

## CASE REPORT

# Aneurysm of the superior vena cava in a bidirectional Glenn procedure with antegrade pulsatile flow

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*We present a 22-year-old male patient with a right bidirectional Glenn procedure history with dyspnea, cyanosis, and chest pain. Images revealed aneurysmal dilation of the right superior venous system. Bidirectional Glenn takedown was performed, resectioning the aneurysmal superior vena cava, and a central systemic-to-pulmonary shunt was created.*

**Key words:** Accessory pulmonary blood flow; Congenital heart disease; Glenn procedure; Superior vena cava, aneurysm.

*Presentamos un paciente masculino de 22 años, con antecedentes de un procedimiento de Glenn bidireccional derecho, con disnea, cianosis y dolor torácico. Las imágenes revelaron dilatación aneurismática del sistema venoso superior derecho. Se realizó desmantelamiento del Glenn bidireccional, resección de la vena cava superior aneurismática y creación de un shunt sistémico-pulmonar central.*

**Palabras clave:** Flujo sanguíneo pulmonar accesorio; Cardiopatía congénita; Procedimiento de Glenn; Vena cava superior, aneurisma.

*Cir Card Mex* 2023; 8(4): 117-119.

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The bidirectional Glenn procedure (BDG) involves the connection of the superior vena cava (SVC) to the pulmonary artery as a palliative procedure in many congenital heart diseases, with a free flow of venous blood through the right and left pulmonary arteries. Complications after this surgery include cyanosis, arrhythmias, protein-losing enteropathy, pulmonary arteriovenous malformations, liver dysfunction, hepatocellular carcinoma, etc. [1].

We present an unusual case of an aneurysm of the superior vena cava after BDG.

### CLINICAL 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 two years with pulmonary artery banding (PAB) and at 17 years underwent right BDG in another institution. He was admitted due to oppressive chest pain, dyspnea, cyanosis, and headache. On physical examination, with 70% pulse oximetry and jugular vein distention. A trans-

thoracic echocardiogram confirmed the diagnosis and reported patent cavopulmonary shunt with a dilated pulsatile flow, PAB with a maximum gradient of 75 mmHg, mean gradient of 52 mmHg, and pulsatile flow. Computed tomography revealed aneurysmal dilation of the right superior venous system (Fig. 1A) (Fig. 1B) that involved the right jugular vein, innominate vein, superior vena cava, azygos, and hemiazygos systems, with increased thoracoabdominal and portosystemic collateral venous circulation; hepatosplenomegaly (Fig. 1C). Cardiac catheterization showed single ventricular end-diastolic pressure between 10 and 12 mmHg, 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 SVC observed an 85 mm aneurysmal dilation and pulsatile flow (Fig. 1D).

Due to cardiac catheterization findings, risk of rupture, and thrombosis with pulmonary embolism, palliative surgical intervention was decided. BDG takedown was performed, resection of the aneurysmal superior vena cava, reconnection of the superior vena cava to the right atrium with a 20 mm Gore-Tex® graft, and creation of a central systemic-to-pulmonary shunt in a horseshoe shape with 10mm ringed Gore-Tex® graft (Fig. 2). Main intraoperative complication was bleeding due to the presence of massive collateral circulation, which led to a torpid evolution dying 25 days after surgery attributable to multiple organ failure.

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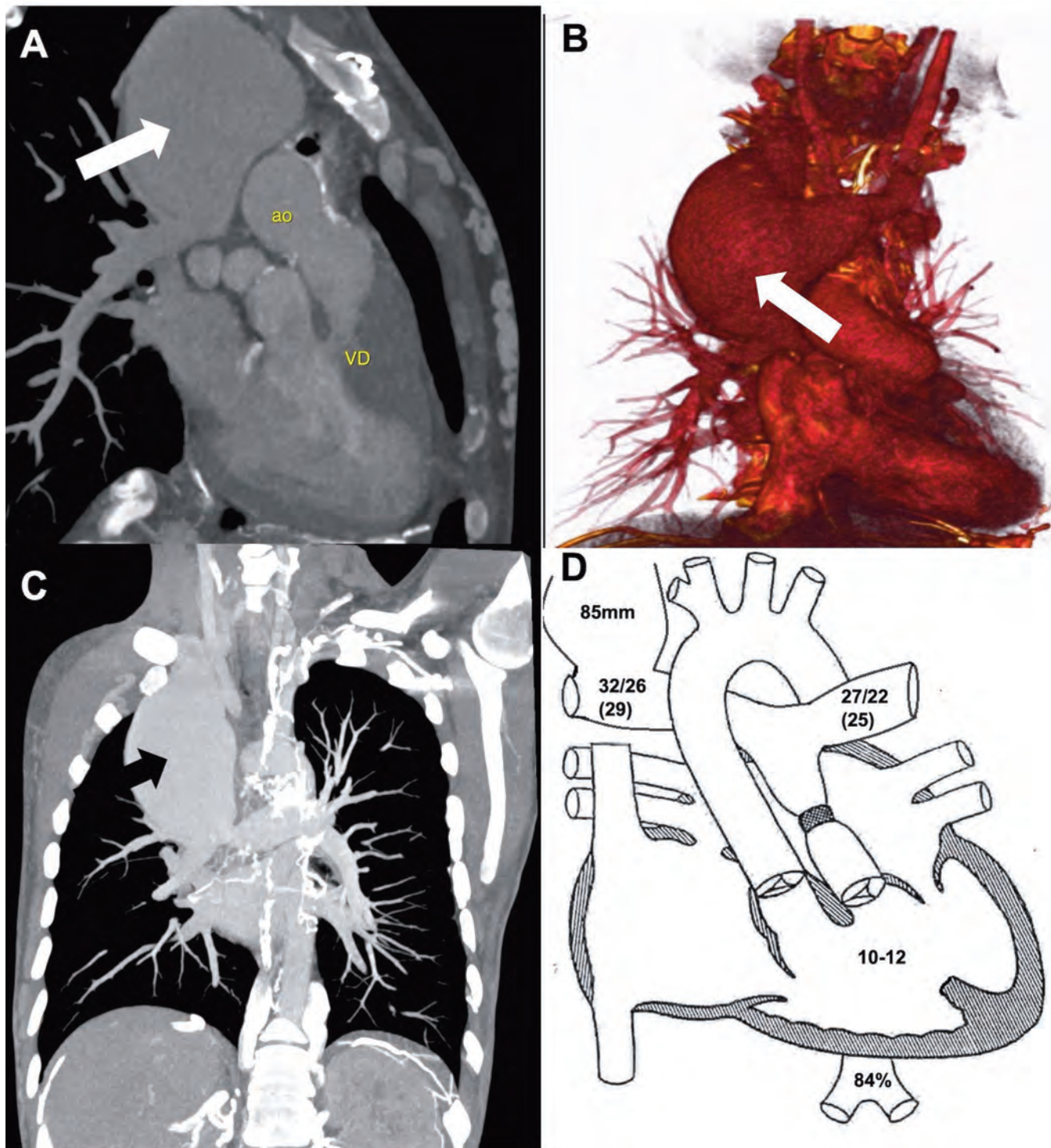


Figure 1. A) Computed tomography and B) Volumetric reconstruction, show superior vena cava aneurysm (arrows), C) Aneurysm (arrow) D) Diagram of cardiac catheterization.

## COMMENT

There is controversy with accessory pulmonary blood flow (APBF); one possibility is to maintain it at the time of BDG to increase systemic saturation or to eliminate it to reduce ventricular volume load. APBF provides more "physio-

logical" oxygen saturation levels, inhibition of arteriovenous malformations, and reduces the risk of developing pulmonary collateral arterial vessels [2]. It can stimulate the growth of the pulmonary artery, leading to better results in candidates for Fontan [2] procedure. However, APBF can have some complications, such as systemic venous hypertension in the upper



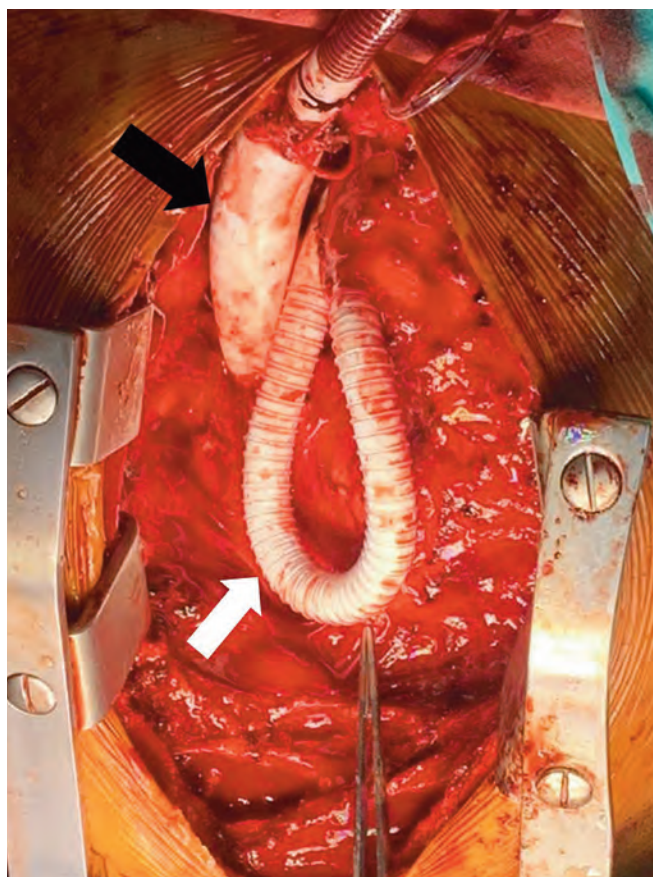


Figure 2. Reconstruction of the superior vena cava (black arrow) and central pulmonary systemic shunt (white arrow).

part of the body, chronic volume overload in the ventricular chamber and secondary effects due to a competitive flow between the SVC and antegrade flow, and 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 [3].

Pathogenesis of venous aneurysms is still unknown [4], finding in histological reports from normal characteristics to a marked disorganization of the middle layer and inflammation of the venous wall. Endophlebohertrophy or endophleboscclerosis is an important factor in the formation of venous aneurysms, whereby arterialization and increased flow in the venous system lead to wall hypertrophy, followed by dilation and sclerosis of it [5]. Endophlebohertrophy is associated with areas of stress, 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 can lead to degenerative changes in the venous wall, which is why trauma has been proposed as a probable etiological factor [5]. APBF in the BDG, along with endophlebohertrophy could contribute to developing venous aneurysms in these patients. Aneurysm of superior vena cava after BDG is rare, with only a few cases reported, so further studies are required to support this hypothesis.

**FUNDING:** None

**DISCLOSURE:** The authors have no conflicts of interest to disclose.

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