

CONSIDERATIONS FOR OPTIMAL PREGNANCY CARE. A case report.

INTRODUCTION

Paroxysmal nocturnal haemoglobinuria (PNH) is a rare acquired haematological disorder caused by mutations in haematopoietic stem cells.

Prevalence is estimated between 0.5-1.5 per million people.

The condition renders hematopoietic cells sensitive to complement-mediated destruction.

Characteristic features include intravascular haemolysis, thrombosis, and variable degrees of bone marrow failure

PREGNANCY AND PNH

PNH leads to increased risks for thrombosis, pre-eclampsia and intra-uterine growth restriction.

In the UK, Eculizumab is the only treatment licenced in pregnancy. It is a monoclonal antibody that targets the complement protein C5.

CASE REPORT

In 2023 a 34-year-old with a diagnosis of PNH booked in her first pregnancy.

Flow cytometry confirmed a large PNH clone- 82.55% of the neutrophils were PNH cells. There was no history of thrombotic episodes, end organ damage, or complement inhibitor treatment.

The patient was on therapeutic low molecular weight heparin (LMWH) and Eculizumab was commenced in the first trimester. Fetal growth was normal throughout her pregnancy. There was no evidence of haemolysis or bone marrow dysfunction.

Induction was planned at 39 weeks, within one week of an Eculizumab dose. An extra dose of Eculizumab was given as the induction process was prolonged. A female infant was delivered by forceps delivery. A PPH of 1.6L was managed with tranexamic acid, an oxytocin infusion and blood transfusion.

LMWH was recommenced postnatally as per guidelines. Eculizumab was restarted on a 2 weekly basis for 3 months postnatally, and the PNH team follow-up was planned.

PRE-CONCEPTION/ANTENATAL CARE

MDT management throughout pregnancy should include midwifery, obstetric, and anaesthetic teams in conjunction with haematologists with experience in managing patients with PNH during pregnancy. A pre-conception consultation is recommended.

The patient's background, including clone size, PNH symptoms, thrombosis, end organ damage, meningococcal vaccination status and previous PNH treatment should be established. Discussions specific to PNH would include the requirement for anticoagulation with LMWH, and Eculizumab treatment.

Baseline blood testing should include FBC, flow cytometry, reticulocytes, LDH, DAT, ferritin and U&Es. The patient should be advised to monitor for symptoms such as anaemia, pain, dyspnoea, fatigue and fever.

INTRAPARTUM AND POSTNATAL

An individualised intrapartum and postnatal plan should be made, with consideration for a planned induction (or caesarean if indicated) to allow for planned Eculizumab administration and a LMWH pause.

Reintroduction of therapeutic LMWH and continuation of Eculizumab needs to be carefully planned and follow-up with the PNH haematology team arranged.

THE NATIONAL PNH SERVICE

The PNH National Service consists of two centres in England, with one based at St James’s University Hospital, Leeds and the other in King’s College Hospital, London. The service provides care and support for patients with PNH throughout England, and support to patients from Scotland, Wales and Northern Ireland.

