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Anesthetic Management of a Patient with Sickle Cell Haemoglobinopathy Undergoing Open Heart Surgery

Essay

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List of Abbreviations

ACE	Angiotensin-converting enzyme
ACT	Activated clotting time
AKI	Acute kidney injury
ARBs	Angiotensin receptor blockers
ВСР	Blood cardioplegia
CABG	Coronary artery bypass grafting
CBC	Complete blood count
ССР	Crystalloid cardioplegia
CMRI	Cardiovascular magnetic resonance imaging
СРВ	Cardiopulmonary bypass
CPV	Catastrophic pulmonary vasoconstriction
CT	Computerized tomography
CVP	Central venous pressure
DDAVP	Desmopressin
2,3-DPG	2,3-diphosphoglycerate
ECG	The electrocardiogram
FPLC	Fast-protein liquid chromatography
Hb	Hemoglobin
HbA	Adult haemoglobin

🕏 List of Abbreviations 🗷

HbF	Fetal haemoglobin
Hct	The hematocrit
HPLC	High-performance liquid chromatography
IABP	An intra-aortic balloon pump
ICU	Intensive care unit
IEF	In isoelectric focusing
KCl	Potassium chloride
LV	Left ventricular
LVH	LV hypertrophy
МСН	Mean corpuscular hemoglobin
MCHC	Mean corpuscular hemoglobin concentration
MCV	The mean corpuscular volume
MR	Mitral regurgitation
NRBC	Nucleated red blood cells
PA	Pulmonary artery
PCR	Polymerase chain reaction
POAF	Post-operative atrial fibrillation
PVR	Increase pulmonary vascular resistance
RBC	The red blood cell
RDW	Red cell distribution width

🕏 List of Abbreviations 🗷

SC	Sickle hemoglobin C disease
SCD	Sickle cell disease
SS	Sickle cell anemia
SVR	Systemic vascular resistance
TEE	Transesophageal echocardiography
UFH	Unfractionated heparin
WBC	White blood cell count
WIC	White blood cell impedance count

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Abstract

Sickle cell disease is a congenital haemoglobinopathy with a high incidence of perioperative complications. Traditional anaesthetic management, based largely on extrapolation from biochemical models, has emphasized avoidance of red cell sickling to prevent exacerbations of the disease.

The primary goal of cardiac surgery is not just minimally acceptable outcome where patient survives without life threating complication or persistant clinically manifest organ dysfunctions or simply hospital survival but healthy productive long term survivor

Keywords: Anaesthesia; complications, acute chest syndrome; complications, sickle cell disease; pain, crisis; surgery, dehydration, infections, hypoxia, inflammatory cascades, and acidosis

Introduction

Sickle cell hemoglobinopathy is a recessively inherited genetic disorder seen worldwide. It results from the mutation at the sixth codon of chromosome 11, ie, the β-globin gene. The condition may present as sickle cell disease (SCD), the severe form of which is the homozygous genotype (Haemoglobin SS), in which the fractional concentration of Haemoglobin S ranges between 70% and 98%, or it can be manifested as sickle cell trait (SCT), which is rather benign and more common among populations as the heterozygous genotype (Haemoglobin AS), in which the fractional concentration of Haemoglobin S is $\approx 50\%$ (*Messent*, 2004).

The classic precipitating factors for sickling include stress, exposure to cold, dehydration, infections, hypoxia, inflammatory cascades, and acidosis (*Firth*, 2005).

The primary goal of cardiac surgery is not just minimally acceptable outcome where patient survives without life threating complication or persistant clinically manifest organ dysfunctions or simply hospital survival but healthy productive long term survivor (*Murphy et al.*, 2009).

Anaesthetic protocols in cardiac surgery are investigated and analized in term of their effect on post operative mortaility and incidence of myocardial infarction following cardiac surgery,post operative troponin release,need for inotropic support,time on mechanical ventilation, icu, and hospital stay (*Landoni et al., 2009*).

Optimal preoperative preparation is required due to concerns of SCD as regarding anaemia, chronic pulmonary disease, pulmonary hypertension, cardiomegaly and heart failure, renal failure, haemolytic transfusion reaction resulting from alloimmunisation and extreme vulnerability to dehydration, acidosis, hypoxia and hypothermia. So, patients should be hydrated, infections controlled and haemoglobin levels should be within acceptable limits (*Lee*, 2009).

It should be noted that above-mentioned predisposing conditions are more common in patients undergoing cardiac surgery. Especially during the operation, CPB itself, as well as aortic cross-clamping, low-flow states, topical or whole-body hypothermia, cold cardioplegia, and use of vasoconstrictive agents, may predispose to the crisis state. Hence, special care should be taken in sickle cell patients who require cardiac surgery to avoid or at least to minimize those risk factors (*Frimpong et al.*, 1998).

Avoidance of hypoxia is crucial for preventing a sickling crisis; thus, oxygen delivery to the CPB circuit should be ensured at all times during surgery (Sachithanandan et al., 2008).

Postoperative oxygen therapy, liberal hydration and normothermia are to be maintained for minimum of 24 hrs, because crisis may occur suddenly postoperatively (*Stephen and Waltom*, 2006).

Aim of the Work

This essay aims to focus on the optimal anesthetic management of a patient with sickle cell haemo-globinopathy undergoing open heart surgery.

Pathophysiology of Sickle Cell Disease

Haemoglobin is a tetramer formed from two globin subunits – two α -globin chains and two β - globin chains in adult haemoglobin (HbA), and two α -globin chains and two γ -globin chains in fetal haemoglobin (HbF)-each bound to haeme Genes encoding the α -globin chains are located within the α gene cluster on chromosome 16, and those encoding the β - and γ -globin chains are in the β gene cluster on chromosome 11 (*Lukens*, 2004).

In addition, <2-3% of haemoglobin in adults is HbA2 which comprises two α -globin chains and two δ -globin chains; δ -globin is encoded by the δ gene, which is located on chromosome 11 between the γ and β genes (*Perkins*, 2004).

In adults, these hemoglobin molecules make up the following percentages of total hemoglobin:

- Hb A: 95% to 98%
- Hb A2: 2% to 3%
- Hb F: 0.8% to 2%