

Anaesthetic Management Of A Patient With Atrio-Ventricular Discordance And Single Ventricle Physiology Posted For Liposuction- A Case Report.

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Abstract- Congenital heart disease is the most common congenital abnormality, accounting for 4-10 cases per 1000 live births. It is approximately one third of all congenital defects. The incidence of congenital heart disease in children has remained constant over the last few decades. However because the therapeutic options have improved the number of adults with congenital heart disease has steadily increased. And these patients presenting for non cardiac surgery has increased over the years.¹ Here we report the anaesthetic management of a 23year old male with atrio-ventricular (AV) discordance and single ventricle who had undergone palliative treatment for congenital heart problem has come for liposuction.

Index Terms- AV discordance, Single Ventricle physiology, Bidirectional Glenn shunt, Fontan physiology, Fenestrated Fontan

I. INTRODUCTION

Patients with AV discordance and single ventricle physiology pose a unique challenge for the anaesthetist as it involves a complex physiology and these patients are constantly at risk of developing cardiac complications like ventricular dysfunction, arrhythmias, protein losing enteropathy and plastic bronchitis.² These patients undergo palliative treatment during childhood for correcting the single ventricle deformity by creating a cavopulmonary shunt and thereby allowing single ventricle for systemic circulation. These patients present for non cardiac surgery and poses challenge for anaesthetist as understanding the physiology behind this deformity is complex. According to ACC/AHA 2008 guidelines treatment of patients should be carried out in regional adult cardiac center and cardiologist consultation prior to surgery is important.³ A clear understanding of the anatomy and physiology in these patients are important in peri-operative management of these patients.

II. CASE REPORT

A 23 year old male coming from Salempur had come to the hospital for liposuction of bilateral gynaecomastia. Patient was diagnosed to have congenital heart disease of AV discordance with single ventricle since birth. He had history of frequent cyanotic spells after birth during crying, feeding and was relieved when the baby was placed in knee chest position. He underwent Bidirectional Glenn procedure when he was 7 year old and Fenestrated Fontan surgery when he was 10 year old. He underwent device closure of Fontan fenestration and coil closure of decompressing vein when he was 11 year old. He had been followed up regularly. Patient was on tablet Warfarin 2.5mg once daily and INR was maintained at 2.5. Medications were stopped 3 years back.

He had history of presyncope for 4months since April 2019 and the episodes were not related to exertion, not associated with palpitations and lasted for few seconds. Patient was investigated with 24 hours Holter monitoring and was found to have sinus tachycardia which settled down without medications.

Preoperatively patient was assessed for his general physical condition and cardiopulmonary status since patient had history of congenital heart disease. Patient gave a history of ability to climb more than two flight of stairs with no discomfort. Airway examination revealed Modified Mallampati Grade II, adequate mouth opening of more than two fingers, no restriction of neck movements and normal thyromental distance. Weight of the patient was 91kgs and height was 179cm with BMI of 28kg/m². Systemic examination revealed Cardiovascular system – S1-normal, single S2 and no murmurs. Respiratory system revealed bilateral air entry was present with no crepitations or wheeze. Room air saturation was 95%, Heart rate – 83/Min, Bloodpressure – 135/71mmHg. On investigation complete blood count, Renal function test, Liver function test and coagulation profile were within normal limits.

ECG showed sinus rhythm with rate of 83/min and T wave inversion in V1, V2, V3, V4 and T wave flattening in V5, V6. ECHO showed Post device closure of fenestration in Fontan, Extra

cardiac conduit from IVC to RPA, well functioning Fontan circuit. No residual fenestration flow, Confluent good sized branch Pulmonary arteries, no AV regurgitation and good ventricular function. 6minute walk test was done which showed %prediction of 76% and Borg scale of 0-0.5 which is no breathlessness to very slight breathlessness.

Anaesthetic consideration for this patient were AV discordance with single ventricle physiology, Pressure dependent pulmonary circulation and intraoperative hypoxia, hypercarbia and acidosis.

III. CONDUCT OF ANAESTHESIA

Patient was kept nil by mouth for 6hours. After receiving the patient in operating room, Infective endocarditis prophylaxis of Inj.Ampicillin2g Iv stat was given 30minutes before the procedure. Standard anaesthetic monitors are connected and baseline readings are measured. Heart rate was 88/min, Blood pressure was 132/63mmhg, Spo2 without oxygen was 98%. Inj.Midazolam 1mg Iv was given 20minutes before induction as a premedicant. Patient was then induced with Inj.Morphine 10mg, Inj.Propofol 160mg and Inj.Vecuronium 8mg and Preoxygenated with 100% oxygen for 3minutes. Patient was then intubated with 8.5 portex endotracheal tube.

Anaesthesia was maintained with oxygen/ Air/ and TIVA with Propofol at 4-12mg/kg/hr infusion titrated to the effect. IV fluid is maintained with Ringer lactate at 2ml/kg/hr. The surgery was completed in 2hours with minimal blood loss. Patient was extubated when fully awake and responding to commands and was observed in 30min in the recovery room. Postoperatively patient was kept in Post anaesthesia care unit. Patient complained of mild-moderate pain during POD-0 and he was discharged to ward on POD-3 with stable hemodynamics.

IV. DISCUSSION

PHYSIOLOGY OF SINGLE VENTRICLE

Patients with single ventricle and AV discordance is associated with complex physiology and these patients require palliative procedures to have a better chance of living and with the advances in the treatment modalities these patients survive till adulthood and there is a high probability of these patients coming for non cardiac surgery. Patients with single ventricle usually have a parallel pulmonary and systemic circulation instead of being a series circulation. This leads to significant cyanosis and ventricular volume overload.⁴

This congenital heart disease is not compliant for a full anatomical correction, which is biventricular repair. Therefore these patients will be palliated by creating a circulation based on single ventricle. The palliative surgery is done to make the parallel circulation into series circulation and to reduce the cyanosis and ventricular volume overload thereby reducing the chance for ventricular failure and death. These procedures are done in a staged manner for better survival of the baby. These patients require a complete cavo-pulmonary anastomosis, so the single ventricle pumps oxygenated blood to the body while blood flows passively to lungs down the pressure gradient from pulmonary artery to left atrium.

Stage I is the Norwood procedure where an aorto-pulmonary shunt is created to connect aorta to main pulmonary artery to provide pulmonary blood flow.⁴ The balance between the systemic and pulmonary blood flow is very weak that makes non cardiac surgeries hazardous. The non cardiac surgeries should be restricted to urgent or emergent conditions. Stage II of palliation involves formation of bidirectional (supplying both right and left lungs) cardiopulmonary shunt that connects superior vena cava to right pulmonary artery and is usually done at 3-5 months of age. Even after this procedure the child remains cyanosed with oxygen saturation of 75-85%.

Stage III is the formation of total cavo pulmonary circulation or Fontan circulation. In this inferior vena cava is being connected to right pulmonary artery and with this procedure the pulmonary circulation is separated from Systemic circulation. This is usually done by 3-5 years of age and it normalise the arterial oxygen saturation. The sole determinant for pulmonary blood flow is the pressure gradient from pulmonary artery to the left atrium.⁵

The factors that determine the outcome of anaesthesia are pulmonary vascular resistance, intrathoracic pressure, positive end expiratory pressure (PEEP), peak inspiratory pressure (PIP), inspiratory time. Spontaneous ventilation increases pulmonary blood flow by negative intrathoracic pressure but the disadvantage is the difficult to maintain normocapnia. Peak inspiratory pressure should be minimized to facilitate pulmonary blood flow.⁵

BIDIRECTIONAL GLENN PROCEDURE

Bidirectional Glenn procedure involves formation of shunt between Superior vena cava and right pulmonary artery and it is a partial cardiopulmonary shunt as only partial venous circulation is diverted to pulmonary circulation. Generally neonates with single ventricle physiology require initial palliation with systemic pulmonary shunt called Blalock Taussig shunt before initiating cavo pulmonary circulation. But this Blalock Taussig shunt has no growth potential and is liable to thrombose which can lead to cyanosis and sudden death.⁶

Though Glenn procedure provides a source of pulmonary blood flow and reduces the severity of shunt dependent circulation and ventricular volume overload it is also associated with mortality and it was found that the cause of death were inadequate pulmonary blood flow and in some cases superior vena cava syndrome.⁶

FONTAN PHYSIOLOGY

In 1971, Fontan and Baudet described a palliative surgery in patients with complex congenital heart disease with single functioning ventricle.⁷ Before the development of Fontan procedure the pulmonary blood flow is usually established by systemic to pulmonary shunts. This improves the life expectancy but the survival rate without cavopulmonary shunt is very less.⁸ In Fontan physiology the systemic venous blood is drained directly into the pulmonary circulation and from there the oxygenated blood drains into the left atrium and then to single ventricle and now single ventricle is solely for the systemic circulation. There are two important physiological considerations. One is the presence of single ventricle and the other is entire systemic venous return enters the pulmonary circulation passively. Pulmonary blood flow is important in maintaining the systemic circulation because the cardiac output is dependent on pulmonary

circulation.⁹ The main determinants are the systemic venous pressure and volume, pulmonary vascular resistance, cardiac rhythm and left ventricular function. Any disturbance in any of these factors can affect the cardiac output and can prove detrimental.¹⁰ Generally these patients have increased central venous pressure and reduced cardiac output and oxygen saturation and so they have adaptations like increased arterial vascular resistance, cardiac output being redistributed to the vital organs and polycythemia.¹¹ Pulmonary vascular resistance is determined by mechanical and biochemical factors like positive pressure ventilation cause decreases pulmonary blood flow and atelectasis causes increase in pulmonary vascular resistance.

For induction of these patients coming for non cardiac surgery, drugs that depress the myocardial contractility like thiopentone and volatile agents should be avoided. Propofol with transient vasodilatation is usually less problematic if normovolemia is maintained. Avoid hypoxia, hypercarbia and inadequate analgesia as these factors increase the pulmonary vascular resistance and it will reduce the pulmonary blood flow and can cause cyanosis, hypoxia, sudden cardiac arrest and death.

V. CONCLUSION

Children with congenital heart diseases are more common and with the advancement in treatment modalities these patients surviving and reaching adulthood and such patients coming for non cardiac surgeries are increasing. Patient with AV discordance and single ventricle is complex and these patients would have undergone multiple palliative procedures. It requires thorough understanding of this complex physiology as there is increased risk of perioperative complications and thorough preoperative assessment of these patients which involves liaison between the surgeon, anaesthetist, paediatrician and cardiologist for successfully outcome in these patients is essential.

These patients coming for non cardiac surgery, the outcome depends on the type of surgery, physiological status of single ventricle post palliative procedures, physical status of the patient, anaesthesia technique and management. Preserving pulmonary blood flow is the priority in patients with Fontan physiology which means avoiding hypoxemia, high airway pressures and avoiding increased pulmonary vascular resistance. In this case, patient has come for liposuction with good physical status and with no complication post palliative procedures. Patient was induced with iv propofol and was maintained with total intravenous anaesthesia (TIVA) with propofol with no significant fluctuations in blood pressure. Volatile agents was avoided in view of myocardial depression and inhibition of protective hypoxic pulmonary vasoconstriction. Ventilation was maintained by controlled mode of ventilation to maintain normocapnia. Post operatively patient was stable and was discharged from post anaesthesia care unit on post operative day 2 with stable hemodynamics and maintaining room air saturation of >96%.

AUTHOR'S CONTRIBUTIONS

Collection of current admission information and previous cardiac surgery informations- G.K.

Review of literature and supervising the final year resident in writing the manuscript- N.M.

Overall supervision of writing the manuscript- K.S.

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