

## Case Report

Zimmermann S and Henningsson R. J Anesth Surg Rep 3: 118.

DOI: 10.29011/JASR-118.100018

# Perioperative Management of Patients with Chronic Inflammatory Demyelinating Polyradiculoneuropathy (CIDP) During Major Peripheral Vascular Surgery

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**Citation:** Zimmermann S, Henningsson R (2020) Perioperative Management of Patients with Chronic Inflammatory Demyelinating Polyradiculoneuropathy (CIDP) During Major Peripheral Vascular Surgery. J Anesth Surg Rep 3: 118. DOI: 10.29011/JASR-118.100018

**Received Date:** 11 June, 2020; **Accepted Date:** 19 June, 2020; **Published Date:** 24 June, 2020

## Abstract

Chronic inflammatory demyelinating polyradiculopathy (CIDP) is a rare autoimmune disease with a high risk of perioperative anesthetic complications. This case report describes a woman with CIDP, who was planned for major vascular surgery. This is to our knowledge, the first described case in vascular surgery with a combination of regional and general anesthesia.

**Keywords:** Anesthesia; CIDP; Vascular surgery

## Introduction

Chronic Inflammatory Demyelinating Polyradiculoneuropathy (CIDP) is a rare autoimmune disease that targets the peripheral nervous system but can also affect bulbar and respiratory muscles [1]. There are very few case reports on the anesthetic management of these patients [2-11], but when possible, nerve blocks seem to be the standard of care due to the increased risk of postoperative respiratory complications [2,11]. Vascular surgery patients per se represent a group of patients with the highest risk of perioperative complications.

## Case Report

The patient herein gave her consent for inclusion in this report. We present a 76 years old woman (152cm, 62 kg) with CIDP diagnosis since beginning of the 1990s. On the INCAT scale [12] she has grade 3 functional reductions in both her arms and legs and needs a walker inside and a wheelchair outside. She experiences no dyspnea at rest but reports on respiratory problems whenever she gets a cold. She has urinary incontinence and orthostatism, but denies problems when swallowing. She is treated with rituximab (monoclonal antibody). Further medical history was a known hypertension, heart failure with left ventricular anteroseptal hypo/dyskinesia with Ejection Fraction (EF) 40%, arteriosclerosis and sleep apnea. She was under investigation for cognitive dysfunction.

The patient was sent to hospital because of aggravating pain in her left leg. Ultrasound and angiography demonstrated multiple arterial stenoses and occlusion of the femoral artery and she was scheduled for a femoral to popliteal bypass. The time of surgery was scheduled to  $6 \pm 2$  hours. On the day of surgery, the patient was administered her daily ASA, gabapentin and beta blockade, but no ACE inhibitor or diuretic. An epidural catheter was placed at the L2-L3 level and an arterial line in the right radial artery. We induced anesthesia using pentothal (300mg - because of lack of propofol due to Covid-19 pandemic), remifentanil and rocuronium and maintained it using desflurane and remifentanil. Tracheal intubation was uneventful. An epidural infusion was started with ropivacaine 2mg/ml at 5 ml/h. A low dose of norepinephrine (0,03-0,07  $\mu$ g/kg x min) considered to counteract the vasodilatory effects of the anesthetics was used to keep Mean Arterial Pressure (MAP) at 70 mmHg or above. The surgery time was 280 minutes and the blood loss was 300ml. She was administered approximately 800ml of crystalloid (Ringer's acetate) during surgery. We administered 200mg of sugammadex to fully restore muscle function and acetaminophen 1000g as well as a bolus of ropivacaine 2mg/ml: 3ml in the epidural catheter for postoperative analgesia. The patient emerged uneventful from the general anesthesia uneventfully and was extubated after confirming adequate spontaneous breathing. The patient was transferred to the recovery ward. The epidural infusion was continued with ropivacaine 2mg/ml at 3ml/h and the patient received no opiates. The postoperative course was without any respiratory complications and she did not need any oxygen to

obtain an oxygen saturation above 93%. Her maximum pain score postoperatively was 1/10. The norepinephrine infusion could be stopped 2 h postoperatively. She was ready to go to the general ward 6 h postoperatively, which is the minimum time for our vascular surgery patients.

## Discussion

Although rare, CIDP is the most common form of chronic inflammatory polyneuropathy, with a prevalence of 0.8-0.9 cases /100 000 and a male dominance of 2:1. [1] It is present in all ages but the average debut age is 50. CIDP is caused by a chronic attack by the immune system at myelin and nodes of Ranvier of peripheral sensory and motor nerves. The pathogenesis is unknown. There are many similarities with Charcot-Marie-Tooth and Guillain-Barrés syndromes [1], but CIDP is seldom preceded by an infection. Obligate criteria for CIDP includes:

- Progressive escape paresis and sensory dysfunction in legs and arms.
- Involvement of proximal muscles in legs and most often also arms.
- Hypo- and areflexia.
- Progressive phase of 8 weeks.
- Demyelination on electroneurography, in at least two different motor nerves.

Standard immunomodulating treatment consists of intravenous immunoglobulin, cortison and plasmapheresis [1].

We can here present the first case report on a CIDP patient during major vascular surgery with a combination of both regional and general anesthesia. Based on this case, a literature study of CIDP and anesthesia was performed by searching PubMed of articles in English from 2000 to April 2020. 10 articles were found, mostly case reports [2-11]. No randomized-controlled studies were found. There is no standard anesthetic care for patients with CIDP. There are more reports on successful regional anesthetic techniques than on general anesthesia. During spinal and epidural anesthesia, it is reported that CIDP patients need reduced doses of local anesthesia at least during cesarean delivery [9], as well as a fear that the block will spread too high cranially to impact the respiratory muscles, which are often weakened in these patients. This is in agreement with our case, where the epidural doses of ropivacaine were lower than usual. There are also reports on longer duration of subarachnoid blocks in patients with CIDP [13]. Baseline worsening of CIDP symptoms in all extremities can also be seen postoperatively due to the perioperative stress response [5] and these symptoms can be difficult to differentiate from effects of blockades if they appear early after the surgery.

During general surgery caution must be taken due to the high risk of postoperative respiratory complications. In a case series of 17 patients under general anesthesia [7] two patients required postoperative mechanical ventilation, one patient had an aspiration one day after surgery and needed reintubation and three patients had worsening of CIDP symptoms. A thorough patient history must be requested for previous respiratory problems as well as preoperative testing with spirometry and/or electrophysiology. Our patient experienced no dyspnea at rest but reported on respiratory problems whenever she got a cold. She denied problems when swallowing.

We used general anesthesia to our patient as a complement to regional because of the long predicted surgery time, and the risk of hypoventilation in patients with respiratory muscle weakness when using sedation with propofol or dexmedetomidine during long procedures (own experiences). A prolonged effect of muscle relaxant in patients with CIDP is known [3], which can increase the postoperative respiratory weakness when train-of-four is not tested before extubation. Our patient with both CIDP and sleep apnea, received no long acting opiates, only short acting volatile agent (desflurane) and sugammadex to receive full muscle strength before extubation, which went uneventful. The six-hour postoperative period was excellent without any need for supplementary oxygen and an oxygen saturation of 95% and above.

## Conclusion

We gave a combination of regional and general anesthesia with short acting agents in combination with sugammadex to a patient with CIDP and sleep apnea during major vascular surgery. This is an approach that we can strongly recommend in these patients.

**Funding:** None

**Conflict of interest:** None

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