When Haemoglobinopathy Meets Neurosurgery: Learning points for the clinical team

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Introduction

Patients with Sickle Cell Disease (SCD) have an increased incidence of neurological complications including Haemorrhagic and ischaemic stroke, silent cerebral infarction and cerebral sinus thrombosis. Patients with SCD who present for neurosurgery present both clinical and logistical challenges to surgical and anaesthetic teams.

Automated Red Cell Exchange (RCE) allows for a rapid and precise reduction in HbS level and is an essential tool in the perioperative optimisation of these patients. Knowledge of this treatment, clinical pathways and logistics are vital for clinicians involved in the care of such patients. Here we discuss two cases which highlight best practice and provide education on RCE and how to access this treatment.

Case 1

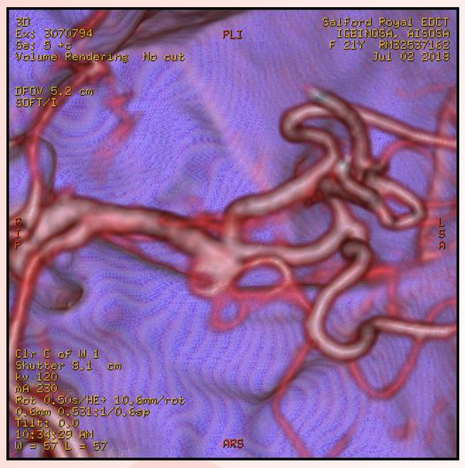


Figure 1. Broad based aneurysm of middle cerebral artery

A 21-year-old female presented with acute subarachnoid haemorrhage which required urgent craniotomy and clipping. She was found to have a broad-based aneurysm of the middle cerebral artery. Due to the high risk of vasospasm, it was felt aneurysmal coiling was unsafe for this patient. There was scarce documentation regarding her diagnosis and treatment.

She presented with an initial Hb S of 84%. Through co-ordination with the regional apheresis service, she was able to have Automated RCE delivered locally and completed within 19 hours of presentation and surgery completed within 34 hours. 9 units of packed red cells were transfused with a post exchange Hb of 98g/dl, HbS 24%.

Case 2

A 65-year-old presented with a pituitary macroadenoma. She was identified as having sickle cell disease during pre-operative workup. (a haemoglobinopathy screen was performed despite the patient not being anaemic and having no history or clinical signs to suggest the condition)

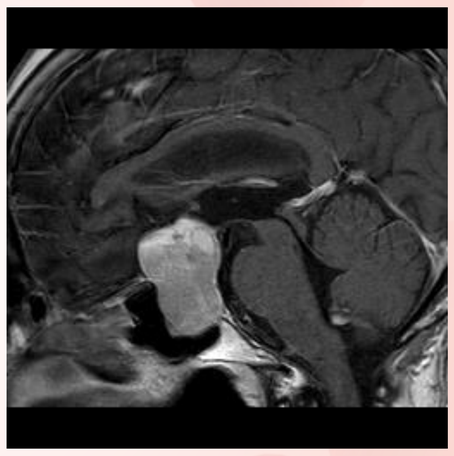


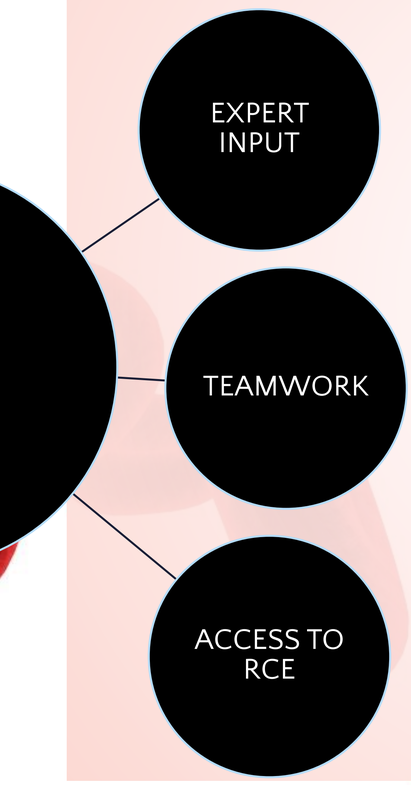
Figure 2. pituitary macroadenoma

Her initial Hb was 128g/dl, HbS 54% and HbF of 35%. Given the location of her tumour she was at increased risk of central thrombotic events, most notably, cavernous sinus thrombosis. Liaison with the regional haemoglobinopathy team allowed urgent confirmation of diagnosis, timely clinical review and automated RCE prior to surgery

Discussion

In both cases liaison with specialist haemoglobinopathy consultant and access to automated RCE was effective at reducing the risk of intraoperative and postoperative complications

Optimal care for these patients requires 3 things:



- Balancing the optimisation of SCD with timely surgery requires expert input
- Lower pre-op HbS and Hb targets may be suggested by haemoglobinopathy specialists for high-risk individuals

- Peri operative diagnosis and management requires effective multidisciplinary teamwork and co-ordination
- The decision to transfuse requires co-ordination with regional haemoglobinopathy centre

- Automated RCE has a vital role in the perioperative optimisation of neurosurgical patients
- Automated RCE is not currently available to all patients requiring neurosurgical intervention. Due to the potential benefits to patient care work is required to improve access for this patient group.

Automated Red Cell Exchange

What is Red Cell Exchange?

Red Cell Exchange involves the removal of the patients' blood and replacement with donor cells

What is the difference between automated and manual RCE?

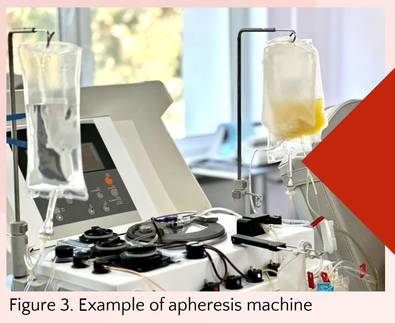
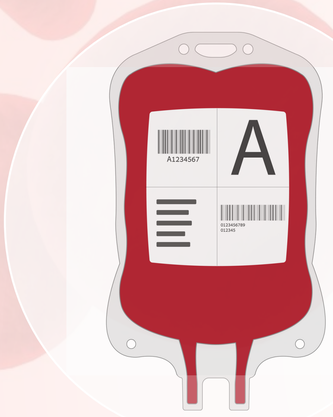


Figure 3. Example of apheresis machine

Manual exchange requires a clinician to perform removal of multiple units of the patients' blood and replace with cross matched units. It is time consuming, exposes the patient to variations in circulating volume, results in loss of some normal transfused red cells and the target Hb is less easily controlled. Automated RCE uses an apheresis system to remove and replace red blood cells.[1]

What are the advantages of automated RCE over manual RCE?



- Specialist Team with single role allocation
- Specialist equipment required
- More precise control of Hb and circulating volume fluctuation
- Quicker
- Less likely to have transfusion errors

How do I get this organised for my patients?

- Early Involvement of your regional haemoglobinopathy specialist Team
- Ideally one member of the team should be the point of contact with the haemoglobinopathy specialist
- Send urgent samples for Hb S level and cross matching
- Your patient may need to be transferred to a specialist centre for RCE and back again for surgery
- Central venous access may be required
- Apheresis requires HDU care with large bore peripheral access
- Exact processes differ by region. Familiarise yourself with local procedures

The northwest has the first regional apheresis service allowing mobilisation of a specialist team to local hospitals to deliver automated RCE.

Acknowledgments

1. Tsitsikas, DA, et al. Automated Red Cell Exchange in the Management of Sickle Cell Disease. J Clin Med. 10(4): 767
2. NICE [MTG28] 2016: Spectra Optia for automatic red blood cell exchange in people with sickle cell disease. Available at https://www.nice.org.uk/guidance/mtg28/

Questions

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