iatrogenic occlusion of bilateral jugular veins, subclavian vein, and superior vena cava after repeated jugular cannulation associated with Arnold-Chiari malformation: Successful endovascular treatment

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ABSTRACT
An Arnold-Chiari malformation is a congenital central nervous system defect. Raised intracranial pressure is commonly observed, and posterior decompression neurosurgery is the treatment of choice. We describe a patient with iatrogenic occlusion of bilateral jugular veins, subclavian vein, and superior vena cava resulting from repeated central venous cannulations. Because of venous hypertension, the patient suffered from neurologic symptoms: headaches, vision disturbances, and marked head edema. Two stents were used to recanalize the right internal jugular vein and superior vena cava. Symptoms subsided, and the patient returned to work. During 24-month follow-up, stents were patent. The patient remains symptom free and continues working. (J Vasc Surg Cases and Innovative Techniques 2020;6:18-20.)

Keywords: Vena cava obstruction; Endovascular treatment; Stenting

An Arnold-Chiari malformation is a congenital defect in which the cerebellum structures are displaced into the upper spinal canal. A diagnosis is based on the patient’s history, careful observation, and neuroimaging (computed tomography [CT] or magnetic resonance imaging), which provides detailed insight into brain structures and especially the cerebellum. Typically, symptoms exhibited by patients suffering from Arnold-Chiari malformation are a consequence of structural congenital defects in the brain and spinal cord. Neurosurgery, the only treatment, is aimed at elimination of the cause; it consists of a posterior fossa decompression, most frequently Pudenz ventriculoperitoneal or ventriculoatrial shunt placement, resulting in a decrease of intracranial pressure and symptom alleviation.

Because of repeated central venous cannulation, patients with Arnold-Chiari malformations are at risk for complications that may lead to occlusion of the jugular veins, subclavian veins, or superior vena cava (SVC). This iatrogenic disease of the venous system caused our patient to develop symptoms suggestive of intracranial hypertension. We obtained the patient’s consent for the publication of clinical data.

CASE REPORT
A 37-year-old man had been diagnosed with Arnold-Chiari I in childhood. The most common symptoms of type I Arnold-Chiari malformation are mild hydrocephalus, displacement of the cerebral tonsils toward the foramen magnum, vertigo, paresis, cranial nerve palsy, problems with balance, poor coordination, urinary incontinence, and sleep apnea. Coughing or sneezing may lead to syncope. In this case, intracranial hypertension resulted from extracranial venous system occlusion because in the course of neurologic and neurosurgical interventions, he had several central catheters placed with resultant occlusion of both internal jugular veins, subclavian veins, left external jugular vein, and SVC. Twenty years before presentation to our department, the patient had received a ventriculoatrial shunt, which after 7 years was replaced with a ventriculoperitoneal shunt. He had also undergone meningioma resection. Despite a well-functioning shunt, severe neurologic symptoms (ie, headaches, head edema, and visual impairment) recurred about a year preceding the treatment. This ultimately led him to visit the neurosurgery outpatient clinic. Following consultation and confirmation of proper shunt function, the patient was admitted to the vascular surgery department for further evaluation including CT angiography with venous phase imaging.

Reports from previous investigations, including 128-slice CT angiography, were thoroughly analyzed. Considering difficulties in interpreting the CT angiograms (slow outflow of contrast material from the head and upper body), venography was performed with administration of contrast material into the aortic arch using left radial artery access to avoid puncture of the jugular veins before the ultimate decision regarding management.
strategy. Contrast material was also injected into the markedly
dilated azygos vein through a right femoral vein access. The
findings helped resolve doubts concerning previously per-
formed CT angiography. The venograms showed occlusion of
the SVC, both subclavian veins, left external and internal jugular
veins, and right internal jugular vein down to the level of the thy-
roid veins (Fig 1). Venous drainage from the brain was by a collat-
eral pathway through the still patent right external jugular and
the patent segment of the right internal jugular vein.

The patient was considered eligible for attempted endovas-
cular treatment. Three vessels were considered as access
evessels—the left radial artery, the right femoral vein, and the
patent segment of the right internal jugular vein. A 7F intro-
ducer was inserted into the femoral vein, a 6F introducer into
the right internal jugular vein, a 6F introducer into the left
radial artery, and a pigtail catheter into the aortic arch. An
intra-arterial bolus of 5000 units of heparin was administered
during the interventional procedure; the patient had been
receiving long-term therapy with acetylsalicylic acid.

Hydrophilic PT2 (Boston Scientific, Marlborough, Mass), Pilot
(Abbott Vascular, Abbott Park, Ill), and AqWire (Ev3) guidewires
were used. After unsuccessful attempts at recanalization through
the azygos vein, a hydrophilic guidewire was navigated across the
right internal jugular vein occlusion and SVC to reach the area of
the right atrium. After it was confirmed that the guidewire tip
was in the right atrium, predilation was performed with a 4-mm
balloon catheter. Contrast-enhanced scans showed venous
outflow from the right jugular vein to the SVC and right atrium.
A dedicated venous stent (Zilver Vena, 14 × 100 mm; Cook Medical,
Bloomington, Ind) was then inserted into the SVC and right jugu-
lar vein. In addition, Wallstent (8 × 40 mm; Boston Scientific) was
inserted into the distal segment of the jugular vein. Consecutive
postdilations were carried out with 8-, 10-, and 12-mm catheter
balloons. A checkup examination at the end of the procedure
confirmed that both stents were in the correct position; adequate
venous outflow from the head was also seen (Fig 2). The postinter-
vention course was uncomplicated; the patient’s condition
improved, head edema decreased, and he was discharged on
postoperative day 3.

During the 30-day observation period, the patient was pre-
scribed therapeutic doses of low-molecular-weight heparin. He
also received dual antiplatelet therapy of aspirin and clopidogrel.
A follow-up appointment on day 30 revealed good general
condition and subsidence of neurologic symptoms and head
edema. Color duplex Doppler ultrasound confirmed venous
flow pattern through both stents. The antithrombotic regimen
was replaced with dabigatran 110 mg plus aspirin 75 mg daily.

The patient continued follow-up and support from a neurolo-
gist and vascular surgeon and returned to work. At 1 year after

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**Fig 1.** Patent part of right jugular vein.

**Fig 2.** Final result after recanalization and stenting.
DISCUSSION
Our patient with an Arnold-Chiari malformation is not among those typically treated with endovascular interventions. The need for endovascular treatment was associated with symptoms of iatrogenic occlusion of the jugular and subclavian veins as well as SVC occlusion revealed on history taking and resulting from multiple intravenous cannulations. The symptoms placed an enormous burden on the patient's everyday life, excluding him from work and causing severe depression. An attempt at endovascular treatment of intracranial hypertension resulting from stenosis or occlusion of the venous system was based on widely available publications regarding the efficacy of such treatment strategies.1-4
Because no clear recommendations are available for covered and uncovered stent placement, we decided to insert an uncovered Zilver Vena, a dedicated venous stent, at the SVC recanalization site. Although Hadad et al5 suggested higher effectiveness of covered stents, they also emphasized the need for further studies in this area. Polytetrafluoroethylene-covered self-expanding or balloon-expandable stents definitely prevent passage of thrombi through the mesh. However, uncovered stents are less rigid and less thrombogenic; they allow better adaptation to vessel anatomy. The increasing use of dedicated venous stents might improve treatment outcomes. The risk of the procedure was considered low as major post-stenting complications (SVC rupture, mediastinal hemorrhage, cardiac tamponade, or pulmonary embolism) are rare. Other complications include chest pain, hemoptysis, and approach-related complications. SVC stent migration into the right atrium is another serious issue; its prevalence may be limited by proper device size and positioning. The most frequent long-term complications are stent thrombosis and in-stent restenosis resulting from thrombus formation or external compression. Fortunately, repeated endovascular treatment is possible.6,7
There is no consensus with respect to effective anticoagulant therapy for prevention of stent thrombosis after SVC recanalization. Oral anticoagulants or antiplatelet agents are typically used for several months. No comparative studies are yet available to analyze both strategies in this group of patients.6,9
The antithrombotic regimen used in our patient during a 30-day follow-up (ie, low-molecular-weight heparin and dual antiplatelet therapy) resulted from our experience in endovascular treatment of arterial disease and thrombosis prevention. Aggressive management was associated with the extent of recanalization and stent length exceeding 120 mm. Long-term antithrombotic therapy (dabigatran 110 mg and aspirin 75 mg daily) seems to have been an effective strategy in our patient.

CONCLUSIONS
Considering literature reports indicating relatively low complication rates after endovascular treatment of iatrogenic and post-thrombotic venous occlusion, we decided to use an endovascular intervention. Early and long-term effects were consistent with literature data; hence, it was concluded that endovascular procedures could be considered the treatment of choice in patients with structural defects and associated conditions.

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