Recent Advances in Acute Chest Syndrome in Sickle Cell Disease

Essay

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List of Abbreviations

ACS	Acute Chest Syndrome.
ANF	*Avascular necrosis of head of femur*
ASSC	Acute Splenic Sequestration Crisis.
ATP	Adenosine Tri Phosphate.
BNP	*Biomarker pro-brain natriuretic peptide*
Ca^{++}	Calcium
CSSCD	The Cooperative Study of Sickle Cell Disease
DNA	Deoxyribonucleic acid
DPG	Diphosphoglyceraie
EDRF	*Endothelium derived relaxing factor*
ENOS	*Endothelium NOS*
EPO	*Erythropoietin*
ET-1	*Endothelin-1*
FDA	Food and Drug Administration
G6PD	Glucose 6 phosphate dehydrogenase
Glu	Glutamic acid
Hb A	Adult hemoglobin
Hb C	Hemoglobin C
Hb F	Fetal hemoglobin
Hb S	Sickle hemoglobin
Hb	Hemoglobin
HBAs	*Heterozygous type of sickle cell disease (sickle cell trait)*
Hbss	*Homozygous type of sickle cell disease (sickle cell anemia)*
His	Histidine
HJB	Howell-Jolly bodies
HPLC	High-performance liquid chromatography
HSCT	*Hematopoietic stem cell transplantation*
Hu	*Hydroxyurea*
IEF	Iso electric focusing
INOS	*Inducible NOS*
Kb	kilo bases
MCH	Main Corpuscular Hemoglobin
MCHC	Main Corpuscular Hemoglobin Concentration
MCV	Main Corpuscular Volume
Mg^{++}	Magnesium
MgSo4	*Magnesium sulphate*
NAD+	Nicotinamide adenine dinucleotide oxidized form
NADH	Nicotinamide adenine dinucleotide reduced form
NO	*Nitric oxide*

NOS	*Nitric oxide synthetase*
OSA	*Obstructive sleep apnea*
PAH	*Pulmonary artery hypertension*
PCR	Polymerese Chain Reaction
RNA	Ribonucleic acid
SCD	*Sickle cell disease*
SCD	Sickle cell Disease
SO2	*Oxygen saturation*
TCD	*Transcranial Doppler*
TIA	Transient ischemic attack
Tyr	Tyrosin
UTRs	Un translated Region
Val	Valin
VCAM-1	Vascular Cell Adhesion Molecule
VOC	*Vasoocclusive crisis*

Aim of the work

The aim of this work is studying the pathogenesis of sickle cell disease , the role of NO in this disease , its complications such as acute chest syndrome and anesthetic implications .

Introduction

Sickle cell disease is one of the most prevalent genetic diseases . Worldwide pulmonary disease , manifested as the acute chest syndrome (ACS) , is a common complication of sickle cell disease , accounting for 25% of premature deaths .
The last decade has witnessed a convergence of research pathways that were leading towards a better understanding of the new possible pathophysiology and therapies for this disease and new data on the effects of nitric oxide (NO) on sickle cell hemoglobin and interaction between them . **(Bunn, 1994)**

The acute chest syndrome is a common form of lung injury in sickle cell disease . When severe , this syndrome is analogous to the acute respiratory distress syndrome . The acute chest syndrome is the second common cause of hospitalization among patients with sickle cell disease and the leading cause of admission to an intensive care unit and premature death. It is found that about 60% of these patients with severe acute chest syndrome (ACS) had pulmonary hypertension and core pulmonale .
The risk for developing an ACS episode appears to be increased following surgery, with an average to the development of ACS of 3 days post surgery.
(Gladwin and Vichinsky, 2008)

The perioperative period can offer a unique insight into the origins of acute and chronic complications of sickle cell disease . An examination of the assumptions and consequences of anaesthetic practice aimed at the prevention and treatment of these complications , similarly can provide a useful distillation of management principles .
(Firth and Head, 2004)

PATHO-PHYSIOLOGY OF SICKLE CELL DISEASE

ROLE OF NITRIC OXIDE IN SICKLE CELL DISEASE

ACUTE CHEST SYNDROME AND CLINICAL FEATURES OF SICKLE CELL DISEASE

ANESTHETIC MANAGEMENT OF SICKLE CELL DISEASE

POST OPERATIVE CARE:

POST OPERATIVE CARE:

It is a crucial time for patient's with sickle cell disease. Skilled supervision is required as anesthetic accidents often occur during the recovery period. The same principles of management employed during surgery should be continued in the post operative period as oxygenation and hydration.

Postoperative analgesia

Postoperative pain management is challenging. Patients with sickle cell disease may have very high perioperative analgesic requirements, and may have tolerance to opioids. A multimodal approach should be used with a combination of opioids where indicated, paracetamol, NSAIDs, and regional anesthesia when possible ***(Goldschneider et al., 2001).***

Pain management strategies that may work in other people may not be as effective in this patient group owing to tolerance to analgesics developed over many years. Increasingly, patients with SCD who experience acute pain crisis are treated with a patient-controlled analgesia pump delivery system for opiates; therefore, it is likely that patients