

Figure 2. Histologic section taken from an allograft shunt 18 months after implantation, showing no signs of inflammation or rejection.

Discussion

Carotid artery allografts offer distinct advantages over synthetic grafts, particularly in small infants. Human tissue demonstrates no oozing of blood or serous leakage from the suture holes and is more resistant to infection than are synthetic materials. The lack of thrombogenicity is also a major advantage when using biologic tissue. In this series, all shunts remained patent, even in the graft that narrowed through a suspected immunologic response. In the absence of a local tissue bank, the availability of human carotid

arteries is problematic. So far, the potential uses of donated blood vessels are underestimated. For example, we use descending aortic tubes to improve the safety of left ventricular assist device cannulation and ascending aorta or pulmonary artery wall as patch material where eventual stiffening and calcification is unimportant. Accordingly, we obtain as much healthy vascular tissue as possible when harvesting aortic and pulmonary valves from cadaveric donors. The risk of immunoreactivity has not proven to be of importance in the short term between shunting and the corrective procedure. Bogats and colleagues² and Tam and associates³ have reported the use of saphenous vein allografts as an alternative for modified Blalock shunts. In their series there was no fibrosis or calcification seen in 4 patients up to 55 months.²

In conclusion, the use of carotid allograft facilitates the microvascular anastomoses in infants with cyanotic congenital heart disease. These conduits have a very low propensity for thrombosis, and the potential for immunogenicity does not preclude their use.

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Unusual systemic venous return with absence of superior caval veins

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The absence of the right superior vena cava in situs solitus is a rare anomaly. It is usually associated with persistence of the left superior vena cava or other cardiac abnormalities.¹ We report here an extremely uncommon case of bilateral absence of the superior vena cava, with no

associated cardiac anomaly, evaluated through a high-resolution computed tomography (CT) scan.

Clinical Summary

A 14-month-old boy was admitted to our department because of a cardiac murmur discovered during a routine pediatric evaluation. On physical examination, a systolic heart murmur of grade 1/6 to 2/6 was found. No chronic heart failure or syndromic dysmorphism had been found. The electrocardiogram showed sinus rhythm and no evidence of atrioventricular arrhythmia. A transthoracic echocardiogram found a situs solitus, levocardia with atrioventricular and ventriculoarterial concordance, normal-sized ventricles, normal intracardiac anatomy, and the inferior vena cava draining normally into the right atrium. No superior caval vein could be seen.

We performed cardiac CT scanning to confirm the presumptive diagnosis: 64-slice CT scan (LightSpeed VCT; General Electric, Milwaukee, Wis), slice thickness 0.625 mm, speed of rotation 0.4 second, pitch 0.9, 80 kV, milliamperes modulated during the acquisition, and peripheral injection of contrast agent (iohexol 300

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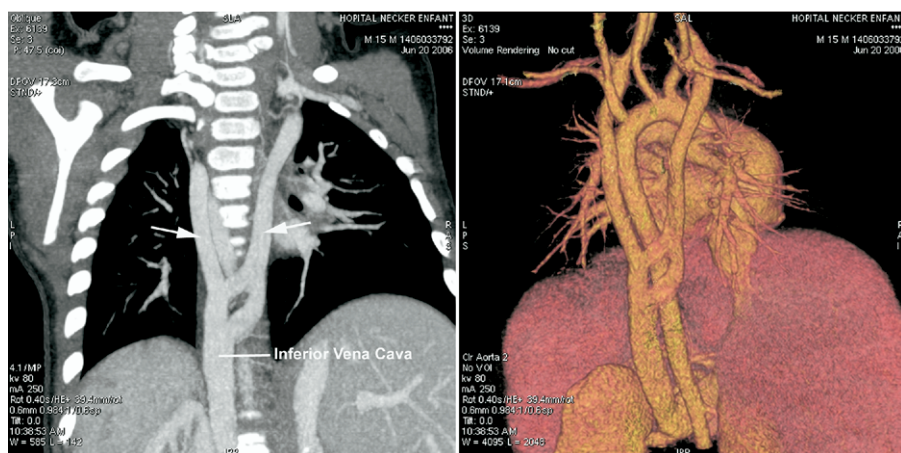


Figure 1. Coronal maximum-intensity projection (*left panel*) and volume-rendered (*right panel*) computed tomographic images showing the two separate azygos and hemiazygos veins (*arrows*) that join retrocardially before continuing into the inferior vena cava.

mgI/mL; volume 1.5 mL/kg; flow rate of the injection 0.5 mL/s). The acquisition lasted 1.3 seconds and the entire examination lasted approximately 15 minutes.

A very unusual type of systemic venous return was found (Figure 1). Both the right and left jugular and subclavian veins drained into two dorsal veins without communication with the right atrium. Indeed, the two jugular veins continued into azygos and hemiazygos veins, respectively. Then, the latter joined retrocardially above the diaphragm and connected to the inferior vena cava. The hepatic segment of the inferior vena cava was present and terminated normally in the right atrium. No upper caval vein or the equivalent of a bridging innominate vein was seen. The pulmonary veins were normal, draining into the left atrium.

Discussion

We report here an extremely uncommon case of complete absence of superior caval veins with no associated cardiac defect. Blood from the head, upper limbs, and upper torso drained to the right atrium through both the azygos and hemiazygos veins and the inferior vena cava.

The superior vena cava starts to develop in the fourth week of gestational age from the right common cardinal vein and the right anterior cardinal veins. The right superior vena cava is formed by the confluence of the left and right brachiocephalic veins and drains into the right atrium, while the left part, for a not yet clear mechanism during development, disappears. Absence of the right superior vena cava is a clear pattern, usually associated with persistence of the left superior vena cava and rhythm disorders or other cardiac defect. Bilateral superior vena cava is also a widely known entity; the left one, which usually drains into the right superior vena cava via the innominate vein or directly into the coronary sinus or the inferior vena cava via a cardinal vein, usually persists.¹

Bilateral absence of the superior vena cava was first reported by Hussain and associates² in a patient with complete atrioventricular block. In the setting of situs solitus, unequivocally lateralized atria, and normal cardiac anatomy, the absence of the superior vena cava is rare. In addition, azygos continuation of the inferior vena cava

is commonly associated with absence of the hepatic segment of the inferior vena cava in the setting of left atrial isomerism and is usually unilateral. Cases of the present condition in isolation are rare, especially inasmuch as patients are asymptomatic, with the diagnosis being made only on the basis of radiologic findings.³ The condition also can be found incidentally⁴ or can be revealed by a congenital chylothorax, by symptoms of superior vena cava obstruction that appear in the fetus, or by symptoms of an obstructed drainage of the thoracic duct at birth.⁵ In our patient, the upper half of the body drained by azygos and hemiazygos accessory veins that flowed into the inferior vena cava, suggesting a complete involution of the anterior cardinal venous system or lack of development with bilateral persistence of the supracardinal venous system.

Absence of upper caval veins should be considered in the differential diagnosis when a patient presents with radiologic evidence of superior vena cava obstruction, especially if the patient is symptom free with no sign of chronic superior vena cava obstruction. As long as there are doubts about systemic venous drainage, noninvasive CT or magnetic resonance imaging should be performed as a complement to echocardiography-Doppler and as a substitute for conventional angiography.

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