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Fatal intraoperative thrombosis in contemporary adult thoracic aortic surgery requiring deep hypothermic circulatory arrest: Observations from the literature, 1993–2006

John G. T. Augoustides, MD, FASE, Philadelphia, Pa

Fatal intravascular coagulation in the setting of thoracic aortic surgery with deep hypothermic circulatory arrest (DHCA) was highlighted in a case series (n = 20) that identified exposure to aprotinin as a significant risk factor.¹ The findings in this case series have since been attributed to inadequate heparinization because it was only appreciated thereafter that aprotinin prolongs the celite-based activated clotting time (ACT).² However, even in the presence of adequate heparinization by kaolin-based ACT or heparin concentration, case reports of fatal thrombosis in thoracic aortic surgery with DHCA have persisted (n = 5, 1998–2006).^{2–5} The purpose of this brief communication is to review this case series as a platform from which further research can be planned. The literature search was conducted with PubMed (last entry: May 21, 2007) with the following search terms: thrombosis and DHCA; aprotinin and thrombosis in DHCA; aminocaproic acid and thrombosis in DHCA; and tranexamic acid and thrombosis in DHCA.

As a group, the published case reports of adult DHCA thrombosis can be summarized as follows^{2–5}:

1. The 5 patients were all older than 60 years (2 men and 3 women).
2. All underwent thoracic aortic procedures with DHCA.
3. Factor V Leiden was identified in 2 of the 5 patients.^{4,5}
4. All patients had antifibrinolytic therapy (3 with aprotinin and 2 with aminocaproic acid).
5. Anticoagulation was titrated with heparin to maintain the ACT at longer than 500 seconds, the heparin concentration

at greater than 2.7 U/mL, or both. The kaolin-based ACT was always used in the presence of aprotinin.

6. Circulatory collapse caused by intravascular thrombosis began in all cases within 60 minutes of separation from cardiopulmonary bypass and protamine administration.
 7. The circulatory collapse was life-threatening and resistant to pharmacologic support. In all cases it necessitated return to cardiopulmonary bypass.
 8. The degree of thrombosis was massive, involving the heart chambers, thoracic aorta, pulmonary arteries, and great veins. In 4 of 5 patients, the disseminated thrombus was evident intraoperatively. In the fifth patient autopsy revealed extensive thrombosis within the small pulmonary arteries that was responsible for fatal intraoperative right ventricular failure.
 9. The syndrome caused intraoperative death in all 5 patients.
- Of course, this small case series has the usual inherent limitations. There are many more questions than answers. However, the following observations might serve to direct further investigation of this lethal syndrome to improve its management:
1. A worldwide case registry would allow a greater case number for further in-depth analysis.
 2. Hypercoagulable states, such as Factor V Leiden, might have a role in the pathophysiology of this syndrome.
 3. In the presence of an antifibrinolytic, standard-of-care anticoagulation with heparin for adult DHCA does not protect all patients against serious intraoperative thrombotic complications.
 4. Aprotinin is not the only antifibrinolytic associated with this syndrome. To date, no cases of this syndrome have been reported in association with tranexamic acid.
 5. When a case occurs, the following considerations might improve clinical care:
 - a. Blood samples from the patient should be analyzed in consultation with a hematologist to identify possible mechanisms.
 - b. The index patient and family should be screened for underlying hypercoagulable states.
 - c. Patient autopsy might be required to determine the mechanism of death, especially if the thrombosis is not evident intraoperatively.

From the Cardiothoracic Section, Anesthesiology and Critical Care, Philadelphia, Pa.

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Address for reprints: John G. T. Augoustides, MD, FASE, Cardiothoracic Section, Anesthesiology and Critical Care, Dulles 680, HUP, 3400 Spruce St, Philadelphia, PA 19104-4283 (E-mail: yiandoc@hotmail.com).

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Despite the major advances in thoracic aortic surgery with DHCA, there are still major clinical challenges, including this thrombotic syndrome in the face of standard-of-care anticoagulation. Further collaborative research should advance our understanding and management of this thrombotic syndrome in the future.

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An innovative technique for the relief of right ventricular trabecular cavity obliteration in endomyocardial fibrosis

Ana Olga Mocumbi, MD,^a Daniel Sidi, MD, PhD,^b Pascal Vouhe, MD, PhD,^a and Magdi Yacoub, FRS,^a London, United Kingdom, Maputo, Mozambique, and Paris, France

Endomyocardial fibrosis (EMF) mainly affects persons from Africa, South America, and Asia.¹ The pathogenesis of this condition remains unknown. In advanced forms, EMF produces marked disability and carries a poor prognosis. Although there is consensus about the potential value of surgical intervention in symptomatic patients, there is still debate regarding the exact timing and the surgical technique to be used.^{2,3}

EMF is characterized by endocardial fibrosis affecting the inflow tract and the apex of 1 or both ventricles, commonly involving the atrioventricular valves. The right ventricle is affected in most cases.⁴ In severe forms, marked reduction of ventricular volume is thought to be due to the presence of a plug of fibrous tissue involving both the trabecular part and the apex.⁵

We here describe a new mechanism for apical obliteration of the right ventricle in EMF. The concept was used to evolve and apply a new surgical technique to increase ventricular volume, improve contractile function by releasing the myocardium and making use of viable myocardium in the obliterated area, and correct the tricuspid regurgitation.

This technique was conceived after detailed echocardiographic examination showed a layer of endocardial fibrosis forming an artificial floor to the right ventricle and isolating the trabecular part from the rest of the ventricular cavity (Figure 1, A).

After establishing cardiopulmonary bypass with moderate hypothermia, cold blood antegrade cardioplegia was used for myocardial protection. A wide longitudinal right atriotomy was performed to expose the tricuspid valve, which was usually markedly dilated, allowing wide access to the right ventricle. The distribution and extent of pathologic involvement of the tricuspid valve apparatus and right ventricular cavity were carefully defined.

In patients with obliteration of the trabecular part of the right ventricle described in this article, white glistening fibrous tissue 2 to 3 mm in thickness was found lining the inflow tract and extended down into the cavity of the right ventricle to the level of the junction of the inflow and trabecular parts. The fibrotic process produced a relatively thin layer that formed an "artificial floor" to the right ventricle, separating the inflow cavity from the obliterated muscular trabecular part. The fibrotic process spared the tricuspid leaflets and chordae.

Endocardial resection was started near the tricuspid annulus by retracting the leaflets of the valve. If the latter was fused in some areas, it was mobilized if at all possible. After the development of a cleavage plane by means of sharp dissection, a combination of sharp and blunt dissection was used to excise the thick, fibrous endocardial lining. This process was continued into the ventricular cavity, ensuring preservation and mobilization of the tricuspid valve chordae and papillary muscles. The membrane covering the entry into the trabecular part was removed (Figure 1, B), exposing the fused muscular tissue underneath it. This was followed by recreating a cavity inside the trabecular part by mainly separating the fused trabeculae but, if necessary, also excising some muscular tissue, taking care not to perforate the ventricular wall.

From Imperial College, London, United Kingdom, and Instituto do Coração, Maputo, Mozambique^a; and Hopital Necker Enfants Malades, Paris, France, and Instituto do Coração, Maputo, Mozambique.^b

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Address for reprints: Magdi Yacoub, FRS, Heart Science Centre–Harefield, Middlesex UB9 6JH, United Kingdom (Email: m.yacoub@imperial.ac.uk).

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