



Autopsy and Case Reports

ISSN: 2236-1960

São Paulo, SP: Universidade de São Paulo, Hospital
Universitário

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Autopsy and Case Reports, vol. 9, no. 3, e2019118, 2019, July-September
São Paulo, SP: Universidade de São Paulo, Hospital Universitário

DOI: 10.4322/acr.2019.118

Available in: <http://www.redalyc.org/articulo.oa?id=576062230009>

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Superior vena cava obstruction after the Glenn procedure

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How to cite: Jonas MC, Ramalho FS, Vicente WVA, Manso PH. Superior vena cava obstruction after the Glenn procedure. Autops Case Rep [Internet]. 2019 Jul-Sep;9(3):e2019118. <https://doi.org/10.4322/acr.2019.118>

ABSTRACT

Acute obstruction of superior vena cava anastomosis right after the Glenn procedure may lead to tragic consequences. We describe the case of a one-year-old child with tricuspid atresia and a previous Blalock-Taussig shunt procedure, who presented severe low cardiac output syndrome right after the Glenn procedure and died forty-four hours after the procedure. The autopsy showed obstruction of the superior vena cava anastomosis. Patients that present superior vena cava syndrome and low cardiac output right after the Glenn procedure should have the surgical anastomosis revised immediately.

Keywords

Heart Disease, Congenital; Heart Bypass, Right, Tricuspid Atresia

INTRODUCTION

The Glenn procedure (superior vena cava to right pulmonary artery anastomosis) is one of the stages of palliative care indicated for children diagnosed with univentricular heart. The postoperative mortality depends on preoperative and postoperative factors,¹ such as thrombosis, arrhythmias, and superior vena cava syndrome.²

The superior vena cava syndrome in patients submitted to Glenn surgery stems from inadequate flow from the superior vena cava to the pulmonary artery due to factors like pulmonary hypertension, vessel lumen narrowing at the surgical anastomosis, and pulmonary artery or vein stenosis. As a result, the patient can present facial and upper limb edema, which can lead to cerebral edema, low cardiac output syndrome, and death.

Herein, we report on a child with the diagnosis of tricuspid atresia who was submitted to the Glenn procedure and evolved to death because of severe superior vena cava syndrome right after the procedure.

CASE REPORT

A one-month-old girl with postnatal diagnosis of tricuspid atresia, concordant ventricular-arterial connection, and patent ductus arteriosus, without pulmonary outflow tract obstruction, was submitted to a modified Blalock-Taussig shunt, ductus arteriosus ligation, and pulmonary artery amputation, with excellent postoperative recovery. The patient had unremarkable evolution, and the Glenn procedure was proposed when she was one year old.

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Surgical Findings

Huge technical difficulties emerged due to various thoracic adhesions, thin and friable right pulmonary artery at the topography of the Goretex tube old insertion, and large thymus with hardened consistency of the superior lobe adhered to the superior vena cava. There was severe bleeding during the procedure, which lasted 10 hours and required cardiopulmonary bypass for 245 minutes.

The patient arrived in the intensive care unit with anuria, hypotension, and acidosis. Low cardiac output syndrome was initially managed with the use of inotropes and volume expansion. In the immediate postoperative period, a transthoracic Doppler echocardiogram was performed after the initial management and showed normal ejection fraction and normal atrioventricular (AV) valve function, with no insufficiency. However, edema, open chest, and surgical bandages prevented proper evaluation of the Glenn anastomosis. A cardiac catheterism was requested, but the patient could not be transported to the cath lab because she was seriously ill and presented refractory cardiogenic shock, superior vena cava syndrome, and several cardiac arrest episodes. Twenty-eight hours after the Glenn procedure, she presented fixed mydriasis and signs of brain death. Death was confirmed forty-four hours after the Glenn procedure.

Autopsy Data

The anatomic diagnosis of tricuspid atresia was confirmed; a large ventricular septal defect, thickened left ventricle wall, and hypoplastic right ventricle were also evident. A metal probe showed that the anastomosis between the superior vena cava and the right pulmonary artery was obstructed (Figure 1).

DISCUSSION

Bidirectional cavopulmonary anastomosis (the Glenn procedure) as the first stage of palliative care for univentricular patients reduces the morbidity and the mortality of children to whom the Fontan procedure is indicated. Mortality regarding cavopulmonary anastomosis in diverse patient cohorts varies from 1 to 13%,³ and factors such as age, cardiopulmonary bypass time, and need for re-intubation have been identified as risk factors for mortality and morbidity.⁴

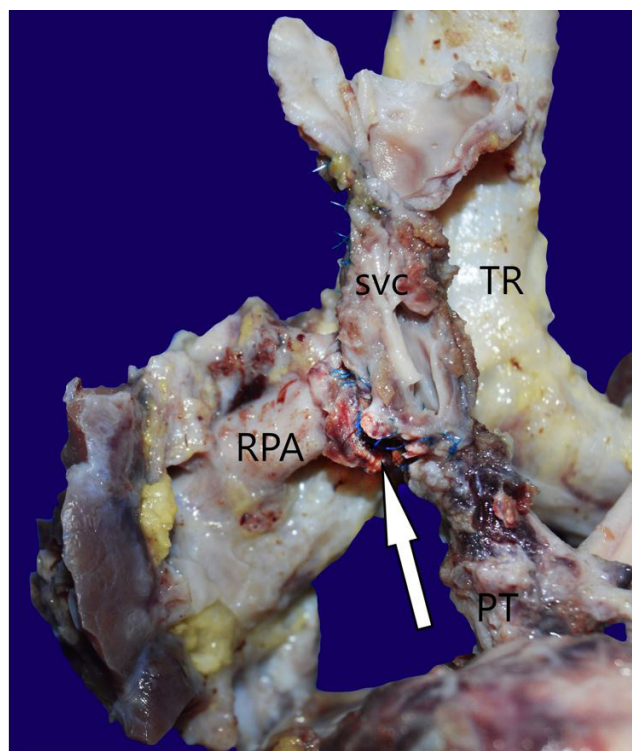


Figure 1. Necropsy specimen (anterior view) showing the anastomosis (arrow) between the superior vena cava (SVC) and the right pulmonary artery (RPA); PT- pulmonary trunk; TR - trachea.

High pulmonary artery resistance, ventricular dysfunction, AV valve regurgitation, and obstruction of the anastomosis must be ruled out in patients with hypoxemia or low cardiac output syndrome.

In the case reported herein, technical limitations during the surgery increased the cardiopulmonary bypass time. Therefore, a functional anastomosis was hard to achieve. The Glenn procedure functional failure culminated in superior vena cava syndrome, low cardiac output, and death.

Although ventricular dysfunction and AV valve regurgitation were excluded, the patient presented no clinical conditions for a cardiac catheterism. Our patient presented a total obstruction of the anastomosis, rapidly became hemodynamically unstable, developed superior vena cava syndrome, and died.

In conclusion, if a patient has unsatisfactory evolution after the Glenn procedure, the surgical anastomosis must be investigated.

ACKNOWLEDGEMENTS

The authors thank to Cynthia Manso for her kind revision of the English language.

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Author contributions: PHM and MCJ conceived and designed the study. PHM, MCJ, FSR, WVAV were involved in the data collection. PHM and MCJ drafted the manuscript, with significant contributions from FSR and WVAV. All authors read and approved the final manuscript.

A family member (next of kin) signed the consent declaration for the clinical autopsy. The authors hereby state that the manuscript is in accordance with the institution's ethics committee policy.

Conflict of interest: None

Financial support: None

Submitted on: June 26th, 2019

Accepted on: August 2nd, 2019

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