

CONGENITAL HEART DISEASE

The Role of Cardiac Magnetic Resonance in the Diagnosis of Anomalous Pulmonary Venous Return with Subsequent Amplatzer Device Treatment

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ABSTRACT

We report the case of a young girl with a mixed total anomalous pulmonary venous return (cardiac and supracardiac) treated sequentially by partial neonatal surgery, and then catheterization at age 19 with installation of an Amplatzer device as a treatment of the remaining anomaly. We describe the usefulness of magnetic resonance imaging in both the diagnosis and follow-up of this anomaly.

INTRODUCTION

Total anomalous pulmonary venous return (TAPVR) is a rare congenital cardiac anomaly, the major type being supracardiac (1). In the English language literature, very few cases of mixed supracardiac and cardiac TAPVR have been reported.

Treatment usually requires surgery (2), and, until now there have been no published cases where treatment, even partial, involved transcatheter procedures and Amplatzer prosthetic devices. Magnetic resonance imaging is an accurate method to diagnose and follow congenital cardiopathies (3–5) and is starting to be used for monitoring treatments involving transcatheter installations of prosthetic devices (6). The case we report describes the exceptional occurrence of mixed TAPVR, the possibility of endovascular treatment for this anomaly and the role of MRI in its diagnosis and follow-up.

CASE REPORT

An infant girl, aged one month, was referred to our institution for cardiac failure (difficult feeding and diaphoresis,

with no cyanosis or failure to thrive). Ultrasound and angiography exploration showed a mixed total anomalous pulmonary venous return (supracardiac and cardiac) with interatrial communication.

This anomaly was complex, since both right pulmonary veins and the lower left pulmonary vein were draining into the coronary sinus via a venous collector, whereas the upper left pulmonary vein (ULPV) was draining into the left innominate vein via a vertical vein. At that time, we tried but could not demonstrate the presence of any communication between the ULPV and the common pulmonary vein (of the other three pulmonary veins).

Partial corrective surgery was performed, i.e., anastomosis of the common pulmonary vein to the left atrium via a coronary sinus fenestration, as well as closure of the interatrial communication. Since the left vertical vein was inaccessible through median sternotomy, it was not ligated and another intervention was scheduled for around age 4 to 5 years. A decision was made to follow her both clinically and with echocardiography. The decision to delay the surgical option was reinforced by the discovery at age 3 of a moderate left-right shunt ($Qp/Qs = 1.4$).

During adolescence, there was gradual dilatation of the right cardiac cavities and, finally, an ultrasonography follow-up at age 17 showed more significant dilatation of the right cavities with paradoxical ventricular septum movements. Furthermore, there was a strong suspicion of an actual communication between the ULPV, and the collector tube anastomosed to the left atrium. A cardiovascular MRI showed the anatomy of the vertical vein and unequivocally confirmed the communication between the LUPV and the common pulmonary vein (Fig. 1A–D), which

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Figure 1. Successive, three-dimensional, native coronal slices showing the communication between the left pulmonary vein and the collector tube (A and B, white arrow), and between the left pulmonary vein and the vertical vein (C and D, white arrow).

presented with localized narrowing but no obstruction. It was therefore decided to treat this residual anomaly.

After multidisciplinary discussion, and given the institution's expertise in interventional procedures, a non-surgical treatment with the installation of an Amplatzer device was proposed to occlude the vertical vein. After angiographic confirmation of the anatomical MRI findings and the non-obstructive nature, the procedure was performed without complication (Fig. 2). Hospitalization lasted for 24 hours.

Follow-up showed quick regression of the dilatation of the right cavities, normalization of ventricular septum movements and absence of residual shunt. The Qp/Qs ratio measured by

isotope scan 10 days after the procedure was normal, with no perfusion deficit.

A follow-up MRI 9 months after the procedure demonstrated that the device was in a satisfactory position to occlude the vein (Fig. 3A–B). There was no obstructive stenosis of the flow noted in the innominate vein or in the ULPV.

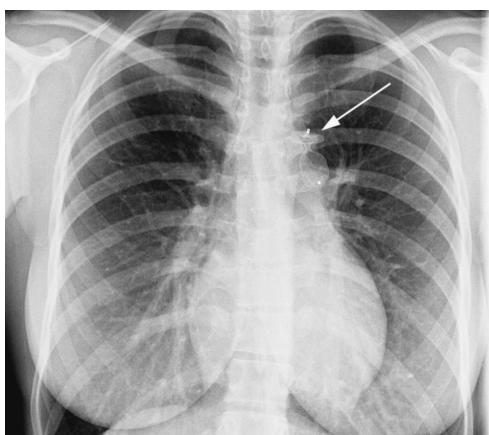


Figure 2. Antero-posterior chest x-ray after the procedure showing the positioning of the Amplatzer device (white arrow).

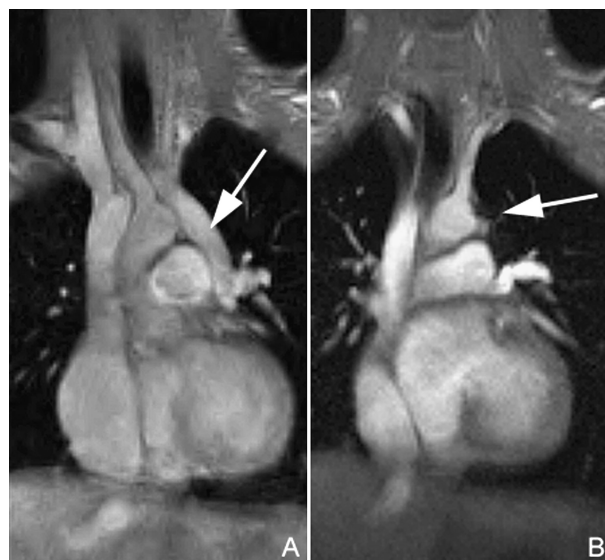


Figure 3. Oblique coronal dynamic sequence in the plane of the vertical vein showing its permeable appearance before the procedure (A, white arrow) and the absence of signal of the in-place device nine months after (B, white arrow).

The patient is now age 20 and, two years post-procedure, is completely asymptomatic, and her anomaly is considered fully corrected.

DISCUSSION

Total or partial anomalous pulmonary venous returns are rare. Total anomalies (TAPVR) represent approximately 2% of congenital cardiopathies and are always associated with patent foramen ovale or interatrial communication. They are mainly supracardiac (55%), most often with a connection towards the innominate vein via a vertical vein (1), although the connection can also be via the superior vena cava or the azygos vein. In the cardiac forms (30%), the anomalous vessel(s) drain(s) into either the coronary sinus (80% of cases) or the right atrium. In the infracardiac type, the venous drainage is most often into the portal vein or, very rarely, the ductus venosus or left gastric vein. Although our case describes a mixed type, this is very exceptional; only 9 cases of mixed TAPVR have been reported in the literature (7, 8). One could argue that connections between pulmonary veins and other structures do not usually just form after infancy by themselves, and that this connection was in our case probably there all along but was either missed or present as a "recruitable" connection. Consequently, the lesion could also be classified as a TAPVR to a common pulmonary vein draining to the coronary sinus with a persistent levocardinal vein draining to the innominate vein.

Echocardiography remains the primary diagnostic tool in pediatric cardiology. However, showing the points of entry of the four pulmonary veins into the left atrium can be challenging, related to the depth of the acoustic window.

Cardiac catheterization is itself a benchmark diagnostic tool. However, it has its limitations. It is invasive, there is exposure to ionizing radiation, and iodine contrast products are used. Angiography shows the drainage of the pulmonary veins, but indirectly, i.e., by selective pulmonary arterial catheterization, which can be poorly tolerated. It also enables the collection of numerous hemodynamic data.

MRI is now known to play a key role in pediatric cardiology (3) not only in situations where cardiac ultrasonography is technically limited but also because it enables non-invasive anatomical and functional exploration of increasing quality. Moreover, it does not have the limitations of cardiac catheterization, and its contribution, especially for direct anatomical visualization of congenital anomalies of the thoracic veins, is well established (4, 5). Some papers even claim it is superior to other techniques in the specific case of anomalous pulmonary venous return (9, 10). The various MRI pulse sequences accurately show the venous pathways and direction of flow; in particular, MRI enables unlimited study of the left retroatrial area. The Qp/Qs ratio can also be assessed by MRI. In addition, the introduction of nitinol devices enables follow-up with MRI, since they generate no significant artifacts and appear as a complete absence of signal (11).

Therapeutic catheterization is a rapidly expanding branch of pediatric cardiology, and we are currently witnessing an

extraordinary interest in transcatheter closure of numerous types of congenital defects: ductus arteriosus since the late 1980s (12), interatrial communications (13), interventricular communications (14), and coronary artery fistula (15, 16). Of course, the list of defects treated by interventional route also includes congenital thoracic venous anomalies, especially the left superior vena cava (17–19).

The treatment of total anomalous pulmonary venous return is traditionally surgical (20). However, when the anatomical malformation, even complex, is favorable, we believe it is possible, safe (absence of sternotomy and brief hospitalization) and appropriate to offer a transcatheter procedure. This case represents the first report in this type of anomaly.

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