See Article page 275.



# Commentary: Managing catastrophic antiphospholipid syndrome—do we have a way out?

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**CENTRAL MESSAGE**

Catastrophic antiphospholipid syndrome is a rare, underdiagnosed and highly devastating clinical entity that warrants systematic, multidisciplinary involvement to improve cardiac surgery outcomes.

Catastrophic antiphospholipid syndrome (CAPS) is a rare yet devastating autoimmune disorder that is characterized by antiphospholipid antibody-related clinical complications such as recurrent arterial and venous thromboembolisms that occur in the setting of progressive microvascular thrombosis. CAPS is triggered by surgery, infection, or changes in anticoagulation therapy.[1] About one-third of patients with primary APS and CAPS have cardiac sequelae that usually manifest as valve masses (nonbacterial vegetations) or thickening, which lead to valve dysfunction.[2,3] Although cardiac surgery is often considered in select cases to prevent recurrent embolization from these valvular lesions, most commonly the mitral valve, patients often develop progressive multiorgan failure and mortality.[1-3] Current literature on CAPS following cardiac surgery with de novo diagnosis of APS is limited.

El-Dalati and colleagues[4] highlight an unusual case of a 58-year-old patient presenting with culture-negative mitral valve bileaflet vegetations who subsequently underwent an uneventful mitral valve repair with annuloplasty ring and bileaflet valve debridement. Unfortunately, the patient's postoperative course was complicated by unclear thrombocytopenia, multiple arterial thrombi, and a deep vein thrombosis. Postoperative testing was positive for APS, which was negative before surgery. Despite extensive treatment with anticoagulation, steroids, immunoglobulins, and even plasma exchange, the patient's clinical condition declined, eventually leading to multisystem organ failure with bowel ischemia and pulmonary embolus that resulted in the patient's death.

Although this case demonstrates a devastating clinical course of a patient with de novo CAPS, it provides some food for thought as we seek to better understand the disease pathophysiology in the setting of cardiac surgery. Although the current literature is not well elucidated, the increased risk of thrombosis is believed to be due to withdrawal of preoperative anticoagulation, intraoperatively due to inadequate anticoagulation during cardiopulmonary bypass, or postoperatively before the achievement of adequate anticoagulation.[1,3,5] Catastrophic exacerbation of APS is most concerning, but is often underdiagnosed in postoperative settings because it can resemble other syndromes such as heparin-induced thrombocytopenia or disseminated intravascular coagulation.[3,5]

It is thus vital for teams to establish multidisciplinary efforts with hematology or vascular medicine early on in preoperative settings for patients who present with nonbacterial thrombotic endocarditis, although nonbacterial thrombotic endocarditis diagnosis is often difficult due to the underlying disease (eg, cancer, autoimmune disorders, or HIV) and relies on strong clinical suspicion at the time of surgery given its

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rarity. Likewise, it is equally important for cardiac surgery teams to establish prompt consultations with hematology or vascular medicine specialists postoperatively in patients who develop de novo thrombosis after cardiac surgery.[3] These multidisciplinary approaches may help tailor treatment strategies that minimize the dire consequences of APS or CAPS. At the same time, we believe that further research in creating an international registry will open a broader window to provide further insight and answers that would help predict and optimize management of CAPS after cardiac surgery. Until then, there seems to be no definite way out without compromising on patient outcomes.

## References

1. Weiss S, Nyzio JB, Cines D, Milas BL, Narula W, Floyd TF. Antiphospholipid syndrome: intraoperative and postoperative anticoagulation in cardiac surgery. *J Cardiothorac Vasc Anesth*. 2008;22:735-9.
2. Hojnik M, George J, Ziporen L, Shoenfeld Y. Heart valve involvement (Libman-Sacks endocarditis) in the antiphospholipid syndrome. *Circulation*. 1996;93:1579-87.
3. Koniari I, Siminelakis SN, Baikoussis NG, Papadopoulos G, Goudevenos J, Apostolakis E. Antiphospholipid syndrome; its implication in cardiovascular diseases: a review. *J Cardiothorac Surg*. 2010;5:101.
4. El-Dalati S, Suffoletto M, Rivosecchi RM, Kaczorowski D. Catastrophic antiphospholipid syndrome in the setting of mitral valve repair. *J Thorac Cardiovasc Surg Tech*. 2021;10:275-7.
5. Pengo V, Ruffatti A, Legnani C, Gresele P, Barcellona D, Erba N, et al. Clinical course of high-risk patients diagnosed with antiphospholipid syndrome. *J Thromb Haemost*. 2010;8:237-42.