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Case report

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

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ARTICLE INFO

Article history:
Received 3 July 2015
Received in revised form
11 August 2015
Accepted 13 August 2015
Available online 4 September 2015

Keywords:
Pulmonary venous anomalies
Atrial septal defect
Adult congenital heart patients

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.

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 reports 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

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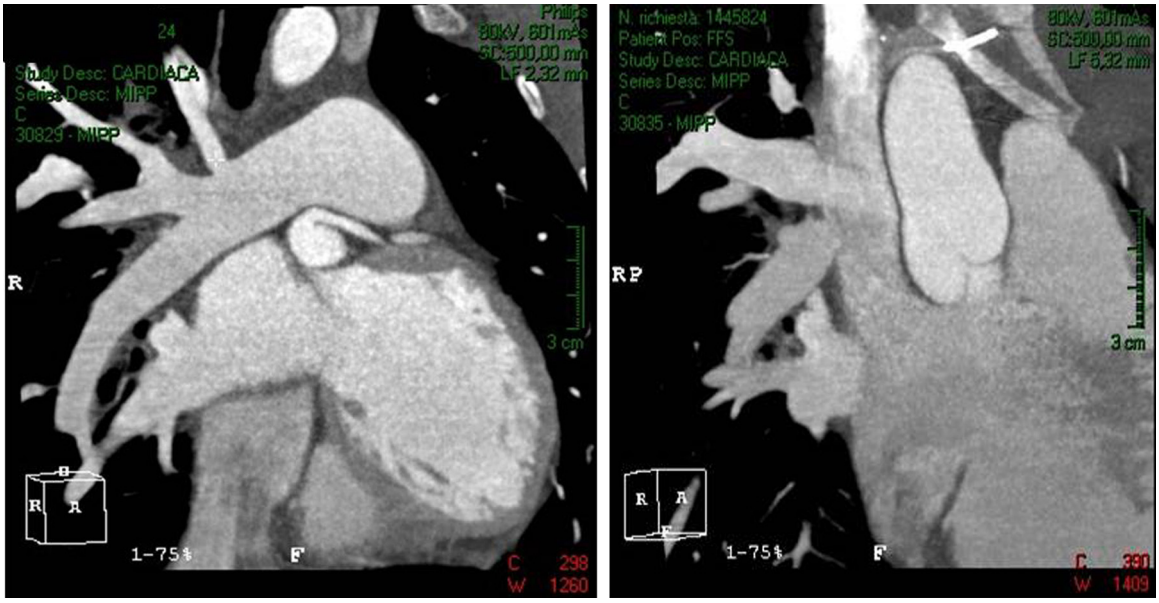
http://dx.doi.org/10.1016/j.crvasa.2015.08.004



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

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, 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

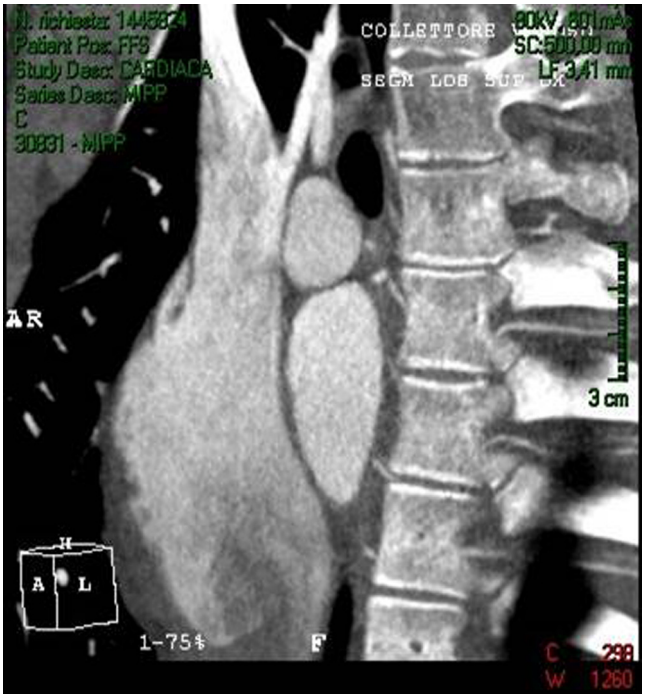


Fig. 3 – MRI scan lateral view with the anomalous pulmonary vein at the SVC-RA junction.