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Case Report

Perioperative Management of a Case of Homozygous Hemoglobin C Disease During Cardiac Surgery with Cardiopulmonary Bypass

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Abstract

Background: Hemoglobin C disease is a rare hemoglobinopathy mainly observed in black people and considered less severe than sickle cell disease. However, its homozygous phenotype predisposes patients to hemolysis and complications during cardiopulmonary bypass, driven by increased blood viscosity, erythrocyte fragility under shear stress, and fluctuations in oxygenation. Case Presentation: We describe the case of a 49-year-old male with homozygous Hemoglobin C Disease admitted for severe aortic regurgitation at the University Hospital of Tengandogo, Burkina Faso. The patient underwent aortic valve replacement under cardiopulmonary bypass. Due to the risk of hemolysis and hemoglobin crystallization, a tailored strategy was implemented: partial exchange transfusion with packed red blood cells and fresh frozen plasma, strict control of temperature, oxygenation and acid-base balance, avoidance of vacuum-assisted venous drainage. The intraoperative course was uneventful, with stable hemodynamics and no evidence of hemolysis. Postoperatively, the patient was extubated within six hours and required no additional transfusion. He was discharged from the intensive care unit after 48 hours. Conclusion: This case highlights the importance of individualized perioperative management in patients with Hemoglobin C Disease undergoing cardiac surgery. Understanding the pathophysiological implications of this hemoglobinopathy is essential to mitigate risks associated with cardiopulmonary bypass.

Keywords

Hemoglobin C Disease, Cardiopulmonary Bypass, Aortic Valve Replacement, Perioperative Management, Hemoglobinopathy, Burkina Faso

1. Introduction

Hemoglobin C (HbC) disease is a generally benign autosomal recessive genetic disorder caused by a point mutation in the beta globin gene [1]. It is predominantly prevalent in individuals of African descent, particularly in the Voltaic Plateau region (Burkina Faso, Ghana, Ivory Coast) [2]. The diagnosis is made by hemoglobin electrophoresis, and the blood smear shows a

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characteristic appearance of crystals and target cells with anisopoikilocytosis. Heterozygotes for HbC (AC) are generally asymptomatic with microcytosis, while homozygotes for HbC (CC) most often present compensated Chronic hemolysis with splenomegaly [2]. Cardiopulmonary bypass (CPB) in patients with hemoglobinopathies, particularly sickle cell disease, carries a high risk of vaso-occlusive complications due to hypoxia, acidosis, and hypothermia [3]. Although often considered more "benign" than sickle cell disease (HbSS), hemoglobin CC also presents critical rheological challenges during cardiopulmonary bypass (CPB) procedures. Unlike hemoglobin S (HbS), which polymerizes in the absence of oxygen, HbC reduces the solubility of oxygenated hemoglobin, forming intraerythrocytic crystals. This increases blood viscosity and decreases red blood cell deformability [4]. Furthermore, the inherent mechanical fragility of HbCC erythrocytes [5] exacerbates the risk of hemolysis during transit through pump rollers and oxygenators. The low prevalence of HbC in Western countries has led to a lack of specific guidelines for this type of hemoglobinopathy in cardiac surgery. We report the case of perioperative management of a patient with CC hemoglobinopathy who underwent aortic valve replacement under cardiopulmonary bypass at the Tengandogo University Hospital in Burkina Faso.

2. Case History

A 49-year-old patient weighing 82 kg was admitted to the cardiovascular surgery department at Tengandogo University Hospital for treatment of severe aortic regurgitation. After confirmation of the diagnosis by ultrasound, the multidisciplinary team decided on replacement of the aortic valve with a mechanical valve. During the preoperative anesthesia consultation, the patient's medical history revealed homozygous type C hemoglobinopathy. His medical history was negative for vaso-occlusive or painful crises. The clinical examination revealed no splenomegaly or jaundice. Laboratory tests performed 1 week before surgery showed a hemoglobin level of 12 g/dL and hematocrit 35%. The free bilirubin concentration was 3 mg/L and the conjugated bilirubin concentration was 5 mg/L before the procedure. Hemoglobin electrophoresis confirmed hemoglobinopathy homozygous C with 98% of hemoglobin C.

The patient was cleared for surgery following a pre-anesthetic evaluation. Induction was performed with sufentanil (3µg/kg) midazolam (0.1µg/kg), propofol (1µg/kg), and rocuronium (0.6 µg/kg). The patient remained hemodynamically stable. An internal jugular central venous line and a radial arterial catheter were placed, as well as an oropharyngeal temperature probe for temperature monitoring. Aortic cannulation and a single two stage venous cannulation were performed for CPB after administration of 300 IU/kg of sodium heparin. The bypass circuit was primed with two units of packed red blood cells (RBC) from a donor, two units of fresh frozen plasma (FFP) from a donor, 500 ml of 5% albumin, 750 ml of Ringer lactate solution. It was supplemented with 10,000 IU of sodium heparin, 20 mg of furosemide, 8 mg of dexamethasone, 100 ml of 20% mannitol, and 80 ml of 8.4% sodium bicarbonate. After the circuit was primed, the fluid was circulated through the oxygenator to achieve adequate oxygenation and warming to 36 ℃.

At the start of CPB, 1200 ml of patient's blood was collected from the venous line before cardiotomy. One reservoir bag was connected with a Y-connector to the venous line of the CPB system to enable the perfusionist to sequester the requested blood volume before CPB reaches full flow. At the same time the prime fluid was infused to maintain hemodynamic stability. It was also agreed not to use Vacuum-Assisted Venous Drainage (VAVD) during CPB and to keep the arterial oxygen pressure below 300 mmHg. CPB was performed at a normal temperature of 36 ℃. The CPB blood flow rates was 3 to 4.4L/mn for a mean arterial pressure of 65 mmHg. Cardioplegia was performed using 1000 ml of modified Delnido cardioplegia solution mixed with 200 ml of RBC from a donor and cooled to 28 ℃. It was administered directly through the coronary ostium at a pressure of 120/80 mmHg in 5 minutes and in a single dose. During CPB, the objective was to maintain metabolism at values close to normal conditions. The Table 1 summarize the changes in blood gas metabolic parameters during the different step of surgery.

Table 1. *Summary of arterial blood gases during the surgery.*

	ABG* before CPB	ABG at the beginning of CPB	ABG before removal of the cross clamp	ABG after CPB
Ph	7.35	7.41	7.44	7.37
PCO_2 (mmHg)	44	37.9	38.7	47.9
PO_2 (mmHg)	273	293.3	300.7	204.9
HCO_3- (mmol/L)	24	24.3	26.5	27.7
Hemoglobin (g/dL)	11.3	9.2	10.7	10.6
Hematocrit (%)	33	27	31	31

	ABG* before CPB	ABG at the beginning of CPB	ABG before removal of the cross clamp	ABG after CPB
Lactates (mmol/L)	0.83	2	1.48	1.54

*Arterial blood gas

A size 27 mechanical valve was implanted. After removing the aorta cross clamp, cardiac activity restarts spontaneously. The patient was successfully weaned from bypass without vasopressor. Cardiopulmonary bypass lasted 106 minutes, aortic cross clamp time 77 minutes with a support time of 22 minutes.

At the end of CPB, urine output was 3000 ml. The urine was clear, indicating no significant hemolysis. A Bair Hugger™ continuous air warming system was used to keep the patient warm until he was transferred to the intensive care unit. The patient received multimodal analgesia with paracetamol (15mg/kg every 6 hours), continuous morphine infusion (20 µg/kg /hour), and hydration maintained at 1.5L/24 h. His recovery in intensive care was uneventful. He was extubated within 6 hours of surgery. No further blood products were administered in the ICU. Free bilirubin concentration was 4mg/L 24 hours after the surgery. The patient was discharged to the hospitalization ward after 48 hours.

3. Discussion

In cardiac surgery Although it ensures circulation and ventilation, CPB is not a physiological system. It causes pathological reactions that can be much more significant in the presence of pathological hemoglobin. Hemoglobin S is the best described in cardiac surgery. In an oxygen-depleted, acidic, or cold environment, it polymerizes, leading to vaso-occlusive crises [6-9]. Unlike hemoglobin S, whose polymerization is induced by oxygen depletion, hemoglobin C has a tendency to crystallize preferentially in the oxygenated state (R-state) [4]. This form can increase blood viscosity and reduce the deformability of red blood cells. Conditions of cardiopulmonary bypass, combining shear stress and variations in oxygenation, can therefore lead to major destruction of these pathological red blood cells [4, 5].

The initial objective for this patient was to perform a partial exchange transfusion during surgery, collecting the patient's blood before it entered the venous reservoir and replacing it with healthy donor blood, thereby lowering the pathological hemoglobin level. The use of packed red blood cells with priming fluid not only provided the patient with healthy hemoglobin but also lowered the hematocrit to a reasonable level without major hemodilution due to blood loss [10, 11]. The target was an hematocrit between 27 and 30 per cent in order to limit destruction of RBC. The addition of albumin to the priming also slowed the decline in colloid osmotic pressure. We also avoided hyperoxia by trying to maintain a PaO_2 below 300 mmHg to reduce the risk of hemoglobin crystallization. The use of donor's RBC in the cardioplegic solution was intended to limit the risk of red blood cell crystallization in the coronary arteries. Metabolism was kept within narrow limits by performing surgery in normothermia and strict acid-base balance.

High-pressure suction is a primary driver of hemolysis, thrombocytopenia, and systemic inflammatory response [12-14]. In this context, with fragile red blood cells, we avoid aggressive suctions and prohibit the use of VAVD. Maintaining a MAP of at least 65 mmHg to avoid blood stasis during CPB [15].

4. Conclusions

Hemoglobinopathies are a challenge in cardiac surgery with cardiopulmonary bypass. Homozygous hemoglobinopathy C requires precautions to be taken during cardiopulmonary bypass to limit morbidity related to the patient's condition. A good understanding of the pathophysiology of this disease is necessary for safe CPB.

Abbreviations

hbC	Hemoglobin C
hbS	Hemoglobin S
CPB	Cardiopulmonary Bypass
RBC	Red Blood Cells
ICU	Intensive Care Unit
HbCC	Homozygous Hemoglobin C Disease

Author Contributions

Pingwinde Farid Belem: Conceptualization, Resources, Writing – original draft, Writing – review & editing

Nestor Palingwende Judicael Kabre: Data curation

Tewende Ouedraogo: Methodology

Armel Marie Jose Kinda: Methodology

Adama Sawadogo: Methodology, Supervision

Conflicts of Interest

The authors declare no conflicts of interest.

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