Review Article

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Glenn shunt: Anaesthetic concerns for a non cardiac surgery

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ABSTRACT

Patients with single ventricle physiology have single chamber for pulmonary and systemic venous return that is supplied in parallel leading to cyanosis and ventricular volume overload. Palliative surgeries in the form of superior cavo-pulmonary anastomosis, also known as Bidirectional Glenn shunt (BDG) and complete cavo-pulmonary anastomosis, also known as Fontan procedure are done to divert blood from superior vena cava and inferior vena cava respectively into pulmonary circulation. As a result systemic and pulmonary blood flow run in series and forward flow is dependent on the relationship of systemic vascular resistance and pulmonary vascular resistance. Patients who have undergone Glenn shunt usually have peripheral oxygen saturation in the range of 75-85 % as a consequence of non-diversion of blood from inferior vena cava into pulmonary circulation. Fontan procedure is done in a staged manner to avoid sudden unloading of ventricle leading to failure. Anaesthesiologists encounter patients with Glenn shunt for non-cardiac surgery before they have undergone completion Fontan. A thorough understanding of Glenn and single ventricle physiology is required to deal with such patients and meticulous approach to anesthesia management is required after discussing with the surgeons and cardiologist, regarding the type of non-cardiac surgery, and to know the functional status of Glenn shunt. The present review article aims to discuss the anaesthesia concerns, and search of terms such as ‘anaesthetic management’, ‘Glenn shunt’, ‘cavopulmonary anastomosis’, ‘Fontan procedure’ was carried out in KKH eLibrary, Medline, PubMed, and Google scholar focusing on current research, randomized control trials, review articles and editorials.

Key words: Cavopulmonary Anastomosis, Fontan Procedure, Glenn Shunt, Non-Cardiac Surgery

INTRODUCTION

Bidirectional cavo-pulmonary anastomosis is an important step in staged palliation of patients with physiologically univentricular heart[1]. Glenn procedure was first introduced by William Glenn in year 1958 and its modifications were done by Dr Azzolina in 1973[1,2]. It is the second procedure preceding the major Fontan procedure[3] done to treat patients with univentricular heart. The ultimate goal is to achieve optimal oxygen delivery for as low as systemic venous pressure as possible.

BIDIRECTIONAL GLENN SHUNT

The Bidirectional Glenn (BDG) is a type of palliative procedure done in infants via sternotomy to drain blood from systemic venous return (superior vena cava) from head and neck region to the pulmonary circulation. It is usually undertaken after 6 months of age because at that time pulmonary vascular resistance has decreased to a point where pulmonary blood flow can be maintained with systemic venous pressure being the driving force for forward

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blood flow. This procedure allows the de oxygenated blood from the upper limb and head to drain passively into lungs. The original description of Glenn procedure described the end to end connection between divided end of Superior Vena Cava (SVC) to right pulmonary artery whereas the modified technique involved the end to side connection of SVC to right pulmonary artery. The azygous vein is ligated unless the Inferior Vena Cava (IVC) is interrupted so that SVC does not decompress retrograde to IVC thereby reducing the quantity of blood delivered to pulmonary arteries[1]. Any previous aorto-pulmonary shunt like Blalock Taussig (BT) shunt is also taken down.

FONTAN PROCEDURE

Fontan is a definitive procedure in which total cavo pulmonary anastomosis is done usually after four years of age. In this procedure, in an already created BD Glenn shunt, the inferior vena cava blood is also diverted to pulmonary artery (Completion Fontan) or sometimes without previous Glenn shunt, both SVC and IVC blood is diverted to pulmonary arteries in a single sitting which is known as Primary Fontan[3].

WHY ARE GLENN AND FONTAN PROCEDURES STAGED IN TWO STEPS?

Glenn is done after 6 months of age whereas Fontan is done after 4 years of age, because in a newborn, there is relative volume overload because heart has to pump to both lung and body, and are not in perfect shape. Staging the Fontan prevents sudden unloading of the ventricle which could have led to sudden changes in mass volume ratio and ventricular failure. Also it provides adequate time for the lungs to adapt to changes of not having blood pumped but rather passively receive via cavo-pulmonary anastomosis, thus allowing adequate compensation for future Fontan procedure[4,5].

WHY PERIPHERAL SATURATIONS ARE 75-85 % AFTER BDG?

In BDG, the deoxygenated blood flow from SVC is drained directly to pulmonary arteries to lungs, whereby it is oxygenated and returned to heart, whereas blood flow from lower limb via inferior vena cava is drained directly to heart and mixing of deoxygenated from lower limb and oxygenated blood from the lungs takes place. Therefore blood being pumped to the whole body is not fully oxygenated and the usual oxygen saturation for a child with BDG is around 75-85 % only[6].

UNDERSTANDING SINGLE VENTRICLE PHYSIOLOGY

The term single ventricle is a misnomer because one of the ventricles is seldom completely absent but infect it is hypoplastic so the word single ventricle physiology is more appropriate. The dominant ventricle can be morphologically left, right or indeterminate. Balancing pulmonary/systemic blood flow (Qp/Qs) is the primary goal of palliation in single ventricle physiology in infancy which is achieved by aorto-pulmonary shunt in case of decreased Qp or pulmonary artery banding in case of increased Qp[6,7]. This is followed by Glenn and Fontan operations.

The goal of Fontan procedure is to convert parallel circulation into series and decrease the volume overload, on single ventricle, thus delaying its failure. Blood flow across the Fontan circuit depends on trans-pulmonary gradient and trans-pulmonary resistance. Trans-pulmonary gradient is difference between caval pressure and pulmonary artery pressure. Trans-pulmonary resistance is dependent on pulmonary vascular resistance, integrity of anastomosis, or any valves in the conduit[7]

FACTORS WHICH DETERMINES THE FLOW OF GLENN SHUNT

Arterial saturation in a patient with single ventricle physiology depends upon the relative volumes and saturation of systemic and pulmonary venous blood that have mixed and reached the aorta. This is summarized in Table 1 as:

Table 1: Arterial saturation in a patient with single ventricle physiology
Arterial saturation (SAO2) = [systemic venous saturation × total systemic blood flow] + [pulmonary venous saturation × total pulmonary blood flow]/[total systemic + total pulmonary venous blood flow]

1. Therefore a patient with higher QP/QS ratio: will result in higher arterial saturation as there is greater proportion of mixed blood getting oxygenated than that of deoxygenated blood in systemic circulation.
2. Systemic venous saturation: for a given QP/QS and pulmonary vascular saturation, a decrease in systemic venous saturation, will result in decrease in arterial saturation. Decrease in systemic saturation can occur as a result of decrease in systemic oxygen delivery or increase in systemic oxygen consumption. Systemic oxygen delivery is

dependent on arterial oxygen content and systemic blood flow. Arterial oxygen content is indirectly dependent on hemoglobin concentration.

Figure 1: Computed tomography angiogram showing patent Glenn Shunt.

3. Pulmonary venous saturation: for a given systemic venous saturation and QP/QS ratio, pulmonary venous saturation will result in a fall of arterial saturation. Pulmonary venous saturation should be close to 100% in the absence of large intra pulmonary shunt or V/Q mismatch or in the presence of pulmonary parenchymal disease.

PREOPERATIVE ASSESMENT OF PATIENTS WITH BDG

The anesthesiologist investigating the case should thoroughly ask the parents and close relatives regarding the pathological lesion of heart, when was the Glenn procedure done. Preoperative evaluation should pertain to the history related to recent respiratory tract infection, which could have a deleterious effect on pulmonary hypertension or cavo-pulmonary anastomosis. Decision to delay the surgery in the event of recent respiratory tract infection should be discussed with the surgeon so as to weigh risk benefit ratio. Previous surgery, any associated complications, whether the child was extubated uneventfully, prolonged ICU stay in the post-operative period, prolonged intubation should be enquired as they may suggest any associated further risk or subglottic stenosis respectively. The parents should be enquired about the weight gain of the child, attainment of milestones, intelligence and play full nature of the child. Assessment of patency of cavo-pulmonary shunt, ventricular function and pulmonary vascular resistance is important. Drug history related to the shunt and associated disease should be sought and enquired. These patients may be on diuretics/antihypertensive/aspirin/anti-arrhythmic/warfarin/Angiotensinogen inhibitors/clopidegrel etc. Cardiologist consultation should be sought regarding the stoppage of any major anticoagulants before surgery and also regarding the functional status of Glenn shunt. In current medical practice, all cardiac medications are usually continued till morning of surgery except major anticoagulants/antiplatelets, which should be discussed with the cardiologist and stopped accordingly. ACE inhibitors are stopped only in anticipation of major surgery and anticipated major fluid shifts/blood loss. Patients with high pulmonary flow may present with tachypnea, tachycardia, irritability, cardiomegaly and hepatomegaly. Associated non-cardiac anomalies including syndromic association/musculo-skeletal defects/neurological defects/genitourinary abnormalities should be ruled out and discussed with pediatrician. Nil per oral instructions are followed according to standard norms, but dehydration should be avoided in these patients, as their cavo-pulmonary blood flow is dependent on adequate preload. These patients should preferably have a wide bore cannula in place so that fluids should be on flow. Sympathetic stimulation should be avoided, and anxiety should be avoided by giving a sedative benzodiazepine, midazolam/alprazolam night before the surgery so as to avoid any increase in oxygen consumption or myocardial demand[8,10].

PREOPERATIVE INVESTIGATIONS

Complete blood count, coagulation profile, renal function tests should always be requested for. Anemia should be avoided for improved oxygen carrying capacity; similarly, polycythaemia should be avoided as it increases the blood viscosity leading to risk of thrombotic events. Serum electrolytes are required as they are put on diuretics. Coagulation abnormalities can be seen due to platelet dysfunction, hypofibrinogenemia and anticoagulant effects. Echocardiography and catheterization can provide information of ventricular function, magnitude of shunt and cardiac anatomy[8]. Echocardiography and Magnetic Resonance Imaging (MRI) can visualize ventricular size and coarctation of the aorta but may fail to visualize pulmonary artery distortion. Cardiac catheterization is required as suggested by Nakanishi et al., to measure pulmonary resistance. Abnormal aortopulmonary collateral vessels may develop after the stage II procedure and echocardiography and MRI may not visualize these vessels. Therefore it is mandatory to get cardiac catheterization done after BDG to know the patency and status of collaterals[9]. Chest X ray reveals the heart size, position, pulmonary vascular markings and possibility of any respiratory tract infection[8].

GOALS OF ANESTHESIA MANAGEMENT IN A CASE OF GLENN SHUNT

After understanding the physiology of BDG and single ventricle, we deduce the anesthetic concerns as summarized in Table 2[10].

Table 2. Goals of anesthesia management (Glenn shunt)
1. Maintenance of adequate preload: to allow uninterrupted flow in cavopulmonary anastomosis,
2. Maintaining low pulmonary vascular resistance and preserving pulmonary blood flow: so as to passively allow blood to flow via cavo-pulmonary anastomosis
3. Maintaining normal sinus rhythm,
4. Maintaining adequate ventricular contractility and filling,
5. Afterload reduction, so as to promote forward blood flow,
6. Preserving Qp/Qs ratio,
7. Avoidance of any sympathetic response,
8. Maintaining coronary blood flow-demand supply relationship.

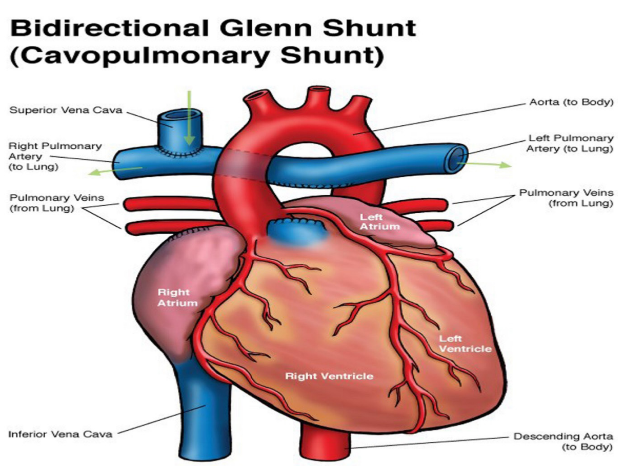

Figure 2. Diagrammatic representation of Glenn shunt.

PREOPERATIVE PREPARATION

All emergency cardiac drugs (adrenaline, calcium gluconate, nor epinephrine, dobutamine, amiadorone, adenosine, nitroglycerine, SNP, esmolol, metoprolol, inotropes and vasopressors) should be made available in Operation Theatre (OT) with defibrillator and anesthesia work station checked, with adequate availability of infusion pumps should be ensured

MONITORING

Monitoring in a case of Glenn shunt is summarized in Table 3[8,10].

Table 3: Monitoring
ROUTINE STANDARD MONITORING Electrocardiogram with rhythm analysis, non-invasive blood pressure, end tidal cardiography, pulse oximeter
SPECIFIC MONITORING Arterial beat to beat monitoring to ensure hemodynamic stability, temperature monitoring, urine output assessment, bispectral index monitoring and neuromuscular monitoring in specific cases is advisable
Central line if placed should be done in the femoral vein because of creation of Glenn shunt in Superior Vena Cava – Pulmonary Artery (SVC-PA).

ANESTHESIA ALLOCATION

Anesthesia can be provided via following subtypes based on the type of surgery, desired hemodynamic goals and associated co-morbidities. Anesthetic management skills are more important than technique of anesthesia which is chosen for:

1. Local anesthesia
2. Regional anesthesia/central neuraxial blockade
3. Combined spinal-epidural anesthesia
4. Combined regional-general anesthesia
5. General anesthesia

1. LOCAL ANESTHESIA

Local Anesthesia (LA) can be given with appropriate sedation, large dosage of LA should be avoided, as it can lead to cardiac dysrhythmias and myocardial depression. Patient should be monitored with ECG, NIBP, and pulse oximeter. Epinephrine with adrenaline should be avoided as it can cause tachycardia. Verbal reassurance and contact is important with the patient. Supplementary oxygen is desirable.

2. REGIONAL ANESTHESIA

Central neuraxial blockade can lead to loss of sympathetic efferent tone leading to rapid hemodynamic deterioration. Sometimes these patients are already on anticoagulants, thereby contraindicating the use of central neuraxial blockade. In a patient with failing heart, who is dependent on sympathetic tone, central neuraxial blockade can precipitate cardiac arrest.

3. COMBINED SPINAL EPIDURAL ANESTHESIA

Literature regarding the use of combined spinal epidural anesthesia in patients with BDG is lacking, as they are usually operated for a definitive Fontan procedure later in life. Monterio R et al. have used low dose combined spinal-epidural anesthesia and epidural analgesia as a mode of anesthesia and analgesia in pregnant females who had undergone Fontan procedure. Depending on the clinical scenario, 7% of all cases underwent caesarean delivery out of case series of 14 patients. A neuraxial catheter was preferred in 86% of all cases, and no delivery was conducted under general anesthesia. 50% of the patients were complicated by post-partum bleeding, and other post-operative complications were arrhythmias (29%), chest pain (14%) and intrauterine growth restriction (57%)[11].

4. COMBINED EPIDURAL AND GENERAL ANESTHESIA

Epidural anesthesia can be combined with general anesthesia for prolonged surgical procedure with anticipated blood loss, if coagulation profile is normal. Combination of thoracic epidural can be done with general anesthesia for upper abdominal, thoracic or vascular surgery. Epidural analgesia both in intraoperative and post-operative period helps suppress pain and improves the regional myocardial blood flow. Kohno et al., have reported a case report of a 32 year old female, who underwent abdominal surgery for incidental retroperitoneal tumor. She had undergone BDG shunt for complex cardiac but did not undergo definitive Fontan procedure owing to insufficient development of pulmonary circulation. Surgery was performed under combined general and epidural anesthesia, after induction she had surges of hypertension and tachycardia and on manipulation of tumor worsened and caused sympathetic surge. Landiolol and nicardipine were used to treat such surges. After the tumor was excised she was managed with low dose nor adrenaline owing to unresponsive hypotension. Later on tumor was found to be extra-adrenal pheochromocytoma[12].

5. GENERAL ANESTHESIA

It is the most common preferred anesthesia for patients with Glenn shunt for non-cardiac surgery and have been used safely by many authors[10–14]. The major advantages of general anesthesia are the avoidance of sympathectomy, hemodynamic stability, blunting the stress responses and airway management in hand in events of crisis situation. Patients with single ventricle pathway with Glenn physiology have already compromised oxygen saturations mostly in range of 75-85%[8–12]. These babies are highly sensitive to changes of systemic and pulmonary vascular resistance, and are at increased risk of arrhythmias. Anesthesiologist should have the availability of all anti-arrhythmic drugs available in OT along with emergency drugs ready, and must diagnose the rhythm first before administering any anti-arrhythmic. All factors leading to increase in pulmonary vascular resistance (hypoxia, hypercarbia, pain, sympathetic stimulation and acidosis) should be avoided. On the other hand increase in pulmonary blood flow will increase oxygenation but will also increase systemic blood pressure, leaving a challenge to the anesthesiologist to maintain a balance between SVR and PVR[1,10–12]. Invasive hemodynamic parameters, inotropic agents and vasopressors thus are an integral part of management of such patients. Adequate preloading with fluids should be done[10–12]. Balance between systemic vascular resistance and pulmonary vascular resistance should be maintained by using induction agents[8,10,11]. Intravenous short acting narcotics like fentanyl, sufentanyl and alfentanyl play an important role in effective control of surges of catecholamine's, provides excellent analgesia, hemodynamic stability and blunting of laryngeal reflexes[12]. As an induction agent, etomidate stands best in terms of maintaining cardiovascular stability and avoids untoward side effects of sudden tachycardia, hypertension as associated with ketamine, and profound hypotension as seen with propofol. Thiopentone is known to reduce myocardial contractility, preload and blood pressure should be administered with caution. Laryngoscopic response to intubation can be prevented with adequate depth of anesthesia, aliquots of fentanyl, short acting intravenous esmolol or iv lidocaine, but one must be cautious enough to avoid any hemodynamic Reponses. Succinyl choline is notorious to cause arrhythmias should be avoided. Pancuronium should also be avoided owing to its tachycardia response. Rocuronium can be considered during rapid sequence induction and vecuronium provides minimum hemodynamic alterations. Drugs causing histamine release like atracurium, mivacurium should be avoided as it may cause hypotension and tachycardia. Nitrous oxide should be avoided, as it may increase pulmonary vascular resistance thus impeding the flow through cavo-pulmonary anastomosis, cause mild depression of myocardial contractility and systemic vascular resistance and potential to cause air expansion[10,11].

Since these patients have intra-cardiac shunts, all care must be taken with utmost care to avoid paradoxical embolism. Maddali MM et al., have reported the embolism of entrained air in a child with single ventricle physiology who had earlier undergone BDG as detected intraoperatively by trans-esophageal echocardiography[10,14].

All efforts should be done to keep the patient warm, as hypothermia may cause shift of oxygen dissociative curve to the left and may increase demand on myocardium.In events of laparoscopic surgeries where intraabdominal pressure should be maintained below 10cmH20 so that cardiac output is not compromised[16,17]. Studies have suggested insufflation pressures less than 8-12 cm H20 did not decrease cardiac output as compared to 15-20 cm H20. Miura H et al., have conducted anesthesia management of a patient who underwent laparoscopic cholecystectomy after BDG for tricuspid atresia, and they had kept insufflation pressure below 8 cm H20 to avoid any hemodynamic changes[16]. Positioning for the surgery have also been found to alter hemodynamic changes: Eg in Trendelenburg position there is an increase in intrathoracic pressure from abdominal compartment leading to increase in pulmonary vascular resistance. Prone position decreases venous return from abdominal compression thereby affecting the forward flow. Reverse Trendelenburg also reduces preload. Lateral positioning leads to an improvement in V/Q mismatch leading to increase in PVR. Fluid administration should be balanced and optimally calculated to avoid any decrease in preload thereby affecting the forward flow and on the other hand overload should be avoided to cause pulmonary edema[10].

VENTILATORY STRATEGY IN PATIENTS WITH GLENN SHUNT

Positive pressure ventilation is known to cause increase in mean airway pressure which can adversely affect pulmonary vascular resistance and ultimately filling of ventricles to maintain cardiac output. Ventilatory strategy includes provision of adequate tidal volume and prolonged expiratory phase with low mean airway pressure simultaneously taking care not to cause hypercarbia. Early institution of spontaneous ventilation improves hemodynamics and also increases Pco2 levels, promoting increase in cerebral blood flow thereby promoting pulmonary blood flow in patients with BDG. Physiological PEEP (3-5 cmH20) is generally tolerated well without compromising Pulmonary Vascular Resistance (PVR) or cardiac output and helps improve oxygenation and ventilation-perfusion mismatch. The most practical approach at the end of procedure and in post-operative period is early extuabtion to allow patient to breathe spontaneously as suggested by Lofland et al., have shown a significant decrease in pulmonary artery pressure and improvement in cardiac output post extubation in comparison to pre-extubation without any change in complication or mortality. If early extubation is not achievable, goals should be to maintain adequate pulmonary blood flow and cardiac output by lowering PVR and intra thoracic pressure. Pain control should be optimum as well while taking care of ventilator strategy[18].

POST OPERATIVE CARE

Patients with BDG operated for non-cardiac surgery, even after extubation and favorable outcome are at risk of complications in post-operative period as well. Adequate spontaneous ventilation is preferable in Glenn physiology, thus safe extubation should be a norm. If for chance, patient is not extubated, all measures should be taken to avoid any increase in PVR. Adequate pain relief should be achieved in order to minimize sympathetic response and increase in PVR. They should be monitored in either High Dependency Unit (HDU) or Intensive Care Unit (ICU) for monitoring of vitals, maintenance of preload, balance between systemic and pulmonary vascular flow should be maintained, and analgesia should be ensured. Cardiac arrhythmias can still persist in the post-operative period and all measures should be done to maintain myocardial demand supply relationship[9,10,15,17].

CONCLUSION

Meticulous planning, formulating plan of management, clear understanding of physiology of Glenn shunt and its implication on non-cardiac surgery is helpful in anesthesia management of such patients in perioperative period.

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