Aneurysm of the superior vena cava in a bidirectional Glenn procedure with antegrade pulsatile flow: a rare complication.

Iris Flores-Sarria[1], Diego Ortega-Zhindón[2], and Jorge Cervantes-Salazar[2]

[1]Instituto Nacional de Cardiología Ignacio Chávez
[2]Instituto Nacional de Cardiologia Ignacio Chavez

March 07, 2024

Abstract

We present a 22-year-old male patient, a history of a right bidirectional Glenn procedure (BDG), with dyspnea, cyanosis, palpitations and chest pain. Transthoracic echocardiography and computed tomography showed aneurysmal dilation of the right superior venous system. BDG takedown was performed with resection of the aneurysmal superior vena cava and reconnection of the superior vena cava to the right atrium and the creation of a central systemic-to-pulmonary shunt. The patient died from complications derived from a massive collateral circulation.

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Iris P. Flores-Sarria, MD[1], Diego B. Ortega-Zhindón, MD[1], Jorge L. Cervantes-Salazar, MD[1].

[1] Department of Pediatric Cardiac Surgery and Congenital Heart Disease, National Institute of Cardiology Ignacio Chávez. Mexico City, México.

Corresponding author: Jorge L. Cervantes-Salazar. Juan Badiano No 1. Colonia Sección XVI, Delegación Tlalpan, Mexico City, México. C.P. 14080. Phone: 5555732911. E-mail:*jorgeluis.cervantes@gmail.com*

Conflict of interest: None.

Funding: None.

Data availability statement: The data that support the findings of this study are available from the corresponding author, [JLCS], upon reasonable request.

Authors' contributions:

Concept/design: Flores-Sarria, MD; Ortega-Zhindón, MD; Cervantes-Salazar, MD.

Drafting article: Flores-Sarria, MD; Ortega-Zhindón, MD.

Critical revision/approval: Flores-Sarria, MD; Ortega-Zhindón, MD; Cervantes-Salazar, MD.

Abstract:

We present a 22-year-old male patient, a history of a right bidirectional Glenn procedure (BDG), with dyspnea, cyanosis, palpitations and chest pain. Transthoracic echocardiography and computed tomography showed aneurysmal dilation of the right superior venous system. BDG takedown was performed with resection of the aneurysmal superior vena cava and reconnection of the superior vena cava to the right atrium and the creation of a central systemic-to-pulmonary shunt. The patient died from complications derived from a massive collateral circulation.

Keywords:

Glenn procedure; blood flow; congenital malformation.

Introduction

The BDG or superior cavopulmonary connection (SCPC) involves the connection of the superior vena cava to the pulmonary artery as a palliative procedure in many congenital heart diseases; where generally the anastomosis is with the right pulmonary artery, with a free flow of venous blood through right and left pulmonary artery. There are complications after this surgery such as: cyanosis, arrhythmias, protein-losing enteropathy, pulmonary arteriovenous malformations, liver dysfunction, hepatocellular carcinoma, etc[1]. We present one case that were approved by the local institutional review board. The approval included a waiver of informed consent because it does not show personal data of patients.

Description of case

We present a 22-year-old male patient, with a history of double inlet left ventricle and transposition of the great arteries, with palliation at 2 years with pulmonary artery banding and at age of 17 years underwent right BDG. He was admitted due to palpitations and oppressive chest pain of moderate intensity, as well as dyspnea, cyanosis and headache. On physical examination with 70% pulse oximetry, jugular vein distention, and systolic murmur in the pulmonary focus and in the aortic focus with radiation to the neck. The transthoracic echocardiogram confirmed the previous diagnosis and reported ventricular septal defect, mild aortic regurgitation and mild right atrioventricular valve regurgitation, patent cavopulmonary shunt with a dilated pulsatile flow extending to the right jugular vein, pulmonary artery banding with a maximum gradient of 75 mmHg, mean gradient of 52 mmHg and pulsatile flow. It was complemented with computed tomography showing aneurysmal dilation of the right superior venous system (**Figure 1A and 1B**) that involved the right jugular vein, innominate vein, superior vena cava, azygos and hemiazygos systems, with increased thoracoabdominal and portosystemic collateral venous circulation in addition to hepatomegaly and splenomegaly (**Figure 1C**). Cardiac catheterization showed single ventricular end-diastolic pressure between 10 and 12mmHg, pulmonary arterial hypertension with mean pulmonary artery pressure of 29 mmHg in the right branch and 25 mmHg in the left branch, selective angiography of the superior vena cava observed a 85 mm aneurysmal dilation with the presence of veno-venous fistulas that had an infra-diaphragmatic connection and pulsatile flow (**Figure 1D**).

Due to the risk of rupture and thrombosis with pulmonary embolism, surgical intervention was decided. The BDG takedown was performed with resection of the aneurysmal superior vena cava and reconnection of the superior vena cava to the right atrium with a 20 mm Gore-Tex® graft and subsequently the creation of a central systemic-to-pulmonary shunt in horseshoe with 10mm ringed Gore-Tex® graft (**Figure 2**). The main intraoperative complication was bleeding due to the presence of massive collateral circulation; the patient had a torpid evolution and multiple organ failure, dying 25 days after surgery.

Discussion

There is controversy with accessory pulmonary blood flow (APBF), where one possibility is to maintain it at the time of BDG to increase systemic saturation or, on the other hand, to eliminate it to reduce ventricular volume load. APBF provides more "physiological" levels of oxygen saturation, inhibition of arteriovenous malformations and potentially reduces the risk of developing pulmonary collateral arterial vessels[2,3]; in addition, it has been proposed that it can stimulate the growth of the pulmonary artery, which leads to better results in candidates for the Fontan[2] procedure. However, APBF can cope with some complications such as: systemic venous hypertension in the upper part of the body, chronic volume overload in the ventricular chamber and secondary effects due to a competitive flow between the superior vena cava and antegrade flow; in addition to an invariable adverse effect on the pulmonary vasculature, since it represents a source of high blood flow pressure that can lead to alterations in pulmonary vascular resistance[4].

The pathogenesis of venous aneurysms is still unknown, finding in histological reports from normal characteristics to a marked disorganization of the middle layer and inflammation of the venous wall. Endophlebo-

hypertrophy or endophlebosclerosis is an important factor in the formation of venous aneurysms similar to the role of arteriosclerosis in the formation of arterial aneurysms, whereby arterialization and increased flow in the venous system lead to wall hypertrophy, followed by dilation and sclerosis of it[5].

Endophlebohypertrophy occurs from birth and is associated with areas of stress, the entrance areas to the veins being tributary and adjacent to the artery, which is why it is suggested that an intraluminal force may affect venous histology with degenerative changes. Finally, the theory of localized inflammation in the vessels or connective tissue disorders can lead to degenerative changes in the venous wall, which is why trauma has been proposed as a probable etiological factor[5,6].

References

1. Pandit AA, Alegria JR, Pandit A, Mookadam M, Mookadam F. Giant superior vena-cava aneurysm after Glenn surgery: a new complication of the Glenn procedure. Heart Lung Circ. 2014;23(5): e136-e138.
2. Mainwaring RD, Lamberti JJ, Uzark K, Spicer RL, Cocalis MW, Moore JW. Effect of accessory pulmonary blood flow on survival after the bidirectional Glenn procedure. Circulation. 1999;100(19 Suppl): II151-II156.
3. McElhinney DB, Marianeschi SM, Reddy VM. Additional pulmonary blood flow with the bidirectional Glenn anastomosis: does it make a difference?. Ann Thorac Surg. 1998;66(2):668-672.
4. Pennati G, Migliavacca F, Dubini G, Pietrabissa R, Fumero R, de Leval MR. Use of mathematical model to predict hemodynamics in cavopulmonary anastomosis with persistent forward flow. J Surg Res. 2000;89(1):43-52.
5. Uematsu M, Okada M. Primary Venous Aneurysms: Case Reports. Angiology. 1999; 50(3):239-244.
6. Panduranga P, Thomas E, Al-Maskari S, Al-Farqani A. Giant superior vena caval aneurysm in a post-Glenn patient. Interact Cardiovasc Thorac Surg. 2012;14(6):878-879.

Figure Legend

Figure 1.

A) Computed tomography and B) Volumetric reconstruction, show superior vena cava aneurysm (arrows), C) Aneurysm (arrow) and enlarged liver and congestion in the thoracoabdominal circulation, D) Diagram of pressures and measurements of cardiac catheterization.

Figure 2.

Reconstruction of the superior vena cava (black arrow) and the central pulmonary systemic fistula (white arrow).