

Perioperative implications of Sickle Cell Disease in neurosurgery and the use of automated red cell exchange as a tool to facilitate time sensitive surgery

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Introduction

Patients with Sickle Cell Disease (SCD) have an increased incidence of neurological complications.

Haemorrhagic and ischaemic strokes, silent cerebral infarction and cerebral sinus thrombosis are all increased.

The rate of neurological complications ranges from 6-30%.¹

Patients with SCD presenting for neurosurgical intervention present a particular clinical and logistical challenge to the surgical and anaesthetic team.

Automated red cell exchange (RCE) was an essential tool in the perioperative optimisation of these patients to facilitate time sensitive surgery.

Emergency Surgery

A 21-year-old presented with an acute subarachnoid haemorrhage (aSAH) requiring urgent neurosurgical intervention. Through excellent co-ordination with the apheresis service this patient was able to have an automated RCE and complete her surgery within 19 hours from initial presentation.

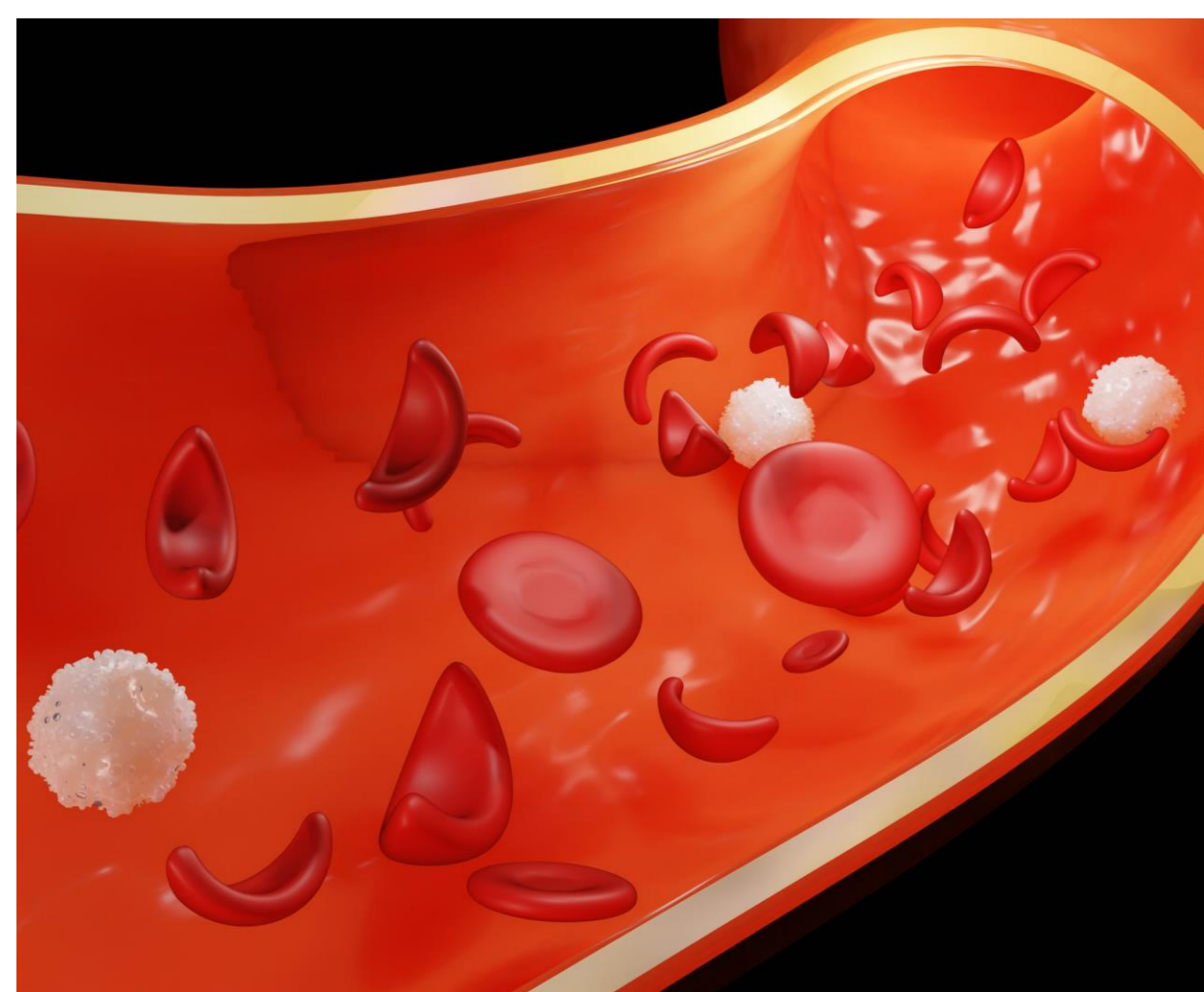


Figure 1. Red cell sickling and pathological changes in intracranial vessels increase the risk of post operative complications.

Patients with aSAH can undergo pathological changes of intracranial blood vessels resulting in focal hypoperfusion.

This patient was at high risk of vasospasm, and it was felt aneurysmal coiling was too unsafe for this patient.

The risk of aneurysmal re-bleeding following aSAH increases with time so automated RCE was necessary to facilitate urgent surgery.

Optimisation of blood flow and oxygen delivery are key along with reducing inflammatory change in cerebral vessels.

Elective Surgery

A 65-year-old presented with a pituitary macroadenoma with associated visual loss. She was found to be sickle cell positive on an incidental screen. She had no prior history or knowledge of this diagnosis. Automated RCE allowed surgery to be carried out in a timely manner.

It is extremely rare for a patient of this age to be unaware of a diagnosis of HbSC and be completely asymptomatic.

Current pre-operative screening criteria³ would not have picked this patient up, the finding was accidental. She had a Hb of 128/dl, HbS 54% with a HbF of 35%.

The anaesthetist and surgeon may not have acknowledged the need to reduce the Hb in this case because of the risk of thrombosis.

Urgent liaison with the specialist haemoglobinopathy team allowed urgent confirmation of diagnosis, timely clinical review and organisation of automated RCE.

Untreated HbSC with a high Hb increases the risk of central thrombotic events, most notably cavernous sinus thrombosis.

KEY MESSAGES

Patients undergoing intracranial neurosurgery are already at risk of both venous and arterial complications secondary to venous thrombosis.

SCD not only increases the chances of patients needing neurosurgical intervention but worsens the potential post operative complications.

HbSC would not have been detected if the standard protocol had been followed.
Are current preoperative screen guidelines sufficient ?

Red Cell Exchange

Automated

Manual

- Time consuming.
- Usually performed by a novice with other responsibilities.
- Unfamiliarity, cognitive load and time factors may increase transfusion errors.

- Specialist team required with single role allocation.
- Specialist equipment required.
- More precise control of Hb and circulating volume fluctuation.
- Reduced risk of transfusion errors.

In both cases automated RCE was effective at facilitating urgent surgery and reduced the risk of both intraoperative and postoperative complications.

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References:

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Figures: istock.com/Love Employee (Figure 1) and Vitalii Dumma (Background). Licensed to use images.

DISCUSSION

Balancing the optimisation of SCD with timely surgery requires expert input.

1 EXPERT INPUT

Lower pre-operative targets may be suggested by haemoglobinopathy apheresis specialists for high-risk individuals and surgeries.

Perioperative diagnosis and management of SCD requires excellent multidisciplinary teamwork and co-ordination.

2 TEAMWORK

The decision to transfuse (and the required modality) requires co-ordination with specialists at the regional haemoglobinopathy coordinating Centre (HCC).

Automated RCE has a vital role in the perioperative optimisation of neurosurgical patients, reducing the perioperative risk of stroke and thromboembolism.

3 ACCESS TO RCE

Access to automated RCE is not universal – further work is required to improve patient pathways and access to automated RCE in both the elective and emergency settings.