

## CASE REPORT

# A Case of Perioperative Management in a Patient with Rare Red Blood Cell Phenotype

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### ABSTRACT

The current focus of perioperative management of anaemia has shifted from allogeneic transfusion to optimising and conserving the patient's blood through the implementation of patient blood management (PBM) strategies. We hereby report a case to illustrate the success of applying PBM strategies in managing a surgical patient with an extremely rare red blood cells (RBC) phenotype. An 80-year-old Malay man was planned for urgent major abdominal surgery following diagnosis of intestinal obstruction secondary to an advanced rectosigmoid tumour. A request of two units packed RBC was made given anticipated blood loss intraoperatively. His pre-operative haemoglobin was 135 g/L. His previous immunohaematological record showed that he had an extremely rare P<sub>2</sub><sup>k</sup> (P1-, P-, P<sup>k</sup>+) phenotype with clinically significant anti-P, anti-PX2, and anti-P1. The elements of PBM strategies were explored and applied. Eventually, the patient successfully underwent a surgical operation without any allogeneic RBC transfusion.

**Keywords:** Patient blood management, Transfusion, Phenotype, Red blood cells

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### INTRODUCTION

Red blood cells (RBC) alloimmunisation is one of the complications in a patient receiving blood transfusion even in non-chronically transfused patients. The presence of alloantibody may complicate and render the search of compatible blood for transfusion. Furthermore, the detection of alloantibody in a pre-operative patient may delay the surgical operation particularly in cases of antibody to rare RBC phenotype; antibody towards high-prevalence antigen; or presence of multiple RBC antibodies. Hence, optimising and conserving patient's blood through the application of Patient Blood Management (PBM) strategies are incorporated in the perioperative management of anaemia. The PBM involves evidence-based and multidisciplinary therapeutic approaches that comprise of optimisation the red blood cell mass, minimisation blood loss and improving patient's tolerance of anaemia (1). We hereby report a case to illustrate the success of PBM strategies in the perioperative management of a surgical patient with an extremely rare RBC phenotype.

### CASE REPORT

An 80-year-old Malayman, with underlying hypertension, presented to the Emergency Department of Hospital Selayang with complaints of severe abdominal pain and distension, altered bowel habit, and vomiting for one week, associated with loss of weight and appetite. Physical examination showed abdominal tenderness and distension with reduced bowel sound. His haemoglobin level was 135 g/L. Computed tomography (CT) scan of abdomen, pelvis, and thorax revealed a constricting sigmoid mass with multiple lung nodules. Histopathological examination confirmed the diagnosis of colon cancer. A diagnosis of intestinal obstruction secondary to advanced rectosigmoid tumour was made. An urgent bowel resection via Hartmann's procedure was planned with the request for two units of packed RBC to standby intraoperatively.

The results of pre-transfusion testing showed blood group O Rh (D) positive with antibody screening using gel card (Coombs Anti-IgG+C3d ID-Card, Bio-Rad, Switzerland) showed strong agglutination (3+) at all three screening cells (DiaCell I+II+III, Bio-Rad, Switzerland). His blood sample was sent to the National Blood Centre (NBC) for further investigation. Previous immunohaematological records at NBC showed that the patient's RBC has a rare

$P_2^k$  (P1-, P-, Pk+) phenotype and his plasma contains clinically significant anti-P and anti-PX2 as well as a trace of anti-P1. These findings were detected during family screening two years ago when his older brother was admitted for robotic prostatectomy secondary to prostate carcinoma. During that time his brother was also identified to have rare  $P_2^k$  (P1-, P-, Pk+) phenotype. Crossmatching the patient's plasma with two donors of null p phenotype (P1-, P-, Pk-) were incompatible (2+ reaction). Due to the extreme rarity of the patient's phenotype and the presence of clinically significant alloantibodies, PBM strategies were employed to minimise allogeneic transfusion. A series of discussions with surgical and anaesthesia teams of the rarity of the patient's RBC phenotype; the difficulty of finding antigen-negative blood; and the possibility of incompatible transfusion were conducted. To optimise the pre-operative erythropoiesis, a single dose of iron sucrose 200 mg was infused without any adverse reactions. His haemoglobin level increased to 141 g/L post-infusion. Perioperatively, unnecessary phlebotomy was also avoided. The initial plan for Hartmann's procedure was abandoned due to family refusal. Instead, colonic stenting placement for the obstructive tumour was attempted. However, the procedure was unsuccessful due to the severely obstructed tumour. Hence, defunctioning colostomy using electrosurgical diathermy was performed by the senior consultant surgeon. The patient was kept normothermia intra- and post-operatively. The operation was uneventful with only minimal blood loss with a postoperative haemoglobin level of 140 g/L. The patient was discharged without any allogeneic blood transfusion.

## DISCUSSION

This case illustrates the success of implementing evidence-based, multidisciplinary therapeutic PBM strategies in managing a surgical patient with an extremely rare anti-PX2, anti-P and less clinically significant anti-P1 alloantibodies. Both anti-PX2 and anti-P are naturally occurring antibodies towards high incidence antigens. They are clinically significant antibodies that are implicated in haemolytic transfusion reaction and haemolytic disease of foetus and newborn. Within P1PK/GLOB blood group system, P1 and P2 phenotypes are the two most common phenotypes with prevalence between 10-90%. As for the patient, his RBC phenotype is  $P_2^k$  which is an extremely rare RBC phenotype with the prevalence is estimated at 1 per million population (2). This phenotype is characterised by the absence of P and P1 antigens due to mutations in B3GALNT1 gene, which lead to dysfunctional of  $\beta$ 1,3-N-acetylgalactosaminyltransferase or P synthase (2). In the absence of a functional P synthase, no PX2 antigen will be formed. Consequently, anti-P, anti-P1, and anti-PX2 are made by these  $P_2^k$  phenotype individuals. This patient's plasma was also found to be incompatible when crossmatched with RBC from null p phenotype

donors. This can be explained by elevated amounts of the PX2 antigen on the RBC of p phenotype individuals which react with the patient's anti-PX2 antibody. Thus, it is recommended that P1k or P2k phenotype blood is selected for transfusion in Pk patients (2).

The PBM strategies are increasingly recommended in managing surgical patients complicated with antibodies towards high prevalence antigens. The PBM aims to identify patients at risk for blood transfusion and provide a suitable management plan that can reduce this need and thus improve patient outcomes (1).

The first pillar of PBM is to optimise the patient's erythropoiesis by identifying and managing the anaemia which is performed in this patient by the administration of iron sucrose pre-operatively. The second PBM pillar which is to minimise iatrogenic blood loss and bleeding were also incorporated into the patient management as there was no unnecessary phlebotomy and minimally invasive surgery using electrosurgery was performed instead of Hartmann's procedure. The last PBM pillar which is to harness and optimise patient physiological tolerance towards anaemia were also included in this patient such as by prevention of hypothermia intra- and postoperatively. Thus, PBM is a comprehensive personalised medicine that specifically tailored to the needs of an individual patient which focused on three pillars and is suitable to be practiced either in perioperative or other clinical settings (1).

Preoperative anaemia is common among colorectal cancer patients and the main cause is iron deficiency anaemia. Anaemia itself is independently associated with a higher risk of morbidity and mortality as well as an increased likelihood of blood transfusion (3). Hence, early identification and treatment of preoperative anaemia is a key focus in PBM. Even though, the preoperative investigation revealed that the patient was not anaemic, intravenous iron was administered to optimise the patient's RBC mass as there is an anticipated substantial blood loss during the major surgery (Hartmann's procedure). Intravenous iron is a safe and effective technique to rapidly increase haemoglobin level and iron supply before surgery.

Another strategy to optimise erythropoiesis is by using erythropoiesis-stimulating agents. However, it was not an option given the proposed erythropoietin's action on tumour angiogenesis and lymphangiogenesis (4). Furthermore, predeposit autologous donation (PAD) and acute normovolaemic haemodilution (ANH) are alternative solutions that can be considered when antigen-negative units are not available. There is no absolute contraindication for autologous donation in advanced age patients as the safety of this procedure had been previously studied and established in elderly patients (5). Nevertheless, both PAD and ANH were not explored for this patient as his surgery was changed

from Hartmann's procedure to electrosurgery which has a very minimal risk of bleeding (6) and thus did not necessitate the need for autologous donation. Besides autologous donation, usage of tranexamic acid and fibrin glue as haemostatic agents, and deliberate induced hypotension had also been shown to reduce blood loss intra-operatively.

For this patient, postoperatively, he was planned for conservative treatment and palliative care because of his advanced age and metastatic cancer.

## CONCLUSION

The supply of allogeneic transfusion in rare RBC phenotype is often a great challenge in a blood transfusion service. Thus, the incorporation of a multidisciplinary and evidence-based approach through PBM strategies is essential to improve the clinical outcomes of such patients.

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