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Mitral Valve Repair in Two Children with Scoliosis

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The Scoliosis Research Society (SRS) defines scoliosis as a lateral curvature of the spine greater than 10°, as measured by the Cobbs method on a standing X -ray of the thoraco - lumbar spine. Scoliosis has a varied aetiology amongst which Idiopathic type is the most common accounting for 75-90% of cases[1]. Congenital scoliosis is yet another type which results from vertebral anomalies that occurs due to an imbalance in the longitudinal growth of the spine[2]. Being a developmental disorder, it is often associated with intraspinal, genitourinary and cardiovascular abnormalities, of which genitourinary is the most common. The incidence of cardiovascular abnormalities in congenital scoliosis is reported to be between 7% and 15.15% in various studies[3]. Mitral valve prolapse is one among them[1]. Here we report the successful management of two patients who presented with severe mitral regurgitation due to mitral valve prolapse and who also had scoliosis of their thoracic spine.

The first patient was an 8 year old female child (figure 1) and the second was a 13 year old female. Both presented with dyspnoea on exertion (grade III–IV), tachypnoea with respiratory rate around 30-40/minute and palpitations for 3–4 years. Both were diagnosed with scoliosis, mitral valve prolapse and mitral regurgitation and were on treatment with Tablets Enalapril and Frusemide. Further clinical details are given in the tabular column (Table 1).

Anaesthetic Management: Both patients underwent mitral valve repair under cardio pulmonary bypass. The anaesthetic management of both patients was more or less the same and included

a) Standard pre induction monitors of electrocardiogram and pulse oximeter
b) Securing wide bore intravenous access and radial arterial line
c) Induction with Etomidate (0.1 – 0.3 mg/kg), Midazolam (0.05 – 0.1mg/kg), Fentanyl (1 -2 µg/kg) and Vecuronium (0.1 – 0.15mg/kg)

d) Central line insertion in right internal jugular vein for central venous pressure monitoring and administration of vasoactive agents
e) Monitoring of nasopharyngeal temperature, arterial blood gas and urine output.
f) Maintenance of anaesthesia was with a mixture of air and oxygen (50% each) and low concentrations of Sevoflurane (0.5 – 2%), and intermittent boluses of Fentanyl, Midazolam and Vecuronium.

PATIENT	AGE (YRS)	SEX	HEIGHT (cm)	WEIGHT (Kg)	CHEST X-RAY	ECG	ECHOCARDIOGRAM	ASSOCIATED ANOMALIES
1	8	F	131	13.4	Cardiomegaly. Cardiothoracic ratio > 0.8. Scoliosis with curvature to right.**(figure 2)**	Normal sinus rhythm . Left axis deviation	Atrial situs solitus. Dextroversion. Severe mitral regurgitation [grade IV to V] due to severe mitral valve prolapse. Large systolic orifice of mitral valve. Dilated left atrium. Mild tricuspid regurgitation. Dilated pulmonary arteries, Pulmonary artery pressure- 65 mmHg	Pectus carinatum **(figure 3)**
2	13	F	157	33.4	Cardiomegaly. Cardiothoracic ratio > 0.9. Scoliosis with curvature to right.	Normal sinus rhythm . Left axis deviation. Right bundle branch block.	Situs solitus. Severe mitral regurgitation [grade III-IV/IV] due to mitral valve prolapse. Small to moderate ostium secundum atrial septal defect. Severe left heart dilatation. Mild right heart dilatation.	Pectus excavatum. Features of Marfans: Arm span > Height, high arched palate, Ectopia lentis.

Table 1: Patient demographic details and investigations details

Surgical Technique:

a) Median sternotomy was done and cardio pulmonary bypass established with aortobicaval cannulation after systemic heparinisation.
b) Patients were cooled to 28°C
c) Aorta cross clamped and mitral valve repair done.
d) Cross clamp released and after adequate re-warming, both patients were weaned off cardiopulmonary bypass with inotropic supports of Dobutamine 10µg/kg/min, Adrenaline 0.1 µg/kg/min and Nitroglycerine 0.5µg/kg/min
e) The second patient also underwent a pectus excavatum repair.
f) After ensuring adequate haemostasis and chest closure, both patients were shifted to intensive care unit for elective post operative mechanical ventilation.
g) The intra operative hemodynamic data and procedural details are given in Tables 2 and 3.

PATIENT	PREINDUCTION			POST INDUCTION				AFTER WEANING OFF CARDIOPULMONARY BYPASS			
	HR (beats/min)	BP (mm Hg)	SpO_2 (%)	HR (beats/min)	BP (mm Hg)	CVP (mm Hg)	SpO_2 (%)	HR (beats/min)	BP (mm Hg)	CVP (mm Hg)	SpO_2 (%)
1	128	100/70	100	100	90/60	5	100	125	110/56	6	100
2	94	84/55	100	95	110/70	7	100	92	116/72	8	100

Table 2: Hemodynamics during the course of surgery

PATIENT	PROCEDURE DONE	CARDIOPULMONARY BYPASS TIME (MIN)	CROSS CLAMP TIME (MIN)	DURATION OF VENTILATION
1	Teflon ring annuloplasty. Commissural fusion. Left atrium reduction.	117	90	13 hours
2	Teflon ring annuloplasty. Commissural fusion. Pericardial patch closure of atrial septal defect. Pectus excavatum repair.	87	54	Reintubated twice and tracheostomy done. Mechanical ventilation for 10 days

Table3: Procedural details and postoperative course

Figure 1: Spine of the patient 1 showing gross scoliosis

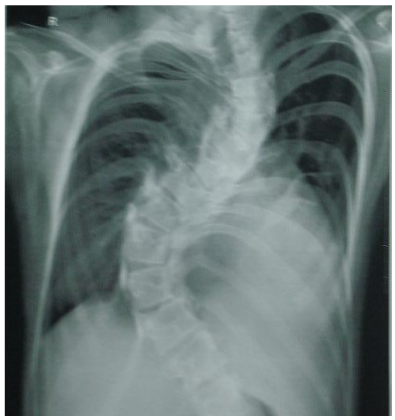

Figure 1: Preoperative chest X ray of patient 1 showing gross scoliosis

Figure 3: Side view of the patient 1 showing pectus carinatum

Post Operative Course:

The first patient had a shorter stay in the intensive care unit. She was extubated on the morning of first postoperative day with duration of ventilation of 13hours. She was weaned off her inotropes by the second postoperative day. She developed mild left ventricular dysfunction on the fourth postoperative day and was treated with Dobutamine 5µg/kg/min and Enalapril 1.25mg bd. Her ventricular function improved and she was weaned off dobutamine on the fifth postoperative day. Sildenafil 5mg was added. She was discharged out of intensive care unit on sixth postoperative day and discharged home on tenth postoperative day.

The second patient had a stormy postoperative course. She was extubated after one day of elective mechanical ventilation. Post extubation, she had mild tachypnoea (RR 38/min). She was reintubated twice due to worsening tachypnoea and desaturation and finally tracheostomy was done with 6.5mm cuffed tracheostomy tube. She developed collapse of right lower lobe. The patient received injection cefuroxime from postoperative day one. She grew pseudomonas aueroginosa in her endotracheal tube culture. Based on culture and sensitivity reports, she was started Inj. Amikacin. She required mechanical ventilator support for a period of 10 days, after which she was observed in the ICU for a further 2 days. She was sent to ward on the 19th POD and discharged home after decannulation of the tracheostomy tube on the 31st POD.

Discussion: Scoliosis, a complex deformity of the spine, results in lateral curvature and rotation of vertebrae, and deformity of rib cage[1]. It is usually associated with secondary involvement of the respiratory, cardiovascular and neurologic systems. The prevalence of scoliosis in the general population varies between 0.3–15.3%[4]. The prevalence is less than 3% for curves more than 10° and less than 0.3% for curves more than 30°. The incidence of scoliosis is also found to be higher in patients with congenital heart disease than in normal subjects[5].

Scoliosis by itself poses various challenges to the anaesthesiologist, which includes –

1. Airway difficulties - especially if it involves cervical/upper thoracic vertebrae.

2. Respiratory system - The abnormality in ribcage in patients with scoliosis leads to a restrictive type of ventilator effect. The severity of pulmonary involvement is influenced by the severity of scoliosis (>70°), number of vertebrae involved (7 or more), and a cephalad location of curvature. In our patients, pulmonary function test was not done as they were not able to do the procedure satisfactorily. The factors associated with post operative mechanical ventilatory requirement includes – pre existing neuromuscular disease, severe restrictive pulmonary dysfunction with a vital capacity less than 35%, associated congenital heart defects, right ventricular failure and obesity. Of these, our patients had one risk factor (associated congenital heart defect) and we could not grade the severity of their pulmonary dysfunction. While one of our patients (patient 2) required prolonged ventilator support the other had a short duration of ventilation.

3. Cardiovascular system - the cardiovascular system involvement in scoliosis may be of two types –

a) Associated abnormalities, among which MVP is the most common, and which was seen in both of our patients and

b) Secondary to scoliosis - This includes alterations due to changes in the structure of mediastinum and secondary to the effects of chronic respiratory insufficiency, leading to chronic hypoxemia and pulmonary hypertension. Our patients had severe pulmonary hypertension, which probably was partly due to mitral regurgitation and partly aggravated by scoliosis.

4. Neurologic system - A detailed neurologic examination and documentation is required for medicolegal purposes.

5. Positioning of patient – Positioning of the patient on the operating room table for surgery or invasive line insertion could be difficult, though we didn't encounter any difficulties in either of our patients[4].

Mitral valve prolapse is defined as the billowing of mitral leaflets into the left atrium during systole. Utilizing the current echocardiographic criteria for diagnosing mitral valve prolapse (valve prolapse of 2mm or more above the mitral annulus in the long axis para sternal view), the prevalence of this entity is 1–2.5% of general population[6]. Mitral valve prolapse occurs in two phenotypic patterns:

1. An anatomic form characterised by thickened, billowing leaflets and
2. A functional form, due to dynamic systolic expansion of the mitral annulus.

The anatomic form occurs in 15 – 20% of patients with mitral valve prolapse and represents those who experience progressive valve deterioration and significant mitral regurgitation and those who require valve repair/replacement[7]. The anatomic form has been found to be associated with other cardiac abnormalities, which includes ostium secundum atrial septal defect, hypertrophic cardiomyopathy, Ebsteins anomaly and WPW syndrome[6]. Extra cardiac association include connective tissue disorder like Marfan's syndrome and Ehlers–Danlos syndrome and thoracic abnormalities like pectus excavatum, pectus carinatum and scoliosis. Among these, both of our patients had thoracic deformity in the form of scoliosis with pectus excavatum/carinatum. Our second patient had an ostium secundum atrial septal defect and features of Marfans syndrome also. Though a variety of cardiac arrhythmias are associated with mitral valve prolapse, both of our patients had a normal sinus rhythm preoperatively.

The natural history of mitral valve prolapse is a heterogeneous, and varies from benign with normal life expectancy to adverse with significant mortality and morbidity. Kolibash et al[8], in their follow up study of 86 patients with severe mitral valve prolapse and mitral regurgitation showed that, on an average, it was 25 yrs from the diagnosis of mitral valve prolapse until severe mitral regurgitation developed, but once significant mitral regurgitation developed, mitral valve surgery was required within 1 year in almost all of the patients. But in both of our patients, there was a very rapid progression to severe mitral regurgitation. Patients with mitral valve prolapse that progresses

to require mitral valve surgery typically have pulmonary artery pressures (PAP) greater than 30mmHg[7]. It has also been found that chronic mitral regurgitation is associated with pulmonary hypertension in as many as 76% of cases, even in those with preserved left ventricle function. Both of our patients had severe pulmonary hypertension (PAP ~60mmHg) with preserved left ventricle function.

Anaesthetic considerations in patients with mitral regurgitation includes–

1. Avoiding sudden decrease in heart rate – which can cause an abrupt left ventricle volume overload

2. Avoiding sudden increases in systemic vascular resistance – which causes a decrease in forward flow and an increase in regurgitant flow

3. Avoiding drug induced myocardial depression[9].

It is for these reasons that Etomidate has been suggested to be an attractive choice for induction of anaesthesia, which was used in both of our patients. Maintenance of anaesthesia with low concentrations of inhalational agent is suggested to increase forward flow by their vasodilatory effect and at the same time, avoid depression of systolic myocardial function. The use of nitrous oxide in patients with pulmonary hypertension may lead to a further increase in pulmonary artery pressure and reduce right ventricle ejection. All of these were followed in the anaesthetic management of our patients.

Conclusion: Successful management of patients with diseases involving various systems of the body requires thorough evaluation, optimal preoperative preparation and meticulous planning. In our patients, there was a compromise in both respiratory and cardiovascular systems, imposed by two different disease processes – mitral regurgitation (due to mitral valve prolapse) and scoliosis. Both also belonged to the paediatric age group. While one of the patients had a smooth intra operative and postoperative course, the second patient had a stormy post operative period requiring prolonged ventilatory support. We report these two cases to highlight the variations in early outcome in the management of such patients presenting with more than one disease process involving multiple systems of the body.

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