

## A case report of postpartum hemolytic uremic syndrome.

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### Abstract

**Postpartum Hemolytic Uremic Syndrome (PHUS) is a rare disease characterized by spontaneous renal failure occurring immediately after delivery to 10 weeks postpartum. The clinical manifestation associated with PHUS includes acute microangiopathic hemolytic anemia, low platelet levels, and acute renal failure. Here, we report a case of a 29-year-old woman with PHUS after the delivery by caesarean section.**

**Keywords:** Postpartum hemolytic uremic syndrome, Microangiopathic hemolysis, Renal failure, Treatment prognosis.

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### Introduction

PHUS is rare, life-threatening disease caused by some potential factors including surgical stimulation, infection, severe preeclampsia, and placental abruption. Diagnosis of PHUS can be difficult and need to recognize clinical symptoms, and also detect hemolytic anemia, thrombocytopenia, and clinical performance for acute renal failure at an early stage. Combination of plasma exchange and hemodialysis at an early stage plays a key role for successful treatment of PHUS, which can contribute the survival rate of 84% [1], while diagnosis of this disease and seizing the rescue time would greatly reduce the mortality and late complications in patients, as well as improve the survival rate of patients.

### Case Report

A 29-year-old women (gravida 1, para 0) at 38 weeks and two days gestation with in vitro fertilization (IVF) was admitted to the Labor & Delivery unit in the hospital in January 11, 2016 and waiting for delivery. The platelet level of the patient was around 51-83 g/L during the pregnancy. Laboratory tests showed a Hemoglobin (HB) level of 122 g/L, platelet count  $50 \times 10^9/L$ ; Alanine transaminase (ALT) 62 U/L, aspartate aminotransferase (AST) 60 U/L, thiobarbituric acid level 4.0 mmol/L, urea level 7.37 mmol/L, uric acid 665  $\mu\text{mol/L}$ ; urine protein test indicated negative and white blood cells was 3+ (Table 1).

**Table 1.** Routine blood and urine, liver and renal function-test results 5 days before caesarean section.

Items	Quantity
HB(g/L)	122
Platelet count(L)	$50 \times 10^9$
ALT(U/L)	62

AST(U/L)	60
Thiobarbituric acid level(mmol/L)	4
Urea level(mmol/L)	7.37
Uric acid( $\mu\text{mol/L}$ )	665
Urine protein	negative
White blood cells	3+

\*Note: HB: Hemoglobin; ALT: Alanine Transaminase; AST: Aspartate Aminotransferase

At 12:10 AM in January 16, 2016, caesarean section was performed for the patient after receiving ten units of type A, Rh positive platelet, oxytocin and carboprost tromethamine was also administrated for the patient during the surgery for the prevention of postpartum hemorrhage. Nine hours (22:00 PM) after C-section, the reading of the patient's blood pressure was 160/86 mmHg, and it dropped to 125/61 mmHg at one and half hour later. At 7:00 AM in January 17, 2016, the patient presented with thirst, blurred vision, oliguria, dark brown urine, and blood pressure was 115/71 mmHg. Further examination revealed hemoglobin level 64 g/L, platelet count  $19 \times 10^9/L$ , thus the patient received a symptomatic treatment. At 11:55 AM, the patient presented with dizziness, weakness, and xerostomia with a fluctuating blood pressure of 128-139/78-95 mmHg, no urine output. The lab examination revealed that her urine protein was 3+, occult blood 3+. The hospital made a critically ill notice to the family and the patient was treated with furosemide intravenously. At 12:55 AM, the patient complained blurred vision, but the other conditions were getting better. Her lab parameters showed blood pressure 130/88 mmHg, no urine output, ALT 180 U/L, AST 343 U/L, protein 42 g/L, serum albumin 22.5 g/L, urea level 14.91 mmol/L, creatinine 305.4  $\mu\text{mol/L}$ , creatinine clearance 15.7 ml/min, uric acid 674  $\mu\text{mol/L}$ ,  $\text{CO}_2$  15.9 mmol/L, potassium concentration 6.72 mmol/L, HB 62 g/L, PLT  $20 \times 10^9/L$  (Table

2). The patient received 1.5 units of type A, Rh positive red blood cells and was treated for acidosis and hyperkalemia. At 15:10 PM, the patient's condition further deteriorated, no urine output with a fluctuating blood pressure of 130-141/87-93 mmHg.

**Table 2.** Routine blood and urine, liver and renal function-test results 1 day after caesarean section.

Items	Quantity
ALT (U/L)	180
AST (U/L)	343
Serum albumin (g/L)	22.5
Urea level (mmol/L)	14.91
Creatinine ( $\mu\text{mol/L}$ )	305.4
Creatinine clearance (ml/min)	15.7
Uric acid ( $\mu\text{mol/L}$ )	674
Potassium concentration (mmol/L)	6.72
HB (g/L)	62

\*Note: ALT: Alanine Transaminase; AST: Aspartate Aminotransferase; HB: Hemoglobin

The possibility of postpartum hemolytic uremic syndrome was considered according to the clinical manifestations of the patient including progressive decline in platelet count, hemolysis, and acute renal failure. The patient was transferred to the Division of Nephrology at West China Hospital. During hospitalization, the highest blood pressure ever recorded on the patient was 150/100 mmHg, the lowest level of hemoglobin was 37 g/L, the highest reticulocyte count was  $0.1089 \times 10^{12}/\text{L}$ , and the highest white blood cell count was  $22.6 \times 10^9/\text{L}$ . A progressive decline in platelet count was also observed and the lowest level was  $18 \times 10^9/\text{L}$ . The direct Coombs test showed negative, and the highest level of LDH, serum creatinine, and urine was 3168 IU/L, 611  $\mu\text{mol/L}$ , 24 mmol/L, respectively. In addition, glomerular filtration rate (GFR) was 8.86 ml/min/1.73 m<sup>2</sup> (decreased), the lowest level of serum total protein and albumin was 36.8 g/L and 23.9 g/L, respectively. The prothrombin time was increased and showed 65.5 seconds. International normalized ratio (INR) was 5.84, Activated partial thromboplastin time (APTT) was also increased 90.6 seconds, Prothrombin Time (PT) was prolonged to 30.5 seconds, Fibrinogen level was increased and showed 1.03 g/L, whereas Antithrombin III (ATIII) was 79.4%. Fibrinogen degradation product (FDP) was higher than normal level and was 27.4 mg/L, D-dimer was also increased and showed 15.98 mg/1FEU. Other decrease parameters include immunoglobulin G (IgG) (5.05 g/L), IGA 638 mg/L, complement component C3 0.376 g/L, complement component C4 0.107 g/L, Properdin factor B (PFB) 167 mg/L, haptoglobin <58.3 mg/L. Additionally, there were no obvious morphological abnormalities of mature erythrocytes. Pro b-type natriuretic peptide (Pro-BNP) level was >35000 pg/ml, which was also increased. Urinary protein was 3.0(3+) g/L and

granular casts were observed. A further lab test showed increased blood levels of amylase and lipase. After multidisciplinary consultation, the patient was treated with a combination of plasma exchange with intermittent flow (5 times) and intermittent hemodiafiltration (4 times) therapy, and also received an intermittent infusion of type A, Rh positive red blood cells of 8 unit, and 600 ml of frozen plasma and Irradiation platelet, along with others specific treatments including fasting, somatostatin treatment by sustained micro-pump, intravenous glucocorticoids, injection of cefoperazone Sodium/sulbactam Sodium, omeprazole therapy, oxytocin for maintaining homeostasis, and infusion of red blood cells and platelets. The patient's condition was improved eventually and discharged from hospital in February 18, 2016, and followed up every two weeks.

**Table 3.** Routine blood and urine, liver and renal function-test results 2 days after caesarean section.

Items	Quantity
Fibrinogen (g/L)	1.03
ATIII	0.794
FDP (mg/L)	27.4
D-dimer (mg/1FEU)	15.98
IgG (g/L)	5.05
IgA (mg/L)	638
PFB (mg/L)	167
Haptoglobin (mg/L)	<58.3
Pro-BNP (pg/ml)	>35000
Urinary protein (g/L)	3.0(3+)

\*Note: ATIII: Antithrombin III; FDP: Fibrinogen Degradation Product; PFB: Properdin Factor B; Pro-BNP: Pro b-type Natriuretic Peptide

## Discussion

PHUS belongs to the group of thrombotic microangiopathies (TMA) characterized by aggregation and reduction of platelets in the renal and/or systemic circulation, and mechanical damage to the red blood cells. It was first reported in 1968 and was estimated to occur at a rate of 1 in 25,000 births [2]. Most cases of PHUS occur following the normal delivery, but Liu et al. reported one case that the patient developed PHUS after artificial abortion [3]. The reason causing PHUS is complicated and also has a diverse pathogenesis, which can finally lead to microvascular endothelial cell injury and microthrombosis [4].

This was the first case of PHUS in a patient diagnosed in our hospital, and the patient's syndroms meet the diagnostic criteria for PHUS. The laboratory examination for the patient showed normal level of hemoglobin (Hb) and serum creatinine (SCr), but low platelet count before delivery. One day after delivery, the patient showed central nervous system symptoms and a series of other symptoms including elevated

transaminase, high blood pressure, dark brown urine, from declined urine output to no urine output, urinary protein 3+, progressive decline in platelet count and hemoglobin, and abnormal levels of urea nitrogen (BUN), creatinine, bilirubin. Because there was no bleeding from superficial sites in the skin and mucous membranes in the patient, it should be differentiated from severe preeclampsia, HELLP syndrome, thrombotic thrombocytopenic purpura. Different from HELLP syndrome that is associated with liver failure PHUS mainly affects kidney function and results in renal failure which supports the diagnosis of the patient in our report. In addition, the patient received ten units of platelet before surgery and was routinely administrated by oxytocin and carboprosttromethamine injection during the surgical delivery. It would be necessary for clinicians to evaluate the feasibility for using the platelet transfusion and uterotonics [5,6]. Once kidney failure has been diagnosed, dialysis should be preferably carried out as soon as possible, while combination of plasma exchange is recommended for avoiding the possibility of obstetric hemorrhage and improving the survival rate of patients [7]. The patient in our report received the treatment of combination of plasma exchange and hemodiafiltration therapy after transferring to a higher level of care center, and the condition was improved eventually, which further indicated that a combination of plasma exchange and hemodiafiltration therapy is still a priority for treating PHUS once the symptom is diagnosed. Additionally, glucocorticoid can be used to reduce hemolysis process and help restore kidney function [8]. Moreover, active treatment of obstetric complications and anticoagulation therapy should also be considered.

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