JACC: CASE REPORTS VOL. 4, NO. 10, 2022

CASE REPORT

ADVANCED

CLINICAL CASE

Multimodality Approach to a Complex Scimitar Syndrome

How Advanced Diagnostics Can Guide Therapeutic Strategies

Alberto Clemente, MD,[a] Gaia Viganò, MD,[b] Luigi Festa, MD,[c] Ettore Remoli, MD,[a] Chiara Marrone, MD,[c] Duccio Federici, MD,[b] Vitali Pak, MD,[b] Dante Chiappino, MD,[a] Giuseppe Santoro, MD,[c] Lamia Ait-Ali, PhD[d]

ABSTRACT

We report an unusual association of scimitar syndrome with right diaphragmatic hernia, left-sided aortic arch with "aberrant right subclavian artery" in a 2-year-old boy who underwent stepwise transcatheter occlusion of a significant aortopulmonary collateral followed by surgical treatment for the repair of the diaphragmatic hernia and esophageal compression. (**Level of Difficulty: Advanced.**) (J Am Coll Cardiol Case Rep 2022;4:596-603) © 2022 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

HISTORY OF PRESENTATION

A 2-year-old 15.3-kg boy was referred to our institution with a suspect of scimitar syndrome (SS) and right supradiaphragmatic mass.

On admission, at physical examination, a weak breathing sound was observed in the upper right chest and a thick breathing sound in the left chest. No pathologic cardiac murmurs were audible. He had normal systemic pressure with no hepatomegaly.

LEARNING OBJECTIVES

- To understand the role and the limitations of the different imaging modalities used to evaluate complex cases of SS in order to choose the best treatment option.

PAST MEDICAL HISTORY

He had history of recurrent episodes of pulmonary infections and respiratory distress, dysphagia for semisolid food.

DIFFERENTIAL DIAGNOSIS

Differential diagnosis included pulmonary sequestration, congenital cystic adenomatoid malformation, bronchogenic cysts, infection, tumor, round atelectasis, and bochdalek hernia.

From the [a]Department of Radiology, CNR (National Council of Research)/Tuscany Region "Gabriele Monasterio" Foundation (FTGM), Massa, Italy; [b]Pediatric Cardiac Surgery, CNR (National Council of Research)/Tuscany Region "Gabriele Monasterio" Foundation (FTGM), Massa, Italy; [c]Pediatric Cardiology and GUCH Unit, CNR (National Council of Research)/Tuscany Region "Gabriele Monasterio" Foundation (FTGM), Massa, Italy; and the [d]Institute of Clinical Physiology, National Research Council, Pisa, Italy.

The authors attest they are in compliance with human studies committees and animal welfare regulations of the authors' institutions and Food and Drug Administration guidelines, including patient consent where appropriate. For more information, visit the Author Center.

Manuscript received November 22, 2021; revised manuscript received March 17, 2022, accepted March 28, 2022.

ISSN 2666-0849 https://doi.org/10.1016/j.jaccas.2022.03.027

INVESTIGATIONS

Chest x-ray showed cardiac dextroposition, bilateral increased pulmonary vascular markings, and a rounded opacity of the right lower hemithorax just adjacent to the diaphragm edges.

Abdominal ultrasound demonstrated a right-sided diaphragmatic hernia.

Transthoracic echocardiogram demonstrated dextroposition of the heart with good biventricular function, and an abnormal right pulmonary vein with high velocity/turbulent flow draining into the inferior vena cava just above the cavoatrial junction. A persistent left superior vena cava and a dilated coronary sinus, mild dilated pulmonary trunk, and an hypoplastic right pulmonary artery were also reported.

Cardiac magnetic resonance (CMR) was performed under deep sedation on a 3-Tesla scanner (Ingenia; Philips Medical Systems) according to standard clinical protocols including 4D-flow CMR sequences. To confirm small vessel abnormalities, and pulmonary and liver relationship, a cardiac computed tomography (CCT) was also performed under deep sedation.

CMR and CCT clearly illustrated the diagnosis of a very rare association of a total anomalous right pulmonary venous return with the following anomalies:

- Dextroposition of the heart with right pulmonary lung hypoplasia (**Figure 1**)
- Right congenital diaphragmatic hernia (CDH) with ectopic liver and hepatopulmonary fusion (HPF) (**Figure 2**, Video 1)
- Persistent left superior vena cava draining into dilated coronary sinus (**Figure 3**)
- Pulmonary venous return draining through a scimitar vein, into the inferior vena cava just above the cavoatrial junction (**Figure 4**, Video 1)
- Hypoplastic right pulmonary artery with reduced right pulmonary flow (14% of the pulmonary flow) (**Figure 5**)
- A large aortopulmonary collateral (APC) from the celiac trunk supplying the right lung coursing around the site of HPF (**Figure 6**)
- Left-sided aortic arch with aberrant right subclavian artery (ARSA) and esophageal compression (**Figure 7**)
- Multiple contiguous vascular arterial and venous structures, both pulmonary and hepatic, seen at the level of the HPF
- Anomalous tracheobronchial tree suggesting for bilobate right hypoplastic lung

MANAGEMENT

After multidisciplinary discussion, the patient underwent a cardiac catheterization, via femoral accesses. Baseline hemodynamic analysis showed

ABBREVIATIONS AND ACRONYMS

APC = aortopulmonary collateral

ARSA = aberrant right subclavian artery

CCT = cardiac computed tomography

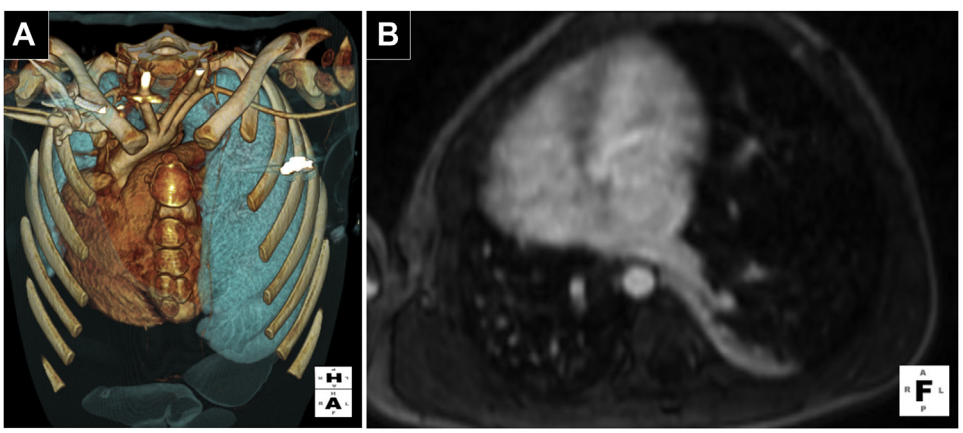
CDH = congenital diaphragmatic hernia

CMR = cardiac magnetic resonance

HPF = hepatopulmonary fusion

SS = scimitar syndrome

FIGURE 1 Preprocedural Imaging of the Heart and Lungs Relationship



Virtual reality-computed tomography scan **(A)** and cardiac magnetic resonance axial view **(B)** showing right lung hypoplasia and dextroposition of the heart.

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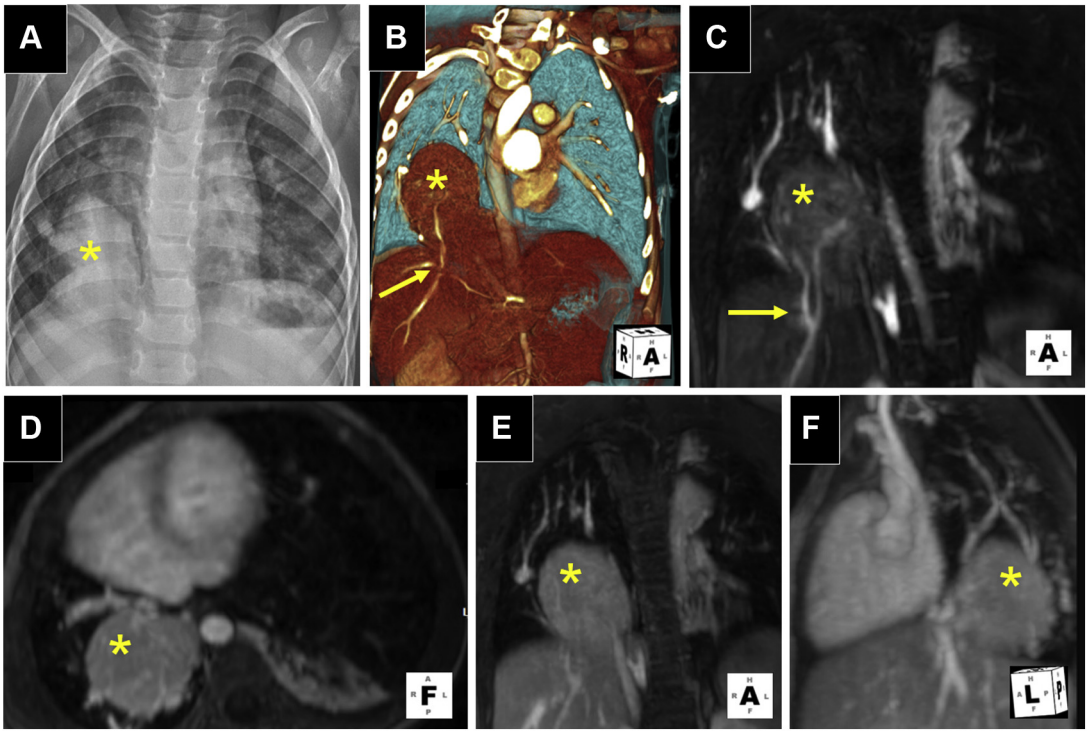
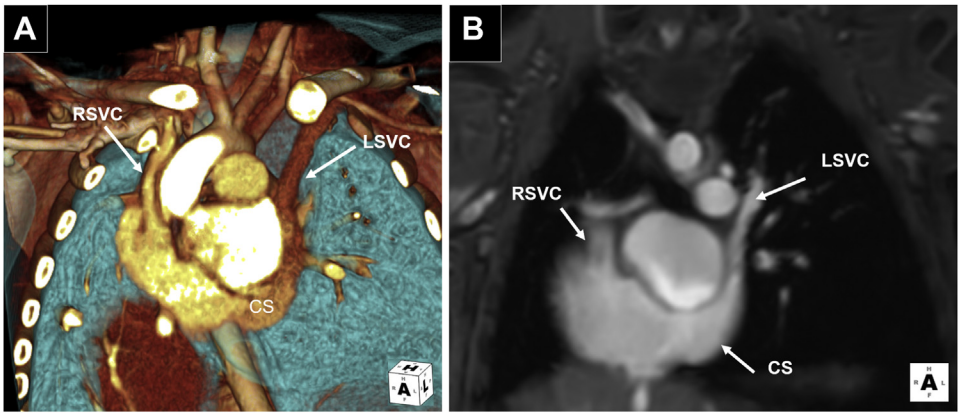
FIGURE 2 Preprocedural Imaging of the Liver and Lung Relationship



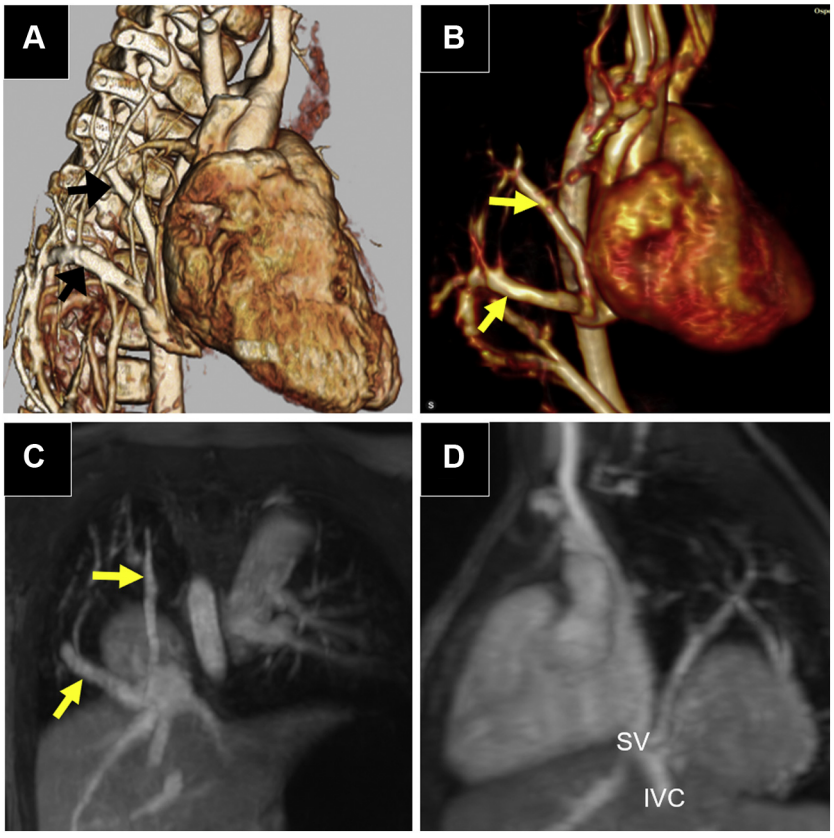
Chest x-ray **(A)**, virtual reality-cardiac computed tomography cut-plane coronal view **(B)**, and cardiac magnetic resonance volumetric angiography **(C to F)** showing congenital diaphragmatic hernia with ectopic liver **(asterisk)** and corresponding portal vein **(arrow)** in the right thorax.

FIGURE 3 Preprocedural Imaging of Superior Venae Cavae



Virtual reality-cardiac computed tomography cut-plane oblique view **(A)** and SSFP-cardiac magnetic resonance sequence **(B)** showing persistent left superior vena cava (LSVC) to a dilated coronary sinus (CS). RSVC = right superior vena cava.

FIGURE 4 **Preprocedural Imaging of the Scimitar Vein**



Virtual reality-cardiac computed tomography cut-plane oblique view **(A)**, cardiac magnetic resonance angiography volumetric reconstruction **(B)**, and multiplanar reconstructions **(C and D)** showing the scimitar vein (SV) draining the right lung veins **(arrows)** into the inferior vena cava (IVC) just above the cavoatrial junction.

severe left-right shunt (pulmonary-systemic flow ratio [Qp/Qs] 2.06) and slightly increased pulmonary pressure with normal pulmonary resistance. X-ray angiography confirmed CMR and CCT findings.

Embolization of the APC was performed using multiple vascular plugs (Figure 8) without signs of pulmonary infarction.

After 10 days, he underwent a combined 2-stage surgery to address the associated malformations.

Right thoracotomy was performed through the sixth intercostal space to repair the CDH with Gore-Tex mesh. Numerous abnormal arterial collaterals to the right lower lobe were sectioned with clips to allow mobilization of the ectopic liver tissue. There was no distinct plane between the lung and the liver; therefore, an electrocautery was used to separate the structures. Then the esophageal compression was released by simple side-biting division of the ARSA at the junction with the aorta and reimplantation to the right carotid artery through supraclavicular approach.

The postoperative course was uneventful.

DISCUSSION

Few reports have described isolated cases of SS and CDH.[1,2]

One case of an SS associated with diaphragmatic hernia, ARSA, and urinary anomalies is reported; the patient underwent classic surgical repair.[3]

The combination of anomalous venous drainage of the right lung, right CDH and HPF was also reported in 1 patients who did not survive the surgery.[4]

Although SS should be suspected at chest x-ray, usually echocardiography is considered the first-line tool for the diagnosis of congenital heart disease, even if limited in evaluating extracardiac anomalies.

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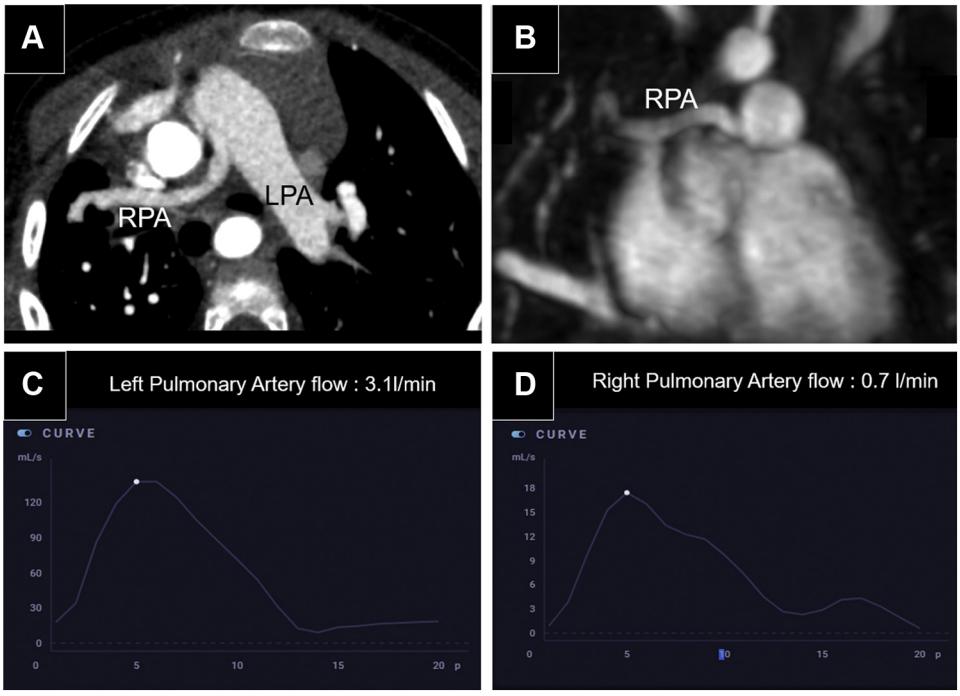
FIGURE 5 Preprocedural Anatomic and Functional Pulmonary Arteries Evaluation



Right pulmonary artery (RPA) hypoplasia and right lung hypoperfusion. **(A)** Axial computed tomography image. **(B)** Cardiac magnetic resonance angiography volumetric reconstruction. **(C and D)** Pulmonary arteries flow. LPA = left pulmonary artery.

Other noninvasive modalities (eg, CCT, CMR) are required to confirm the diagnosis and evaluate other potential associated anomalies, as in our reported case.[5]

The management of SS is challenging. Accurate anatomic evaluation, including associated extracardiac anomalies and the quantification of the shunt is mandatory besides the clinical presentation.

When indicated, active management usually includes reimplantation of the anomalous vein into the left atrium baffling of the anomalous vein, and in selected cases catheter rehabilitation of the stenotic scimitar vein.[6]

In the present case, the transcatheter occlusion of the systemic arterial supply to the lung followed by surgical repair of the CDH was decided with a favorable outcome.

Recently, multiple reports have shown that transcatheter intervention (including embolization of the APC and closure of cardiac defects) may improve symptoms, decrease pulmonary arterial pressure, and postpone or even eliminate the need for surgical correction.[7,8]

In our case, after the embolization of the APC, the Qp/Qs almost normalized and no cardiac surgery was performed.

FOLLOW-UP

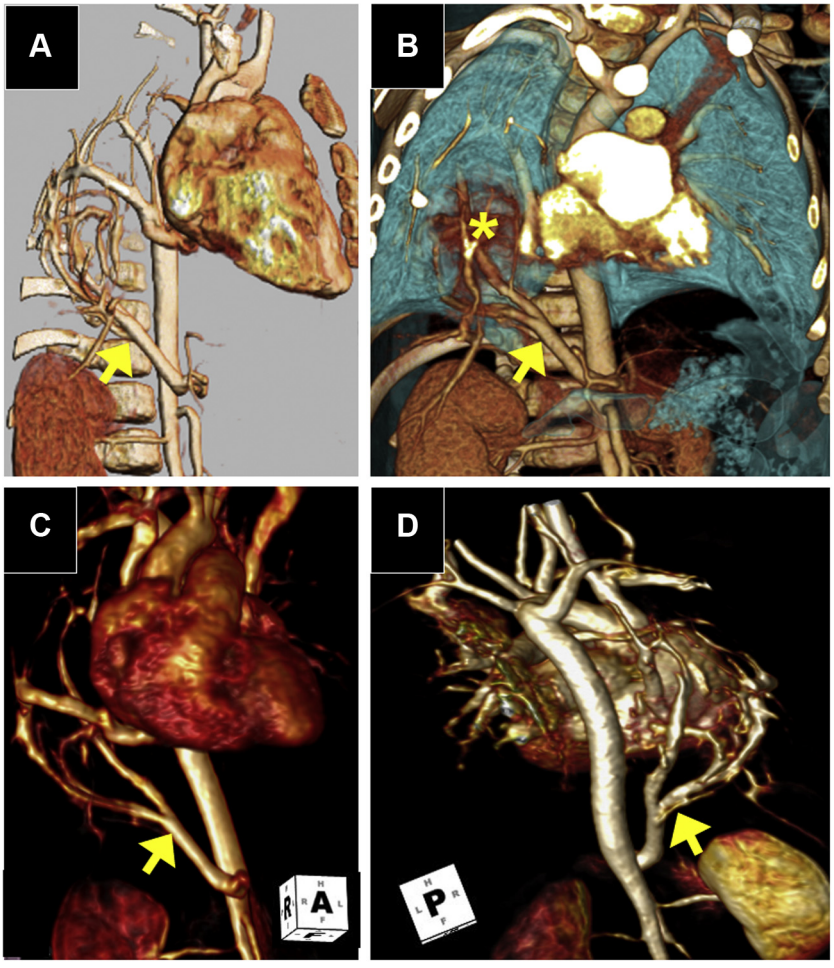
One month after the procedure, the lung was expanded with no recurrence of the diaphragmatic hernia and echocardiogram revealed normal right ventricular pressures and good biventricular systolic function. No more dysphagia.

CONCLUSIONS

SS is a rare congenital complex anomaly with multiple associated findings and a wide variety of presentations. We report a unique case of a 2-year-old boy with SS, right CDH, and retroesophageal ARSA who underwent successful transcatheter embolization of the APC followed by reimplantation of ARSA to release the esophageal compression and surgical repair of the CDH with favorable outcome.

This case highlights the importance of assessing anatomy and associated findings in patients with

FIGURE 6 **Preprocedural Hepato-Pulmonary Fusion Evaluation**



A large aortopulmonary collateral **(arrow)** arising from the celiac trunk for the right lung surrounding the hepatopulmonary fusion **(asterisk)**. **(A and B)** Virtual reality-computed tomography angiography cut-plane oblique view. **(C and D)** Virtual reality-cardiac magnetic resonance angiography.

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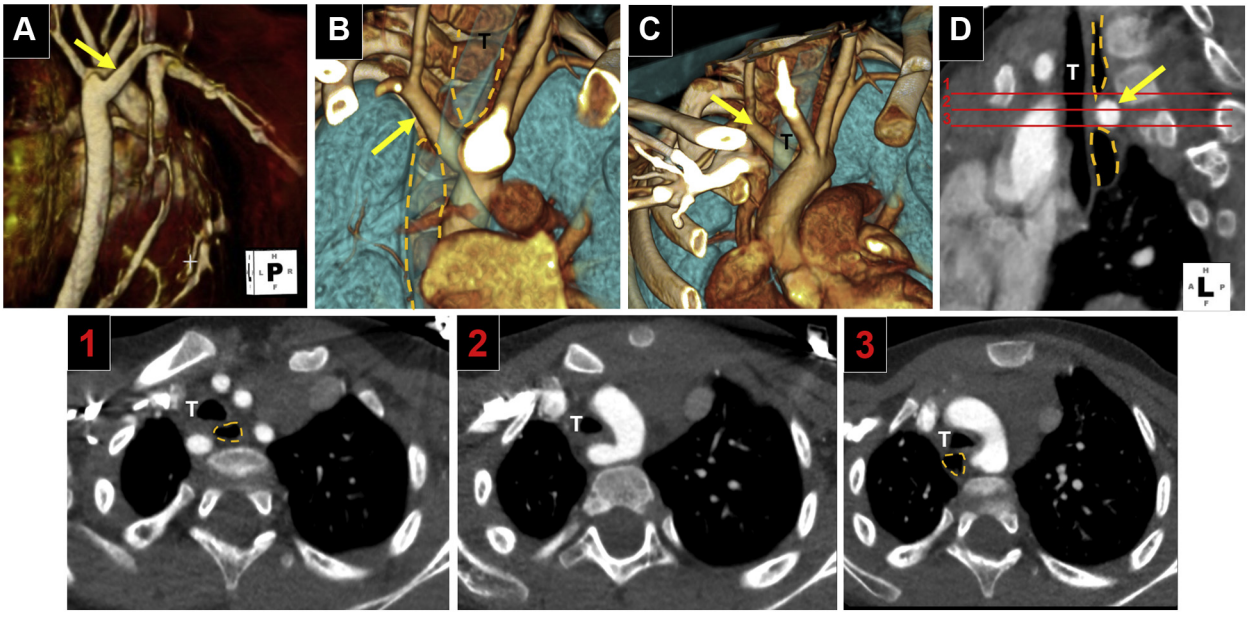
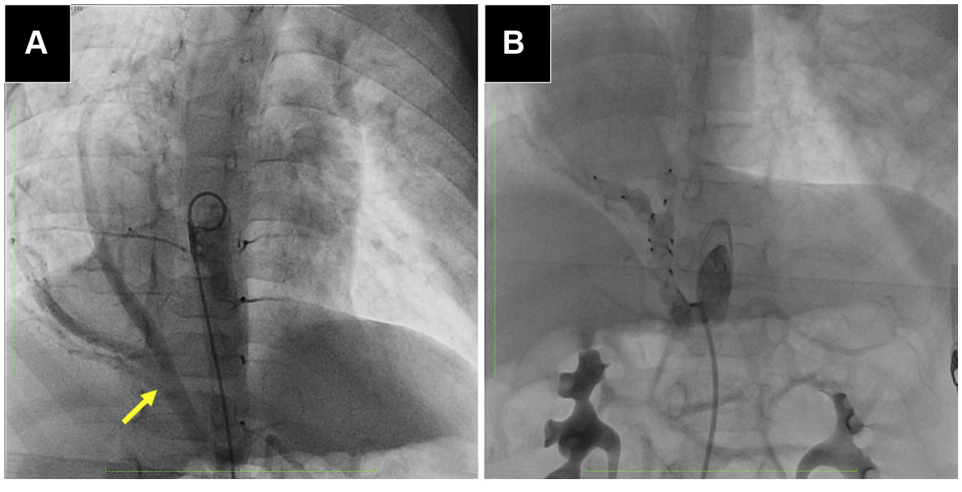
FIGURE 7 Preprocedural Imaging of the Aberrant Right Subclavian Artery



(A) Posterior virtual reality-cardiac magnetic resonance angio view showing the retroesophageal aberrant right subclavian artery **(arrow)**. **(B and C)** Anterior and oblique virtual reality-cardiac computed tomography view showing aberrant right subclavian artery **(arrow)** with normal tracheal (T) while the esophagus is compressed **(dotted yellow line)**. **(D)** Cardiac computed tomography lateral view showing the esophagus compressed between the trachea (T) and the aberrant right subclavian artery **(arrow)**.

FIGURE 8 Angiographic Evaluation and Treatment



X-ray angiography confirmed the presence of a large aortopulmonary collateral **(yellow arrow)** before **(A)** and after **(B)** endovascular exclusion with multiple vascular plugs.

SS, as this influences the treatment options and outcomes.

FUNDING SUPPORT AND AUTHOR DISCLOSURES

The authors have reported that they have no relationships relevant to the contents of this paper to disclose.

ADDRESS FOR CORRESPONDENCE: Dr Gaia Viganò, CNR/Tuscany Region "Gabriele Monasterio" Foundation, Via Giuseppe Moruzzi, 1, 56124 Pisa PI, Italy. E-mail: gaiavigano85@yahoo.com. Twitter: @gaiavigano85, @Alb3rtoClemente.

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KEY WORDS aberrant right subclavian artery, anomalous pulmonary venous return, congenital heart disease, diaphragmatic hernia, scimitar syndrome

APPENDIX For a supplemental video, please see the online version of this paper.