We report a rare complication of massive aneurysm of the proximal ligated end of the main pulmonary artery which occurred in the setting of a patient with a functionally univentricular heart and increased pulmonary blood flow undergoing superior cavopulmonary connection. Awareness of this possibility may guide others to electively transect the pulmonary artery in such a clinical setting.

Key words: Antegrade pulmonary blood flow; Bidirectional Glenn; Pulmonary artery

INTRODUCTION

Ideally, patients undergoing superior cavopulmonary connection for a functionally univentricular heart should have an antegrade pulmonary blood flow to maintain pulsatility and circulation of hepatic factor into the pulmonary arterial system to prevent pulmonary ateriovenous fistula formation. However, a subset of patients with increased pulmonary blood flow and raised pulmonary artery pressures in the preoperative period require the interruption of the antegrade pulmonary blood flow to prevent the development of superior vena cava (SVC) syndrome. The interruption of the pulmonary blood flow through the main pulmonary artery (MPA) may be accomplished either by MPA ligation or division. Till date, published literature does not address the superiority of one over the other. We report a postoperative case of superior cavopulmonary connection with complete interruption of the antegrade pulmonary blood flow by ligation who subsequently had developed a massive aneurysm of the right ventricular outflow tract (RVOT). The extreme rarity of this complication, the use of angiocardiography and contrast echocardiography for diagnosis, and a brief review of the medical literature forms the basis of this communication.

CASE REPORT

A 4-year-old boy, weighing 15 kg underwent superior cavopulmonary connection with concomitant ligation of the MPA for a functionally univentricular heart with increased pulmonary
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Blood flow and raised pulmonary artery pressure under normothermic cardiopulmonary bypass without cardioplegic arrest. Echocardiographically, the atrial septal defect was large and nonrestrictive. The child had been followed up regularly at our cardiac outpatient department. Despite optimal medical management, by 8-year of age, the child had recurrent hemoptysis and persistent precordial pain. Repeat echocardiogram and cardiac catheterization were performed for detailed hemodynamic evaluation. Echocardiography revealed mild systemic ventricular dysfunction and a properly functioning superior cavopulmonary connection. There was no evidence of antegrade pulmonary blood flow. However, a large echo-free space in the area of RVOT was noted. Computerized tomographic angiography revealed a large aneurysm of the MPA and RVOT measuring about 5–6 cm in diameter compressing the left main bronchus [Figure 1a and b]. The mean pulmonary venous wedge pressure was 17 mmHg. The child was taken up for MPA aneurysmectomy with concomitant completion Fontan procedure.

Surgical technique

Given the complexity of the situation, the resternotomy was performed under femoral arteriovenous bypass. The aneurysmal MPA was located proximal to the ligature, measuring about 8 cm × 5 cm in size involving the RVOT and was densely adherent to the overlying sternum. Identification and mobilization of the aneurysmal wall was extremely difficult and the aneurysm had to be laid open under profound hypothermia for intra-aneurysmal obliteration of the RVOT [Figure 2a-d]. The aneurysm was resected. The RVOT opening was closed using a polytetrafluoroethylene patch (WL Gore and Associates, Arizona, USA). A lateral tunnel, total cavopulmonary connection was performed using a fenestrated (4 mm) polytetrafluoroethylene patch. The patient had a stormy postoperative course with persistent Fontan failure with the features of low cardiac output unresponsive to inotropes and pulmonary vasodilator therapy. The child died on the 18th postoperative day due to septicemia. Permission for autopsy was denied.

DISCUSSION

Children born within the anatomic matrix of a univentricular heart mandate a wellprotected pulmonary vascular bed to optimize their suitability for definitive univentricular repair.[1-5] In contrast to the western world, many patients in the developing countries present for medical help in late childhood.[6]

Several investigators have demonstrated the difficulties inherent in using a grading system of the pulmonary vascular changes to evaluate the severity of the disease and predict postoperative outcome of patients with a functionally univentricular heart with increased pulmonary blood flow undergoing univentricular type of repairs.[7-10] As yet, the safe upper limit of pulmonary artery pressure and pulmonary vascular resistance in the setting of a net left-to-right shunt in patients with a functionally univentricular heart with increased pulmonary blood flow undergoing superior cavopulmonary connection remains unknown. Pulmonary artery hypertension has long been found to be a risk factor for patients undergoing univentricular-type of repairs.[5-10] This includes both the Fontan procedure and superior cavopulmonary connections.

In our previous investigation, we had subjected 82 patients with a functionally univentricular heart and

**Figures 1:** (a and b) Images from axial computerized tomographic angiography and reconstructed maximal intensity projection showing the aneurysmal main pulmonary artery. The right pulmonary artery and left pulmonary artery are getting filled from the bidirectional Glenn circuit

**Figure 2:** (a-d) Operative views of the techniques used to resect the aneurysmal main pulmonary artery and closure of the opening of right ventricular outflow tract using a polytetrafluoroethylene patch
postpulmonary artery band to second stage superior cavopulmonary connection. This subset of patients was considered less than ideal candidates for a complete cavopulmonary connection. These patients underwent bidirectional Glenn under at least one of the following conditions: (i) The mean pulmonary artery pressure was >20 mmHg with a net left-to-right shunt, and (ii) pulmonary vascular resistance >3.0 (and <4.5) Wood units/m² but reactive to vasodilators including nitric oxide. The primary aim for conversion to bidirectional Glenn was to reduce ventricular pressure and volume load, and possibly to improve candidacy for future completion Fontan.[6]

It is noteworthy that a substantial number of this subset of patients required complete interruption of antegrade pulmonary blood flow to reduce the Glenn pressure to <20 mmHg to prevent SVC syndrome.[6] In order to avoid the transection of the pulmonary artery and subjecting the transected proximal pulmonary arterial end to systemic ventricular pressure, we have been ligating the pulmonary artery to achieve our objective.

The current case serves to emphasize that the interruption of the antegrade pulmonary blood flow in the setting of superior cavopulmonary connection should always be done by pulmonary artery transaction and not pulmonary artery ligation. Interruption of the pulmonary artery would avoid subjecting the proximal ligated pulmonary artery stump to increased systemic ventricular pressure, thereby avoiding/eliminating the development of this complication. In order to avoid bleeding complication, we would recommend re-enforcement of the transected proximal pulmonary arterial stump using pledgeted sutures.

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Conflicts of interest
There are no conflicts of interest.

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