

# A Case Report of Epidural Labour Analgesia Management In Congenital Methemoglobinemia Type I

Case Report

Volume 4 Issue 2- 2024

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## Article History

Received: November 10, 2024 Accepted: November 13, 2024 Published: November 14, 2024

## Abstract

Congenital methemoglobinemia is a rare condition that impairs oxygen transport. During pregnancy, it may affect the fetus with intrauterine growth restriction, preterm birth, and neonatal death. The use of common anesthetic molecules in pregnant patients with methemoglobinemia may be challenging. We present a case report of epidural labour analgesia management in a 24-year patient diagnosed during early pregnancy with congenital methemoglobinemia type 1. Close monitoring of methemoglobinemia levels during labour permitted safe and efficient analgesia in obstetric patients.

### Glossary of Terms:

- a)MetHb – methemoglobinemia
- b)CS- cesarean section
- c)MB-methylene blue
- d)CYB5R3- cytochrome B5 reductase gene
- e)PEPD-peptidase D gene
- f)CBC-complete blood count
- g)ABG- Arterial blood gases
- h)PCEA-patient controlled epidural analgesia.
- i)CEI- continuous epidural infusion
- j)PreEpi – Before Epidural

## Introduction

Methemoglobinemia is a rare condition, defined as an abnormal level of methemoglobin (MetHb) (>1%). MetHb is a form of hemoglobin (Hb) in which iron persists in the ferric state (trivalent) and is unable to return to the ferrous state (bivalent) [1]. MetHb cannot carry oxygen

and shifts to the left oxygen-hemoglobin dissociation curve, leading to hypoxia and cyanosis. MetHb can be either acquired (most commonly) or inherited. Clinical severity correlates with MetHb level, clearance, and rising rate. The main clinical signs include cyanosis, dark blood, dyspnea, dizziness, fatigue, syncope, metabolic acidosis, dysrhythmias, seizures, delirium, and death [2]. Pregnancy may be a triggering



factor in otherwise asymptomatic patients, in whom commonly used obstetric and anesthetic drugs may be at risk. There is evidence in the literature that the use of lidocaine, benzocaine, procaine, prilocaine, bupivacaine, nitroglycerin, nitrous oxide, and even fentanyl has an oxidizing effect on Hb, and these molecules are best avoided [3]. There is very little evidence of labor epidural analgesia in patients affected by this condition to assess the safety of this approach. The most reported cases are either emergency cesarean sections (CS) [4] managed under general anesthesia or patients in whom methemoglobinemia was diagnosed postoperatively. A recent report presented the case of a patient with congenital methemoglobinemia in which labor analgesia was administered with intravenous remifentanyl [5].

Methylene blue (MB) is the primary antidote for methemoglobinemia. MB should be used during pregnancy only if its benefits outweigh its risks<sup>6</sup>. It might be teratogenic depending on gestational age (ileal atresia or premature birth). In newborns, complications include hyperbilirubinemia, hemolytic anemia, methemoglobinemia, respiratory distress, skin photosensitization, and desquamation. If there is an absolute contraindication, inefficacy, or worsening of the MetHb level after MB treatment, an urgent total blood exchange transfusion might be necessary.

Written consent has been obtained from the patient for the publication of this case report and Ethics Committee approval for this study was provided by the Ethical Committee of the Erasme Hospital (n. P2023/296), Brussels, Belgium, on 29 August 2023.

## Case Report

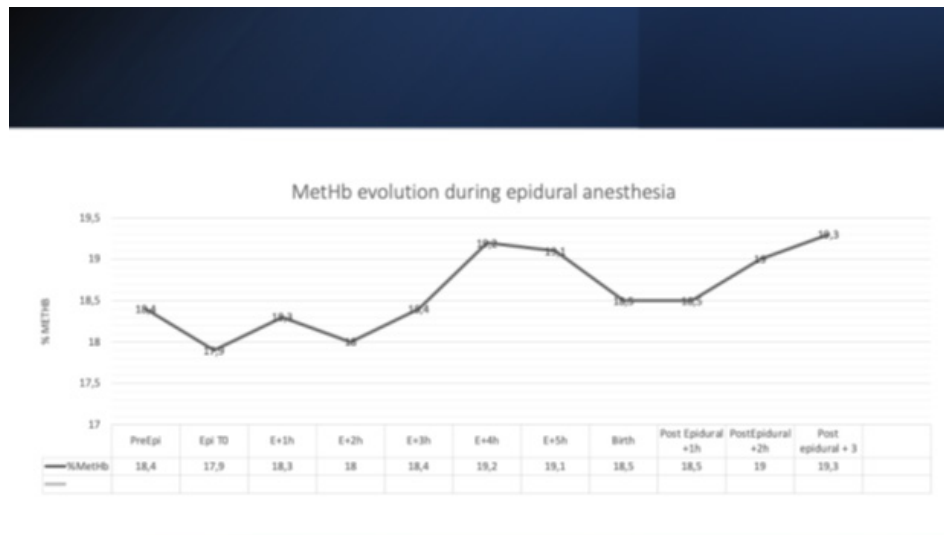
A 24 years old, multiparous patient was diagnosed in early pregnancy with congenital methemoglobinemia type 1. Genetic analysis revealed a homozygous mutation in CYB5R3 and heterozygous mutation in PEPD. Clinically, she presented persistent cyanosis, moderate dyspnea, poor effort capacity, and a stable SpO<sub>2</sub> of 91%. No signs of fetal hypoxia were observed. The MetHb level at diagnosis was 13.7%, which stabilized at 16% during pregnancy after administration of vitamins C and B. As only medical history, she presented one previous spontaneous vaginal birth under epidural analgesia (lidocaine/

ropivacaine) with persistent desaturation (91%-94%), unresponsive to oxygen. No further investigations were conducted at that time and the births were uneventful.

During the pre-anesthetic visit at 32 weeks of pregnancy, the risks of MetHb were discussed, and the indications, benefits, side effects, and complications of the different analgesic drugs and techniques were explained. Despite alternative analgesic methods for labor and general anesthesia in the case of cesarean section, the patient claimed epidural analgesia and refused any other techniques. After a multidisciplinary team discussion including anesthesiologists, obstetricians, hematologists, and midwives, epidural analgesia with levobupivacaine was administered, with regular monitoring of MetHb levels.

The patient was scheduled for induction at 38 weeks with intravenous prostaglandins, and 12h later, she went into active labor. On hospital admission, complete blood count (CBC), coagulation panel, and arterial blood gases (ABG) were performed, and six units of packed red cells were prepared. MetHb level was 18.4 %, SpO<sub>2</sub> 94%, ABG normal. MB (1mg/kg) was readily available in cases of MetHb > 20% or clinical deterioration. Upon maternal request, an epidural catheter was placed at 5 cm cervical dilation after radial arterial catheterization, ECG, SPO<sub>2</sub>, and oxygen supplementation. The epidural catheter was topped-up with a fractionated bolus of levobupivacaine 0,0625 % + sufentanil 0,25 %.

Epidural anesthesia maintenance was levobupivacaine 1.25%+ sufentanil 0,25mcg/ml patient controlled epidural analgesia with continuous epidural infusion (PCEA+CEI) protocol (5ml/h continuous infusion, bolus 6ml, lockout 15min.). The total duration of labor with epidural infusion was 4hours, and a total volume 66ml of levobupivacaine 1.25mg/ml was infused. The quality of analgesia was judged to be satisfactory (EVA scores varied between 1/10 and 6/10, and a global positive experience of the birth process). MetHb levels were assessed hourly and only slightly increased (Figure 1). SpO<sub>2</sub> remained stable at 94%. The Spontaneous vaginal birth was uneventful, with was healthy baby (MetHb < 2 %). 2days postpartum, the patient was discharged without complications.



**Figure legend:** Methemoglobin blood concentration (%) evolution during epidural anesthesia.

## Discussion

This is a case report of the successful management of labor anesthesia with epidural levobupivacaine in a patient with congenital methemoglobinemia type 1. Epidural analgesia was chosen upon maternal request before referral to our hospital, while refusing any other anesthetic and analgesic alternative. This informed choice was

supported by her medical history, all monitoring possibilities, and availability of a multidisciplinary team. The use of levobupivacaine and ropivacaine for this pathology is controversial [6]. Very few cases of regional anesthesia for emergent CS have been reported. Modern obstetric anesthesia uses a lower anesthetic concentration, which potentially increases safety. In the present case, MetHb concentration was monitored and showed only a slight increase over several hours



of infusion (and active labor). MetHb never reached the critical value of 20% (Figure 1), which was the predetermined value to start the treatment with MB suggested by our referent hematologist and current guidelines. Clinical tolerance and satisfaction of the mother and baby were excellent.

Congenital methemoglobinemia is a rare but life-threatening condition that can complicate pregnancy. This adds complexity to the management of labor analgesia and represents a challenge for anesthesiologists. In this clinical setting, we were able to provide efficient labor epidural analgesia and a safe and positive birth experience thanks to a tailored anesthesia plan, multidisciplinary approach, and close biological and clinical monitoring.

## Acknowledgements

i. Assistance with the article: none declared.

ii. Financial support and sponsorship: none declared.

iii. Conflicts of interest: none declared. The authors declare that they have no affiliations with or involvement in any organization or entity with any financial interest in the subject matter or materials discussed in this manuscript.

iv. Presentation: none declared.

v. Ethics: Ethical approval for this study (Ethical Committee P2023/296) was provided by the Ethical Committee Erasme Hospital,

Brussels, Belgium (Chairperson Prof J.-M Boeynaems) on 29 August 2023.

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