Case Report

Recived on 27.09.2016, **Accepted on** 12.10.2016

Journal of Cardiovascular Medicine and Surgery
Volume 2 Number 2, July - December 2016
DOI: http://dx.doi.org/10.21088/jcms.2454.7123.2216.3

Recurrent Thrombosis in Bidirectional Glenn Shunt

Makhija Neeti*, Dhawan Ira, Kiran Usha*****

Authors Affiliation
* Professor, ** Senior Resident,
***Professor & Head, Department of Cardiac Anaesthesia, Cardiothoracic Centre, CNC, All India Institute of Medical Sciences, Ansari Nagar, New Delhi-110029, India.

Reprints Requests
Neeti Makhija, Professor, Department of Cardiac Anaesthesia, Room No. 9, 7th Floor, Cardiothoracic Centre, All India Institute of Medical Sciences, Ansari Nagar, New Delhi—110029, INDIA.
E-mail: neetimakhija@hotmail.com

Abstract

Thrombus formation post Blalock-Taussig shunt and Fontan surgery is well known. Thrombus formation following Glenn surgery has only limited case reports. Early detection and treatment of thrombosis is important as it may lead to neurological complications, may render patient unfit for completion Fontan surgery and can even lead to mortality. We report a rare case of post Glenn procedure in a patient with double outlet right ventricle with ventricular septal defect, who developed recurrent thrombi in both right and left pulmonary arteries.

Keywords: Bidirectional Glenn; Thrombosis.

Introduction

Glenn procedure is an end to side cavo-pulmonary connection between superior vena cava and the right pulmonary artery. It is a first stage procedure prior to completion Fontan. With the witnessed success of Glenn procedure and its completion to Fontan surgery, Glenn procedure has now become a very commonly performed surgery in patients with univentricular physiology to improve oxygenation without increasing ventricular load.

Performing Glenn procedure during infancy remains a controversial issue but Warrier et al [1] in his study concluded that Bidirectional (BD) Glenn shunt can be performed in infants more than 2 months of age safely. Reddy et al [2] also demonstrated the feasibility of the cavopulmonary shunt operation in 1-4 months old as a primary surgery, except in those with very small pulmonary arteries. BD Glenn helps to avoid an additional step in the form of systemic arterio-pulmonary shunt, thus avoiding associated pulmonary artery distortion and the ventricular volume overload.

BD Glenn is associated with complications like pleural effusion, pulmonary arteriovenous fistula and thrombosis. We present a rare case of recurrent thrombi formation in both right and left pulmonary arteries and its successful management by surgery and in cardiac catheterization laboratory.

Case Report

An 8 months old female child presented in emergency of a tertiary care hospital with cyanosis, apnea, desaturation followed by bradycardia. The child was intubated, resuscitated, and mechanically ventilated for 7 days. There was previous history of cyanosis since 1 month of age and one episode of cyanotic spell requiring hospitalization one month prior to this episode. On echocardiography, child was diagnosed with situs solitus, levocardia, double outlet right ventricle (DORV), peri-membranous ventricular septal defect (VSD), 3.5 mm ostium secundum atrial septal defect (OS-ASD), malposed great arteries, pulmonary atresia, adequate sized pulmonary arteries, patent ductus arteriosus (PDA) associated with tight stenosis at its distal end, with major aorto-pulmonary collateral arteries (MAPCAs), along with right pulmonary artery (RPA) thrombosis.

Patient was taken up in cardiac catheterization laboratory for PDA stenting as a palliative procedure to improve saturation, which was attempted but deferred due to stenosis at its distal end. Patient was put on unfractionated heparin (UFH) infusion and

was operated upon for BD Glenn with RPA thrombectomy and left pulmonary artery (LPA) dilation with hegar of full size. UFH and warfarin were continued postoperatively. Patient was extubated post surgery, but had to be reintubated as he developed respiratory distress. Patient had persistent desaturation upto 68%, therefore MAPCA coiling was done. Post coiling saturation improved. On ventilatory support patient's saturation was 77% but failed 3 trials of extubation.

Due to inability to wean from ventilator, patient was again taken into catheterization laboratory. Angiography showed thrombus in LPA with no flow to left lung (Figure 1). Right internal jugular vein (IJV) was then cannulated using 5Fr sheath and fielder wire was negotiated into the LPA (Figure 2), followed by balloon dilatation (Figure 3). Good flow was established. A central venous catheter was then placed into the LPA over a guide wire and infusion Streptokinase (STK) started at the rate of 1000U/kg/hr for 24hrs after giving a bolus of 2000U/kg. This was followed by heparin infusion. Patient got extubated a day later with saturation of 80%.

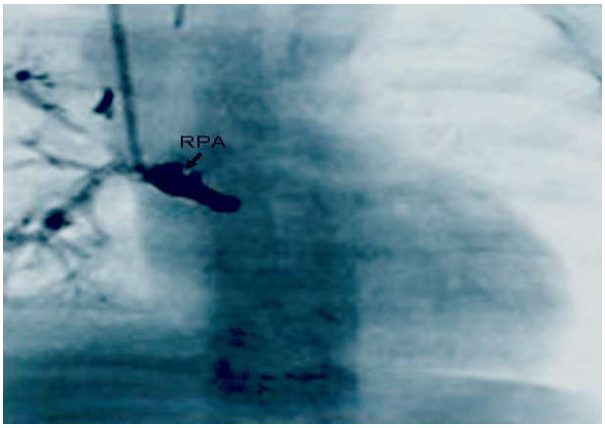

Fig. 1: Angiography showed flow in RPA with thrombus in left pulmonary artery and no flow to left lung. RPA: Right pulmonary artery

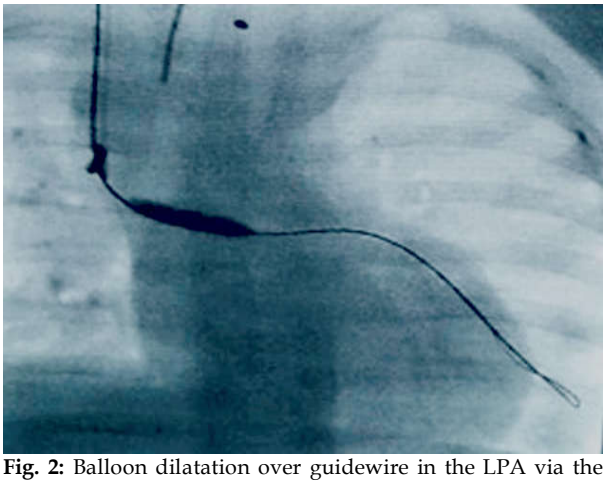

Fig. 2: Balloon dilatation over guidewire in the LPA via the cannulation of right internal jugular vein

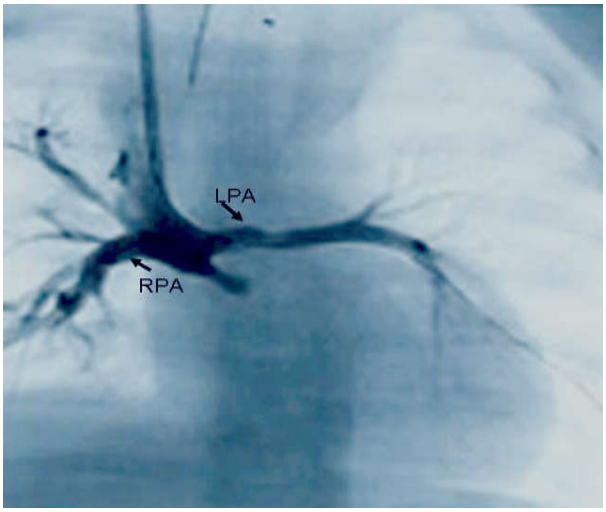

Fig. 3: LPA with good flow established. LPA: Left pulmonary artery, RPA: Right pulmonary artery

The patient remained stable for few days but developed desaturation which was again due to thrombus formation in both LPA and RPA. The patient underwent both LPA and RPA thrombectomy and plasty on cardiopulmonary bypass with deep hypothermic circulatory arrest and shifted to intensive care unit. The child was extubated on first post operative day with gradual tapering of inotropes and discharged after a week.

Discussion

Thrombosis following Glenn procedure is relatively rare. There are several reports and studies on thrombosis in Fontan surgery, but only few case reports [3-6] of thrombus formation following Glenn procedure are described in the literature. Thrombosis is more commonly encountered in females and with increasing age. It is commonly associated in those who have elevated right atrial and ventricular end-diastolic pressure before surgery, or an elevated superior vena cava pressure and poor ventricular function after surgery [3]. Decreased ventricular function, low antithrombin –III levels and increased tissue plasminogen activator are also associated with higher incidence of thrombus formation following palliative surgery for single ventricle physiology.[7] Our patient presented with thrombosis and was immediately started on heparin and warfarin.

It is crucial to address thrombus formation in a patient with Glenn since unattended thrombus in Glenn circuit may render the patient unfit candidate for future Fontan. If pulmonary thrombus leads to increase in the pulmonary vascular resistance, it becomes a contraindication for Fontan surgery

leaving the patient with no option other than heart transplant [8]. Hence, this problem needs to be paid heed upon and managed.

The options available for prevention of thrombosis are heparin followed by aspirin, or heparin followed by warfarin therapy. Few do not believe in prescribing any anticoagulation regime [8].

In our patient in spite of heparin and warfarin continuation post surgery, patient developed thrombus in pulmonary arteries. As an institutional protocol a 22G cannula is inserted in IJV that measures the Glenn shunt pressure during surgical procedure which is removed postoperatively as soon as the patient is hemodynamically stable. This is done in order to prevent thrombus formation.

Conclusion

Thrombosis in cyanotic congenital heart disease is common but recurrence in spite of anticoagulation is rare and can present with multiple challenges. The choice between performing a Blalock–Taussig (BT) shunt versus Glenn procedure, both of which are at risk of developing thrombus can be tricky. Retrospective and prospective data on thrombosis secondary to BT shunt and Fontan surgery are available. The need of the hour is to collect data on incidence, causes, methods of prevention and treatment of thrombosis secondary to bidirectional Glenn shunt.

References

1. Warrier G, Dharan BS, Koshy S, Kumar S, Krishnanaik S, Rao SG. Bidirectional glenn operation in infancy. Ind J Thorac Cardiovasc Surg, 2004; 20:159–163.
2. V. Mohan Reddy, John R. Liddicoat, Frank L. Hanley. Primary bidirectional superior cavopulmonary shunt in infants between 1 and 4 months of age. Ann Thorac Surg 1995; 59:1120–25.
3. Kozicka U A, Kowalski M, Hoffman P. Thrombosis within Glenn anastomosis. Journal of Rare Cardiovascular Diseases 2015; 2(3):89–91.
4. Pennington D, Nouri S, Ho J, et al. Glenn Shunt: long-term results and current role in congenital heart operations. Ann ThoracSurg 1981; 31:532–9.
5. Kopf G, Laks H, Stansel H, et al. Thirty-year follow-up of superior vena cava-pulmonary artery (Glenn) shunts. J ThoracCardiovascSurg 1990; 100:662–70.
6. Koutlas T, Harrison J, Bashore T, et al. Late conduit occlusion after modified Fontan procedure with classic Glenn Shunt. Ann Thorac Surg 1996; 62: 258–61.
7. Tzanetos DRT, Yu C, Hernanz- Schulman M, Barr FE, Brown NJ. Prospective study of the incidence and predictors of thrombus in children undergoing palliative surgery for single ventricle physiology. Intensive Care Med.2012; 38:105-112.
8. P. Monagle . Thrombosis in children with BT shunts, Glenns and Fontans. Progress in Pediatric Cardiology. 2005; 21:17–21.