



Anaesthetic Management of Scimitar Syndrome: A Case Report”

Ajmer Singh¹, Neeraj Sharma²

1 Senior Consultant, 2 Associate Consultant

Department of Cardiac Anaesthesia

Fortis Escorts Heart Institute, Okhla Road, New Delhi-110025

Corresponding Author: Dr. Ajmer Singh (ajmersingh@yahoo.com)

About the Author: Dr Ajmer Singh did his MD (Anaesthesia) in 1993. He did senior residency in Cardiac Anaesthesia from AIIMS. He joined Escorts Heart Institute in 1996 and is working there as a Senior Consultant. His areas of interests are Anaesthesia for Paediatric and Congenital heart diseases.



Abstract: Scimitar syndrome, a rare congenital cardiopulmonary anomaly, consists of partial anomalous pulmonary venous connection of the right lung to the inferior vena cava, right lung hypoplasia and anomalous systemic arterial supply to the right lung. A 5 month old child with scimitar syndrome and pulmonary hypertension, underwent stenting of anomalous pulmonary vein, and coil embolization of systemic collateral artery to the right lung. Surgical ligation of large patent ductus arteriosus was performed to further decrease pulmonary artery pressure. Child was discharged after a successful combined therapy of surgical and transcatheter intervention.”

Case Report: A 2.5 kg, 38 week gestation female infant, delivered through a caesarean section for cephalopelvic disproportion after an uncomplicated pregnancy, was noticed to have suprasternal retraction since birth by her mother. The baby did not undergo any detailed workup or investigation. At two months of age, child had an episode of respiratory infection and required treatment with antibiotics and decongestants. At 5 month of age, child presented with severe respiratory distress. On examination she weighed 3.5 kg, had a pulse rate of 126/min, blood pressure of 82/56 mmHg, respiratory rate of 48/min with suprasternal retraction and cyanosis (room air saturation 84%). Breath sounds were diminished in right side of chest and heart sounds were shifted to the right with grade III/V murmur. Liver was palpable 3 cm below right costal margin. Rest of the systemic examination was unremarkable. Child was given supplemental oxygen. Decongestive therapy in the form of digoxin & furosemide was started and nasogastric feeds were initiated. Patient was admitted for stabilization and further evaluation.

Chest radiograph showed normal sized heart with cardiac silhouette shifted to right, prominent bronchovascular markings on the left side and right lung hypoplasia. 2-Dimensional Transthoracic echocardiogram revealed anomalous right pulmonary veins draining into sub diaphragmatic part of IVC, fossa ovalis atrial septal defect (ASD) of 5 mm with right to left shunt, large patent ductus arteriosus (PDA) of 5 mm with bidirectional shunt and severe PAH. An obstruction was noted at the junction of the right pulmonary venous confluence and the IVC. A diagnosis of Scimitar Syndrome was made. For further evaluation a 16 slice computerized



tomography (CT) angiogram of pulmonary tree was performed using omnipaque (350 mg/ml 1.2 cc) which revealed anomalous pulmonary venous drainage on the right side. Left pulmonary veins were draining into left atrium. An arterial collateral arising from descending aorta was seen supplying the posterior basal segment of lower lobe of the right lung, suggestive of sequestration of lung. Pulmonary sequestration was defined as segment of lung not connected to tracheo-bronchial tree. CT pulmonary angiography also confirmed presence of ASD, PDA, bilateral superior vena cava, hypoplastic right pulmonary artery and hypoplastic right lung.

The management plan comprised of cardiac catheterization, stenting of the pulmonary venous obstruction and coil embolisation of the arterial collateral to sequestered right lung, followed by surgical correction at later stage if symptoms persisted. After obtaining informed consent from parents, child was premedicated with chloral hydrate 50mg/kg orally and atropine sulphate 10 µg/kg intramuscularly, 45 minutes before the procedure. In cardiac catheterization laboratory heart rate, electrocardiogram, digital pulse oximetry and radial artery pressure were monitored. Anaesthesia was induced with midazolam 0.05 mg/kg, fentanyl 5 µg/kg and ketamine 1 mg/kg intravenously. Trachea was intubated with oral endotracheal tube (3.5 I.D, Portex), facilitated by vecuronium 0.1 mg/kg. Mechanical ventilation was initiated with respiratory rate of 30/min, tidal volume of 40ml/min, positive end expiratory pressure (PEEP) of 5 cm of H₂O and inspired oxygen concentration (FiO₂) of 0.6. Anaesthesia was maintained with oxygen: air, midazolam infusion 1µg/kg/min and ketamine infusion 50 µg/kg/min intravenously. Cardiac catheterization data are shown in table1.

Table 1: Catheterization Data	S. No	Site	Pressure(mm Hg)	Saturation (%)
RA: Right Atrium	1.	RA	a ⁹ v ⁴ (6)	90%
RV: Right Ventricle	2.	RV	58/6 (25)	90%
PA: Pulmonary Artery	3.	PA	58/28 (42)	96%
PV: Pulmonary Vein	4.	PV	37/17 (20)	100%
	5.	Aorta	70/33 (47)	100%

Child had high pulmonary artery pressure (PAP) 58/28 (42) mmHg and pulmonary venous pressure was 37/17 (20) mmHg. Stenting of the pulmonary venous obstruction (at junction of IVC) was done with Palmaz Genesis stent (6mm x 15 mm, Cordis Europa N.V. Netherlands) mounted on balloon. Coil embolization of systemic collateral artery from descending aorta to the right lung was done with Cook's coil (0.038"x 4cm x 4 mm. M. Reye embolization coil, Williams Cook Europe, CE). The procedure was uneventful, child was shifted to intensive care unit (ICU) and ventilated electively.

In ICU , child could not be weaned off the ventilator for 3 days, because of persistently high peak and mean airway pressure, high PCO₂ (more than 50 mmHg) and frequent episodes of



pulmonary hypertensive crises despite inodilators (milrinone 0.4 µg/kg/min) and vasodilator (nitroglycerine 1.0 µg/kg/min) therapy. Pulmonary hypertensive crisis was suspected when child had bradycardia, systemic hypotension (means BP less than 45 mmHg) and high peak airway pressure. Transthoracic echocardiography showed suprasystemic PAP to confirm the diagnosis. Sedation and analgesia were ensured with intravenous midazolam 1µg/kg/min and morphine 10µg/kg/hr infusion. Boluses of fentanyl (3-5 µg/kg intravenously) were used to prevent PAH crisis before doing any procedure such as endotracheal suction or change of posture.

It was decided to ligate PDA surgically on 3rd day of the admission, to further reduce the PAP. Anaesthesia was maintained on oxygen: air: isoflurane (1% end tidal concentration) and intermittent doses of midazolam, fentanyl and vecuronium. During PDA ligation, systemic arterial pressure was 78/30(45) mmHg and PAP decreased from 50/20(32) to 32/15(20) mmHg. There was a decrease in peak and mean airway pressures and PCO₂ became normal. The trachea was extubated after two days of PDA ligation. After extubation room air saturation was 91%. Rest of the postoperative period was uneventful. Child was discharged on 8th day after admission. Follow up echocardiogram done after 3 months showed unobstructed pulmonary venous flow to IVC with no evidence of stenosis and mild PAH. Future therapeutic plan was surgical reimplantation of scimitar vein into left atrium and closure of ASD.

Discussion: First described in 1836, Scimitar Syndrome is a rare constellation of congenital cardiopulmonary anomalies consisting of a partial anomalous pulmonary venous connection of the right lung to the inferior vena cava (IVC), right lung hypoplasia, dextroposition of the heart and anomalous systemic arterial supply to the right lung^{1,2} The term Scimitar Syndrome was coined because of radiographic appearance of anomalous vein, which appears as a tubular opacity paralleling the right cardiac border resembling a Turkish sword or Scimitar.³

It has been reported most widely in adults and older children and is usually found during a workup for dyspnoea, recurrent respiratory infections, or as an incidental finding on a routine chest radiograph. This adult form of Scimitar Syndrome usually is not associated with pulmonary artery hypertension (PAH) and typically has a benign prognosis. A second, infantile group of patients become symptomatic soon after birth. Their course is often complicated by severe PAH and cardiac failure, making management difficult and mortality high.⁴ Scimitar Syndrome occurs in approximately 1-3 per 100,000 live births.⁵ A triad of respiratory distress, right lung hypoplasia and dextroposition of heart should alert the clinician to the possibility of this syndrome.⁶ Surprisingly, Scimitar Syndrome is almost exclusively a right sided anomaly.

This case is being reported because of paucity of information regarding anaesthetic management of Scimitar Syndrome in literature. Diagnosis of Scimitar Syndrome is based on echocardiography findings, which may delineate scimitar vein, systemic arterial supply to the lung, and associated cardiovascular anomalies. CT pulmonary angiogram is helpful in visualizing the anomalous pulmonary vein. Cardiac catheterization and angiography are probably the most useful procedures for confirming the diagnosis and clarifying the exact degree of PAH.



The decision to tackle the problem by an interventional method was taken as surgical correction in neonates and small infants have shown a mortality rate as high as 50 percent.⁵ Interruption of anomalous arterial supply helps to avoid or defer definitive surgery during infancy. It also helps these patients to grow until their vein are bigger, at which stage a more generous anastomosis could be made, which is expected to stay patent longer.

A combination of midazolam and ketamine is frequently used technique for anaesthetizing children undergoing cardiac catheterization. Midazolam is an effective sedative hypnotic agent, which maintains cardiovascular stability and has a wide margin of safety. Potential advantages of ketamine include sympathetic stimulation with support of blood pressure and heart rate, excellent sedation and analgesia. Malviya et al.⁷ in a retrospective review of 184 interventional cardiac procedures found ketamine infusion as the most frequently used technique. Singh et al.⁸ also found ketamine infusion as a simple, safe and effective method for anesthetizing children in the cardiac catheterization laboratory for interventional cardiac procedures. Oklii et al⁹ in a prospective randomized study (n = 41) compared haemodynamic effects of propofol with ketamine during paediatric catheterization and found that ketamine produced no changes in PAP, systemic and pulmonary vascular resistances, while propofol produced decrease in systemic vascular resistance, increase in right to left shunting and arterial desaturation. Recently William's et al¹⁰ also found no significant differences in mean blood pressure, mean PAP, systemic and pulmonary vascular resistance indices, cardiac index, pH and PCO₂ after ketamine infusion in spontaneously breathing children with severe PAH. The key anaesthetic goals in these patients include prevention and management of PAH crisis. This is ensured by smooth induction, avoidance of hypoxia and hypercarbia during and post procedure. Adequacy of sedation and analgesia must be ensured by using appropriate drug therapy. Additional boluses of narcotic such as fentanyl should be used before performing any manipulation such as endotracheal suction.

Infants with Scimitar Syndrome deteriorate mainly from anomalous systemic arterial supply, significant intracardiac shunt and pulmonary vein stenosis.¹¹ Other cardiovascular malformations such as a large PDA (as observed in our case) or reduction of the pulmonary vascular bed, are also main causes of PAH. Gao et al¹² have described balloon angioplasty of scimitar vein followed by placement of a balloon- expandable stent to reduce the degree of PAH. Surgical ligation or catheter embolization of the aortopulmonary collateral often produces dramatic improvement from cardiac failure in infants without significant associated cardiovascular anomalies. Oshima et al¹¹ found a dramatic improvement in clinical symptoms and decrease in left to right shunt ($Q_p/Q_s = 1.9$ from 4.1) after performing transcatheter coil embolization of anomalous systemic artery. Wang et al¹³ have also reported coil embolization of the abnormal feeding arteries in one out of five cases. Muta et al¹⁴ performed coil occlusion of collateral arteries which resulted in improvement of symptoms of CHF, followed by surgical correction three months later. Uthaman et al¹⁵ in a series of 23 patients, found a decrease in left to right shunt and degree of PAH in more than 90% cases after interruption of anomalous arterial supply. Definitive surgery was deferred for optimal results for a mean period of eight months.



The postoperative course of Scimitar Syndrome is often characterized by episodes of acute pulmonary hypertension. Hypercarbia and residual left to right shunt (large PDA in this patient) are important causes of pulmonary hypertension. PDA ligation was performed to reduce pulmonary artery pressure and this helped in successful weaning from the ventilator. Inhaled nitric oxide may be useful in patient with pulmonary hypertension, but its nonavailability precluded the use.

In conclusion, a successful combined treatment by surgical and transcatheter intervention in an infant with Scimitar Syndrome is reported. These patients are prone to PAH crisis, which must be anticipated and prevented by employing appropriate drug therapy.

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