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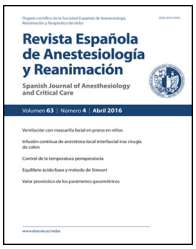

CASE REPORT

# Femoral and sciatic nerve block for knee arthroscopy in a patient with acute intermittent porphyria

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**KEYWORDS**
Acute intermittent porphyria;
Peripheral plexus block;
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**Abstract** Acute intermittent porphyria is an autosomal dominant disorder that results from a partial deficiency of porphobilinogen deaminase and that causes very severe symptoms. Attacks may be triggered by a series of drugs and by other factors that the anesthesiologist should be aware of in order to reduce morbidity and mortality. Our objective is to review anesthetic considerations in acute intermittent porphyria.

We present the case of a patient diagnosed with acute intermittent porphyria who was scheduled for knee arthroscopy. The anesthetic technique used was a femoral and sciatic nerve block under sedation with an infusion of remifentanil. The surgery proceeded without incident and the patient was discharged home after 24 h.

We consider the use of a peripheral plexus block of the lower limb to have been the safest anesthetic technique for this patient.


**PALABRAS CLAVE**
Porfiria aguda intermitente;
Bloqueo plexo periférico;
Consideraciones anestésicas

**Bloqueo nervioso femoral y ciático para una artroscopia de rodilla en paciente con porfiria aguda intermitente**

**Resumen** La porfiria aguda intermitente es una enfermedad autosómica dominante que resulta de un déficit de porfobilinógeno deaminasa y que causa síntomas muy severos. Los ataques se pueden desencadenar por fármacos y otros factores que el anestesiólogo debe conocer para reducir la morbilidad y la mortalidad. Nuestro objetivo es revisar las consideraciones anestésicas en la porfiria aguda intermitente.

Presentamos el caso de una paciente diagnosticada de que porfiria aguda intermitente programada para una artroscopia de rodilla. La técnica anestésica realizada fue bloqueo nervioso

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femoral y ciático bajo sedación con perfusión de remifentanilo. La cirugía transcurrió sin incidencias y la paciente fue dada de alta a domicilio a las 24 h.

Consideramos que el bloqueo nervioso periférico de la extremidad inferior es la técnica anestésica más segura para esta paciente.


## Introduction

Acute intermittent porphyria (AIP) is an infrequent autosomal dominant hereditary disease caused by a deficiency of the enzyme porphobilinogen (PBG) deaminase, which catalyzes the third step in heme synthesis. Its incidence is estimated at 0.13 per million inhabitants per year,[1] and is more frequent in women. An acute attack of AIP is characterized by acute dysfunction of the central, autonomic, and peripheral nervous systems.[2] The clinical picture is associated with an abrupt overproduction of the heme precursors PBG and delta-aminolevulinic acid (D-ALA) in the liver, following the induction of ALA synthase-1 (ALAS1) gene expression. AIP is the form of porphyria that produces the most severe symptoms.[3] The most common of these is abdominal pain, sometimes accompanied by psychiatric and/or neurological symptoms that may progress to bulbar palsy. Acute attacks are triggered by exposure to certain medications and by other precipitating factors.[4]

We present the case of a patient diagnosed with AIP scheduled for knee arthroscopy.

## Case report

We present the case of a 34-year-old patient with an allergy to latex, ASA physical status III. She was diagnosed with AIP following an emergency caesarian section performed at 31 weeks of gestation due to the premature rupture of membranes secondary to chorioamnionitis. She presented syndrome of inappropriate antidiuretic hormone secretion, epileptic seizure, and predominantly proximal tetraplegia with very high levels of PBG and D-ALA in urine, which required admission to the intensive care unit and for which she has been receiving chronic therapy with intravenous hemin (150/d mg for 3 days) every 2 weeks (the last dose prior to surgery was 9 days before the operation). The patient gave written permission for the authors to publish the report.

Knee arthroscopy was scheduled for the repair of a lateral meniscus tear that was causing the patient pain in the left knee. Written consent was given for the procedure. A few days before surgery, intravenous hemin was ordered from the hospital pharmacy in order to ensure that the drug would be available immediately in case of an attack of AIP during the perioperative period. The patient was admitted to our hospital the day before surgery and an infusion of 10% glucose (200 mg/d) was initiated in order to avoid prolonged fasting. As the patient was allergic to latex, surgery was performed in the early morning, following the latex-allergy protocol. In the operating room, electrocardiogram, oxygen saturation determined by pulse oximetry, and noninvasive arterial blood pressure were monitored. The patient was premedicated with intravenous midazolam (4 mg) and a continuous infusion of remifentanil was started at 0.1 μg/kg/min. Subsequently, ultra sound-guided femoral and sciatic nerve blocks were performed using a specific nerve-stimulating needle for each (Locoplex, Vygon, Ecouen, France; 20G, 17° bevel and 50-mm length for the femoral nerve block and Sonoplex, Face tip, Pajunk®, Norcross, GA, USA; 22G and 100 mm length for the sciatic nerve block), connected to a neurostimulator (Plexygon®, Vygon, Ecouen, France). The femoral and sciatic nerves were located with no paresthesia or discomfort of any kind (patella twitch and plantar flexion, respectively, elicited at an intensity of 0.4 mA, a frequency of 2 Hz and a pulse duration of 100 μs. A total volume of 40 mL of bupivacaine 0.5% plus dexamethasone 8 mg was injected without incident and antibiotic prophylaxis with intravenous amoxicillin–clavulanic acid (2 g) was administered. The surgery was uneventful. The patient remained in the postoperative recovery room for 1 h and was then transferred to the ward. Postoperative pain was controlled with paracetamol and subcutaneous morphine on demand. Motor function and sensation returned to the limb 5 hours after the puncture. The postoperative period was without complications and she was discharged home 24 h after surgery.

## Discussion

Porphyrias are metabolic disorders resulting from deficiencies in the enzymes involved in the synthesis of porphyrins of the heme group. AIP is the most frequent of the acute porphyrias, which are rare diseases that have implications for anesthesia. Some of them may produce no symptoms.[5] Attacks may be brought on by hypothermia, fasting, surgical stress, infection, dehydration, pregnancy, or certain drugs whose metabolism is dependent on cytochrome P450 (Table 1). Treatment of the attacks involves treating the symptoms and the early administration of intravenous hemin, which represses ALAS1.

During the pre-anesthesia visit, all porphyria patients scheduled for surgery should undergo careful evaluation, which should include a neurological examination to rule out peripheral and/or autonomic neuropathy, cardiovascular and respiratory assessment, as well as assessment of hydration status and an ionogram.

Pre-anesthetic management should include adequate anxiolysis[6] and a reduction of fasting time to a minimum, as both stress and caloric restriction may trigger attacks.

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