Comment

At present, there is no universally accepted definition for vasoplegic syndrome [1, 3]. From a survey of trials using methylene blue for vasoplegic syndrome, accepted parameters for defining vasoplegic syndrome are a systemic vascular resistance of less than 800 dyne · s · cm⁻², an MAP of less than 60 to 65 mm Hg, a cardiac index of greater than 2.5 to 3 L · min⁻¹ · m⁻², and a requirement for at least one or more high-dose pressors (ie, norepinephrine >0.05 μg · kg⁻¹ · min⁻¹). If one or more of these conditions are met either during cardiopulmonary bypass or within 24 hours after cardiopulmonary bypass, then it is generally accepted that this constitutes vasoplegic syndrome [1].

The most widely accepted theories on the mechanism of vasoplegic syndrome involve dysregulation of nitric oxide homeostasis [1, 3]. It has been demonstrated that methylene blue is capable of binding nitric oxide (a reactive oxygen species), inhibiting both constitutive and inducible nitric oxide synthase, and inhibiting soluble guanylate cyclase [2, 3]. Unfortunately, methylene blue has now been shown to precipitate serotonin syndrome when administered to patients taking a serotonergic antidepressant as a result of a direct inhibitory effect of methylene blue or its metabolite on monoamine oxidase activity [2, 4].

Clinically, serotonin syndrome is characterized by the triad of altered mental status, neuromuscular hyperactivity, and autonomic instability [5]. Many patients with heart failure are also taking an antidepressant. Switching antidepressants or stopping them altogether for several days before undergoing cardiopulmonary bypass can decrease the risk of serotonin syndrome. Our patient was taking citalopram. An alternative therapy for the treatment of vasoplegic syndrome was, therefore, needed.

Hydroxocobalamin is one of four forms of vitamin B₁₂ found in the body. High-dose hydroxocobalamin has been used for more than 40 years in Europe for the treatment of cyanide poisoning. No significant adverse events have been reported, even with doses as high as 30 g within 24 hours [6]. In recent years hydroxocobalamin has been approved for use in the United States for the same indication (cyanide poisoning) and is currently marketed under the trade name Cyanokit. Cyanokit is supplied as a kit, typically containing a single 5-g vial of powdered hydroxocobalamin, which is then reconstituted with 200 mL of a diluent (not included in the kit). This may be either normal saline solution, lactated Ringer’s solution, or dextrose 5% in water for a final concentration of 25 mg/mL. This may be infused during a period of 15 minutes to 2 hours, and may be immediately followed by a second 5-g dose if necessary. There are currently no contraindications to its use, and it is not known to interact with methylene blue.

A side effect of Cyanokit administration is a rapid, sustained, and significant increase in blood pressure. It has been demonstrated that this is caused by direct binding of nitric oxide and direct inhibitory effects on nitric oxide synthase and soluble guanylate cyclase [7, 8]. Unlike methylene blue, hydroxocobalamin is not known to have any direct effects on the serotonergic pathway and has not been associated with an increased risk of serotonin syndrome.

This case is important because it represents the first report on the use of high-dose hydroxocobalamin for vasoplegic syndrome. More importantly, it also represents a viable alternative to methylene blue for the treatment of vasoplegic syndrome in patients taking a serotonergic antidepressant. The use of hydroxocobalamin in this patient population presents a novel solution to this problem, and may have additional clinical benefits beyond its effects on blood pressure, which will require further evaluation.

References


Thoracoabdominal Aortic Repair in a Patient With Ehlers-Danlos Syndrome

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Type IV Ehlers-Danlos syndrome is a life-threatening inherited disorder of connective tissue associated with multiple aneurysm formation. Thoracoabdominal aortic repair in these patients has rarely been performed.

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We report the case of a 13-year-old patient with Ehlers-Danlos syndrome who had multiple aortic operations from the ascending aorta to the thoracoabdominal aorta.

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Type IV Ehlers-Danlos syndrome is a life-threatening inherited disorder of the connective tissue associated with multiple aneurysm formation, spontaneous rupture, and dissection. Here we report the case of a patient with Ehlers-Danlos syndrome who had multiple aortic operations from the ascending aorta to the thoracoabdominal aorta.

A 7-year-old boy whose elder brother had died at 7 years of age because of a ruptured thoracic aortic aneurysm had chest pain and was admitted to a hospital. Computed tomography (CT) revealed a type A acute aortic dissection. He was referred to our hospital to receive an ascending aorta and aortic arch replacement using a 20-mm Hemashield branched graft (MAQUET Holding GmbH & Co KG, Rastett, Germany). His postoperative course was uneventful. Microscopic examination of a skin biopsy sample confirmed the diagnosis of type IV Ehlers-Danlos syndrome in this patient. He was followed at our outpatient clinic after discharge, and follow-up CT was performed annually.

Five years after the patient’s first operation, CT demonstrated progressive enlargement of his descending thoracic aorta. The thoracic aorta, now 41 mm in diameter, had been 33 mm in diameter 1 year before. Because of its rapid growth, we decided to perform descending thoracic aortic replacement using a 22-mm Triplex graft (Terumo Corp, Tokyo, Japan). A proximal anastomosis was made to the previous prosthetic graft and a distal anastomosis to the native aorta above the diaphragm was performed. No intercostal arteries were reattached. Postoperatively, chylothorax was observed, and repeated thoracotomy was performed to ligate the thoracic duct. He was discharged 3 weeks after that operation without any other problems.

One year later, follow-up CT demonstrated a dilated aorta extending from above the celiac artery to above the inferior mesenteric artery. The thoracoabdominal aorta was 47 mm in diameter. We planned a repair of the thoracoabdominal aorta using partial cardiopulmonary bypass. The right femoral artery and vein were exposed for cardiopulmonary bypass. The previous prosthetic graft and thoracoabdominal aorta were exposed through a left thoracoabdominal incision. The proximal aortic clamp was placed at the previous graft and the distal aortic clamp was placed approximately 3 cm distal to this. After aortic clamping, partial cardiopulmonary bypass was initiated to preserve the viscera and thoracolumbar spinal cord. The aorta was divided between clamps and proximal end-to-end anastomosis to the previous graft was performed using a 22-mm Vascutek branched graft (Terumo Corp, Tokyo, Japan), after which another distal aortic clamp was applied just above the celiac artery. The aneurysm was opened longitudinally, and the 10th and 11th intercostal arteries were separately anastomosed to the 10-mm tube grafts. The tube grafts were anastomosed proximally to the Vascutek graft, and perfusion of the intercostal arteries was resumed. After the clamp was moved to the distal aorta, the celiac, superior mesenteric, and bilateral renal arteries were immediately cannulated and perfused. Each visceral branch was separately reconstructed. Finally, the distal end of the Vascutek graft was anastomosed to the aorta above the inferior mesenteric artery. The aortic clamp was removed and the patient was taken off cardiopulmonary bypass. He had an intracranial hemorrhage postoperatively; however, it resolved with no neurologic dysfunction. Postoperative CT demonstrated patency of all branches (Fig 1).
Comment

There are few reports of thoracoabdominal repair in patients with Ehlers-Danlos syndrome [1]. To the best of our knowledge, this is the first case report of a thoracoabdominal repair in a child with Ehlers-Danlos syndrome.

Arterial aneurysms in children with Ehlers-Danlos syndrome usually involve the aorta, whereas in adults, multiple aortic and peripheral artery aneurysms have been reported [2, 3]. Oka and colleagues [4] reported 4 graft replacement procedures, including the aorta and peripheral arteries, in the same patient. Patients with type IV Ehlers-Danlos syndrome are prone to rupture of large arteries through dissection or trauma, and the fragility of the arterial wall makes surgical repair extremely difficult [1, 5]. Our patient did not have translucent skin or hypermobility of joints; however, he had a family history of vascular death and his clinical presentation suggested a connective tissue disorder.

In the present case, branches of the aorta were repaired separately. In particular, 2 intercostal arteries were individually anastomosed to the interposed tube grafts. We believe that compared with reconstruction with an aortic cuff inclusion procedure, individual reconstruction of the intercostal arteries prevents further aneurysmal dilatation at the reconstructed site in patients with connective tissue disorders, even though the patency rate of intercostal artery reconstruction using an interposed graft has been reported to be inferior to that achieved with the aortic cuff technique [6]. In the present case, the patency of the intercostal arteries was confirmed on postoperative CT.

In conclusion, thoracoabdominal aortic repair in a child with Ehlers-Danlos syndrome is rare and was safely performed. Close follow-up should be considered in these patients, because multiple aortic aneurysms may be detected.

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References


Fragmentation Injury to the Innominate Artery in a Three-Year-Old Child

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The management of vascular trauma in pediatric patients presents numerous challenges, especially in an austere environment. We present the case of a 3-year-old girl who sustained multiple fragmentation injuries to the right chest and right upper extremity as a result of combat activity in Iraq. This resulted in an occult pseudoaneurysm of the innominate artery identified during exploration of her right chest for a persistent air leak from the right side of the chest. Computed tomography angiography delineated the injury, which was surgically repaired. This report demonstrates the type of challenging cases encountered in a combat zone and illustrates the need for a national database of such injuries in pediatric patients to better inform surgical decision making.


We present the case of a 3-year-old girl who presented to our level 3 combat facility after an explosion that resulted in multiple fragmentation injuries to her right arm and chest. On initial evaluation, she was found to have soft tissue injuries to her right upper chest and right arm as well as a right pneumothorax. No other injuries were identified on tertiary survey or computed tomography.