Acta Biomed 2022; Vol. 93, Supplement 1: e2022110 DOI 10.23750/abm.v93iS1.10864

Case report

Multi-modality imaging evaluation of a rare and complex case of single ventricle physiology; the important role of cardiac MR

Cesare Mantini[1], Domenico Mastrodicasa[2], Valeria Di Mascio[1], Luca Procaccini[1], Marzia Olivieri[1], Michele Scarano[3], Fabrizio Ricci[1,4,5], Filippo Cademartiri[6]

[1]Department of Neuroscience, Imaging and Clinical Sciences, "G. d'Annunzio" University of Chieti-Pescara, Italy; [2]Department of Radiology, Stanford University School of Medicine, Stanford, CA, USA; [3]Cardiology Unit, Emergency Department, Hospital "Madonna del Soccorso", San Benedetto del Tronto, Italy; [4]Department of Clinical Sciences, Lund University, Malmö, Sweden; [5]Casa di Cura Villa Serena, Città Sant'Angelo, Pescara, Italy; [6]Department of Radiology, Fondazione Monasterio/CNR, Pisa, Italy

Abstract. Congenital heart diseases (CHD) represent a major clinical and diagnostic challenge for correct abnormality identification and subsequent successful therapy; even more challenging is following-up patient health after multiple post-interventional corrections often required in complex cardio-vascular abnormalities. We describe a multi-modality imaging evaluation of a complex congenital cardio-vascular diseases, underlining the relevance of cardiac magnetic resonance to non invasively solve some issues related to postsurgical changes. (www.actabiomedica.it)

Key words: CMR, cardiovascular magnetic resonance, cardiac magnetic resonance, single ventricle, multimodality imaging, congenital heart disease

Introduction

Congenital heart diseases (CHD) represent a major clinical and diagnostic challenge for correct abnormality identification and subsequent successful therapy; even more challenging is following up patient health after multiple post-interventional corrections often required in complex cardio-vascular abnormalities. In most of the cases a single tool is unable to show the wide spectrum of congenital heart disease and the result of multiple post-interventional corrections attempting to restore an almost normal physiology. Furthermore, multiple post-interventional corrections raise up the possibilities of different, and often very particular, collateral effects.

Showing this case, we'd like to highlight the importance of a multi-modality approach; further more we'd like to underline the relevance of cardiac magnetic resonance (CMR) for assessment of complex anatomy, function and post-surgical changes, in particular in excluding a graft occlusion with a non-invasive tool.

Case presentation

We report a case of a 28-year-old woman born with a complex cyanotic CHD characterized by: 1) dextrocardia in the absence of situs viscerum inversus, 2) common atrium with insufficient atrio-ventricular valve, 3) large ventricular septal defect resulting in effective single ventricle, 4) displacement of great arteries with double outlet left ventricle, 5) pulmonary stenosis (Figure 1), 6) absence of inferior vena cava (IVC) with azygous continuation and polysplenia (Figure 2).

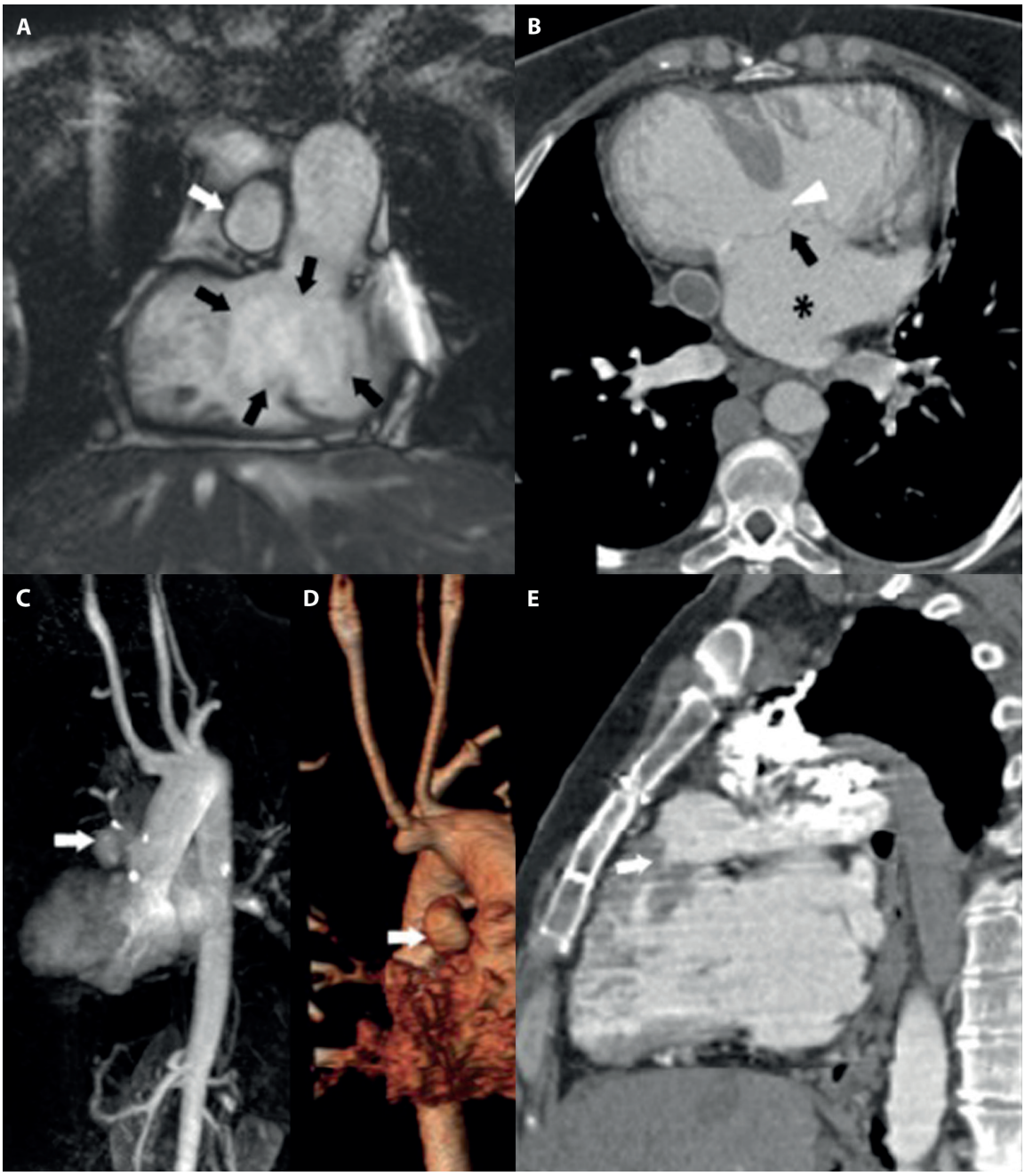

Figure 1. Coronal b-SSFP image (a) shows the sing le ventricle and the common atrioventricular valve (black arrows). Axial CT scan (b) shows the large ventricular septal defect (arrowhead) and the common atrium (*) with common atrioventricular valve (black arrow). Maximum intensity projection (MIP) (c) and volume rendering techniques (VRT) (d) reconstruction of CE-MRA and sagittal CT scan (e) show the double outlet left ventricle with ascending aorta and origin of pulmonary artery (white arrow in a,c,d,e).

Moreover, patient exhibited Siewert syndrome, i.e., chronic sinusitis, bronchiectasis and dextrocardia (Liver and spleen were normally placed). "Single ventricle physiology" is seen in a variety of complex CHD; a single ventricle supports both systemic and pulmonary circulation. It is amenable to limited therapeutic options, carrying significant morbidity, mortality and often an abbreviated life span. At age of 16 months,

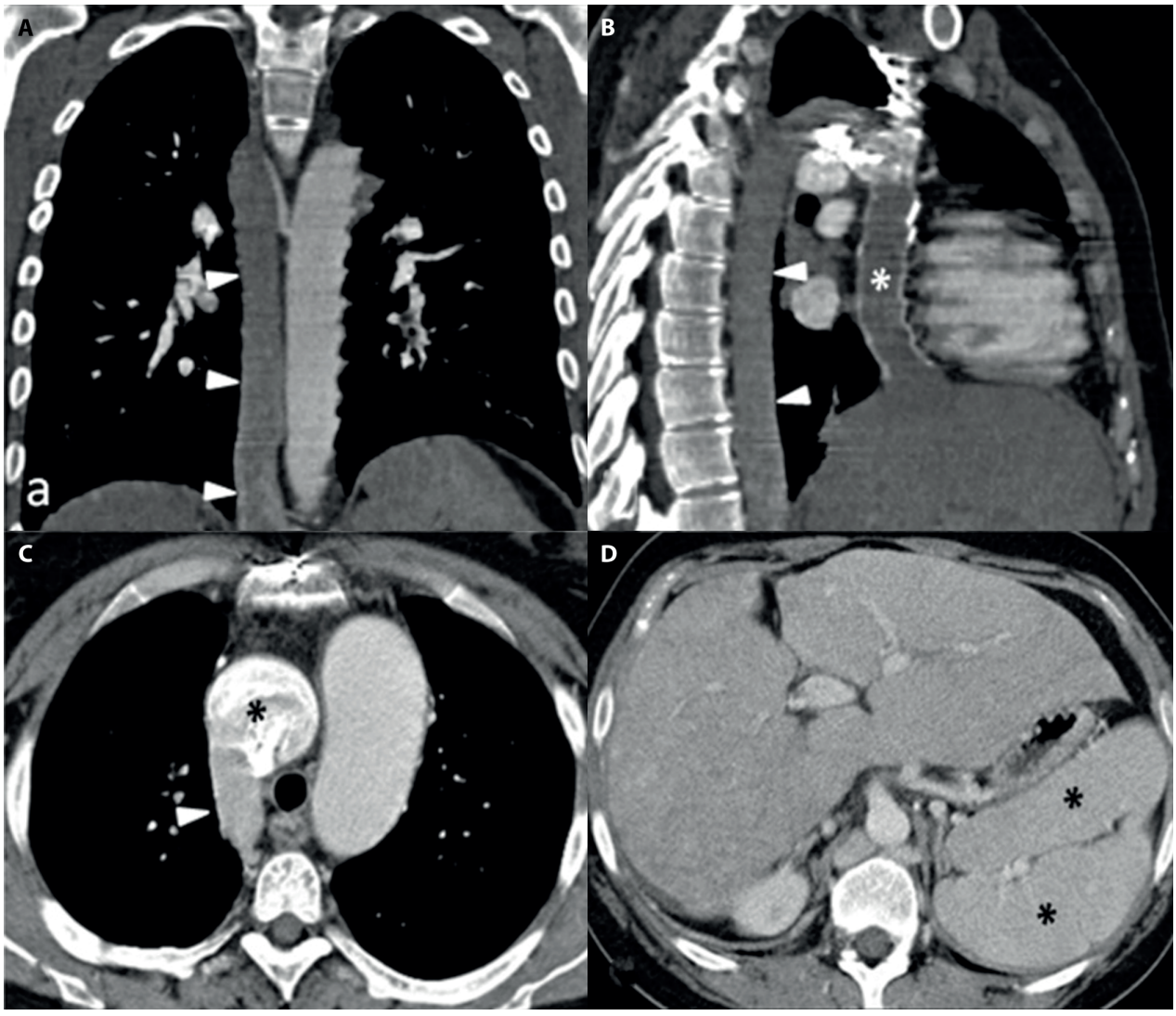

Figure 2. Coronal (a), sagittal (b) and axial (c) CT scans show the presence of azygous continuation (arrowhead) to an enlarged superior vein cava (* in c). Upper abdominal CT scan (d) depicts the presence of two spleens (*).

as first step to deal with cyanosis, patient was treated with a right Blalock-Taussig shunt consisting in the anastomosis of right subclavian artery to right pulmonary artery, resulting in increased low oxygenating blood delivered to lungs (1). At age of 2 years, patient had Kawashima procedure, applied to CHD with single ventricle physiology and interrupted IVC (2); the superior vena cava (SVC) was anastomosed to right pulmonary artery creating a superior cavopulmonary connection for directing body venous flow to lungs, except for hepato-cardiac venous and coronary sinus flow. At the age of 10 years, patient underwent a surgical procedure with the aim of decompressing venous flow coming from liver; a 18 mm Goretex graft was interposed between hepatic veins and right pulmonary artery (Figure 3).

At the age of 28 years, patient was referred to our Department for acute respiratory failure investigation. Postero-anterior chest radiographs showed an ill-defined patchy opacity in the mid portion of the left lower pulmonary lobe, projecting at the level of the left border of cardiac silhouette; then, for unilateral lower limb pain and high D-dimer levels, she was referred for multidetector computed tomography (CT) pulmonary angiography with the suspicion of pulmonary embolism. The CT pulmonary angiogram revealed no

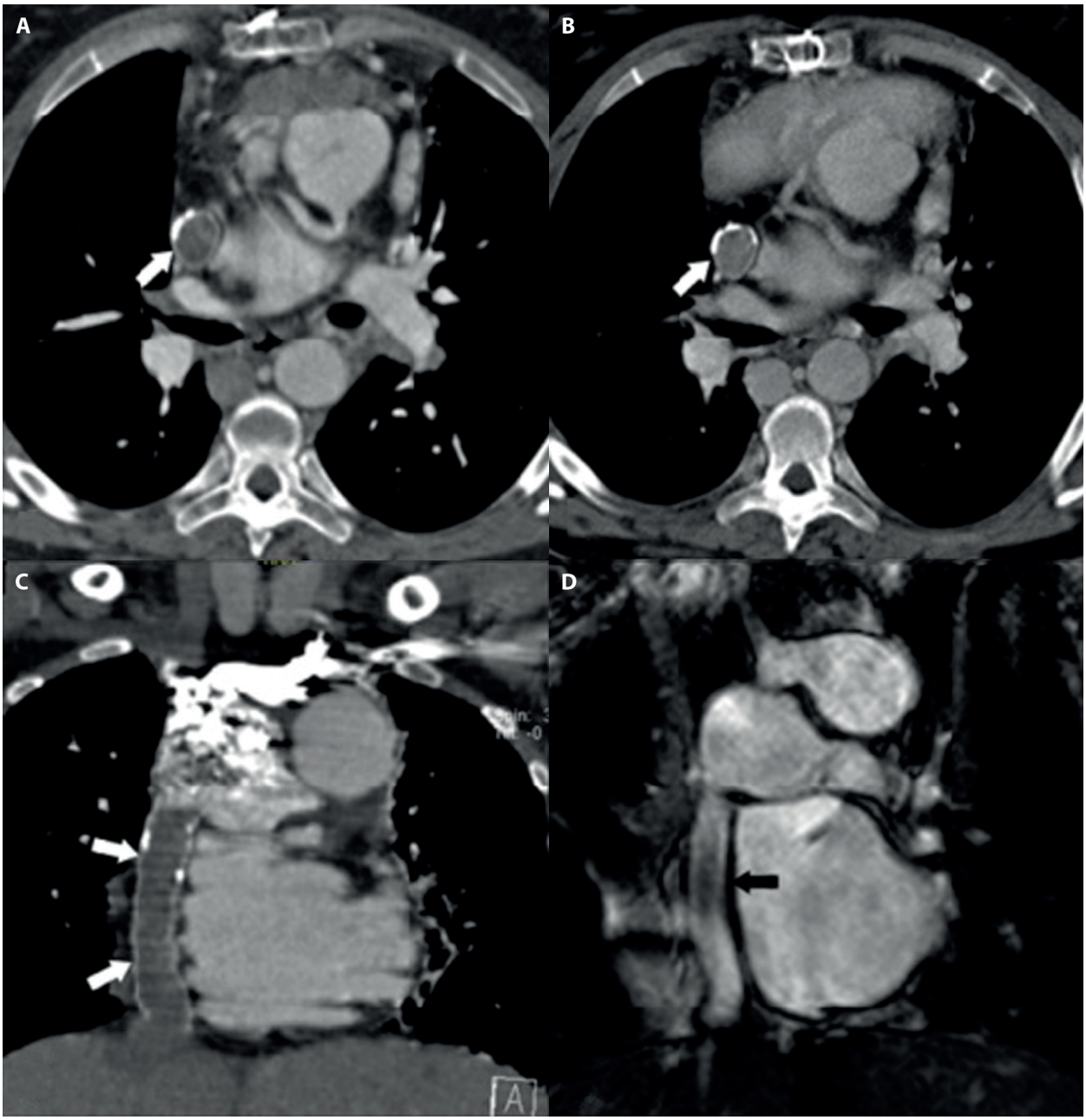

Figure 3. CT scans acquired during arterial (a,c) and late phase (b) did not show a clear opacization of the prosthesis (white arrows). Bright-blood coronal b-SSFP image (d) shows the presence of a signal void within the prosthesis (black arrow).

evidence of pulmonary embolism but it was not able to clearly show opacification of Goretex graft during arterial and late phase, thus raising the suspicion of graft thrombosis (Figure 3a, c); in addition, axial-CT images, with lung window settings, revealed the presence of multiple cylindrical bronchiectasis in the lingula and in the left lower lobe with retained secretions in the lumen of the most severe bronchiectasis associated to diffuse bronchial wall thickening. These findings correlated retrospectively to the opacity seen on chest radiograph. To better evaluate graft patency and for a comprehensive evaluation of complex anatomy, patient was referred for CMR (3). Balanced steady-state free precession (b-SSFP) and 2D breath-hold cine phase

contrast (PC) sequences showed the presence of an eccentric systolic atrio-ventricular valve regurgitant jet (quantitative assessment: 30% regurgitant fraction). b-SSFP (Figure 3d), and mainly PC sequences (Figure 4), acquired in a coronal and in a perpendicular plane through the prosthesis, clearly showed the presence of flow within the graft thus non invasively excluding graft thrombosis.

To complete the assessment of the complex vascular anatomy, 3D contrast-enhanced MR angiographic (CE-MRA) sequence was planned. A Coronal 2D-BolusTrak sequence, able to display contrast bolus arrival, was acquired for triggering 3D-high resolution CE-MRA acquisition start. Tracking sequence showed an unexpected very early enhancement of prosthesis lumen. The contrast bolus arrived firstly in the SVC and

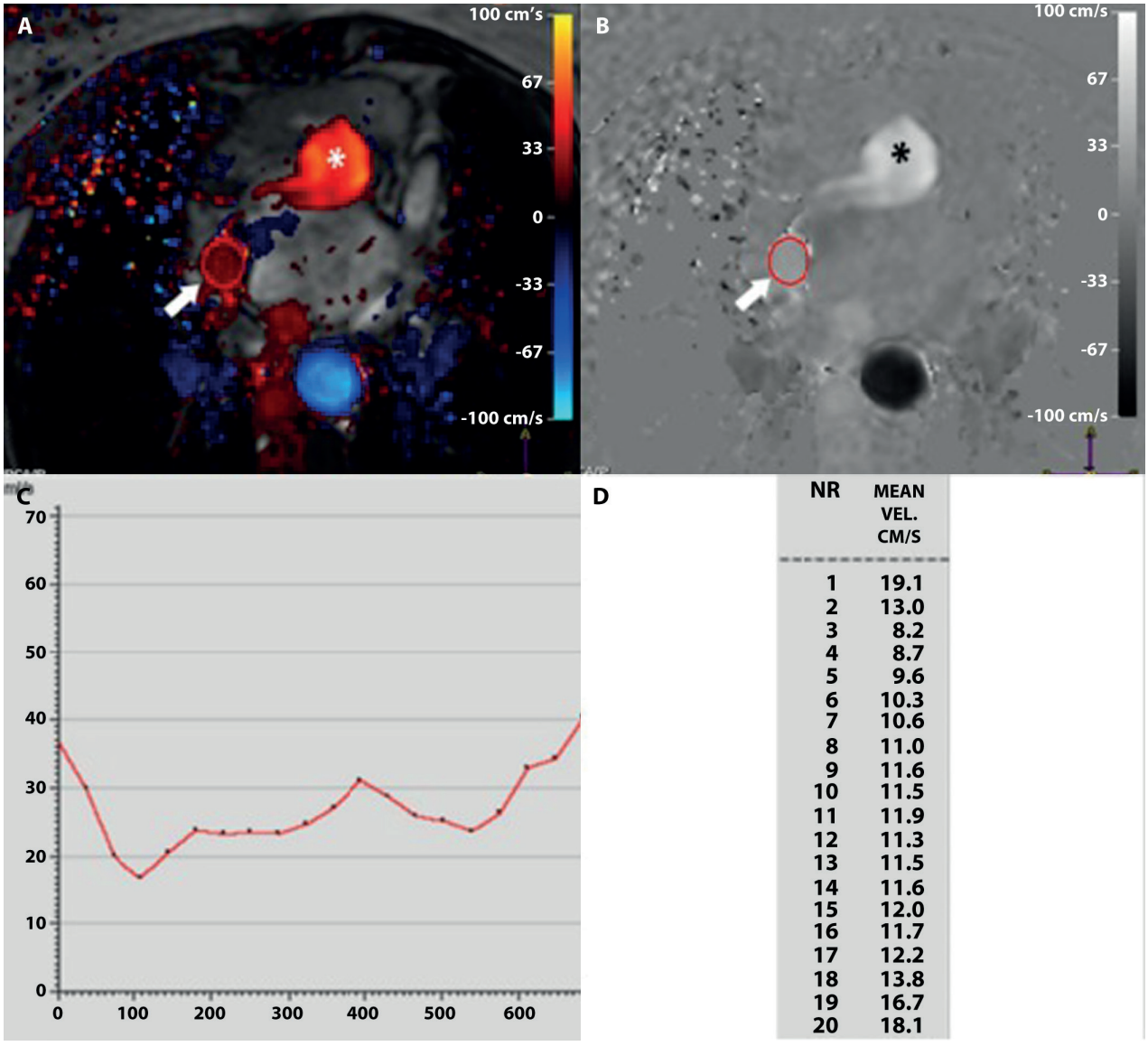

Figure 4. Axial PC sequence with magnitude (a) and phase image (b) shows the presence of flow within the Goretex graft (arrow) directed cranially (red in a and white in b) like the ascending aorta (*) and opposite to the descending aorta (blue in a and black in b). This quantitative flow images were analyzed drawing regions of interest (ROI) on each of the 20 phases of magnitude images and corresponding flow map (c) and mean velocities (d) were obtained.

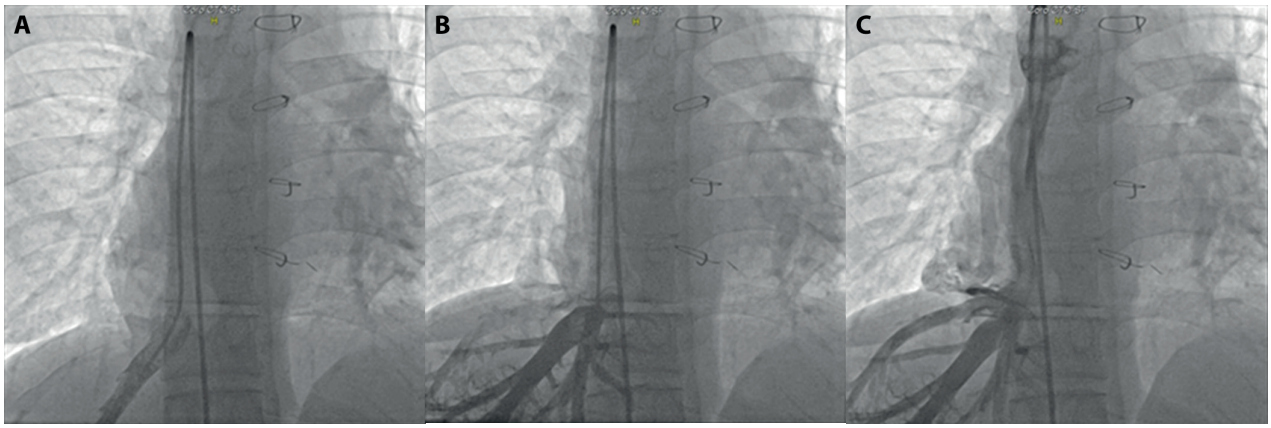

Figure 5. Cardiac catheterization confirms the presence of a slow venous flow from the hepatic veins to the right pulmonary artery.

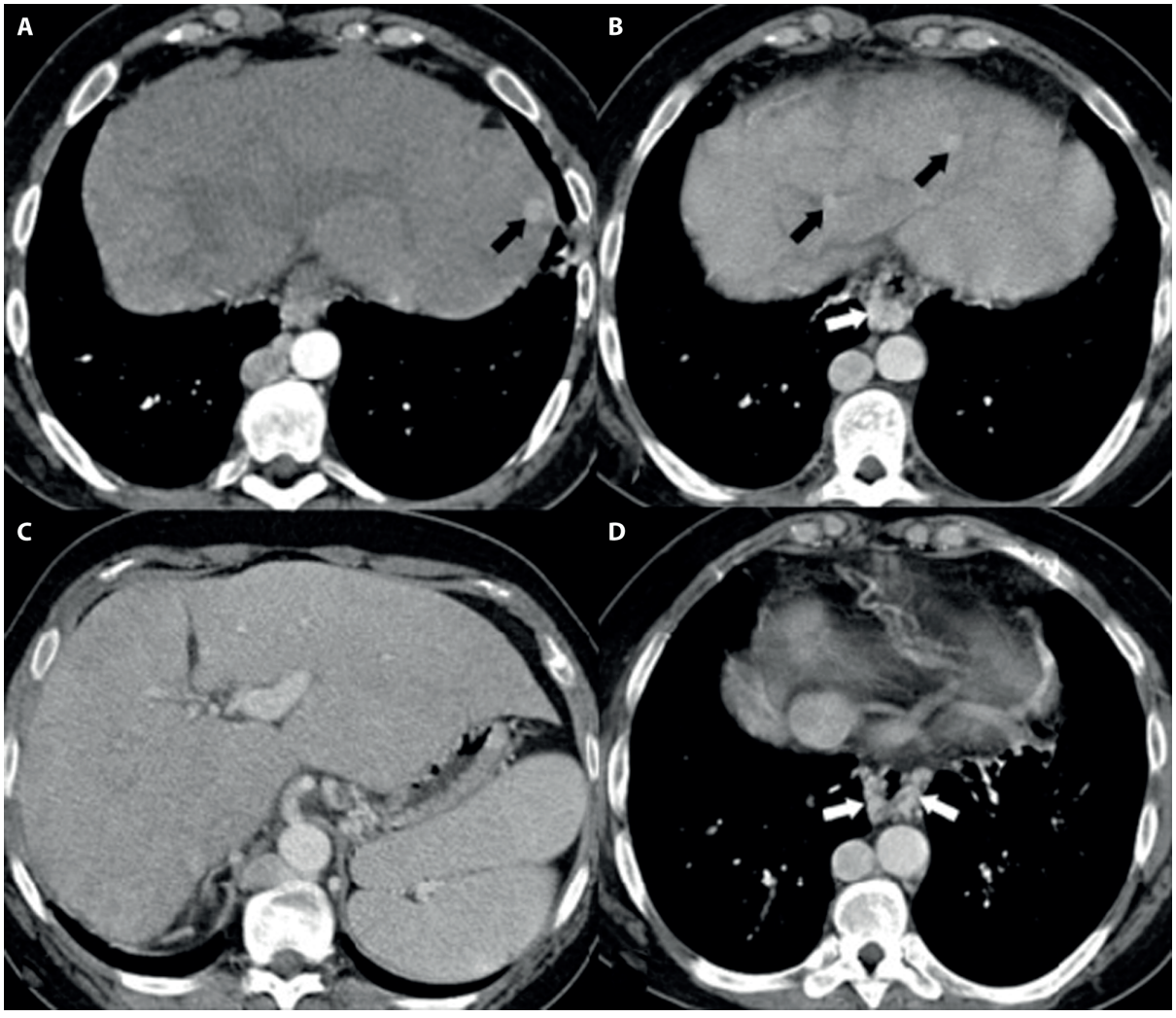

Figure 6. Upper abdominal CT scans show hepatomegaly with caudate lobe hypertrophy, patchy pattern of parenchymal enhancement, hypervascular nodules (nodular regenerative hyperplasia) (black arrows) and esophageal varices (white arrows).

then flowed inside the prosthesis up to the hepatic veins and against venous flow; most probably the low pressure venous flow within the prosthesis was exceeded by the pressure applied by contrast injection system push. The unexpected early enhancement of prosthesis lumen, clearly showed by bolus tracking, did not permit us to catch the right instant for 3D-CE-MRA acquisition thus missing an optimal opacification on this sequence. Six months later, patient underwent cardiac catheterization to evaluate atrial and ventricular pressures and oxygen saturation; this examination invasively confirmed the presence of slow venous flow from hepatic veins to right pulmonary artery via the graft (Figure 5).

In addition, an upper abdominal CT scan (Figure 6) revealed signs of Budd-Chiari syndrome (hepatomegaly, caudate lobe hypertrophy, patchy pattern of parenchymal enhancement and nodular regenerative hyperplasia) related to the absence of the IVC with compromised flow in the hepatic veins.

Discussion

Integration of different techniques is often crucial for a correct diagnosis in complex cases such as a CHD. Over the past two decades, there has been a marked increase in the use of CMR for the anatomical and functional evaluation and post-procedural follow-up of patients with CHD (4). It complements echocardiography, provides a non-invasive alternative to x-ray angiography, avoids the ionizing radiation exposure of CT, and overcomes many of the limitations of these modalities. Besides tissue characterization and clarification of anatomy and function (including anomalous vessels, connections, shunts, stenoses, abdominal situs and possible polysplenia) the strengths of CMR include versatility and comprehensive cardiac, mediastinal and upper abdominal coverage (5) using stacks of contiguous transaxial cine images and dynamic contrast-enhanced angiography, and the relatively accurate measurements of ventricular function and volume flow. These are particularly useful in the assessment and follow-up of adults after Fontan operations in which the systemic and pulmonary vascular beds are surgically connected in series with one another downstream of the single effective ventricle, thus eliminating shunting at the cost of a critically elevated systemic venous pressure that maintains flow through the lungs. Patency of the cavo pulmonary pathways, ventricular and valvular function and possible causes of shunting require a periodic and noninvasive evaluation through CMR.

Conflict of interest: Each author declares that she or he has no commercial associations (e.g. consultancies, stock ownership, equity interest, patent/licensing arrangement etc.) that might pose a conflict of interest in connection with the submitted article

References

1. Blalock A, Taussig HB. The surgical treatment of malformations of the heart in which there is pulmonary stenosis or pulmonary atresia. JAMA 1945; 128:189-202.
2. Kawashima Y, Kitamura S, Matsuda H. Total cavopulmonary shunt operation in complex cardiac anomalies. A new operation. J Thorac Cardiovasc Surg. 1984; 87(1):74-81.
3. Pamela K. Woodard, Sanjeev Bhalla, et al. Cardiac MRI in the management of congenital heart disease in children, adolescents, and young adults. Curr Treat Options Cardiovasc Med. 2008; 10:419–424.
4. Marcotte F, Poirier N, Pressacco J, et al. Evaluation of adult congenital heart disease by cardiac magnetic resonance imaging. Congenit Heart Dis. 2009; 4:216–30.
5. Mantini C, Mastrodicasa D, Bianco F, et al. Prevalence and Clinical Relevance of Extracardiac Findings in Cardiovascular Magnetic Resonance Imaging. J Thorac Imaging. 2019 Jan;34(1):48-55.

Correspondence
Received: 1 October 2020
Accepted: 28 October 2020
Prof. Dr. Filippo Cademartiri, MD, PhD, FESC
Chairman of Radiology - Fondazione Toscana "Gabriele Monasterio" - Heart Center
Ospedale "San Cataldo" di Pisa - Area della Ricerca CNR (Consiglio Nazionale delle Ricerche)
via Giuseppe Moruzzi, 1 - 56124 - Pisa (PI) – Italy
Tel. +39 +39 050 621 2817 (direct)
E-mail: fcademartiri@ftgm.it
www.monasterio.it