

Case Report

Percutaneous Intracardiac Baffle Stenting After a Scimitar Vein Correction

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A typical scimitar vein was surgically corrected in a female patient of 35 years old. The postoperative course was uneventful and in the first months after the functional capacity normalized. Six months after the correction, the patient developed progressive dyspnea. Kinking and distal stenosis of the intracardiac baffle which connected the scimitar vein to the left atrium were diagnosed. Balloon dilatation and stenting could normalize the blood flow through the baffle. The patient remains symptom-less up to six months after the procedure. © 2012 Wiley Periodicals, Inc.

Key words: congenital heart disease; anomalous pulmonary venous return; stent; transcatheter intervention

INTRODUCTION

The number of adult congenital heart disease patients is continuously growing. As the consequence of repair in childhood, some residual lesions persist and sequels may become more important at older age. Also, due to advanced imaging techniques and better awareness of underlying structural anomalies, some congenital defects are merely diagnosed later in life. In several of these cases, surgery is indicated for prognostic reasons. However, redo surgery or repair at older age is more complex and holds a higher risk for complications. Therefore, when surgery did not succeed, a less invasive transcatheter procedure might still belong to the possibilities. This is one of the reasons for the growing interest in hybrid procedures. We present a case report, where a patient had surgery for a congenital heart defect, the short-term result was not optimal, but a transcatheter procedure could offer a therapeutic solution.

CASE REPORT

A 35-year-old female patient was referred to our hospital for the correction of a scimitar vein, an anomalous pulmonary venous return of the entire right lung into the inferior vena cava. She was treated conservatively for years, but became recently symptomatic. Transthoracic echocardiography showed enlarged right heart cavities and right heart catheterization revealed a pulmonic to systemic cardiac output ratio of 1.7/1 without pulmonary hypertension. There were no medi-

cal antecedents except bronchial asthma, which was treated with inhalation corticosteroids and bronchodilators. The surgeon created an intracardiac baffle with an autologous pericardial patch between the scimitar vein and the left atrium, as indicated in Fig. 1.

The corrective procedure went easily, the postoperative course was uneventful, and the patient became completely asymptomatic. However, after 6 months, she started to complain from shortness of breath, even more than before the surgical correction. The physical examination, the electrocardiogram, and the chest X-ray were all normal. Transthoracic echocardiography showed normalized dimensions of the right heart, however, a distal baffle stenosis was observed on transoesophageal echocardiography (Fig. 2). A CT scan of the chest showed also a kink in the intracardiac baffle at the level of the diaphragm (Fig. 3).

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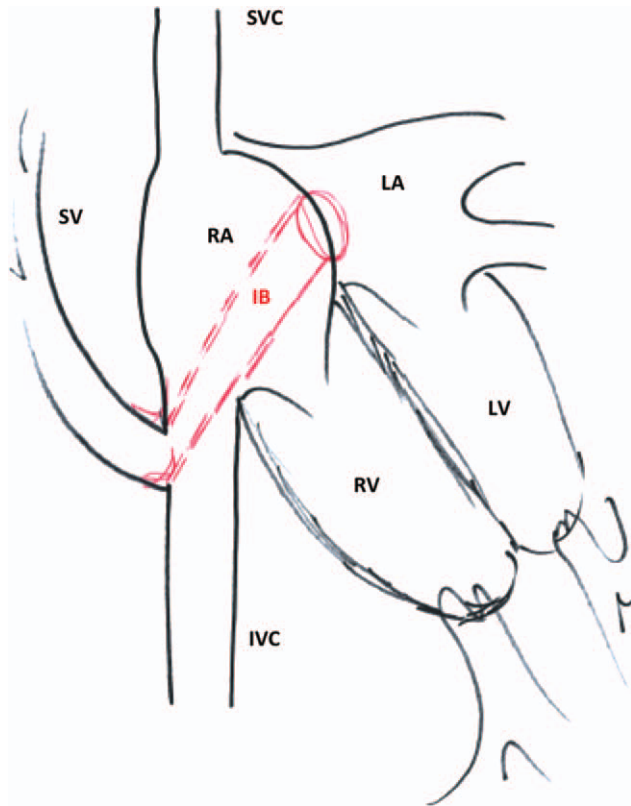


Fig. 1. Schematic drawing of the surgical intervention showed the intracardiac baffle, which was created between the scimitar vein and the left atrium. IB: intracardiac baffle, IVC: inferior vena cava, LA: left atrium, LV: left ventricle, RA: right atrium, RV: right ventricle, SV: scimitar vein, SVC: superior vena cava.

The patient was sent to the catheterization lab to preserve the patency of the baffle. General anesthesia was applied, and after venous access through the right femoral vein and a transseptal puncture, the left atrium was reached. With a Terumo guide wire (Leuven, Belgium), we were able to advance an angiography catheter retrogradely into the obstructed baffle. The angiogram confirmed the diagnosis of the kink and the stenosis at the distal part of the baffle (Fig. 4). A minimal left to right shunt from the baffle to the right atrium was visualized, but was calculated to be insignificant. It was decided to implant a self-expandable stent (Cook Medical ZIV6-80-14-6.0) and to dilate the distal part of the tunnel with a 14-mm balloon (Cordis Maxi LD 416-1440). After the stent implantation, the run off of contrast normalized (Fig. 5). The patient became asymptomatic and a CT scan of the chest at 6 months showed a normal patency of the stent.

DISCUSSION

The scimitar syndrome is a rare congenital heart defect, which is mainly characterized by an anomalous

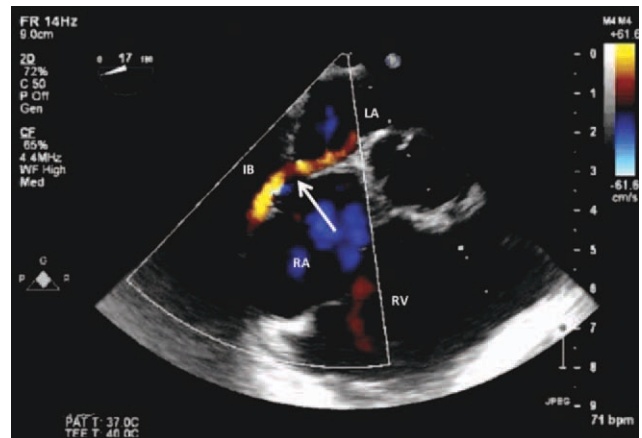


Fig. 2. On transoesophageal echocardiography (color Doppler) a stenosis on the distal part of the intracardiac baffle was observed (white arrow). IB: intracardiac baffle, LA: left atrium, RA: right atrium, RV: right ventricle.

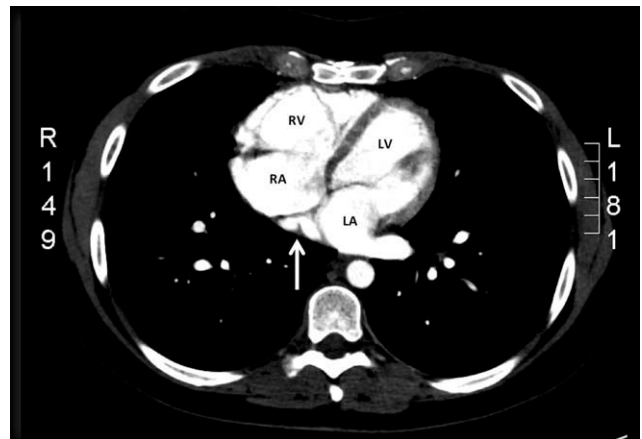


Fig. 3. CT scan of the chest documented a kinking in the intracardiac baffle between the scimitar vein and the left atrium at the level of the diaphragm (white arrow). LA: left atrium, LV: left ventricle, RA: right atrium, RV: right ventricle.

pulmonary venous return of the entire right lung to the inferior vena cava (IVC), one of the hepatic veins, or the vena porta [1]. The name is based on the typical appearance of a curvilinear shadow adjacent to the right heart border that looks like a curved Turkish sword [2]. It is not uncommon that a scimitar vein is associated with an atrial septal defect, and, however less frequently, with a ventricular septal defect, coarctation of the aorta, tetralogy of Fallot, or a persistent arterial duct. Other manifestations of the syndrome are hypoplasia of the right lung and right pulmonary artery, abnormalities of the trachea-bronchial architecture, anomalies of pulmonary lobation, sequestration, diaphragmatic hernia, and rarely an absent IVC [3]. In general, surgical treatment is required when the ratio of the pulmonic tot systemic blood flow is greater than

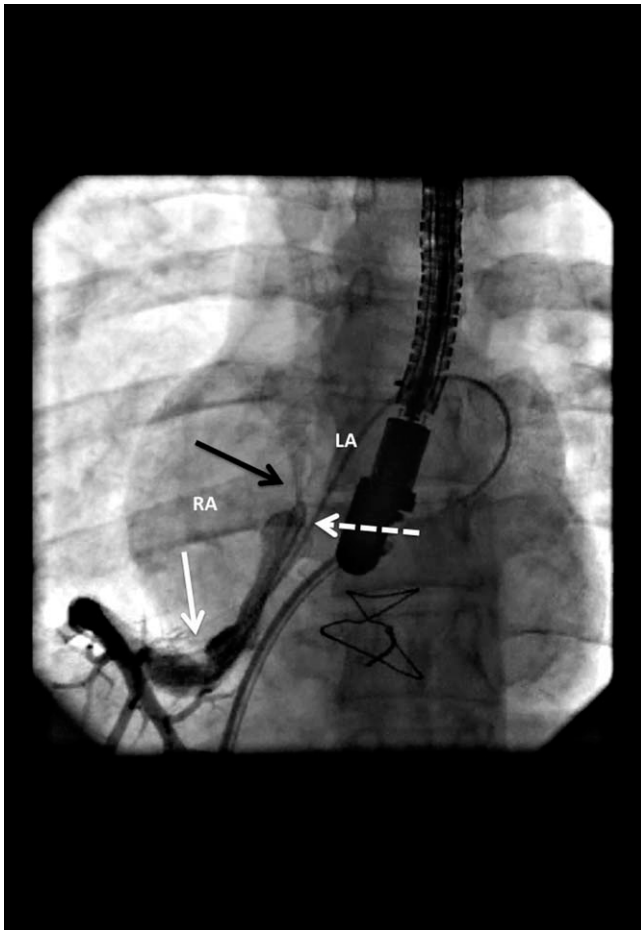


Fig. 4. Face angiography showed the kink (white full line arrow) and the stenosis (white dotted arrow) of the baffle. A small left to right shunt could be visualized, however, calculated to be insignificant (black arrow). LA: left atrium, RA: right atrium.

2/1 [4], and includes redirection of the scimitar vein to the left atrium, and correction of the associated anomalies [5,6]. Although the pulmonary to systemic blood flow reached only 1.7/1, we decided to correct because of the patient's symptoms and the enlarged right heart on transthoracic echocardiography. The deep position into the lung tissue of the scimitar vein let the surgeon decide to create an intracardiac baffle from where the scimitar vein entered the IVC, through the diaphragm, up to the left atrium. Unfortunately, scimitar correction is frequently associated with pulmonary vein obstruction [7,8]. Vida et al. found that that scimitar vein stenosis was lower for patients who underwent an intracardiac baffle repair compared with those who had a re-implantation (18% versus 50%) [1]. They also suggested that direct re-implantation of the scimitar vein into the left atrium carried a higher incidence of post-operative complications. The kink in the baffle was probably caused by the sharp angle between the vein

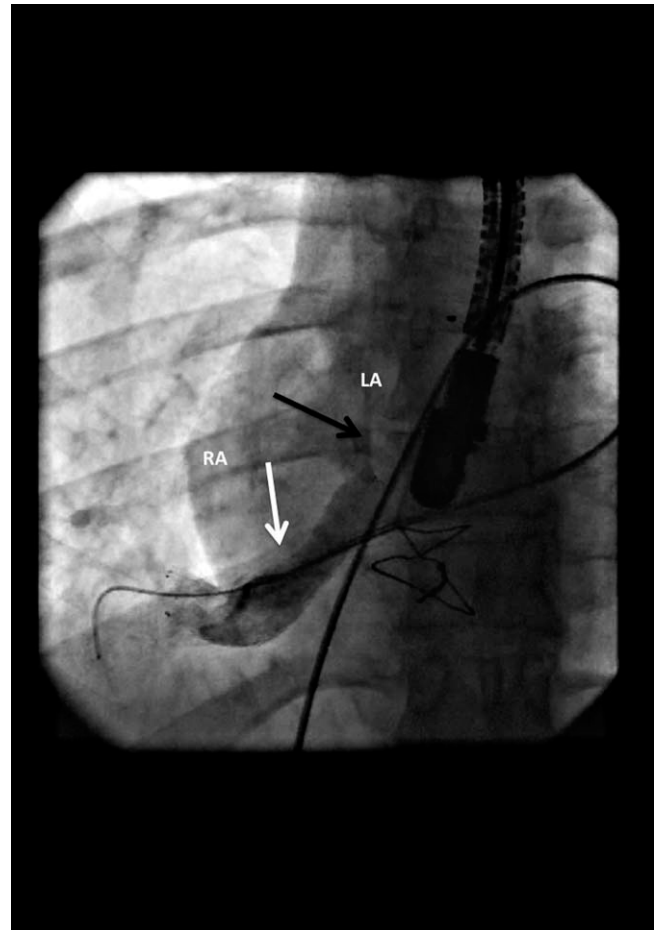


Fig. 5. Face angiography documented the fully deployed stent (Cook Medical ZIV6-80-14-6.0) in the baffle (white full line arrow). Run off of contrast normalized. The small left to right shunt persists (black arrow). LA: left atrium, RA: right atrium.

and the baffle. We decided to balloon dilate and to stent the intracardiac baffle through a retrograde approach. The symptoms regressed and the stent is patent at 6 months after the intervention.

Therefore, retrograde stenting of the intracardiac baffle is feasible and efficient. However, the long-term results of this new approach remain unknown so that systemic follow-up is mandatory.

CONCLUSION

Scimitar repair in adults is a rare surgical intervention. Tunnel stenosis might occur due to kinking or scar tissue. Retrograde stenting the tunnel is feasible and effective.

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