INTRODUCTION

William Glenn performed first the cavopulmonary anastomosis in 1958 to divert systemic venous blood to the pulmonary circulation in a case of tricuspid atresia with pulmonary stenosis. It was an end-to-side anastomosis of the right pulmonary artery after complete transaction at its confluence to superior vena cava (SVC) with side clamping of SVC without using cardiopulmonary bypass (CPB). Since the late 1980s, when the bidirectional Glenn shunt (BDG) was first introduced, it had emerged as an intermediate modality in the repair of single ventricle and pulmonary stenosis complex. It involves an end to side anastomosis of SVC to the ipsilateral pulmonary artery where the continuity of branch pulmonary arteries is preserved. It is also performed as a part of one and a half ventricle repair in patients with hypoplastic right ventricle and for lesions like Ebstein's anomaly to reduce volume overload. Ultimate treatment is quite similar for most of these anomalies where biventricular repair is not possible and all generally undergo staged reconstructive procedures ultimately resulting in a “Fontan circulation”. Usually performed with cardiopulmonary bypass (CPB) it can also be performed without CPB in selected patients. The conduct of BDG without CPB can be associated with significant elevation of the proximal SVC pressure that may lead to neurological damage. Many authors have reported the safety of this procedure earlier, using various techniques to drain the SVC blood during clamping. Using a venoatrial shunt to decompress the SVC is safe, easy to perform and rational. At the same time, avoiding CPB gives protection from its inherent complications and reduces the cost of the procedure when no oxygenator, a heart-lung machine and a perfusionist is required.

To determine the efficacy of bidirectional Glenn shunt (BDG) without cardiopulmonary bypass (CPB).
decision to conduct the procedure without CPB was made after complete evaluation with echocardiography. Cardiac catheterization was done in only 6 patients where anatomical details had to be elaborated. All these patients had good sized branch pulmonary arteries (Macgoon's index > 1.4) and PA pressures < 16 mm Hg (measured on table with a needle attached to a transducer). Exclusion criteria were intracardiac pathologies requiring CPB for their repair and significant AV valve regurgitation.

Cardiopulmonary bypass (CPB) was defined as an artificial system used in open heart surgery to take over the function of heart and lung. Single ventrical anomalies were defined as a group of complex congenital cyanotic heart defects sharing the common feature of having only one ventricle of adequate functional size. Fontan circulation was defined as a diversion of systemic venous blood flow to the pulmonary arterial circulation without being propelled by the right ventricle. Macgoon's index was defined as the ratio of the sum of the sizes of branch pulmonary arteries to the size of aorta at diaphragm. Accessory source of pulmonary blood flow was defined as any systemic to pulmonary communication other than BDG like patent Ductus Arteriosus and Blalock Taussig's shunt. Antegrade pulmonary flow was defined as the direction of blood flow from the right ventricle to the main pulmonary artery.

Bidirectional Glenn shunt procedure was performed through median sternotomy. Superior vena cava pressure was monitored by a catheter placed in the internal jugular vein. Pressures (PA) were recorded by a needle attached to a transducer (mini-catheterization) before administration of Heparin. BDG procedure was performed only if the mean PA pressure was < 16 mm Hg. The SVC and pulmonary arteries were fully mobilized and the azygos vein was ligated and divided. In two cases of interrupted inferior vena cava (IVC), the azygos vein was kept intact because it represented the IVC in this morphology. By cannulating the SVC - innominate vein junction and the right atrium after systemic heparinization (300 IU/kg), a shunt was established between the SVC-innominate vein junction, to the right atrium with standard right-angled cannulae which were carefully de-aired and connected to each other (Figure 1). After establishing the temporary shunt, the SVC was clamped and divided just above the cardiac end avoiding the SA node. The cardiac end of the SVC was over-sewn. The right pulmonary artery was side-clamped at its superior aspect and opened here, and the distal end of the SVC was anastomosed to the pulmonary artery from end to side. In case of persistent left superior vena cava (LSVC), separate cannulation for LSVC was done and left anastomosis was completed first. Then LSVC cannula was used on the right side and that anastomosis completed. The clamp was removed as soon as the anastomosis was finished. The temporary shunt was disconnected in the middle, and blood in the cannulae was allowed to drain into the SVC and right atrium. Then the cannulae were removed and heparinization was neutralized if required. Extra sources of pulmonary blood supply (PDA and BT shunt) were disconnected at the end of the procedure.

Statistical analysis was performed using the SPSS 12.0 for windows. Categorical data are expressed as percentages and continuous data as mean ± standard deviation. Comparison with the means was done with the student t-test. Statistical significance was defined as p < 0.05.

**RESULTS**

There were 18 males and 13 females with the mean age of 5.01 ± 4.62 years ranging from 5 months to 18 years and mean weight of 13.12 ± 6.60 kg ranging from, 5 to 27 kg. All patients presented with dyspnoea on exertion and obvious cyanosis. Haemodynamic status of all the patients was a functional single ventricle with pulmonary arterial stenosis. Diagnoses were: tricuspid atresia (n=15), double inlet left ventricle (n=07), double outlet right ventricle (n=03), transposition of the great arteries (n=03), unbalanced AV canal defect (n=02) and heterotaxi syndrome (n=01). Mean pre-operative SpO2 on air was 68.52% ± 12.29% ranging from 32% to 80%. Gradients across the pulmonary valve measured on echocardiography ranged from 50 to 120 mm. Mean pulmonary artery pressures (PAP) were 12.62 ± 2.30 mm Hg (ranging from 8 to 16 mm Hg) as measured on the operation table. Three (9.67%) patients had a functioning right modified Blalock Taussig (BT) shunt and 17 (54.83%) had patent Ductus Arteriosus (PDA) as accessory source of pulmonary blood flow. Two patients (6.45%) had previously done PA band at less than 3 months of age for unrestrictive pulmonary blood flow. Two patients presented with interrupted IVC and required BDG as a complete procedure (Kawashima).
All patients survived. Twenty seven (87.09%) patients received BDG and 04 patients (12.90%) received bilateral BDG due to persistent left SVC. Atrial septectomy with inflow occlusion was performed in 05 (16.12%) patients. Mean pressures during SVC clamping were 23.90 ± 5.95 mm Hg (ranging from 14 to 36 mm Hg). The time taken for cavopulmonary anastomosis (SVC clamping) ranged from 18 to 45 minutes (mean = 26.81 ± 6.05 minutes). Antegrade pulmonary blood flow was left in 24 (77.41%) patients. There was significant improvement in SpO2 post-operatively (mean = 90.48 ± 5.05 mm Hg) as shown in Table I. Other postoperative parameters are given in Table II. All the patients were in sinus rhythm except one who remained in nodal rhythm for initial few hours. There were no postoperative neurological complications as per the clinical examination. Sepsis occurred in 2 patients who ultimately recovered. One patient had chylothorax which stopped after 3 days in ICU. Copious amount of pleural drainage (1100 ml) was actually the drainage of chyle in this case. Discharge echocardiography showed functioning Glenn shunts without any obstruction at the anastomosis. Patients were in stable physical and mental state and their exercise tolerance had markedly improved.

### Table I: Haemodynamic status and oxygen saturation (SpO2) during the operation.

<table>
<thead>
<tr>
<th>Time</th>
<th>HR (beats/min)</th>
<th>BP (mm Hg)</th>
<th>SVCP (mm Hg)</th>
<th>SpO2 (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Before SVC clamping</td>
<td>106.67±13.55</td>
<td>81.87±11.43</td>
<td>6.58±2.10</td>
<td>71.87±14.87</td>
</tr>
<tr>
<td>SVC clamping</td>
<td>124.70±12.22</td>
<td>70.96±11.95</td>
<td>23.90±5.95</td>
<td>64.19±8.34</td>
</tr>
<tr>
<td>After SVC clamping</td>
<td>101.12±8.41</td>
<td>90.48±11.35</td>
<td>13.35±2.0</td>
<td>90.48±5.05</td>
</tr>
</tbody>
</table>

HR=heart rate; BP=systolic blood pressure; SVCP=superior vena cava pressure.

### Table II: Postoperative parameters.

<table>
<thead>
<tr>
<th>Ventilation hours</th>
<th>Median 03 hours (range, 1 to 8)</th>
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<tbody>
<tr>
<td>Duration of inotropes</td>
<td>Median 24 hours (range, 0 to 90)</td>
</tr>
<tr>
<td>ICU stay</td>
<td>Median 40 hours (8 to 140)</td>
</tr>
<tr>
<td>Complications:</td>
<td></td>
</tr>
<tr>
<td>Chylothorax</td>
<td>01</td>
</tr>
<tr>
<td>Sepsis</td>
<td>02</td>
</tr>
<tr>
<td>Neurological</td>
<td>Nil</td>
</tr>
<tr>
<td>Mortality</td>
<td>Nil</td>
</tr>
</tbody>
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### DISCUSSION

Bidirectional Glenn shunt without using CPB has been performed safely all over the world. The classical Glenn shunt used to be performed through a thoracotomy with side clamping of SVC without CPB. In 1990, Lamberti and associates first reported a technique for BDG without CPB by establishing a temporary veno-atrial shunt between the SVC and right atrium. Later on, Murthy and colleagues described the technique of veno-pulmonary shunt in 1999. Actually, the risk of decreased cerebral perfusion with the clamping of SVC has been a concern behind all these shunts techniques. Jahangiri and associates in 1999 and then Hussain and associates in 2007 proved that no temporary shunt is essentially required for this purpose. Their SVC pressure during clamping ranged from 19 to 65 mm Hg with median of 26 mm Hg but they did not report any neurological complications in their series. Liu and colleagues suggested that BDG without CPB is reasonably safe if SVC pressure after clamping remains at less than 30 mm Hg and clamping time is less than 30 minutes. In the present study, mean SVC clamping time and SVC pressure after clamping had remained at less than 30 minutes and 30 mm Hg respectively without any neurological complication.

Both veno-atrial and veno-pulmonary temporary shunts have been used with equal beneficial results. Veno-pulmonary shunt functions as a modified Glenn’s shunt while SVC is clamped for anastomosis and SpO2 is actually increased during this period. Veno-pulmonary shunt is obviously difficult to use in patients with main pulmonary artery atresia or hypoplasia. Secondly, veno-pulmonary shunt can cause distortion of the pulmonary artery rendering it unsuitable for future Fontan repair. In this study a veno-atrial shunt was used on all the patients and it was found that the technique was easy to perform and allowed a good operative field exposure, although there was a mild SpO2 decrease while the SVC was clamped (Table I).

CPB apparatus was always kept ready to function in case of emergency. Twice the procedure had to be converted on pump because of dysrrhythmias and haemodynamic instability just after clamping SVC - RA junction. In both of those patients no particular reason for haemodynamic instability could be found. Off pump BDG should be done with caution when there is a history of dysrrhythmias, or in patients having hypoplastic pulmonary arteries, severe volume overload and even mild AV valve regurgitation. Similarly, terminating extra sources of pulmonary blood supply (PDA and BT shunt) prior to the construction of BDG may lead to severe hypoxaemia which we encountered in one case after closure of PDA. PDA and BT shunt should be closed at the end of the procedure.

Antegrade pulmonary blood flow was left intact in 24 patients rendering the flow pulsatile. Different authors have reported the usefulness of pulsatile BDG. Preserving additional pulsatile pulmonary blood flow provides additional oxygen for the growth of branch pulmonary arteries. At the same time, there is a disadvantage in the terms of relatively higher Glenn pressures. The MPA was ligated routinely in patients with borderline pulmonary artery pressures (mean > 15 mm).

Surgical atrial septectomy under normothermic caval inflow occlusion is very helpful for patients who have a restrictive atrial septum and require an atrial septectomy. The advantage of avoiding CPB in this setting is in large part related to the effects of CPB on
pulmonary resistance. The procedure requires surgical experience and involvement of a good anaesthetist. Otherwise, a brief period of CPB for performing the procedure is not an unreasonable alternative.

In general terms, the postoperative management of these patients after off pump BDG had been similar to that after BDG with CPB. That is, all treatment should aim at decreasing the pulmonary vascular resistance and accelerating the SVC return. In our experience, these patients generally do well in terms of ventilation time, requirement of inotropes and perioperative mortality and morbidity (Table II). The estimated procedural cost of CPB for a single case was roughly Pak Rs. 50,000. The cost may further increase if CPB related complications occur leading to prolonged hospital stay and consumption of more hospital resources. We haven’t actually compared patients with off pump BDG with those done on CPB in this study, in terms of morbidity and mortality, as this issue needs a separate discussion. By and large, less was spent on patients with off pump BDG by saving the cost of CPB and avoiding complications related to it.

CONCLUSION

Off pump BDG is an economical and safe procedure and avoids complications related to CPB and blood transfusion. If patients are carefully selected and a valid self-bypass shunt is established, off-pump BDG is easy to perform and the risk of possible neurological complications is also avoided.

REFERENCES


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