

A rare anatomic variant of a single-conduit supraclavicular cephalic arch draining into the external jugular vein presenting with recurrent arteriovenous fistula stenosis in a hemodialysis patient

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The cephalic arch is a common location of stenosis, especially in brachiocephalic fistulas. The cephalic arch has a number of anatomic variations. Cephalic arch stenoses are often resistant and have poor primary patency. Here we describe an unusual case of a hemodialysis patient with a single-conduit supraclavicular cephalic arch draining into the external jugular vein presenting with recurrent cephalic arch stenoses and external jugular vein stenosis. In our view, extrinsic compression by the clavicle may contribute to the high rate of recurrence, the lack of complete dilation of even high-pressure balloons, and a theoretically heightened risk of rupture when cutting balloons are used. (*J Vasc Surg Cases and Innovative Techniques* 2017;3:20-2.)

CASE REPORT

A 61-year-old man undergoing hemodialysis (HD) through a left brachiocephalic fistula (BCF) was noted to have high venous pressures during HD and prolonged bleeding after HD. Fistulography was performed, which showed severe juxta-anastomotic stenosis as well as a severe cephalic arch stenosis. The single-conduit cephalic arch was found to follow an aberrant supraclavicular course and to drain into the external jugular vein (EJV; Fig 1). The stenotic lesions were dilated with a 6-mm REEF balloon (Bard, Murray Hill, NJ) with improvement in flow and partial resolution of the cephalic arch stenosis. Two months after the first procedure, patient was noted to have persistently high venous pressures and prolonged bleeding. He was thus referred to our service. Clinical examination revealed a hyperpulsatile BCF suggestive of outflow obstruction. The cephalic arch was found to be visible superficially, passing over the clavicle. Doppler ultrasound examination confirmed the presence of a single-conduit supraclavicular cephalic arch, which was found to be restenosed.

Angiography was repeated subsequently, which confirmed these findings and also revealed stenosis of the EJV at its caudal portion near the confluence with the subclavian vein. Contrast material was found to reflux into the cranial portion of the EJV, suggesting hemodynamically significant EJV stenosis. The EJV was successfully dilated on first pass using a 7-mm balloon. For the cephalic arch stenosis, complete effacement of the high-pressure balloon (Conquest, Bard) could not be achieved

despite repeating dilation at high pressures (up to 30 atm). A cutting balloon was not used because of perceived increased risk of rupture in view of the underlying bone. Postangioplasty images, however, revealed satisfactory dilation and improved flow. Postprocedural flow was found to be improved, and HD was uneventful. The patient was observed again 3 months later with recurrence of high venous pressure on dialysis. Angiography was repeated, which confirmed severe restenosis of the cephalic arch and EJV (Fig 2). Repeated angioplasty was performed, with both the EJV and cephalic arch dilated with 7-mm balloons (Fig 3), and adequate postprocedural flow was achieved. The patient's consent was obtained for publication of this case report and the associated images.

DISCUSSION

The cephalic arch has been described as the "final arch before the cephalic vein enters the axillary vein to form the subclavian vein."¹ Whereas the cephalic arch typically passes inferior to the clavicle, turning sharply to pierce the clavipectoral fascia and drain into the axillary vein, some anatomic variants may be present.¹ The bifid double arch represents one of the anatomic variants described in the literature in which the arch bifurcates.² Both limbs of the bifid cephalic arch may drain into the axillary vein, or one of the limbs drains into the EJV and may take a supraclavicular course. Trifid circulations with complex collaterals have also been discovered.³ Lau et al described a case of supraclavicular cephalic arch that drained into the subclavian vein during permanent pacemaker implantation.⁴ Yeri et al described a cadaveric case of the cephalic vein joining the EJV, albeit taking an infraclavicular course.⁵ Here we report a unique case of a *single-conduit* supraclavicular cephalic arch draining into the EJV in the context of dialysis vascular access. Furthermore, the patient was found to have recurrent cephalic arch and EJV stenosis. To our knowledge, this has not been reported, especially in the vascular access literature.

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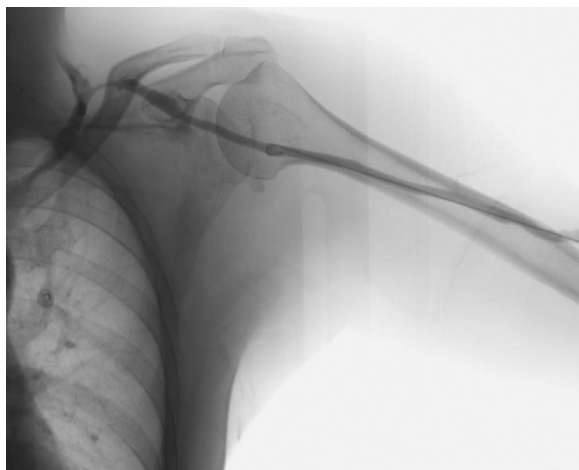


Fig 1. First angiogram showing cephalic arch stenosis in a single supraclavicular cephalic arch.



Fig 3. Postangioplasty image showing dilation of both cephalic arch stenosis and external jugular vein (EJV) stenosis with improved flow. The *arrows* indicate areas of stenosis.



Fig 2. Angiogram showing cephalic arch stenosis in the supraclavicular single-branch cephalic arch draining into the external jugular vein (EJV) with associated EJV stenosis. The *arrows* indicate areas of stenosis.

Cephalic arch stenosis is one of the leading causes of arteriovenous fistula (AVF) failure, especially in BCFs.⁶ The prevalence of cephalic arch stenosis may be as high as 39% in patients with a BCF and leads to significant morbidity and mortality due to failure of the AVF.⁷⁻⁹ Although the exact etiology is not known, it has been postulated that hemodynamic factors, such as turbulent flow, intimal injury, and valvular hypertrophy, contribute to making the cephalic arch particularly vulnerable to stenosis.² This is seen more commonly in BCFs because of higher flow and presence of a single outflow channel compared with that of the radiocephalic fistula. It is well recognized to be resistant to conventional angioplasty, often requiring use of high-pressure balloons; it has poor patency and high reintervention rates.⁶ Cephalic arch stenosis is also associated with higher risk of vein rupture

(up to 15%) during angioplasty.¹⁰ Percutaneous transluminal balloon angioplasty with conventional angioplasty balloons is the standard first-line therapy. Alternatives include high-pressure balloons, cutting balloons, stents, and surgery. Surgical options available include cephalic vein transposition, flow reduction surgery/intervention, and creation of an alternative AVF, such as a brachiobasilic fistula. Whereas some studies have shown better outcomes in terms of primary patency through the use of stent grafts and cutting balloons, a systematic review concluded that there is much heterogeneity in the results with small numbers in each study.¹¹ No single procedure was deemed to be superior.¹²

In our patient, three angioplasty attempts were required in a short span of 6 months. In this variant, the extrinsic compression by the underlying clavicle may contribute to the higher risk of recurrence than that observed with other cephalic arches. Extrinsic compression is one of the oft-cited hypothetical causes of cephalic arch stenosis described in many papers and texts.^{6,7,13} Altered hemodynamics in the form of turbulence has been shown to lead to neointimal hyperplasia in all forms of AVF stenosis including cephalic arch stenosis.^{6,14,15} It is therefore plausible that the presence of the clavicle immediately underneath the cephalic arch causes turbulence. This assumption is hypothetical, however, and needs more data for confirmation. The underlying bone is likely to limit adequate dilation and may also limit the use of other modalities, such as cutting balloons and stents. Cutting balloons were not used because of the perceived higher risk of rupture due to the underlying bone.

The EJV is not commonly encountered as the main draining vein for AVFs. The high flow and pressure from the BCF are transmitted to the EJV, causing stenosis. As such, we had to dilate the EJV twice within a span of 6 months. This phenomenon was observed despite presence of

upstream stenosis (cephalic arch stenosis), suggesting that EJV stenosis rates may be even higher otherwise.

In addition, the EJV may be a useful vein for tunneled dialysis catheter placement in patients with challenging vascular access. Its stenosis further limits venous access options.^{16,17}

Detection of such a variant before AVF creation may prompt one to consider alternative access, such as distal radiocephalic arteriovenous graft (AVG), brachiobasilic AVG, or AVF on the contralateral side. This anatomic variation is amenable to identification during Doppler ultrasound vein mapping, a common investigation performed as part of vascular access planning.

CONCLUSIONS

We report a unique variant of cephalic arch—a single-conduit supraclavicular cephalic arch draining into the EJV in an HD patient. This variant anatomy may be prone to a higher rate of restenosis and limit therapeutic options. If it is identified before AVG creation, clinicians caring for vascular access patients may choose to consider alternative vascular access or perhaps a more intensive surveillance strategy.

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