Guillain-Barré syndrome is an acute inflammatory demyelinating polyradiculopathy characterized by progressive motor weakness, areflexia, and ascending paralysis. Guillain-Barré syndrome is extremely rare in pregnant patients, and there are no established guidelines for delivery or safest anesthetic methods. We report a Cesarean delivery in the case of a 32-year old woman who was diagnosed with Guillain-Barré syndrome 18 weeks into gestation. Tracheostomy was performed due to progressive respiratory muscle weakness and respiratory failure, and ventilator support was required in the intensive care unit. The respiratory difficulty was exacerbated by the growth of the fetus, necessitating emergency Cesarean delivery. The delivery was successfully performed under general anesthesia, and the patient recovered without neurological sequelae. (Korean J Anesthesiol 2013; 64: 268-271)

Key Words: Cesarean section, General anesthesia, Guillain-Barré syndrome, Pregnancy, Tracheostomy.

The incidence of Guillain-Barré syndrome has been reported as 0.75 to 2 cases per 100,000 persons per year. Patients with Guillain-Barré syndrome usually have a history of upper respiratory tract infection or gastroenteritis 1 to 3 weeks prior to the onset of the disease [1]. Guillain-Barré syndrome starts with weakness of the extremities and can progress to weakness in the trunk, cervical area, facial muscles, and even respiratory muscles, causing respiratory failure in severe cases. Patients may also present with sensory symptoms, including cranial nerve deficits and autonomic nervous system dysfunction.

Guillain-Barré syndrome during pregnancy is very rare. We report a case of Cesarean delivery under general anesthesia in a 32-year-old woman who was diagnosed with Guillain-Barré syndrome at 18 weeks into gestation, and who eventually required ventilation via a tracheostomy.
Case Report

A 32-year-old, 162 cm, 68 kg, gravida 1, para 1, pregnant woman received cesarean delivery due to failure of labor progression at 33 weeks and 6 days of pregnancy. The patient was diagnosed with gestational diabetes mellitus at 27 weeks of pregnancy with insulin therapy. She had a history of gastroenteritis at 17 weeks from gestation. Dysarthria and diplopia occurred after recovery from the gastroenteritis (18 weeks from gestation) and she was admitted to the emergency room (ER) and was diagnosed with Guillain-Barré syndrome. In the ER, the patient suffered from ongoing respiratory insufficiency (decreased oxygen saturation and CO₂ retention) along with mental decline. She was intubated shortly after arrival, transferred to the intensive care unit (ICU) and received mechanical ventilator care (Pressure controlled ventilation with synchronized intermittent mandatory ventilation mode, pressure support 8 cmH₂O, tidal volume 600 ml, frequency 12 per minute, FiO₂ 0.4). TPN started at 19 weeks from gestation for expecting prolonged fasting period. At 20 weeks from gestation, she eventually received tracheostomy (T-cannula I.D. 7.0) for prolonged ventilator care due to progressive respiratory muscle weakness. At 27 weeks from gestation, mechanical ventilator was converted to home ventilation Bi-level positive airway pressure (BPAP, BiPAP®). The setting was inspiratory positive airway pressure (IPAP) 8 cmH₂O, expiratory positive airway pressure (EPAP) 4 cmH₂O. And gradually started intermittent BPAP during night (29 weeks from gestation). At 33 weeks from gestation patient presented dyspnea and desaturation then continuous BPAP ventilation was applied again until delivery.

She underwent intravenous immunoglobulin (IVIG) therapy twice to prevent disease progression and enoxaparin therapy for the prevention of thrombosis. However, enoxaparin therapy was stopped due to the elevation of liver enzymes at 28 weeks from gestation. The patient suffered from various neurologic symptoms including binocular diplopia, dysarthria, facial diplegia and muscle weakness in upper and lower extremities (both upper and lower extremities: 3 to 4 of 5, Medical research council scale for muscle strength). The nerve conduction study showed electrophysiologic abnormalities suggesting motor-dominant polyneuropathy in bilateral upper and lower extremities. In the cerebrospinal fluid study, albumin level was 13.1 (10–30) mg/dl, and IgG 2.17 (0–8) mg/dl. Magnesium infusion started at 34 weeks from gestation for prevention of eclamptic seizure.

At 34 weeks from gestation the attending obstetrician judged that it was no longer possible to maintain the pregnancy due to further mental decline, elevation of liver enzymes, and respiratory difficulty. At first, induction of delivery was tried, but emergency cesarean section was decided to be performed with retardation of fetal heart rate. Preoperative evaluation was within normal limits, except elevation of liver enzymes (AST/ALT 372/458) and sinus tachycardia (120 beats per minute) on electrocardiogram.

Without premedication the patient was transferred from ICU to the operating room with manual ventilation via tracheostomy. Standard monitoring including electrocardiogram, pulse oxymeter and noninvasive blood pressure measurement was started. Upon arrival at the operating room, the blood pressure was 124/90 mmHg, heart rate was 118 beat per minute with pulse oxygen 98%. A bispectral index (BIS) was placed on the patient’s forehead to monitor depth of anesthesia and the leads for the nerve stimulator were placed at the ulnar side of the patient’s left forearm to monitor muscle relaxation. Under gentle manual assisted ventilation with 100% oxygen, approximately 500 ml of gastric juice was aspirated from the nasogastric tube. After the sterile surgical drape was confirmed, 250 mg of thiopental was injected via the intravenous line and ventilated manually with sevoflurane 1.5 vol% and oxygen. Muscle relaxation was checked with a nerve stimulator on 40mV stimulus. When the train of four (TOF) count of 4/4 was confirmed, 10 mg of rocuronium was administered and mechanical ventilation via tracheostomy was started, after the TOF count was 0/4 and BIS score was 55. Arterial catheter was placed at the patient’s right radial artery with a 20 Gage angiocatheter to monitor arterial blood pressure. Anesthesia was maintained with oxygen, medical air and sevoflurane 1.5 vol% on mechanical ventilation (tidal volume: 500 ml, frequency: 12/min, FiO₂: 0.5). The BIS score and TOF count were serially monitored (Table 1).

The baby (3,390 g, Apgar score: 5–7) was delivered at 8 minutes from the start of the surgery, and the placenta was extracted 2 minutes later. Oxytocin 30 IU (mixed with normal saline 100 ml) was administered intravenously slowly. Arterial blood gas analysis and blood sugar test (glucose 137 mg/dl) were performed 20 minutes after the induction of anesthesia.

| Table 1. BIS and TOF Count during the Surgery |
|----------------|---|---|---|---|---|---|---|---|---|---|---|
| Time (min) | 0 | 5 | 10 | 15 | 20 | 25 | 30 | 35 | 40 | 45 | 50 | 55 | 60 | 65 | 70 | 75 |
| BIS | 56 | 58 | 57 | 59 | 60 | 56 | 45 | 45 | 36 | 37 | 85 |
| TOF | 4/4 | 0/4 | 2/4 | 4/4 | 4/4 | 4/4 | 4/4 | 4/4 | 4/4 |

BIS: bispectral index, TOF: train of four.
Arterial blood gas analysis data and other electrolyte levels were in the normal range (pO₂ 146.3 mmHg, pCO₂ 40.7 mmHg, pH 7.425 at FiO₂ 0.5, EtCO₂ 35 mmHg). Intraoperative hemodynamic changes are shown in Fig. 1. No additional muscle relaxant was administered during surgery. Post-surgery TOF count was 4/4 and atropine 1 mg, and neostigmine 1.5 mg was administered to reverse the muscle relaxation. Fifteen minute after administration of reverse agents, patient’s spontaneous ventilation was recovered and she was sent to the ICU with assisted manual ventilation. The motor score evaluated upon ICU arrival was 3 to 4 in all extremity, similar to that of preoperative values. Total operation time was 55 minutes and anesthesia time was 75 minutes. Total infused crystalloid, colloid, estimated blood loss, and urine output were 200 ml, 500 ml, 500 ml and 100 ml, respectively.

Weaning of mechanical ventilation was tried on postoperative day (POD) 7 and successfully done on POD 25. Abrupt chest pain and respiratory difficulty with desaturation occurred on POD 32. CT angiography showed severe pulmonary thrombosis in both pulmonary arteries and deep vein thrombosis in the left lower extremity. She was diagnosed with pulmonary thromboembolism and treated with warfarin targeted at INR 2−2.5. Two months later, she was discharged and has been on outpatient department follow-up to the department of internal medicine, neurology, and rehabilitation medicine.

**Discussion**

Pregnant Guillain-Barré patients have a higher risk for neurological deficits, a respiratory failure rate of 35%, and a maternal mortality rate of 10 to 35% [2,3]. There is no clear evidence that termination of pregnancy can improve the outcome or facilitate the recovery of the mother, and previous studies have shown that uterine contraction is preserved and normal vaginal delivery is possible in these patients [4,5]. Thus, Guillain-Barré syndrome itself is not an indication for pregnancy termination or for Cesarean delivery. Nevertheless, owing to a lack of previous studies, no guidelines for delivery and anesthetic techniques have been established [2,3]. The appropriate mode of delivery and anesthetic management of the parturient patient with Guillain-Barré syndrome depend on the patient’s clinical condition at the time of delivery [1].

In the present case, an obstetrician judged that the general condition of the patient was severely compromised and that induction of delivery or emergency Cesarean delivery was essential. Louis et al. reviewed the medical records of 30 pregnant Guillain-Barré patients from 1986 to 2002 and found that preterm deliveries occurred in eight cases (34.7%); three had spontaneous labor, and five were iatrogenic premature deliveries due to deterioration of the maternal neurological condition or preeclampsia [4]. In our case, the patient had markedly elevated liver enzyme levels, respiratory difficulty, and mental decline, and the fetal heart rate had decreased, making iatrogenic premature delivery inevitable.

In patients with Guillain-Barré syndrome, both regional and general anesthesia may be performed. It has been reported that there is no superior mode of anesthesia, as administration of both regional and general anesthesia have each been associated with potential risks [4]. For general anesthesia in Guillain-Barré syndrome patients, succinylcholine should be avoided because of its risk of hyperkalemia [1]. Feldman reported that a parturient with Guillain-Barré syndrome had a cardiac arrest due to hyperkalemia that occurred shortly after succinylcholine administration for general anesthesia [6]. Non-depolarizing muscle relaxants should be administrated with caution, because they may result in prolonged neuromuscular block and postoperative mechanical or assisted ventilation [7]. The TOF count should be monitored from the beginning of induction of anesthesia to prevent overdosing of muscle relaxant. In this case, the patient was transfer to the ICU after the operation and full recovery of muscle relaxation was confirmed by TOF count.

There are controversies in regional anesthesia with Guillain-Barré syndrome patients. Steiner et al. [8] reported Guillain-Barré syndrome occurring one to two weeks after epidural anesthesia in three patients who had general surgery or delivery, mentioning that epidural anesthesia may have triggered Guillain-Barré syndrome. Wiertlewski et al. [9] reported a Guillain-Barré syndrome case, with worsening of symptoms after delivery via epidural anesthesia. The patient did not fully recover from the motor block after epidural anesthesia, and neurological symptoms worsened immediately after delivery. However, it would be unreasonable to generalize that regional anesthesia causes Guillain-Barré syndrome or that it makes Guillain-Barré syndrome worsen, solely based on several.
previous reports. Neurologic symptoms after the surgery can be affected by many patient-related, surgery-related and anesthesia related risk factors [10]. In addition, there is no clear evidence that epidural anesthesia causes Guillain-Barré syndrome yet. Several cases have been reported that epidural anesthesia was successfully performed during cesarean delivery of Guillain-Barré syndrome patients, in which all patients are fully recovered after the anesthesia [7,11,12]. Furthermore Hebl et al. reviewed the medical chart of 139 patients with a history of CNS disorder who received neuraxial anesthesia or analgesia from 1988 to 2000, and found no case of new or worsening neurologic symptoms. It was therefore concluded that adverse events after regional anesthesia to patients with CNS disorders are not as frequent as once thought and regional anesthesia should not be considered an absolute contraindication in these patients [10].

Autonomic nervous system instability is another important consideration for Guillain-Barré syndrome patients in pregnancy. Because Guillain-Barré syndrome patients may present with autonomic nervous system instability [13], it is more appropriate to use directly acting adrenergic agents rather than indirectly acting sympathomimetic agents due to unpredictability of the effect [7].

This patient had a pulmonary embolism a month after delivery as a complication of Guillain-Barré syndrome. The incidence of pulmonary embolism in non-pregnant Guillain-Barré syndrome patients has been reported to be between 1 to 13% [14]. Therefore prophylactic anticoagulation treatment is considered as a standard management in immobilized Guillain-Barré syndrome patients [15]. Gaber et al. [15] reported that the incidence of deep vein thrombosis was 7% and pulmonary thromboembolism was 4% despite of prophylactic anticoagulation in Guillain-Barré syndrome patients (including non-pregnant Guillain-Barré syndrome patients). Furthermore as pregnancy itself is a strong risk factor for thromboembolism, early prophylactic anticoagulation should be applied and other supportive care, including physiotherapy, compressive stockings and early ambulation after delivery, is highly recommended [4].

In this case, emergency cesarean delivery was successfully performed under general anesthesia without neurological exacerbation. Although there is no established guideline for delivery and anesthetic technique yet, Guillain-Barré patients with pregnancy should be evaluated individually and carefully anesthetized according to medical judgment by the anesthesiologist.

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