Secondary sclerosing cholangitis following cardiac surgery: An uncommon but deadly gastrointestinal complication

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Gastrointestinal complications following cardiac surgery carry a mortality rate of >30%.^{1,2} Secondary sclerosing cholangitis in a critically ill patient (SC-CIP) is a rare cholangiopathy associated with a grim prognosis that may go unrecognized in patients undergoing cardiac surgery. SC-CIP is a cholestatic process leading to biliary obstruction, with ischemia and biliary destruction as the central pathological features.³ The diagnosis of SC-CIP requires endoscopic retrograde cholangiopancreatography (ERCP), and liver transplantation is the sole curative treatment option.⁴ Contemporary critical care management and the increased use of mechanical circulatory support allows long-term survival of patients with postcardiotomy shock; however, failure to recognize postsurgical complications such as SC-CIP, with high mortality and limited treatment options, may prolong futile resuscitation efforts.¹⁻⁷

In this issue of the *Journal*, Schade and colleagues present a single institution experience of 12 patients who developed SC-CIP following cardiac surgery with the aim of to increasing the awareness of this rare and often fatal complication.⁸ None of the patients had evidence of preexisting liver dysfunction. The diagnosis of SC-CIP was often delayed, underscoring the difficulty in recognizing this complication. Although elevated liver enzyme values are commonly seen in patients in the intensive care unit (ICU; ~30%), the incidence of SC-CIP following cardiac surgery, also preceded by liver enzyme elevations, was low (0.14%). However, the mortality rate in the patients with SC-CIP was sobering, with only a single survivor. All of the patients with SC-CIP had postcardiotomy shock, necessitating prolonged and complicated ICU admission. Notably, 50% of the patients with SC-CIP required mechanical circulatory support with extracorporeal membrane oxygenation or postoperative intra-aortic balloon pump insertion.

Based on their comparative analysis (cases matched 3:1 with controls with similar demographic variables and cardiac procedures), the authors identified several distinct clinical features in the patients with SC-CIP. In addition mechanical circulatory support, these patients required prolonged mechanical ventilation and inotropic administration

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Central Message

Secondary sclerosing cholangitis after cardiac surgery is rare entity with poor survival. Endoscopic drainage of the bile ducts is palliative, but liver transplantation remains the sole therapeutic option.

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(norepinephrine in particular). Reexploration was performed in >80% of the patients. Common indications for reoperation included hemorrhage, wound healing complications, and mediastinitis (median, 3 repeat procedures per patient). Liver enzymes (alkaline phosphatase, gamma-glutamyl transferase, and bilirubin) rose to higher levels in the patients with SC-CIP compared with matched controls; however, despite these marked liver enzyme abnormalities, the diagnosis of SC-CIP was commonly delayed. In most patients, confirmatory ERCP was performed more than 3 weeks after surgery (median, 25 days). ERCP-directed diagnosis and drainage was palliative, with curative treatment in the sole survivor achieved by liver transplantation. The prognosis in patients with SC-CIP was grim, with an overall mortality in this series of 92%. Death was most frequently related to multi-organ system failure after a mean ICU admission of 45 days.

This report adds another condition to the list of lethal gastrointestinal complications that can occur after cardiac surgery. Increased awareness of SC-CIP by cardiothoracic surgeons and intensivists should allow for earlier diagnosis. Palliation is the goal in most cases, with definitive liver transplantation limited to highly selected patients with SC-CIP.

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