Clinical Standard Operating Procedure (SOP)

THROMBOTIC THROMBOCYTOPENIA PURPURA

SETTING	Service-wide
FOR STAFF	All staff
PATIENTS	Patients requiring time critical transfer to the Bristol Royal Infirmary for TTP investigation and management as part of NHS England commissioned service

Introduction

Thrombotic thrombocytopenic purpura is a very rare, complex condition that can present as an acute life threatening disorder that requires prompt diagnosis, early referral and effective immediate multidisciplinary management in a specialist centre.

Following NHS England Highly Specialised commissioning in 2021, University Hospitals Bristol and Weston NHS Foundation Trust (UHBW) has been commissioned to become the specialist centre for the South West.

This SOP covers the Retrieve management of patients with TTP and the coordination of the time critical transfer of patients, including identification of patients who may not require critical care transfer.

TTP overview

TTP is a rare form of thrombotic microangiopathy characterised by microangiopathic haemolysis, thrombocytopenia, neurological abnormalities, fever and renal dysfunction. It has a reported incidence of around 6 per 1,000,000 per year in the UK and is most common in adults, with a peak in the fourth decade of life. It is estimated that there are around 10 confirmed cases in the South West per annum, with up to 20 transfers for suspected cases pending confirmation of diagnosis.

Presentation is very variable, and there may be a prodrome of flu-like illness (including fever, fatigue, malaise and arthralgia). Patients can present with:

- Thromobocytopenia (epistaxis, bruising, petechiae, gingival bleeding, haematuria, menorrhagia, gastrointestinal bleeding, retinal haemorrhage, haemoptysis)
- Neurological sequelae (confusion, headache, paralysis, speech disturbance, visual problems, encephalopathy, coma) – this is often fluctuating
- Fever, pallor, jaundice, fatigue, arthralgia, myalgia
- Proteinuria, micro-haematuria, deranged creatinine and urea
- Cardiovascular instability (chest pain, cardiac failure, arrhythmias, hypotension)
- Abdominal pain

It requires rapid recognition, diagnosis and management – untreated mortality can be as high as 90%. The mainstay of management is plasma exchange (plasmapheresis / PEX) which removes the inhibitory antibodies and should be initiated as soon as possible. The combination of treatments, including those described below, can reduce acute episode mortality to around 20%.

Treatment strategies

The main goal is to provide plasma exchange at the Bristol Royal Infirmary (BRI) within 4 hours (and no longer than 8 hours) of referral, as specified in the NHS England Service Specification and monitored with quality indicators. To deliver this, time critical and safe transfer is required.

Clinical course and initial management

The clinical course of TTP is unpredictable and early deterioration is often subtle. Patients can present looking extremely well and progress to critical illness within hours. ‘

Most patients have a **very low platelet count** but, despite this, bleeding is usually rare (even after invasive procedures such as line insertion). **Do not administer platelets** unless specifically instructed to do so by a Consultant Haematologist as administration will usually exacerbate the microvascular thrombosis.

The medical management of patients for transfer to BRI will be **jointly directed by the Haemostasis Consultant on call in UHBW**. The most effective treatment is rapid transfer to BRI for early plasma exchange, but on occasion may also include:

- 1g methylprednisolone (usually already given before critical care referral)
- Fresh Frozen Plasma (FFP) 15mls/kg – this will depend on timescales of referral and transfer, and may need to be continued en route to the BRI – the preferred preparation is Octaplas (solvent detergent treated)
- CT imaging should ONLY be undertaken on the direct instruction of the Haemostasis Consultant – intracranial bleeding is rare and the initial treatment remains rapid plasma exchange in this instance
- Other organ failures due to widespread microthromboses – most often causing confusion, agitation, seizures (treat with sedation, intubation, ventilation) or hypoxia/ pulmonary oedema/ myocardial failure (if significant then intubate for transfer) – see the flowsheet below.

Early intubation and ventilation, particularly of the patient who is neurologically agitated, is often indicated to facilitate safe transfer. In patients who have signs of cerebrovascular accident or suspected intracranial haemorrhage (including those with low GCS and abnormal pupils), initial management remains plasma exchange. CT imaging should not delay transfer and will be arranged in the BRI as required and may, on a case-by-case basis, be delayed until plasma exchange has been initiated. Patients should receive standard neuroprotective care.

Intravenous and arterial access

Referring hospitals and the Retrieve team should not routinely insert central venous lines. In line with the Retrieve ‘Packaging’ SOP, patients should have two sites of peripheral intravenous access. Invasive arterial blood pressure monitoring is indicated in all critically unwell patients.

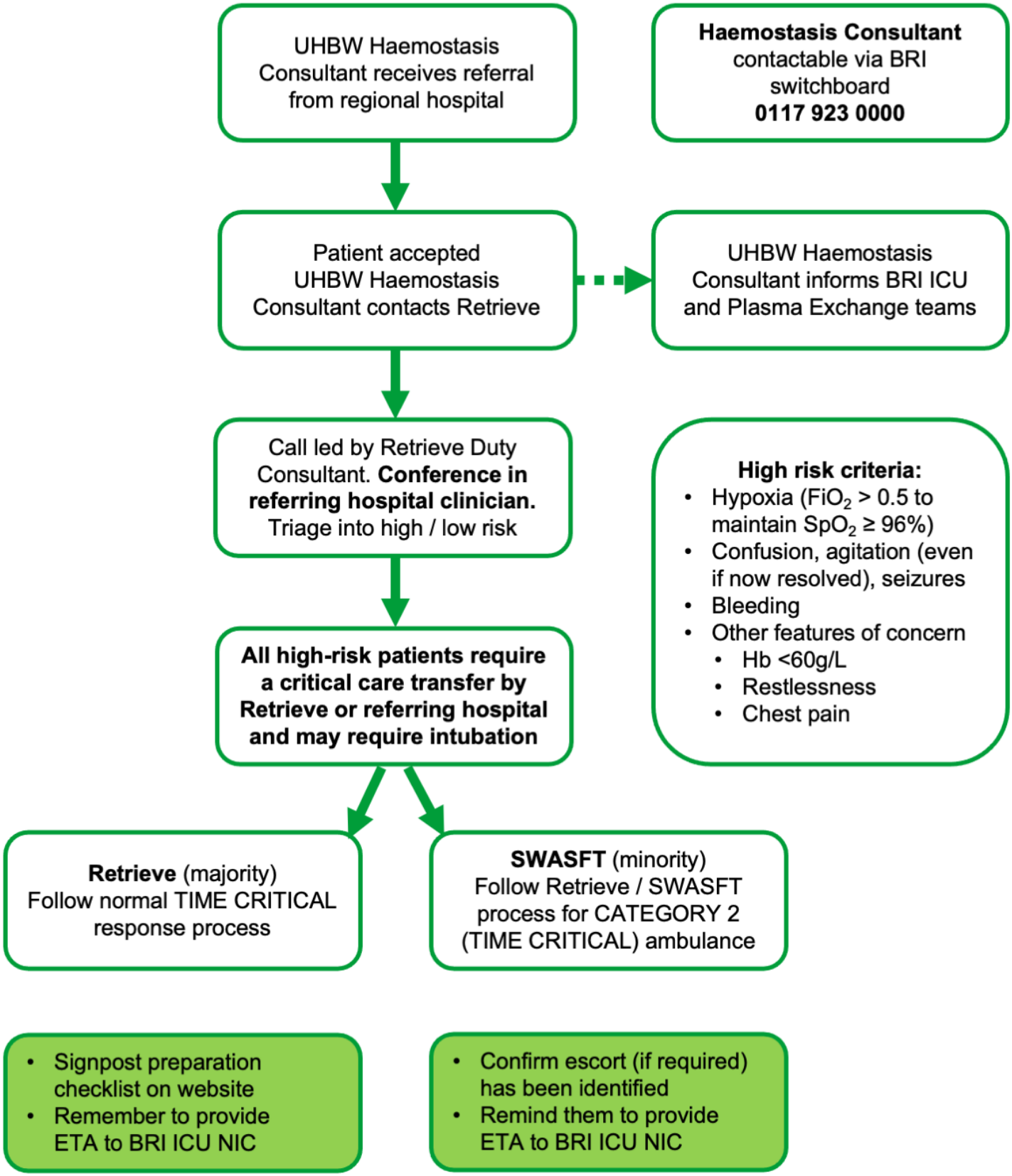
Transfer considerations

The usual Retrieve considerations should be taken for time critical patient transfers. Specific consideration should be given to the risk of patient deterioration if longer distance transfers (particularly those over 60 minutes and all patients from Peninsula to Bristol) are being undertaken.

Specific clinical advice, if required, should be sought from the Consultant Haematologist on-call (number below) at the BRI and/or the BRI Intensive Care Consultant on-call.

Referral pathway

The following process summarises the referral process for TTP patients:



Useful contact telephone numbers

UHBW Haemostasis Consultant

- Rota circulated to regional haematology departments every quarter with direct phone numbers
- Available on mobile via BRI Switchboard 0117 923 0000

BRI ICU

- Referral registrar baton phone 0117 342 7139
- Consultant available via BRI Switchboard 0117 923 0000
- Nurse-in-charge baton phone 0117 342 7238

Document Governance

REFERENCES	1. NHS England Service Specification – Thrombotic Thrombocytopenic Purpura https://www.engage.england.nhs.uk/consultation/thrombocytopenic-purpura/user_uploads/thrombotic-thrombocytopenic-purpura-service-specification.pdf (accessed 26th December 2021) 2. Patient.info Thrombotic Thrombocytopenic Purpura. https://patient.info/doctor/thrombotic-thrombocytopenic-purpura-pro (accessed 26th December 2021)
RELATED DOCUMENTS AND PAGES	
AUTHORISING BODY	Division of Surgery, University Hospitals Bristol & Weston NHS Foundation Trust
SAFETY	
QUERIES AND CONTACT	Retrieve Leadership Team