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EDITORIAL COMMENT

Chylothorax and Central Venous Thrombosis

A Difficult and Challenging Relationship*

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We have read with great interest the paper by Deogaonkar et al (1) in this issue of *JACC: Case Reports* about percutaneous venous reconstruction as therapeutic option for central vein thrombosis associated with chylothorax (CTaC).

The presence of lymphatic fluid (chyle) in the pleural cavity can result from a laceration of the thoracic duct and/or lymphatic vessels. Its causes can be broadly grouped into congenital, traumatic, infectious, and neoplastic. CTaC is a rare clinical entity in pediatric patients. As the authors stated, central vein thrombosis (CVT) directly causes venous hypertension with disruption of the thoracic duct anatomy and function.

The treatment of chylothorax is mainly focused on the following: relief of respiratory symptoms, evacuation of the pleural space most commonly achieved using a chest tube, maintenance of adequate nutritional status and hydration, reduction of chyle flow, and control of associated morbidities. The therapeutic algorithm of chylothorax is not clearly established. Most authors advocate an initial period of conservative treatment, which can range from 2-4 weeks. This is successful in roughly 25%-50% of all patients presenting chylothorax within 2 weeks of treatment. Surgery would be relegated to patients resistant to conventional treatment. The goals of a surgical intervention are reduction in chyle leak, maximizing lung expansion, and obliteration of the pleural space. However, surgery is not without risks and the ideal moment for surgical treatment is still under discussion (2).

Tamai et al (3) have already shown that, although thoracic duct ligation has an excellent overall result for cases of persistent chylothorax, in those patients with associated CVT the risk of treatment failure is high, and consequently so are the associated increased morbidity and mortality experienced by these patients. Therefore, a high index of suspicion for CVT is essential in cases of chylothorax refractory to treatment. A decrease in the number of platelets as well as an increase in D dimer values can help us to establish the diagnosis (3).

If CVT is confirmed, this should be treated. Anticoagulation therapy is the cornerstone of treatment (4). In early stages, anticoagulation alone could achieve good results. The optimal duration of treatment is not clearly established and could have recurrences after stopping treatment. In other cases, as reflected in the article by Deogaonkar et al (1), anticoagulation is not successful, requiring other procedures for symptomatic improvement.

Before the generalization of endovascular techniques, surgical reconstruction of the superior vena cava was the treatment of choice for CVT cases. Doty and Baker reported optimal long-term results using the internal saphenous vein, demonstrating its superiority over the use of polytetrafluoroethylene for the reconstruction of the superior vena cava (5).

According to the article by Deogaonkar et al (1), the increasing development of endovascular techniques

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allows us to treat CVT with less procedure-related morbidity. Among the endovascular techniques, several options can be performed such as thrombolysis, angioplasty, or stenting.

Thrombolysis, which was performed in 2 of 3 cases reported by the authors, is a suitable therapeutic option. This is usually effective if started within 2-5 days of symptom onset and tends to be ineffective if started after the 10th day (6). However, it has a significantly higher risk of bleeding compared with anticoagulation (7). Innovative techniques using pharmacomechanical thrombolysis seems to be promising because these devices achieve elimination of the thrombus, reducing the need for thrombolytic agents, and, therefore, avoiding complications related to fibrinolytics. One of the most popular devices is the AngioJet, which has been designed to remove thrombus with the Venturi-Bernoulli effect, with multiple high-velocity, high-pressure saline jets, which are introduced through orifices in the distal tip of the catheter to create a localized low-pressure zone, resulting in a vacuum effect with the entrainment and dissociation of bulky thrombus. On the other hand, the Trellis Infusion Catheter and the Rotarex and Aspirex equipment use mechanical microfragmentation. Properly protocolized studies will be necessary to evaluate the effectiveness and long-term durability of treatment after the use of these devices.

Studies that have evaluated angioplasty for the treatment of CVT are small and retrospective, with reported technical success between 70% and 90% (8). Depending on the response of the lesion to angioplasty, we can differentiate between inelastic lesions (≥50% improvement in luminal diameter after angioplasty) and elastic lesions (worse response to angioplasty). For elastic lesions, stenting is proposed. Stents can be made of either Nitinol or stainless steel. No significant differences in terms of the patency have been demonstrated between both alternatives. Stenting is considered appropriate for both acute elastic recoil (≥50%) after angioplasty and recurrent stenosis within 3 months after angioplasty. The stent should be 15%-20% larger than the vein. It seems appropriate to consider the use of covered stents in those cases in which a perforation of the vessel has occurred during the venoplasty. In case of CVT secondary to benign causes, the use of uncovered stents would be the most suitable.

Generally, endovascular treatments for recanalization of the central venous system in the event of thrombosis are safe procedures. However, despite the growing popularity of all these endovascular procedures and their considerable safety and efficacy, some minor complications, such as inguinal hematoma or local infection at the puncture site, and major complications, including stent migration, bleeding, pulmonary embolism, cardiac injury, and cardiac tamponade, which can occur as a consequence of perforation of the intrapericardial portion of the superior vena cava during stent deployment, have been reported in the literature (9). Additionally in-stent thrombosis or restenosis after thrombolysis can happen. We must emphasize the necessity of close monitoring of the patient after performing these procedures with angioplasty or venous stenting to maintain patency and prevent long-term occlusion. A repeat endovascular treatment is recommended in cases of restenosis for recanalization of the affected venous axis.

In conclusion, thoracic duct ligation must be considered the surgical treatment of choice in patients with CTaC. However, endovascular techniques could be a valid alternative in selected patients to re-establish the venous and lymphatic flow. As the authors mentioned, we agree that the role of interventional recanalization of CVT for treating CTaC is under-reported despite recommendations for aggressive treatment of CVT in such patients. Hence, we still need more scientific evidence to standardize the use of these endovascular techniques in this specific challenging clinical scenario.

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