

# ANESTHESIA FOR CESARIAN SECTION IN PREGNANT WOMAN WITH ACUTE INTERMITTENT PORPHYRIA AND HYPOTHYROIDISM

## - Case Report\* -

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### Abstract

Acute intermittent porphyria (AIP) is a rare autosomal dominant metabolic disorder<sup>1</sup> affecting the production of heme, the oxygen-binding prosthetic group of hemoglobin<sup>2</sup>. It is characterized by a deficiency of the enzyme hydroxymethylbilane synthase; without this cytoplasmic enzyme, heme synthesis cannot finish, and the metabolite porphobilinogen accumulates in the cytoplasm<sup>3</sup>. Some additional factors must also be present such as drugs, hormones, dietary changes, infections diseases and surgery that trigger the appearance of symptoms, which include neurological disorders, abdominal pain, constipation, and muscle weakness. We present a perioperative course of a pregnant woman with porphyria in association with hypothyroidism and its anesthetic management.

### Case Report

A 30 yr-old, 67 Kg female woman was admitted with history of 42 weeks of gestational age and proposal to hold an emergency cesarean section for cephalo-pelvic disproportion.

Past medical history included diagnosis of AIP (without sequelae and with no history of acute crisis of last 2 years) and hypothyroidism with hormone replacement to 75 mcg of levothyroxine daily. His airway classification was Mallampati 2 and her ASA status was PIII.

After completion of the venipuncture with an 18G catheter and monitoring with ECG, NIBP, pulse oximetry and urine output hourly, carried spinal anesthesia with the patient in the left lateral position with a 27G Quincke tip and, by median, L3-L4 interspace and injection of 12 mg of heavy bupivacaine combined with 50 mcg of morphine. At the fifth minute of the blockade, the sensory level was at T4. After the birth of male fetus (Apgar 9/10), was administered 10 IU of oxytocin. The volume replacement was given with 500 ml of dextrose 5% and 1000 ml of Ringer's lactate; had diuresis of 150 ml during the procedure. The patient remained stable throughout the procedure, which lasted about 75 minutes.

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## Discussion

The AIP management includes high-carbohydrate diet; in severe attacks, a glucose 10% infusion is recommended, which may aid in recovery. Infection is one of the causes of attacks and requires treatment. Pain is extremely severe and almost always requires the use of analgesic drugs and should be treated early as medically possible.

Nausea and vomiting can be severe. Hematin and heme arginate are the drugs of choice and need to be given very early in an attack to be effective. They are not curative drugs, but can shorten attacks and reduce the intensity of an attack. Patients with a history of acute porphyria are recommended to wear an identification at all times in case they develop severe symptoms, a result of which may be that they cannot explain to healthcare professionals about their condition and the fact that some drugs are absolutely contraindicated.

An attack of acute intermittent porphyria may be precipitated by one of the "four Ms": medication, menstruation, malnutrition, maladies. If drugs have caused the attack, discontinuing the offending substances is essential. Patients that experience frequent attacks can develop chronic neuropathic pain. This is thought to be due to axonal nerve deterioration in affected areas of the nervous system. Depression

and seizures often accompanies the disease and is best dealt with by treating the offending symptoms and, the judicious use of anti-depressants and anti-convulsant drugs. Most seizure medications exacerbate this condition. Urine from a person experiencing an acute attack may be red or "port wine" in color because of the presence of porphyrins.

In obstetrics, the main factors triggering the acute crisis of porphyria are hyperemesis gravidarum, fasting pre and post-operative, hormonal changes, infections, surgery and anesthesia. The anesthetic management must include a careful selection of drugs, hydration with glucose solution, diet-based carbohydrates, prophylaxis of bronchial aspiration and evaluation of liver function. Regional anesthesia is preferred, because they are many unknown and unsafe anesthetic drugs, which can be seen in the Table 1. However, regional anesthesia should be avoided in cases where there is a neurological injury suspect or indefinite.

Worsening of porphyria during pregnancy may be between 50% and 90%, with maternal mortality ranging from 2% to 40%, fetal mortality from 13% to 40%. In conclusion, AIP and its association with hypothyroidism in a pregnant patient can be catastrophic. The main complicating factors in this case are the association of exposure to surgical stress, drugs and anesthetic procedure itself.

*Table 1*  
*Common Used Anesthetic Drugs Classification*

DRUGS	SAFE	UNKNOWN	UNSAFE
HYPNOTICS	Midazolam, Propofol, Lorazepam	Ketamin, Diazepam	Barbiturates, Etomidate
INHANALATED	Nitrous Oxide, Desflurane	Isoflurane, Halothane, Sevoflurane	Enflurane
OPIOIDS	Fentanyl, Morfine	Alfentanyl, Sufentanyl	
MUSCLE RELAXANTS	Succinylcholine, Vecuronium	Atracurium, Pancuronium	
LOCAL ANESTHETICS	Bupivacaine	Lidocaine	
ANOTHER DRUGS	Neostigmine		Steroids

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