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Kasuistika | Case report

Modified Warden procedure in adult with partial anomalous pulmonary venous connection after previous atrial septal defect repair

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SOUHRN

Dvacetiletý muž byl hospitalizován v naší nemocnici pro recidivu pleurálního výpotku, dušnost při námaze a počáteční známky selhání pravého srdce. V dětství mu byla stanovena primární diagnóza defektu síňového septa typu secundum a ve věku 16 let absolvoval korekci defektu síňového septa záplatou. Vyšetření magnetickou rezonancí prokázalo anomální návrat dvou plicních žil vysoko do horní duté žíly. Provedli jsme modifikovanou Wardenovu operaci s konduitem z umělého materiálu. Popisujeme výhody této techniky při opakovaném výkonu u dospělého pacienta s vrozenou vadou.

ABSTRACT

A 20-year-old boy was admitted to our Hospital for recurrent pleural effusion, effort breathless and initial signs of right heart failure. He had primary diagnosis, in childhood, of secundum ASD and had undergone, at the age of 16 years, ASD repair with patch. A MRI showed anomalous connection of two pulmonary veins in highest superior vena cava. We performed a modified Warden procedure with a prosthetic conduit. We report the advantages of this technique in reintervention in adult congenital patient.
Learning objective: To give information about the possible surgical technique in adult congenital patient with anomalous pulmonary veins connection who had previously undergone surgical procedure.

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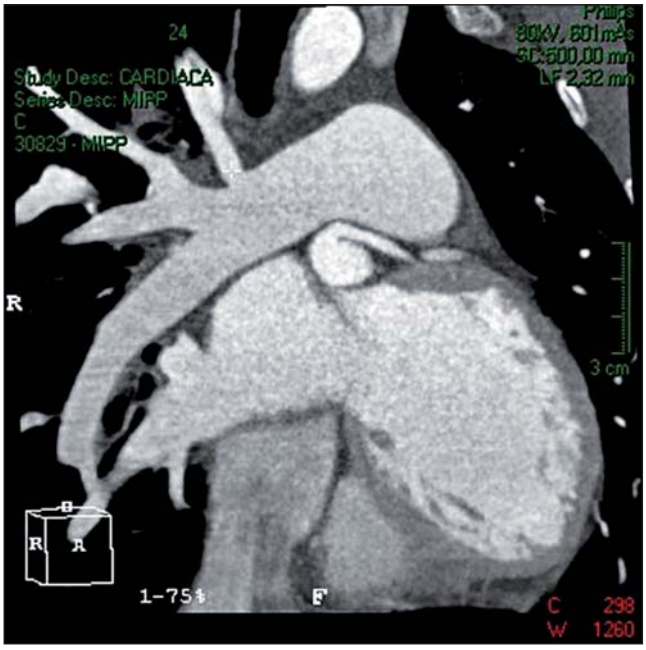
Introduction

Partial anomalous pulmonary veins connections (PAPVC) is a congenital heart defect characterized by the failure of 1–3 pulmonary veins (PV) to drain into the left atrium (LA) [1]. It is often associated to atrial septal defect (ASD), more frequently to sinus venous ASD, rarely to secundum ASD [2–4]. The PAPVC to the superior vena cava (SVC) occurs in 10–15% of all patients [3]. In adult patients the incidence of PAPVC was 5% [5]. Some report showed cases of PAPVC misunderstanding at time of ASD repair that developed symptoms in adulthood, indeed PAPVC has been reported in 0.4–0.7% of autopsies carried out in patients with other congenital heart disease [4,6]. The aim of surgery was to re-route the anomalous pulmonary flow in the LA in an unobstructed way. In 1984, Warden and colleagues [7] reported a caval division technique in which the SVC was separated: the cephalic end was anastomosed to the right atrial (RA) appendage and the caudal end, carrying the anomalous vein, was baffled with an internal ASD patch to the LA. The SVC-RA connection may be achieved directly or by the interposition of prosthetic conduit. The advantage of this technique is to allow a re-routing of pulmonary flow avoiding PV or SVC obstruction and sinus node injury [1,4,8].

Here we report a case of an adult patient, who had undergone secundum ASD repair in childhood, with late diagnosis of PAPVC for persistent invaliding symptoms, and we describe our surgical approach with a modified Warden procedure.

Case report

A 20-year-old boy was referred to our grown-up congenital heart disease clinic for recurrent pleural effusion and shortness of breath on exertion for the past 12 months. He had primary diagnosis, in childhood, of secundum ASD and had undergone, at the age of 16 years, ASD repair with patch in another Hospital. For the past few years he developed several hospital admissions for worsening shortness of breath and recurrent pleural effusion treated with medical therapy. When we first saw him, on physical examination, the child was in discreet general health, eupneic with a mild systolic murmur in the pulmonary area. The ECG showed sinus rhythm with RBBB. We performed an echocardiogram which showed moderate to severe right ventricle dilatation and dysfunction (TAPSE 20 mm), moderate tricuspid regurgitation and initial sign of pulmonary hypertension (mean PAPs 35 mmHg) with no residual shunt at atrial septal level. The transthoracic echocardiogram was not able to clearly assess pulmonary veins connection but we suspected the possible co-existence of PAPVC undiagnosed at the time of ASD repair, so we asked further imaging evaluation. The CT scan highlighted only the presence of azygos accessory lobe and a single anomalous PV to the SVC. Then, according to the recent literature [9], we decide to go further with diagnostic tests, performing a morpho-functional evaluation through a MRI scan. The MRI (Figs. 1–4) showed an anomalous return of the right upper superior vein and of the accessory vein of azygos lobe in the higher part of SVC,



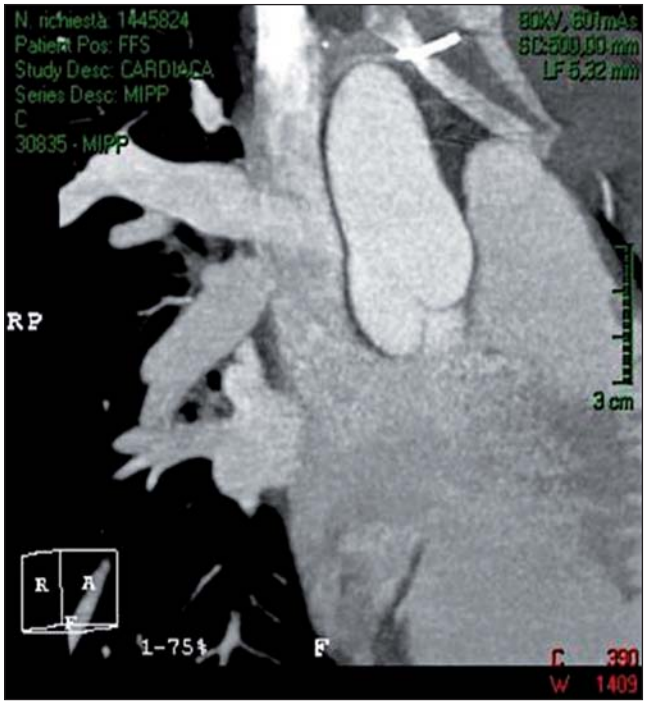


Fig. 1 and Fig. 2 – MRI scan frontal view with the anomalous pulmonary vein.

RA dilatation, a moderate to severe RV dysfunction and a functional Qp:Qs ratio of 1:9 also related to moderate hypoplasia of left lung. After multidisciplinary discussion, a surgical re-routing of anomalous pulmonary vein drainage in the LV was planned. A redo-sternotomy was carried out and the cardiopulmonary bypass was instituted by cannulation of ascending aorta for the arterial output and of inferior vena cava and innominate vein for the venous drainage. After accurate dissection, the SVC and the two anomalous pulmonary veins were identified and encircled with vessel loops. Particularly, care was taken to identify the azygos vein. The aorta was cross-clamped

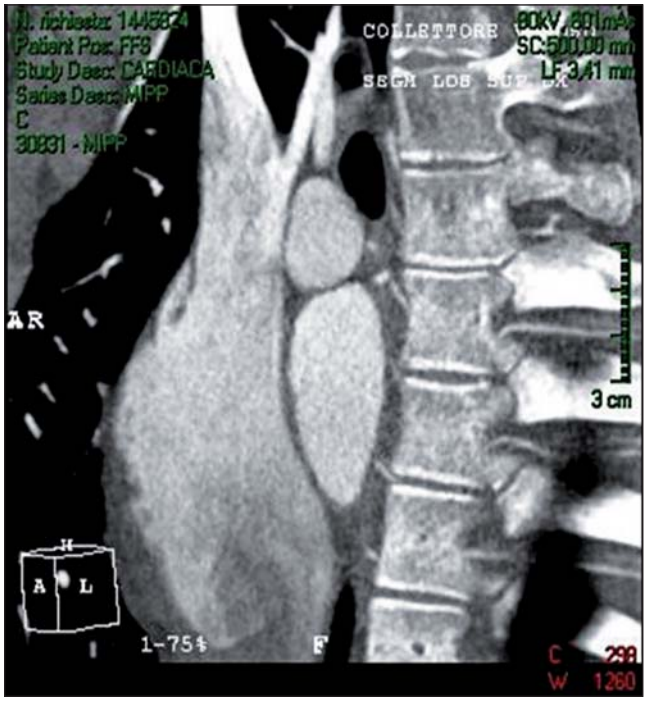


Fig. 3 – MRI scan lateral view with the anomalous pulmonary vein at the SVC-RA junction.

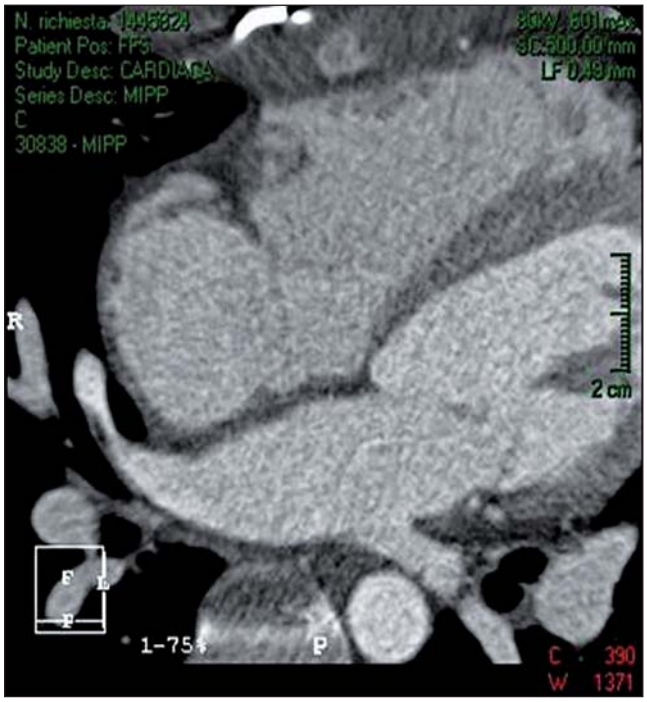


Fig. 4 – MRI transverse section view of the cardiac chambers.

and cardioplegia infused in the aortic root. A right atriotomy was done and the orifice of pulmonary veins in the higher part of SVC was identified. Due to the peculiar position, we decided to perform a modified Warden procedure with a prosthetic conduit. The SVC was divided above the insertion of the highest pulmonary vein. A pericardial patch was used to close the cardiac stump of the SVC to prevent stenosis or obstruction of the pulmonary venous return. The previous ASD patch was removed and the atrial septum enlarged. A heterologous pericardial patch was sutured to the margin of the ASD on to the SVC orifice, baffling the SVC orifice to re-routing the blood flow from the anomalous pulmonary veins into the LA.

The RA appendage was amputated and enlarged and the intra-atrial trabeculations were extensively excised. The upper end of SVC was connected to the RA through a Goretex (W.L. Gore and Associates, Flagstaff, AZ) conduit. The postoperative course was uneventful but one week from the surgery the patient needed surgical revision for pericardial effusion probably due to not well-controlled anticoagulant therapy.

On the 20th postoperative day, the patient was discharged from the Hospital and a follow-up of three months, with echocardiogram, confirmed the good result of surgical repair with patent SVC-RA conduit.

Discussion

Surgical repair for PAPVC to SVC ideally includes complete closure of the septal defect and redirection of the anomalous pulmonary veins into the LA without pulmonary venous or SVC obstruction or injury of the sinus node or its blood supply [4]. Various surgical techniques have been introduced [4,8,10] but, nowadays, the Warden procedure is still considered the gold standard for this type of patients [1,4,6,7]. Our case showed peculiarities both on preoperative evaluation and on intraoperative management. Instead, as previously reported [2,8], despite the association between secundum ASD and PAPVC being uncommon, in each type of ASD, the course of pulmonary veins should be explored either pre- or intraoperative. The high connected pulmonary veins may be missed during surgery in absence of sufficient preoperative information or due to the reluctance of surgeons to extend dissection around the SVC for the associated risk of phrenic nerve injury. Furthermore, since echocardiogram and CT scan can fail to identify the course of pulmonary vein, especially in adult patients, an MRI should be performed when diagnostic suspicion exists in accordance with recent report [9]. Indeed suboptimal echocardiogram acoustic windows may preclude adequate inspection of pulmonary veins or atrial septal defect while the CT scan may fail to give functional information that is often necessary for surgery indication. On the contrary, MRI ensures not only noninvasive volumetric anatomic data and enables evaluation of systemic veins, as well as the number, origin, course, and drainage, but also provides a functional evaluation that allows to define the Qp/Qs shunt ratio [9].

From a surgical point of view, our case suggested that in adult patients the best option to PAPVC repair, especially in the high SVC connection, is the modified Warden procedure with a conduit. Indeed, in the classic Warden procedure to avoid SVC-RA stenosis, the cavoatrial anastomosis must be accomplished without tension and this requires very careful and extensive dissection of the brachiocephalic vein and SVC [4]. In adult patients, especially during reintervention, extensive SVC mobilization requires wide adhesions dissection that may be associated to bleeding and to phrenic nerve injury. Moreover when anomalous pulmonary veins are connected to high SVC,

the cephalic end of the divided SVC becomes more distant from the RA appendage, especially in adult patient, and there is likelihood of development of tension in the cavoatrial anastomosis [1,7] that causes SVC stenosis or obstruction. All these considerations, in accordance with previous report [4], lead us to suggest a modified Warden procedure with a prosthetic conduit in adult reintervention. The risk related to conduit is low and the conduit allows to achieve a good anatomic geometry, without additional risks.

Conclusion

Particular care and appropriate examination are needed to be done to identify the course of pulmonary veins when each type of ASD has to be treated. Moreover, PAPVC must be suspected in cases of previously repaired ASD when signs or symptoms of right heart failure are present with no apparent reason. In our experience, a modified Warden procedure, using a prosthetic conduit is the best option in adult reintervention for PAPVC repair.

Conflict of interest
The authors declare that there is no conflict of interest.

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None.

Ethical statement
I declare, on behalf of all authors, that the research was conducted according to Declaration of Helsinki.

Informed consent
I declare that informed consent requirements do not apply to this manuscript.

Appendix A. Supplementary data
Supplementary data associated with this article can be found, in the online version, at http://dx.doi.org/10.1016/j.crvasa.2015.08.004.

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