

Fatal Autonomic Dysfunction Due to Guillain-Barré Syndrome After Cardiac Surgery

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Guillain-Barré syndrome, a rare peripheral neuropathy, appears to occur more often in patients who have recently undergone surgery than in the general population. However, the pathophysiologic relationship between surgery and Guillain-Barré syndrome is elusive. Few cases of Guillain-Barré syndrome after cardiac surgery have been reported. Autonomic dysfunction, a serious complication of Guillain-Barré syndrome, has not been previously reported after cardiac surgery.

We describe the case of a 71-year-old woman in whom the acute motor axonal neuropathic subtype of Guillain-Barré syndrome developed after mitral valve replacement. Despite plasmapheresis and intravenous immunoglobulin therapy, she died of complications from severe autonomic dysfunction 25 days postoperatively. Recognizing the potential cardiovascular involvement of Guillain-Barré syndrome is important, because patients who undergo cardiac surgery can be vulnerable to autonomic dysfunction in the early postoperative period. (Tex Heart Inst J 2022;49(3):e207439)

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Guillain-Barré syndrome (GBS) is a rare immune-mediated disease that causes acute polyradiculoneuropathy.¹ The estimated annual incidence of GBS is 1 to 2 cases per 100,000 people.² Infection is a well-known trigger of GBS. However, vaccines, immunologic treatments, trauma, malignancy, and surgery may also be associated with GBS.¹

The syndrome usually presents as bilateral, progressive, ascending weakness in the extremities. Neurologic sequelae persist in 15% to 20% of patients who have GBS. In severe cases, the risk of death can exceed 10%.² Death is typically due to complications of prolonged neurologic deficits or severe autonomic dysfunction.

Neurologic complications that occur after cardiac surgery, especially aortic surgery, are chiefly cerebrovascular and rarely peripheral. Guillain-Barré syndrome occurs more frequently in patients who have recently undergone surgery than in the general population.³ However, only a few cases of GBS after cardiac surgery have been reported (Table I).⁴⁻⁹ We report the case of an elderly woman who underwent mitral valve surgery and then experienced life-threatening autonomic dysfunction caused by a variant of GBS.

Case Report

A 71-year-old woman presented at our hospital with dyspnea during daily activity. An echocardiogram revealed severe rheumatic mitral stenosis and severe secondary tricuspid regurgitation. The patient had no history of recent infection or chronic disease other than autoimmune thyroiditis, and her thyroid hormone levels were normal. We performed mitral valve replacement and tricuspid valve repair by means of open surgery. Local dissection was detected in the ascending aorta immediately after decannulation, and the ascending aorta was replaced with use of antegrade selective cerebral perfusion.

On postoperative day (POD) 1, the patient was conscious and alert; however, her motor response to verbal commands was limited. A manual muscle test revealed severe muscular weakness in all extremities that was more pronounced proximally (grade 1/5

in the lower extremities and grade 2/5 in the upper) than distally (grades 2/5 and 3/5, respectively). A deep tendon reflex examination revealed generalized areflexia. No sensorial impairment or cranial nerve dysfunction was detected. Serial radiologic results, including cranial and spinal magnetic resonance images, ruled out pathologic conditions of the central nervous system.

The patient's clinical condition did not improve, prompting an additional neurologic consultation. Progressive muscle weakness was detected in her extremities, and she was placed on a ventilator for respiratory support. The negative imaging results and the patient's clinical state were consistent with GBS; however, this is rare after cardiac surgery, so the diagnosis was confirmed after a nerve conduction study and lumbar

puncture. On POD 5, cerebrospinal fluid test results indicated albuminocytologic dissociation, typical of GBS. Nerve conduction studies revealed low-amplitude, compound muscle action potentials with no demyelination. Together, the patient's clinical condition and test results were consistent with the acute motor axonal neuropathic (AMAN) subtype of GBS.

Five plasmapheresis treatments (250 mL/kg/d of plasma each) substantially improved the patient's condition. By POD 15, her muscular strength had improved to grade 4/5 in the upper extremities and grade 3/5 in the lower extremities. However, 3 weeks postoperatively, her neuromotor status again began to deteriorate. The patient's bilateral muscular weakness progressed rapidly. By POD 21, she was completely quadriplegic, and she

TABLE I. Reports of Guillain-Barré Syndrome After Cardiac Surgery

Reference	Pts. (n)	Age (yr), Sex	Operation Type	Symptom Onset (POD)	Clinical Presentation	Treatment	Outcome
Renlund DG, et al. ⁴ (1987)	1	65, M	Elective on-pump CABG	8	Weakness; paresthesia in legs	Plasma-pheresis	Hospital discharge with sequelae (POD 22)
Hogan JC, et al. ⁵ (1992)	2	60, M	AVR and MVR	23	Progressive weakness in legs	Plasma-pheresis	Substantial improvement 4 wk after operation
		53, M	Elective on-pump CABG	14	Progressive weakness in legs; intubation	Plasma-pheresis	Substantial improvement 6 wk after operation
Punith K, et al. ⁶ (2011)	1	65, M	Elective on-pump CABG	12	Paresthesia; proximal muscle weakness in legs; bilateral facial nerve palsy; areflexia	IVIg	Full recovery 10 wk after operation
Cingoz F, et al. ⁷ (2012)	1	67, M	Elective off-pump CABG	2	Weakness; paresthesia in legs	Plasma-pheresis	Full recovery (POD 10)
Aldag M, et al. ⁸ (2017)	1	50, M	Emergency on-pump CABG	5	Ataxia; left-sided ptosis; weakness; paresthesia in legs; dysphagia; dyspnea (Miller-Fisher syndrome)	Plasma-pheresis and IVIg	Death (POD 9)
Raut MS, et al. ⁹ (2019)	1	32, M	Emergency RSOV aneurysm repair	2	Proximal muscle weakness; absent DTR; bilateral vocal cord paralysis (tracheostomy)	Plasma-pheresis	Substantial improvement (POD 28)
Current case	1	71, F	MVR; tricuspid valve repair; ascending aorta replacement	1	Proximal muscle weakness; absent DTR; autonomic dysfunction	Plasma-pheresis and IVIg	Death (POD 25)

AVR = aortic valve replacement; CABG = coronary artery bypass grafting; DTR = deep tendon reflexes; F = female; IVIg = intravenous immunoglobulin; M = male; MVR = mitral valve replacement; POD = postoperative day; RSOV = ruptured sinus of Valsalva

had fluctuating arterial tension and refractory sinus tachycardia. Diagnostic results ruled out a new-onset cerebrovascular event. Repeat echocardiograms showed a functioning mitral bioprosthesis, minimal tricuspid regurgitation, and a normal left ventricular ejection fraction. Intravenous immunoglobulin (IVIg) therapy (0.4 g/kg/d) was planned for the next 5 days. Despite the first 3 days of IVIg therapy and advanced hemodynamic support, the patient died on POD 25 of persistent vasoplegia.

Discussion

Postoperative GBS is defined as the onset of GBS symptoms within 6 to 8 weeks of a surgical procedure.^{3,10} Some have speculated that postsurgical GBS is triggered by the neuroendocrine response to surgical trauma, transient immunosuppression during operation, impairment of the blood-nerve barrier, immune dysregulation triggered by pharmacologic agents, concomitant malignancy, and underlying infectious conditions.^{3,10,11} Only a few cases of GBS after cardiac surgery have been reported.⁴⁻⁹

Guillain-Barré syndrome is a clinical diagnosis, and a typical finding associated with it is albuminocytologic dissociation (elevated protein levels and normal cell count) in the cerebrospinal fluid. In clinical practice, nerve conduction studies are important for supporting the diagnosis, evaluating the treatment response, and determining the GBS subtype; however, they are not essential for making a diagnosis. The typical GBS variant in Western countries, acute inflammatory demyelinating polyneuropathy (AIDP), usually presents with progressive weakness and mild to moderately severe sensory symptoms. In some regions, especially East Asia, the AMAN subtype is seen in a substantial proportion of patients.¹² This variant is characterized by rapidly progressive ascending tetraparesis and by respiratory system dysfunction. Unlike the demyelinating GBS subtypes, the AMAN variant affects sensory nerves minimally or not at all. Electrodiagnostic criteria are used in classifying variants.¹ In one study,¹³ the incidence of AMAN was higher in patients with postoperative GBS than in patients with classical GBS. We found no other reported case of GBS after cardiac surgery that was clearly defined as the AMAN variant.

The primary treatments for sequelae related to GBS—plasmapheresis and IVIg—have similar efficacy.^{14,15} Approximately 10% of patients with GBS recover initially, but then their condition deteriorates.¹⁶ This phenomenon, called treatment-related fluctuation, has a poor prognosis. Our patient's neurologic symptoms improved after plasmapheresis; however, her status deteriorated drastically when plasmapheresis was stopped. Changing treatments after clinical deterioration does not provide extra benefit¹⁷; however, because

of our patient's hemodynamic alterations, we decided to administer IVIg instead.

In GBS, autonomic dysregulation involving the cardiovascular system may cause morbidity and death. Elevated catecholamine levels, denervation hypersensitivity, and impaired baroreceptor mechanisms may be responsible for autonomic dysfunction. In a recent study, the incidence of autonomic dysfunction in patients with GBS was higher in those who had undergone surgery than in those who had not.¹³ Severe autonomic involvement is more pronounced in the AIDP than in the AMAN subtype,¹³ and life-threatening autonomic dysfunction in the AMAN subtype usually occurs in the presence of severe neurologic deficits.¹⁷ Only one other reported case of GBS after open heart surgery has proved fatal; however, the authors did not emphasize an association with autonomic involvement.⁸ To our knowledge, this is the first report of GBS after cardiac surgery in which a patient presented with cardiovascular involvement caused by autonomic dysfunction.

Conclusion

Guillain-Barré syndrome rarely develops after cardiac surgery. Diagnosis of GBS is clinical and can be challenging. Although severe autonomic dysfunction due to the AMAN variant of GBS is rare, it should be considered in the differential diagnosis because it may cause severe sequelae and even death.

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