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Case Report
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A Modified Warden Procedure for a Partial Anomalous Pulmonary Venous Connection to the Azygos Vein

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Abstract

Partial anomalous pulmonary venous connection to the azygos vein is a very rare condition and only a few cases are described in literature. The main problem in surgical correction is the distance between the pulmonary veins and the left atrium. For this reason, we modified the Warden technique using a conduit between the distal stump of the superior vena cava that was 10 cm away from the right atrium, and the right appendage to restore the systemic venous drainage. A cardiac MR, performed after six uneventful months, documented unobstructed pulmonary and systemic venous drainage.

Introduction

Partial Anomalous Pulmonary Venous Connection (PAPVC) to the AV is a very rare condition and only a few cases are described in literature [1]. One of the problems related with this condition is the distance between the anomalous pulmonary veins and the left atrium. Different surgical solutions have been proposed, but none of them fitted with our case. For this reason, we decided to modify the Warden procedure with interposition of a conduit for Superior Vena Cava (SVC) reimplantation [2].

Case Presentation

An asymptomatic 58-years-old man underwent medical examination for competitive sport. Echocardiographic evaluation disclosed a severe dilation of the right heart with volume overload. A CMR evidenced the right superior and inferior pulmonary veins directly connected to the Azygos Vein (AV). A smaller right pulmonary vein from the middle lobe drained directly in the left atrium, as did the left veins. No interatrial communication was present. Qp/Qs ratio was 1:91. A contrast-enhanced CT scan evidenced a huge AV (max diameter 25 mm), receiving the anomalous pulmonary veins, that drained in the SVC approximately 10 cm from the Right Atrium (RA). A resin 3D printed reconstruction was used to plan the surgical strategy (Figure 1).

Upper body drainage for CPB was obtained with a 12 Fr venous cannula in the innominate vein. A Warden procedure seemed the right solution to preserve the pulmonary venous return. The AV was extensively isolated and ligated just before anomalous pulmonary veins entry. The SVC was interrupted above the AV connection and the proximal stump oversewn. Through a right atriotomy, a generous septectomy was obtained and the SVC, draining the anomalous pulmonary veins, was baffled to the left atrium with a heterologous pericardial hood. The 10 cm distance between the distal portion of the SVC and the RA was considered excessive for a direct connection. A 20 mm armed Gore-Tex conduit (W.L. Gore assoc. Inc, Elkton, MD) about 12 cm long was used to reconnect the distal stump of the SVC to the RA (Figure 2). The patient was discharged after six uneventful post-op days. Pre-discharge echocardiography showed an unobstructed systemic and pulmonary venous flow. A 3-month anticoagulation therapy followed by a lifelong intake of antiplatelets were prescribed. A cardiac ECG synchronized MR at 6 months follow-up evidenced a Qp/Qs=1.04, an unobstructed pulmonary venous return to the left atrium and normality flow pattern inside the SVC-Gore-Tex conduit. A volume reduction of the right heart was evident.

Discussion

Less than 20 cases of PAPVC to AV are reported in literature [1,3]. Our patient was particularly difficult because the AV drainage in the SVC was very far from RA. A direct reimplant of the AV in the left atrium, described essentially for scimitar syndrome and anomalous left pulmonary veins, was considered impossible for the interposition of the right bronchus [4]. The intracaval rerouting of the anomalous flow with a patch, our technique of choice in PAPVC repair, was thought to

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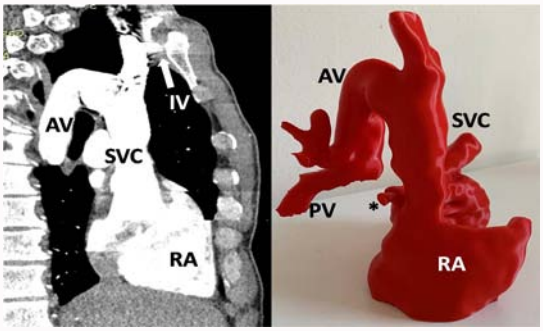

Figure 1: CT scan and 3D reconstruction.
(AV: Azygos Vein, SVC: Superior Vena Cava, RA: Right Atrium, IV: Innominate Vein, PV: Superior and Inferior Pulmonary Veins, *: Median Pulmonary Vein)

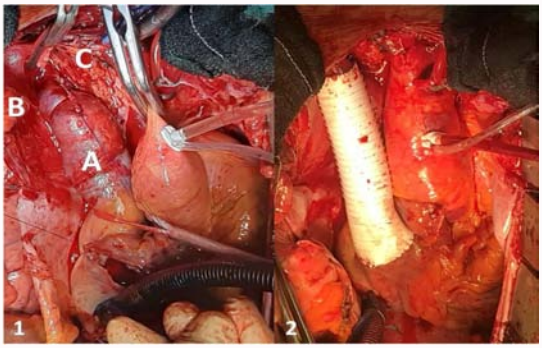

Figure 2: Intraoperative view: 1) The superior vena cava (A) Transected at azygos vein entry level (B) Leaving a short distal stump (C); 2) The interposed Gore-Tex conduit.

be at risk of obstruction because the distance was more than 10 cm [5]. The only option appeared to be the Warden procedure which proved to preserve effectively the abnormal pulmonary venous return to the heart [2]. Unfortunately, the distance between AV and innominate vein drainage in the SVC was minimal and the distal stump was too short and difficult to reimplant to the right appendage without excessive tension on the anastomosis that could cause SVC obstruction as previously described [4]. For this reason, we decided to interpose an armed Gore-Tex conduit between the distal SVC stump and the right appendage. This strategy was already described in modified warden operation for PAPVC in SVC but this is the first time for a PAPVC in AV. This conduit guarantees a perfect laminar flow preserving an optimal fluido-dynamic and avoiding kinetic energy loss. This solution proved to be effective and can be useful in difficult anatomies.

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