Acoustic-Assisted Catheter-Directed Thrombolytic Therapy for Upper-Extremity Phlegmasia Cerulea Dolens
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**ABSTRACT:** Phlegmasia cerulea dolens (PCD) is a rare disease entity involving complete thrombotic occlusion of venous outflow in an extremity, and is considered a vascular emergency. Most cases described encompass the lower extremities; however, we present a rare case of upper-extremity PCD related to the presence of a peripherally inserted central catheter. Endovascular therapy was utilized in the form of acoustic-assisted catheter-directed thrombolysis with complete resolution of thrombus and clinical symptoms.

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**Key words:** deep vein thrombosis, peripheral vascular disease, thrombosis, thrombolytic therapy

**CASE PRESENTATION**
A 57-year-old male with a prior history of coronary artery disease, ischemic cardiomyopathy, and congestive heart failure (CHF) presented for evaluation of acute right upper-extremity swelling and discoloration (Figure 1). The patient awoke with his symptoms earlier that morning and presented 6 hours later once his arm swelling persisted and pain developed. He had a peripherally inserted central catheter (PICC) inserted in his right basilic vein 4 weeks prior for intravenous milrinone therapy in treatment of recurrent systolic CHF. On physical examination, the right upper extremity was markedly edematous with violaceous discoloration from the forearm to axilla. There was mild digital cyanosis and moderate tenderness to palpation with normal capillary refill, 1+ right radial and brachial pulses, and no evidence of venous gangrene. The patient was hemodynamically stable without chest pain or dyspnea. Venous duplex ultrasound demonstrated occlusive thrombus in the right basilic vein extending into the subclavian vein.

The patient was initiated on systemic anticoagulation...
with intravenous heparin and plans made for invasive therapy given the acute onset of symptoms and evidence of PCD. Right common femoral vein 6 Fr access was initially obtained and a JR4 catheter was advanced to the origin of the innominate vein. Selective innominate venogram demonstrated 100% total occlusion of the right proximal subclavian vein (Figure 2). A 0.035˝ stiff-angled glidewire with support catheter was used to attempt to cross the occlusion, but was unsuccessful.

Subsequently, the PICC line was accessed with insertion of a 0.014˝ wire which was advanced into the right atrium. The PICC line was then removed, and a 6 Fr glidesheath inserted into the basilic vein over the 0.014˝ wire. A venogram demonstrated complete occlusion of the brachial and basilic veins (Figure 3). The Angiojet Peripheral Thrombectomy Catheter (Boston Scientific) was initially used for thrombectomy in the right axillary and subclavian veins with minimal debris removed. Ensuing venogram showed no improvement of venous flow. Therefore, an EKOS Catheter (BTG) with a 30 cm treatment length was inserted into the right basilic, axillary, and subclavian veins (Figure 4). Alteplase infusion at 1 mg per hour, and heparin infusion at 500 units per hour, was initiated for a total of 12 hours. The patient was transferred to the medical intensive care unit for observation.

After 12 hours of treatment, the patient noted no further arm pain and improvement in right upper extremity swelling. Repeat venous ultrasound demonstrated no further thrombus in the basilic, axillary, or subclavian veins (Figure 5). Alteplase infusion was discontinued, and EKOS catheter and glidesheath were removed. Full dose intravenous heparin was restarted and the patient bridged to oral anticoagulation with rivaroxaban. An
ACE wrap and thromboembolism deterrent hose were used to provide compression and aid in the reduction of edema in the right arm, and the patient was discharged home 48 hours post lytic therapy.

He returned for a 2-week follow-up visit and noted complete resolution of his symptoms. Clinically the right arm had returned to its normal size and color (Figure 6). He was continued on rivaroxaban for the next 3 months.

**DISCUSSION**

Phlegmasia cerulea dolens of the lower extremities has been well reported and recognized as an endovascular emergency, yet upper extremity PCD can be exceedingly rare and can lead to venous gangrene, hemodynamic collapse, and death. The most common predisposing factors of PCD include cardiac disease, immobility, history of venous thrombosis, and...
hypercoagulable states such as antithrombin III deficiency, malignancy, and estrogen use. Another potential source of DVT can originate from indwelling central venous catheters. In one single center review, upper extremity deep venous thrombosis from the presence of a central venous catheter accounted for over half of the total hospital-acquired deep venous thromboses.

Signs and symptoms of PCD often include profound limb pain, cyanosis, massive edema, and venous gangrene with fluid sequestration leading to significant hemodynamic instability and shock. Further complications include pulmonary embolus, limb amputation, and even death.

Prompt recognition and treatment are critical in achieving favorable outcomes in PCD. A multidisciplinary approach is best suited to treat affected patients including endovascular or vascular specialists and intensivist input. Treatment begins with the initiation of systemic anticoagulation with intravenous heparin to prevent further thrombus propagation. Surgical options include open thrombectomy or fasciotomy in the setting of compartment syndrome, whereas endovascular

Figure 6. Right upper extremity, now normal in size, 2 weeks post ultrasound-assisted catheter directed thrombolysis.
therapies consist of rheolytic thrombectomy or catheter-directed thrombolysis. The Angiojet catheter is a rheolytic device that can often effectively remove large amounts of thrombus in both the venous and arterial space. Using the power-pulse mode of the Angiojet system involves infusing thrombolytics directly into the affected vessel, then subsequently 30 minutes later performing thrombectomy. Power-pulse technique was not chosen for this case, but rather catheter-directed thrombolytic therapy. The EKOS catheter is an FDA-approved porous catheter designed to deliver thrombolytic therapy locally within the clot, with the added component of ultrasonic vibration, or acoustic pulse, to aid in the breakdown of fibrin, thereby decreasing time of thrombus dissolution. Overall, the outcomes of PCD patients are relatively poor with a limb amputation rate of 20% to 50% and mortality rate of 20% to 40%.

Herein, we present a case of central venous catheter associated upper-extremity PCD, promptly recognized and successfully treated with acoustic pulse-assisted catheter-directed thrombolytic therapy.

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