Chronic Occlusion of the Superior Vena Cava Resulting in Cyanosis in an Adult
Unusual Case Highlighting the Value of Collaboration Between Adult and Congenital Cardiology Services

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A 42-year-old man presented to his local hospital with sepsis. He was cyanotic and had finger clubbing with oxygen saturation of 85% to 90%. A computed tomographic pulmonary angiogram led to an initial diagnosis of left-sided superior vena cava and partial anomalous pulmonary venous drainage. He had been generally well but always had limited exercise capacity (New York Heart Association class 2) and mild cyanosis and finger clubbing had been noted previously but had never been investigated. He was reviewed at an adult congenital center, and his symptoms were thought to be inconsistent with partial anomalous pulmonary venous drainage and left-sided superior vena cava. An echocardiogram raised suspicion for abnormal drainage of the SVC, and bubble echocardiography showed early and complete opacification of the left atrium and left ventricle after contrast injection in the left arm. There was late but significant filling of the right atrium and right ventricle, but this was less dense, signifying a right to left shunt. A cardiac magnetic resonance imaging demonstrated complete occlusion of the right SVC (Figure [A]). The brachiocephalic vein and other head and neck veins drained via tortuous venous collaterals to the left upper and right upper pulmonary veins (Figure [B]; Movie I in the Data Supplement). The estimated Qp:Qs was 0.7:1.0 because of right to left shunt.

The initial incorrect diagnosis of partial anomalous pulmonary venous drainage was based on imaging, showing a connection between the left atrium and the innominate vein. Magnetic resonance imaging was helpful to show the exact anatomy, and that flow was in fact from the systemic vein to the left atrium (the reverse flow direction of partial anomalous pulmonary venous drainage). The patient had no previous procedures and no previous medical history, hence the cause of SVC obstruction was unknown. Anticoagulation was not advised as a primary therapy because of the chronic nature of occlusion. Elective SVC recanalization and stenting was attempted.

At cardiac catheterization (Movie II in the Data Supplement), initial attempts to recanalize the SVC were unsuccessful using chronic total occlusion wires, and the SVC stump was finally perforated from below with the sharp end of 0.018” Terumo wire (Terumo Interventional Systems) with a preformed curve. The Terumo wire was exchanged for an 0.014” Fieddler wire, which was snared with 20-mm snare through a 6F snare catheter in the innominate vein. A venovenous circuit was created, and a Super Stiff Amplatz (Boston Scientific) 260-cm wire was exteriorized. The occluded segment of SVC was predilated with a 14-mm Tyshak Balloon (NuMED Inc, Hopkinton, NY) and subsequently with a 12-mm Atlas Balloon (Atlas PTA Catheter-Bard PV-BARD). Two 45-mm covered Cheatham-Platinum Stents (NuMED Inc) premounted on a 14-mm BIB were sequentially implanted via a 12F Mullins sheath (Medtronic). Both stents were post dilated with a 14-mm Atlas Balloon (Atlas PTA Catheter-Bard PV-BARD) to relieve the waist, and then the right atrial end was flared with a Coda balloon (Cook Medical). Also, the top end of the stent was molded toward the innominate vein with 8- and 10-mm Powerflex Balloons (Cordis). Covered stents were used because there was a suspicion of extravastion. Final angiography showed a satisfactory result with restoration of physiological flow and an increase in the oxygen saturation to 96%. The patient was commenced on warfarin therapy to prevent thrombosis. On follow-up evaluation 6 months later, the patient reported being more energetic (New York Heart Association class 1) and had an oxygen saturation of 95%. Three months later, a repeat magnetic resonance imaging scan showed a patent SVC and regression of the pulmonary venous collaterals. (Figure [C] and [D]), as well as improvement of the estimated Qp:Qs to 0.9:1 from 0.7:1.

Chronic occlusion of the SVC is commonly associated with the development of collateral veins to theazygos and hemiazygous systems and internal thoracic and paravertebral veins.1,2 In this case, systemic-to-pulmonary venous
collaterals had developed, draining the head and neck vessels to the left atrium. Although patients with congenital heart disease may develop SVC obstruction because of a variety of reasons and individuals with transvenous pacing may develop SVC obstruction or occlusion related to the leads, there are few reports of symptomatic SVC occlusion in adults who have not undergone previous cardiac interventions. Furthermore, SVC recanalization and stenting is well established in those patient populations. Restoring effective flow through the SVC has led to the regression of the collaterals within a short period of time, leading to a clinical improvement. The optimal management after recanalization and stenting of systemic veins remains unclear. In our case, the patient will remain warfarinised to prevent further occlusion of the SVC and will remain under follow-up with repeated cardiac magnetic resonance imaging. This case underscores the value of a thorough collaborative approach between acquired and congenital cardiologists combined with expertise in both advanced imaging and diagnostic as well as interventional catheterization.

Disclosures
None.

References

Key Words: cardiac catheterization ◼ magnetic resonance imaging ◼ stents ◼ superior vena cava syndrome ◼ thrombosis
Figure. A, Volume-rendered image from contrast-enhanced angiography, right lateral projection. The arrow points at the superior vena cava (SVC) stump (there is no communication with the right atrium [RA]). B, Volume-rendered image from contrast-enhanced angiography, anterior projection. The arrow shows extensive tortuous systemic venous collateralization to the upper pulmonary veins (down pointing on the left shows the systemic venous collaterals draining into the left upper pulmonary vein, whereas the horizontal arrow on the right shows systemic venous collateral draining into right upper pulmonary vein). C, Volume-rendered image from contrast-enhanced angiography, right lateral projection shows regression of systemic venous collaterals (arrow pointing at the regressing collaterals) when compared with B only 1 small collateral left drains into the left atrial through the pulmonary vein. This volume-rendered image does not highlight the patency of SVC. Hence, D shows a turbo-spin echo dual-inversion recovery (black-blood) magnetic resonance images of the SVC (blue arrow showing patent SVC stent), showing very little susceptibility artifact from the stent and indicates patency of this portion of the SVC. LUPV indicates left upper pulmonary vein; and RUPV, right upper pulmonary vein.
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_Circ Cardiovasc Interv._ 2015;8:
doi: 10.1161/CIRCINTERVENTIONS.114.002163

_Circulation: Cardiovascular Interventions_ is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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Print ISSN: 1941-7640. Online ISSN: 1941-7632

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circinterventions.ahajournals.org/content/8/9/e002163

Data Supplement (unedited) at:
http://circinterventions.ahajournals.org/content/suppl/2015/08/18/CIRCINTERVENTIONS.114.002163.DC1

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SUPPLEMENTAL MATERIAL

Video Legends

**Video A** (Volume-rendered video from contrast-enhanced angiography) shows extensive tortuous systemic venous collateralization to the upper pulmonary veins.

**Video B** shows the catheter lab based procedure for Superior Vena Cava recanalization. Images show stepwise approach to creation of veno-venous circuit, predilatation to placement of stents with final successful angiography results.