$, and peak inspiratory pressures; avoided medicines that stimulate the sympathetic system, and normothermia was ensured. Urine output was monitored. The authors prevented hypoxia, hypercarbia, and acidosis; avoided positive end-expiratory pressure (PEEP), air bubbles in IV fluids or medicines, and factors that cause a rise in systemic vascular resistance (SVR) and pulmonary vascular resistance (PVR); all of these are the target goals for anaesthesia management in a patient with congenital kyphoscoliosis and atrial septal defect.

Key Words: *Atrial septal defect, Kyphoscoliosis, Pulmonary hypertension, Anaesthetic management*.

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INTRODUCTION

Kyphoscoliosis is an abnormal curve of the spine in two planes, the coronal plane and the sagittal plane,[1] and a Cobb angle of more than 40 degrees in the thoracolumbar spine. The incidence of idiopathic kyphoscoliosis is around 4 per 1,000 with a male-to-female ratio of 4:1.[2] The three most common idiopathic aetiologies are cerebral palsy, Duchenne muscular dystrophy, and mitochondrial diseases. It can also be secondary to other factors such as degenerative changes, inflammatory changes, trauma-induced changes, neuromuscular weakness disorders, recurrent microtrauma, and thoracic surgeries.[2] If the curvature of the spine is left-sided (left convex), this is associated with congenital conditions involving the spinal cord (20%), genitourinary system (20-33%), and cardiac conditions (10-15%).[3] Preoperative evaluation in kyphoscoliosis patients includes assessment of difficult airway, severity of pulmonary dysfunction due to restrictive lung diseases, cardiac manifestations such as mitral valve abnormalities, atrial septal defect, decreased cardiac output, cardiomyopathies, and right-sided cardiac failure secondary to pulmonary hypertension (PH), neurologic manifestations, and nutrition assessment.[4]

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As kyphoscoliosis corrective surgeries are elective procedures, these patients must be optimised for smooth intraoperative course and postoperative recovery.

Atrial septal defect (ASD) is the most prevalent congenital acyanotic heart disease and constitutes about 10% of acyanotic congenital cardiac defects. Ostium secundum constitutes 70% of ASD cases, with a male-to-female ratio of 1:2.[5] ASD causes left-to-right intra-cardiac shunt with an increase in right ventricular volume load and increased pulmonary blood flow leading to PH with resultant right ventricular hypertrophy and eventually congestive heart failure.[6]

We hereby present the case report of anaesthesia management of a patient with congenital kyphoscoliosis with large ASD who underwent corrective surgery for kyphoscoliosis under general anaesthesia.

CASE REPORT

A 12-year patient (weight = 32 kg) presented for application of growing rods from T5-6 and pedicle screw instrumentation at L3-L4 level for correction of her kyphoscoliosis. She had no history of congenital abnormalities. She had no difficulty in lying straight and no difficult airway parameter was present. X-ray of the spine (Figure 1) showed left thoracolumbar scoliosis (left convex) with Cobb angle of 40 degrees, kyphosis of 55 degrees, and C7 and T12 hemivertebrae. No respiratory distress, exercise intolerance, or shortness of breath (NYHA-II) was present, and no neurologic deficit was found.

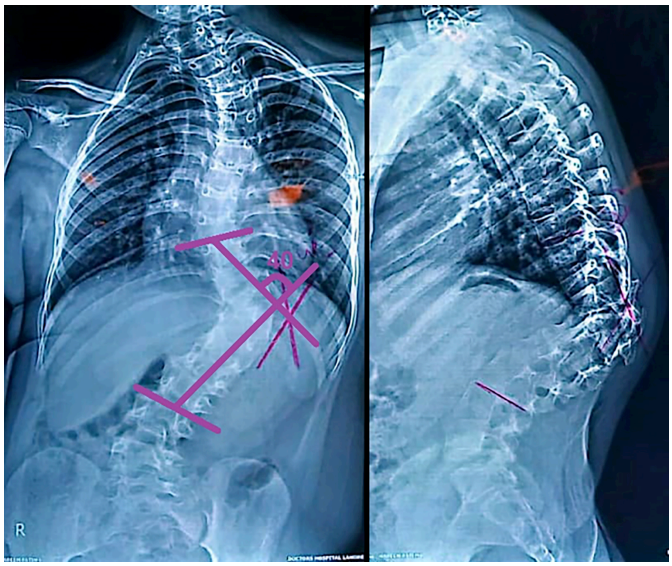

Figure 1: X-ray of the patient, showing a Cobb angle of 40 degrees and kyphosis of 55 degrees. One can also appreciate C7 and T12 hemivertebrae. Cobb angle: A transverse line was drawn from the upper border of the uppermost tilted vertebrae and the lower border of the lower most tilted vertebrae. A perpendicular line was then drawn from these two transverse lines. The angle made between these two perpendicular lines is the Cobb angle.

On systemic examination, she had a systolic murmur on cardiac auscultation. Bedside-focused cardiac ultrasonography (FOCUS) showed left-to-right shunt and left ventricular contractility seemed to be >50% by fractional shortening. A detailed echocardiography was done that showed ejection fraction of 62%, situs solitus with laevocardia, large ASD (secundum) 24 mm in size with left-to-right shunt, dilated right heart with normal right ventricular systolic function, mild tricuspid and mitral regurgitation, and mild PH with mean pulmonary artery systolic pressure (PASP) of 39 mmHg. Her complete blood count showed haemoglobin of 14.1 g/dl, platelets of 430,000 cells/mm^3, and white blood count of 7300 cells/mm.[3] Her renal function tests showed creatinine of 0.81 mg/dl. After obtaining written consent from the family, securing wide-bore intravenous access and ensuring completion of nil-per-oral status, the patient proceeded for surgery with a cardiologist on board.

After attaching the monitors as per American Society of Anesthesiologists (ASA) standard-1 monitoring, induction of anaesthesia was done with propofol (2-2.5 mg/kg) in titrated doses until the effect (total: 80 mg) and atracurium 20 mg (0.5 mg/kg), and the airway was secured with 6.0 mm reinforced endotracheal tube. Invasive blood pressure monitoring was done after induction, foley's catheter was passed, bispectral index (BISS) monitor was attached to monitor the depth of anaesthesia (target: 40-60), and an upper oesophageal temperature probe was placed. Prone positioning was then done. Anaesthesia was maintained with total intravenous anaesthesia (TIVA) spine surgery protocol of our department. The latter includes two infusions of 50 ml volume at 0.33 ml/kg/hour. Infusion 01 has Propofol 40 mg, Xylocaine 45 mg, and ketamine 30 mg, while infusion # 02 has fentanyl 150 µg, Dexmedetomidine 30 µg, and midazolam 5 mg. When these infusions are started at 0.33 ml/kg/hour, the individual drug doses are propofol @3 mg/kg/hour, ketamine @0.2 mg/kg/hour, xylocaine @0.3 mg/kg/hour, fentanyl @1 µg/kg/hour, midazolam @0.05 mg/kg/hour, and dexmedetomidine @0.2 µg/kg/hour. Infusion # 02 is to be held 30-45 minutes before skin closure, and infusion # 01 is to be halved when muscle closure starts and held when skin closure is completed. Somatosensory evoked potentials (SSEP) and motor evoked potentials (MEP) were monitored throughout the surgery. Goal-directed fluid therapy was the target. The patient's lungs were ventilated with oxygen and medical air with a target FiO_2 of 60% making a total flow of 2 L/min, ventilated with volume control mode with tidal volumes of 300 ml/breath, respiratory rate of 16/min, positive end-expiratory pressure (PEEP) 0 cmH_2O, monitoring of peak inspiratory pressures, and targeting the $EtCO_2$ between 32-35 mmHg.

Total procedure time was 150 minutes, and it went uneventfully with stable haemodynamics, adequate urine output, and intact SSEP and MEP throughout the procedure. The patient was then extubated in the supine position, making adequate tidal volume, fully awake, and responding to vocal commands. Total estimated blood loss was 200 ml, urine output was 220 ml (70 ml/hour), and fluid given was 800 ml. The patient was then shifted to the post-anaesthesia care unit (PACU) with stable haemodynamics and no sensory or motor weakness. She was pain-free (VAS score 1/10). Oral intake was encouraged. No respiratory distress was noted, and she was mobilised during her PACU stay. She was then shifted to the ward after 88-hour stay in PACU and discharged on the second postoperative day with stable vitals and normal neurology.

DISCUSSION

Patients with kyphoscoliosis have a loss of chest wall elasticity and lung compliance, increasing the work of breathing. This will result in a reduction in vital capacity, functional residual capacity, and total lung capacity, while residual volume remains normal, and these patients have compensatory increased respiratory rate.[7] Long-term presence of these changes causes chronic hypoxemia, hypercarbia, infections, and pulmonary vasoconstriction, resulting in irreversible pulmonary vascular changes leading to PH, and eventually right-sided congestive heart failure and cor pulmonale. The severity of cardiopulmonary impairment is determined by the degree of Cobb angle.[2] The pulmonary impairment severity also depends on the number of vertebrae involved, cephalad location of the curvature, speed of onset of scoliosis, and aetiology of neuromuscular weakness.

The authors did not perform pulmonary function tests as the patient had no dyspnoea or shortness of breath while running and had a good cough reflex. Chest x-ray had no major respiratory abnormality. The above investigations are required in any patient with dyspnoea at rest or shortness of breath on mild activity and Cobb angle of more than 65 degrees, patients with a vital capacity of less than 40% are not fit for elective correction of kyphoscoliosis.

As in this patient, a systolic murmur was present, so after doing FOCUS, detailed echocardiography was ordered which showed a

large ASD with left-to-right shunt. ASDs are grouped as septal defects of less than 3 mm (closes spontaneously), defects between 3-8 mm in size (spontaneously close 80% of the time), and defects >9 mm (usually fail to close).[7] This patient had an ASD of 24 mm, much larger than 9 mm. However, this patient had no signs of exercise intolerance or shortness of breath. The left-to-right shunt depends on the size of the defect, and right ventricular failure and increased pulmonary vascular resistance (PVR) develop as the shunt progresses.[8] The patient had mild PH, so the authors avoided the use of nitrous oxide and ketamine; and targeted good anaesthetic depth, as the goal of anaesthetic consideration in a patient with PH is to avoid factors causing an increase in pulmonary and systemic vascular resistances. Nitrous oxide is avoided to prevent a rise in PVR. Hyperventilation, factors causing sympathetic nervous system stimulation, and acidosis should be avoided. Core temperature should be kept as close to normal values as possible, and intrathoracic pressure should be minimised.

ASD is a left-to-right shunt condition, but due to a rise in either the PVR or SVR, it may convert into a right-to-left shunt, which raises the concerns of paradoxical cerebral embolism. The risk of this complication is increased by PEEP in mechanical ventilation.[9] Therefore, the authors did not apply PEEP in this case and remained very vigilant in not allowing any air bubbles to enter intravenous fluids.

Left-to-right shunts usually have minimum or no change on the induction speed of inhalational agents. While patients with right-to-left shunts show slow induction speed with inhalational agents with greater impact on less soluble inhalational anaesthetic agents.[10,11]

Prone positioning concerns should be looked upon and all the pressure areas should be covered. The abdomen and genitals should be free of any pressure. Arms should be positioned properly. Facial structures should be checked frequently for any pressure, and the endotracheal tube should be secured very tightly to prevent dislodgment.

In conclusion, monitoring of haemodynamic parameters, $EtCO_2$, peak inspiratory pressures, maintaining normothermia, urine output monitoring, avoiding hypoxia, hypercarbia, and acidosis, avoiding PEEP, avoiding air bubbles in IV fluids or drugs, and avoiding factors that cause a rise in SVR and PVR are the target goals for anaesthesia management in a patient with congenital kyphoscoliosis and ASD.

PATIENT'S CONSENT:
Informed consent was obtained from the patient's father to publish the data concerning this case.

COMPETING INTEREST:
The authors declared no conflict of interest.

AUTHORS' CONTRIBUTION:
SUR: Conception of the study.
AM: Drafting of the work for the important intellectual content.
AR: Drafting of the work for important intellectual content.
KA: Proofreading.
All authors approved the final version of the manuscript to be published.

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