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Obstructions of the inferior and superior vena cava

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Anatomy and pathophysiology

Obstruction of caval veins is rare, and usually iatrogenic. Any surgical procedure involving the caval veins, including cannulation for bypass, may be complicated with caval vein obstruction. It was a common complication of the Mustard operation during early and long-term follow-up, with restrictive venous pathways in the majority of the patients.[1,2] It has been reported after a Senning-type procedure, after a Glenn shunt or other cavopulmonary connections, after transplantation with stricture of the anastomosis,[3] or after repair of abnormal pulmonary venous return with subdivision of the superior vena cava (SCV).

Multiple pacing leads or other long-term catheters may lead to progressive narrowing, especially during growth. Thrombosis of a caval vein may occur after endothelial damage during traumatic puncture for central lines, after a long period of low cardiac output with hypertonic IV fluids, or after recurrent and long-term central venous line,[4] infections. Any hypercoagulable state may exacerbate this problem (protein C or S, antithrombin 3, Leiden factor, etc.).

Congenital membranous obstruction of the inferior vena cava (ICV) at the junction with the right atrium or a restrictive Eustachian valve has been described.[5] An obstructed ICV may present as Budd–Chiari syndrome.

External compression by a tumor (lung cancer, lymphoma), aneurysmal dilation of the ascending aorta, pseudoaneurysm of a venous coronary graft,[6] goiter, mediastinal fibrosis, constrictive pericarditis, bile bladder distention, polycystic kidneys, hydatid cyst, and hematoma after blunt liver trauma have been reported. Vasculitis such as Behcet's disease may lead to shrinkage and obstruction of the caval veins.[7]

## Clinical symptoms, indication for treatment, and alternatives

Clinical symptoms will depend on onset and obstruction rate, the development of collateral flow, and the functionality of the other caval vein.

Obstruction of the SCV may clinically result in superior caval vein syndrome: congestion, swelling, and cyanosis of the head and the upper limbs, headaches or cerebral venous hypertension, (pre)syncope, cough, and airway obstruction. Pemberton's sign involves jugular vein distension in upright position, which progresses to cyanosis and facial edema while keeping both arms elevated. Retrograde congestion of the thoracic duct may lead to leakage of chyl into the gut (protein losing enteropathy), into the pleural space (chylothorax[8] or chylopericardium), or into the bronchial tree (plastic bronchitis).

Obstruction of the ICV may lead to abdominal congestion, chronic hepatic congestion leading to fibrosis, varices, exercise intolerance, fatigue or swelling of the legs, renal insufficiency with proteinuria,[9] or Budd–Chiari syndrome.[10,11]

If inflow to the heart is severely limited from all sides, this will result in decreased cardiac output, which can be very difficult to detect clinically. The heart will typically have no preload reserve; tachycardia or exercise will decrease stroke volume, which may result in hypotension, vertigo, syncope, or sudden death.

Alternative treatment to interventional catheterization depends on the etiology: mass resection or debulking, thrombolysis, anticoagulation, treatment with anti-inflammatory, antibiotic, or oncologic drugs, or radiation treatment may result in fast relief.
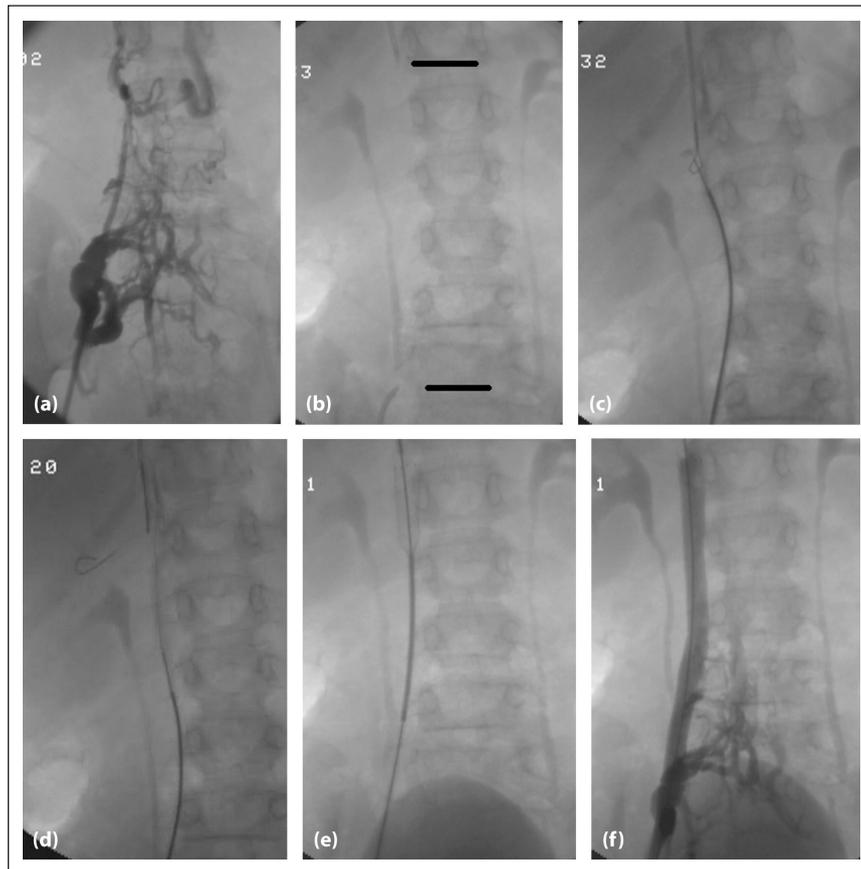
Figure 80.1 (a) A ten-year-old patient with obstruction of inferior caval vein between iliac vein and insertion of renal veins. (b) The distance to cover between catheter in iliac vein and catheter from jugular vein down to the obstruction (solid lines). (c) A 10-mm snare was used as target for the Brockenbrough needle within a 6 F long sheath; (d) A 0.014" wire was inserted through the Brockenbrough needle and grasped by the snare. (e) Partial opening of a 10/80 SMART Cordis self-expandable stent, which was lengthened with a 10/60 SMART Cordis stent. (f) Final result after balloon dilation with an 8-mm balloon; good patency and runoff were documented 8 years after this procedure.

History of the procedure

Surgery has for a long time been the therapy of choice for caval vein obstruction; however, it is not well tolerated with significant morbidity. Stents have changed the treatment strategies enormously; it is currently the technique of choice. The procedure has evolved from bailout for significant stenosis or obstruction, to electively altering flows to the heart, where currently percutaneous Fontan completion with rerouting of the ICV to the SCV or hemi-Fontan is being evaluated.

Precatheter imaging/ assessment, indications

A high clinical suspicion for caval obstruction is mandatory in patients with previous caval vein surgery. Because of sharp angles of Mustard patches when entering the pericardium, a subobstruction can easily be missed. Similarly, Fontan conduits may be difficult to visualize. Even good clinicians may clinically miss obstruction of major caval veins, as this may not result in retrograde congestion, but in low flow cardiac output.

Prior to the catheterization, the interventionalist should know which vessels are open and can be punctured, and whether a thrombus in either SCV or ICV is present. All information usually can be obtained with echo or preferably computed tomography/magnetic resonance (CT/MR). If a recent thrombus is present, thrombolysis should be given followed by anticoagulation, as any manipulation near or through the thrombus may cause multiple (paradoxical) embolizations.

Anesthesia/supporting imaging

Interventions of the SCV or ICV can best be approached from the femoral and/or jugular vein. After cannulation, a distal angiogram through the sheath should be made to exclude thrombi. If the caval vein is completely obstructed, both the femoral and/or jugular vein should be cannulated, as this will allow the interventionalist to visualize
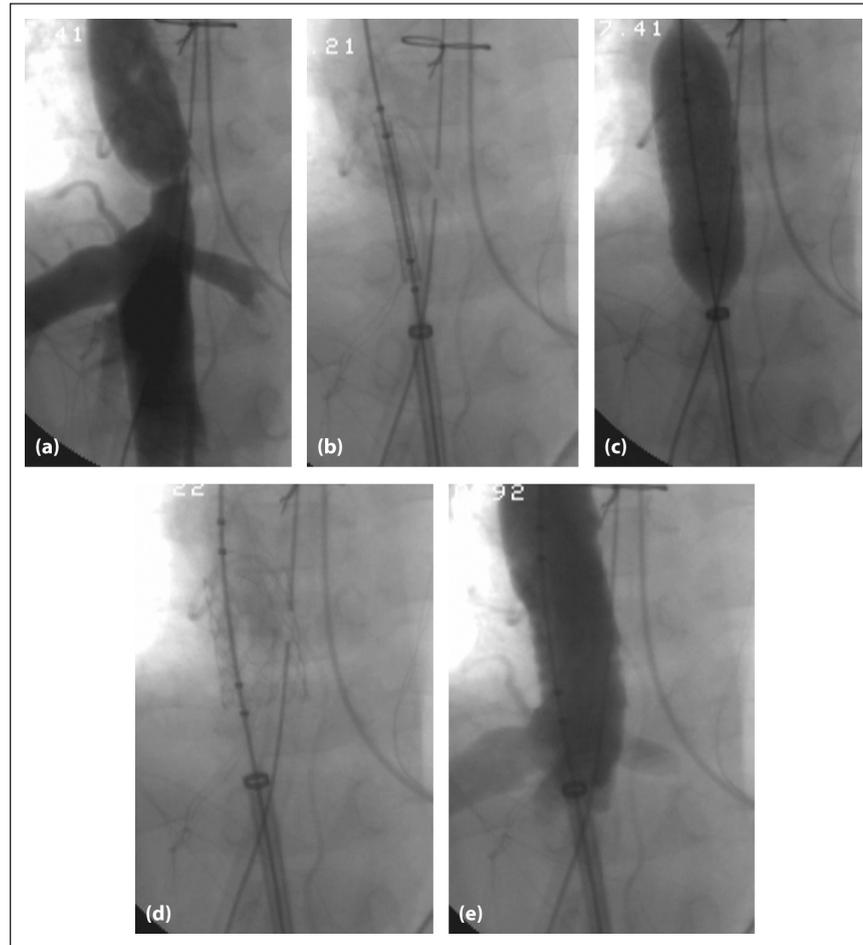
Figure 80.2   (a) A four-year-old patient 1 h after TCPC Fontan completion: a significant stenosis at the caudal junction of the inferior caval vein with an 18-mm Goretex conduit is demonstrated. (b) A 25-mm Genesis stent (Johnson and Johnson) mounted on a 14-mm BIB balloon (Numed) is positioned through an 11F long Mullins sheath (COOK). (c) Full inflation of outer BIB balloon. (d) Stent well deployed. (e) Cavogram through the sheath demonstrating good relief of the stenosis. This stent was fully expanded up to 18 mm 6 months later.

the "target" from both ends; occasionally, this may reveal "hidden" hypoplastic but patent pathways.

Stenting the ICV has been reported to be performed under echographic guidance only from centers with no or limited access to radiographic equipment.[12]

Protocol of hemodynamic assessment

Gradients across obstructions can be obtained, but because of collateral circulation with low flow and low cardiac output, the clinical significance of the obstruction can be severely underestimated.

Angiography catheter selection

Good angiographic visualization in standard perpendicular planes proximal and distal to the stenosis obstruction

is important. If the vessel is no longer patent, the lesion should be approached from both ends (cannulation of groin and neck vein). This will allow accurate determination of the length of the obstruction and the desired diameter of the final stent.

Catheter/wire interchange for delivery of balloon/stent/device

After visualization of the (sub)obstruction, a wire must be positioned across the lesion. If the caval vein is still patent, this is usually easy. If a segment has thrombosed or is atretic, a new route must be made.

Frequently, a mini vein may bridge partially the thrombosed distance; this vein should be probed with a thin wire, preferably a microcatheter system (Prograde: 0.018″ hydrophilic wire in 2.4 F coaxial catheter system, used through a 4 F end-hole catheter).[13] A final segment can be
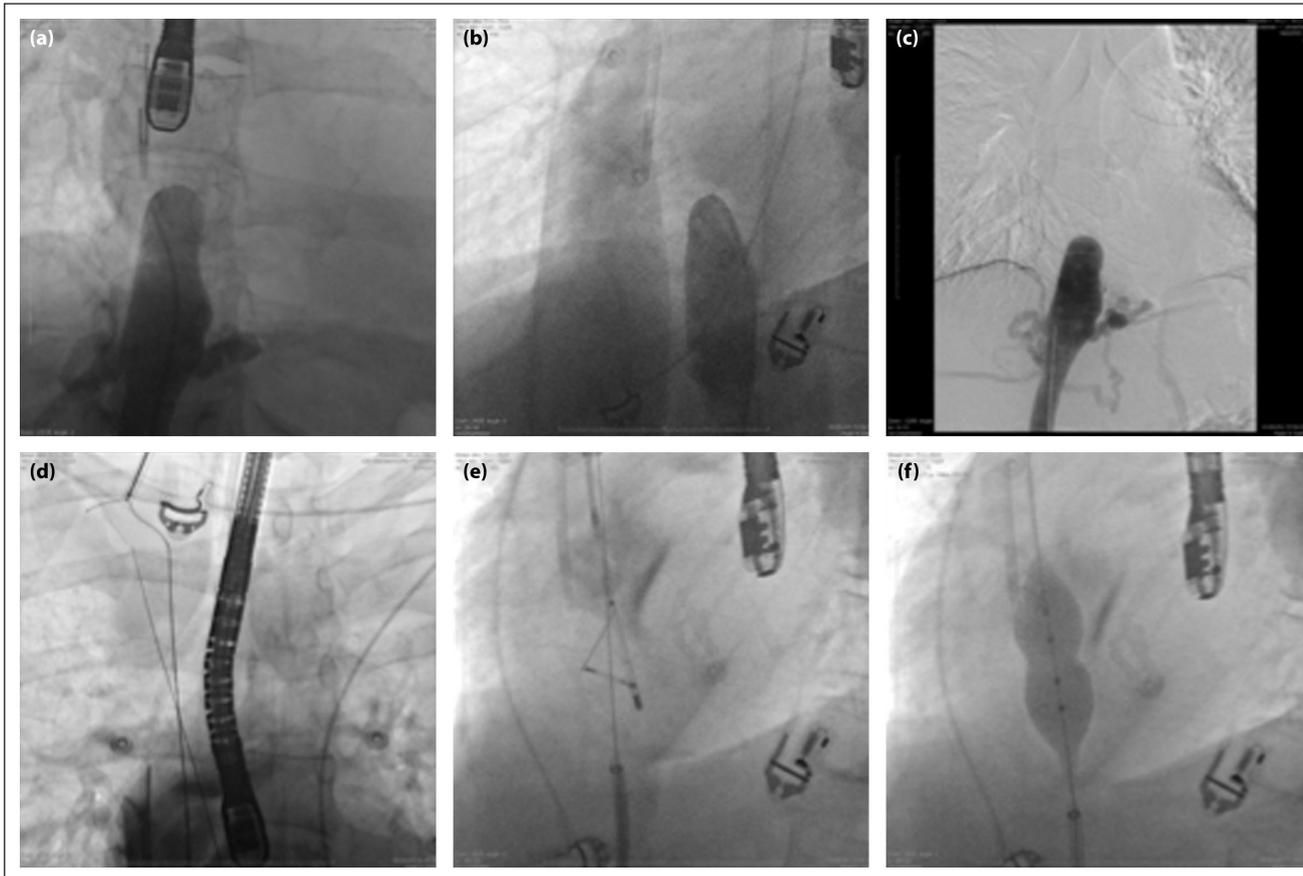
Figure 80.3   (a,b) Frontal and lateral views of cavogram showing complete obstruction of ICV by Eustachian valve. (c) Extensive collateral circulation. (d) Under TEE guidance, the Eustachian valve was punctured with a Brockenbrough needle and a wire was snared in the RA and exteriorized through the jugular vein. (e) Enlargement of opening with blade. (f) Tearing the opening with a 25-mm Mullins balloon to get an unobstructed connection from ICV to the right atrium.

completely obliterated, making a new route necessary. The most common technique is puncturing with a straightened Brockenbrough needle within a 6 or 8 French dilator transseptal sheath. It must be determined from which cannulation point (groin or neck) the puncture will be easiest. Preference will be given to the side that allows a straight route. When puncturing from one end, it is wise to provide a target at the other side: deployment of a 5 or 10 mm snare perpendicular to the needle direction provides a vessel-centered and radiopaque marker; it also allows it to snare and exteriorize the wire once grabbed creating a veno-venous loop. Alternatively, a new route can be made with radiofrequency ablation.

Once the lesion is crossed, an extra stiff wire should be used, to give optimal support and steering of the balloon during deployment.

Balloon dilation alone may occasionally give good relief,[14] however, with frequent early recurrence of obstruction.[15,16] Most interventionalist therefore will prefer stent implantation. Predilation with small balloons may be indicated in order to get the stent or delivery system in place. Predilation or low-pressure balloon interrogation with big balloons may facilitate assessment of the contours of the stenotic site, the stretchability, and the recoil.

A choice must be made from two different types of stents: self-expanding or balloon-expandable.

A self-expanding stents is more flexible; it will continuously push radially to reach its nominal value; it will reexpand after external compression (resuscitation, blunt thoracic trauma). A self-expanding stent is limited in maximal diameter, and cannot be dilated beyond nominal value. Such stents are good for long lesions, not ideal for short discrete lesions (obstruction in Mustard repair).[17]

Balloon-expandable stents have high radial strength, and are ideal for short lesions. External force may deform such stents, thereby decreasing or obliterating the lumen; this is rarely a problem in the SCV, but any stent in the ICV may be compressed by the liver.

Both types of stents can come with a cover (graft stents). Covered stents are indicated if rupture to adjacent cavity vessel is likely or has occurred, such as pulmonary pathway (in Mustard with patch leak, rupture to pleura), or if endothelial reaction or tumor invasion is likely.

Multiple case reports or small series can be found in the world literature.[18–31]

Post "deployment" protocol

Angiography post deployment of the stent must be made prior to removal of the wire (injection through sheath, or via multitrack (Numed)). If extravasation of contrast is observed, a covered stent can still be positioned and deployed. If blood loss is significant, gentle balloon occlusion may temporarily obliterate the tear while preparing the covered stent.

Pitfalls, problems, complications

Dilation of the caval veins may be complicated by rupture to the pleura with hemothorax, tear to the pericardium with tamponnade, tear to the pulmonary pathway allowing right–left or left–right shunting, tear to the ascending aorta, compression or damage of phrenic or vagal nerve, compression or elongation of the sinus node artery with loss of stable sinus rhythm, compression of the thoracic duct, or compression of the ureter.

Caval veins can significantly stretch and may show some contractility—peristalsis; this may lead to stent migration within minutes or hours after deployment, and embolization to the right ventricle or into the pulmonary artery. Self-expanding stents can be recaptured with a lasso at one end, and refolded into a big sheath[32]; balloon-expandable stents are much more difficult to retrieve, and should be parked-expanded-left in the circulatory system, or retrieved surgically.

Recurrent stenosis or thrombosis may occur; appropriate anticoagulation should be given. However, large stents in large veins in patients with good cardiac output have a low tendency to thrombose. When in doubt, it is safer to give antiaggregation or anticoagulation, at least early after the procedure allowing the endothelium to cover most of the bare metal.

During long-term follow-up, a stent may fracture, or may be compressed by a blunt external trauma. Radiographic control in two perpendicular dimensions or CT will easily reveal this complication.

When deployed next to pacing wires, the pacing lead may be submitted to concentrated movements in one limited region, resulting in metal fatigue and lead fracture; damage to the insulation may cause a current leak of the pacemaker leads with dysfunction of the pacing system.

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