
Procedure

Referral procedure for patients with suspected or confirmed acute Thrombotic Thrombocytopenic Purpura (TTP) from the East of England Network

Key messages

- Thrombotic Thrombocytopenic Purpura (TTP) is a life-threatening condition that requires immediate admission and treatment.
- Cambridge University Hospitals (CUH) is the regional TTP centre for the East of England.
- This procedure outlines the referral pathways for patients to CUH, to enable Plasma Exchange (PEX) within 4-8 hours of referral.

Summary

Acute TTP has a mortality of 90% untreated. With prompt therapy (PEX), which must be within 4-8 hours when the diagnosis is suspected, this falls to 10-20%. Care of patients with acute TTP in the East of England is delivered at CUH, which is the commissioned regional TTP centre. This document summarises the referral pathway for referring hospitals to CUH. This document aims to meet the standard that patients with suspected TTP start treatment with PEX within 4-8 hours of referral to the CUH TTP centre, as specified in the national service specifications and standards, and to support timely transfer of patients to CUH.

1 Scope

For Trust use and sharing with East of England referring hospitals, for which CUH provides TTP services.

2 Purpose

To describe the process for patient referral to CUH with suspected or confirmed TTP from referring hospitals. This does not cover the management of TTP which is found in [Acute treatment of thrombotic thrombocytopenic purpura \(TTP\)](#). This document aims to meet the standard that patients with suspected TTP start treatment with PEX within 4-8 hours of referral to the CUH TTP centre, as specified in the national service specifications and standards, and to support timely transfer of patients to CUH.

3 Definitions

ADAMTS13 a Disintegrin and Metalloproteinase with a Thrombospondin Type 1 motif, member 13

CCU coronary care unit

CUH Cambridge University Hospitals NHS Foundation Trust

ED emergency department

EPIC emergency physician in charge

GP	general practitioner
ICU	intensive care unit
PEX	plasma exchange
RRT	rapid response team
TMA	thrombotic microangiopathy
TTP	thrombotic thrombocytopenic purpura
UCLH	University College London Hospitals

Where TTP is discussed in this document it refers to suspected or confirmed cases.

4 Introduction

TTP is a rare life-threatening medical emergency. It has an incidence of 6-10 per million people. It has a mortality of 90% untreated which can be reduced to 10% with modern treatment. National TTP centres were commissioned in 2022 and it is a requirement that each centre has local treatment guidance for the condition.

Prompt referral and initiation of treatment from referring hospitals to CUH is therefore critical and life-saving – it is also stipulated in the commissioning service specification for specialist TTP centres. CUH was commissioned as a specialist TTP centre in 2022.

5 Referring hospitals/Trusts within the East of England TTP centre network

- West Suffolk Hospital
- Ipswich Hospital
- Queen Elizabeth Hospital
- James Paget Hospital
- Norfolk and Norwich University Hospital
- Bedford Hospital
- Lister Hospital
- Princess Alexandra Hospital
- Peterborough City Hospital and Hinchingsbrooke Hospital (North West Anglia Foundation Trust)
- Royal Papworth Hospital

6 Sending ADAMTS13 activity samples to CUH specialist haemostasis laboratory

The CUH non-malignant haematology consultant on-call must be called if a referring centre is considering sending an ADAMTS13 sample for testing. Prior to sending a sample, review a blood film as the initial diagnosis of TTP usually can be made where there is severe thrombocytopenia with red cell fragments in the absence of other causes, rather than relying on ADAMTS13 testing which is time consuming and may lead to treatment delays (see flow chat in section 8 for more details).

A specimen request form for all first-time or urgent ADAMTS13 samples is required to be sent with the sample and is available in Appendix 1. This form is not required for samples from known TTP patients requiring a non-urgent ADAMTS13.

Medical haematology

Division B

Once the referring clinician has discussed with the CUH non-malignant haematology consultant on call, in working hours (Monday-Friday, 0900-1700) the referral laboratory should contact the CUH specialist haemostasis laboratory (01223 274 722) to arrange delivery. Out of hours the CUH non-malignant haematology consultant will liaise with the specialist haemostasis laboratory biomedical scientist to facilitate the sample transfer.

To speak with the on-call haematology consultant call 01223 245 151 (CUH switchboard) and ask for the 'on-call non-malignant haematology consultant' via mobile phone.

7 Process for patients in the community with TTP who are outpatients with a suspected acute episode

7.1 Patients known to CUH with TTP or from the CUH GP catchment area

The CUH non-malignant haematology consultant on-call should be called. If possible, the patient should be advised to attend the CUH ED. The ED reception and EPIC must be informed by the CUH consultant.

If the patient is 'out of CUH GP catchment area' and too sick to attend CUH ED they should go to a local ED for transfer and stabilisation and discussions had about hospital transfer (see later in this procedure). The CUH non-malignant haematology consultant must be informed as must the relevant referring hospital haematology consultant should be contacted about the patient.

To speak with the on-call haematology consultant call 01223 245 151 (CUH switchboard) and ask for the 'on-call non-malignant haematology consultant' via mobile phone.

7.2 Patients out of the CUH GP catchment area and not previously known to CUH with TTP

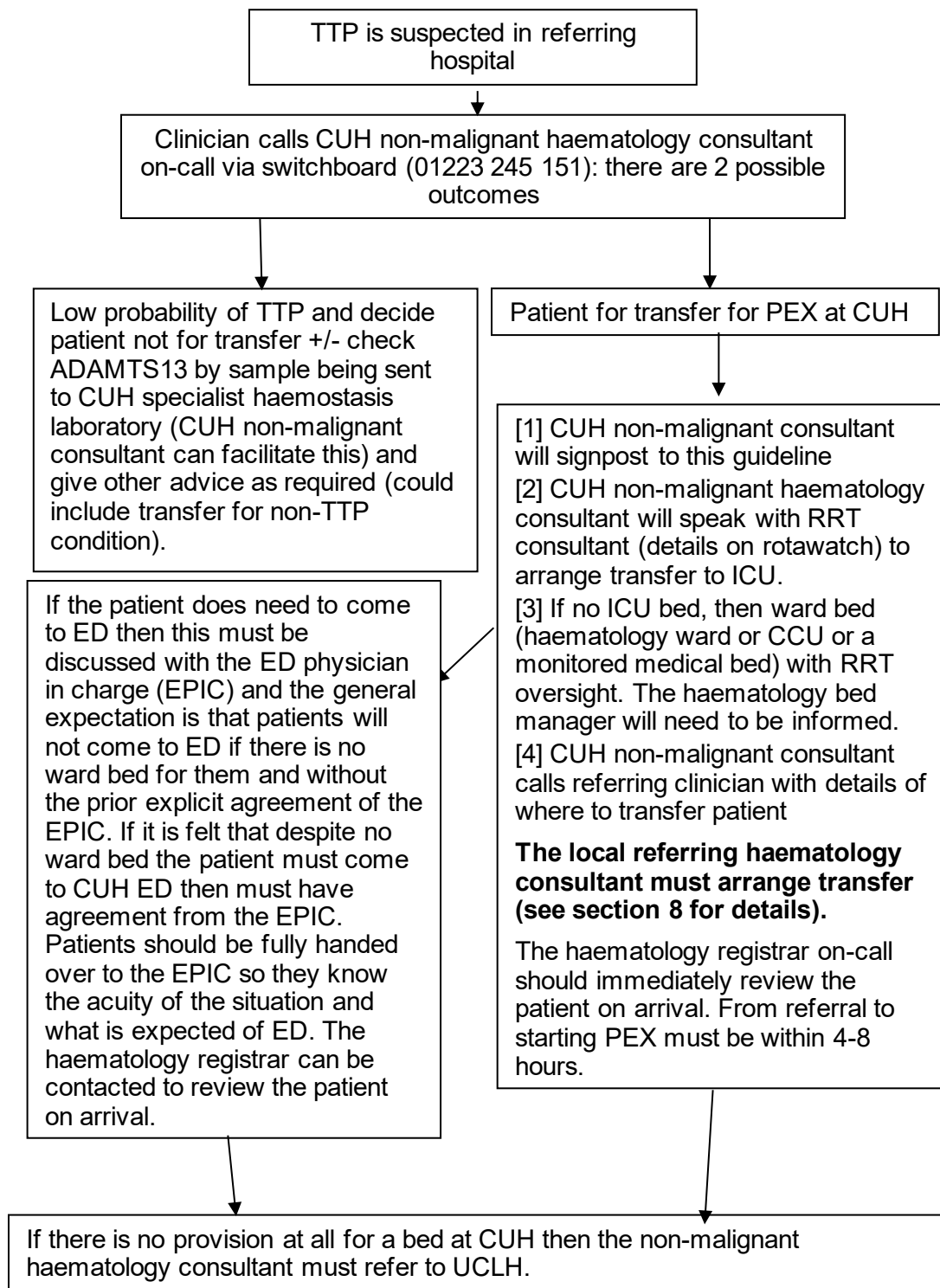
The referring hospital clinician should call the CUH non-malignant haematology consultant. Inpatients must not be referred to ED without discussion and agreement from the CUH non-malignant consultant (generally an inpatient-inpatient/ED-inpatient transfer is preferred rather than inpatient-ED or ED-ED). If a transfer to CUH ED is considered this must be discussed by the CUH haematology consultant with the EPIC (see section 7 for more details).

For outpatients if TTP is felt likely and if possible, they should be advised to attend CUH ED. The ED reception and EPIC must be informed, by the CUH consultant. If the patient is 'out of area' and too sick to attend CUH they should go to a local ED for transfer and stabilisation and discussions had about hospital transfer (see later in this procedure).

To speak with the on-call haematology consultant call 01223 245 151 (CUH switchboard) and ask for the 'on-call non-malignant haematology consultant' via mobile phone.

8 Process for patients with TTP who are in a DGH and require transfer

The TTP commissioning service specification has an ‘automatic acceptance’ and ‘non-refusal’ of referrals stipulation. The process for a referral from a referring hospital clinician is outlined in the flowchart below:



9 Practicalities of arranging urgent ambulance transfers

[1] **First line option.** The East of England Adult Critical Care Transfer Service (ACCTS) is the first line for TTP patient transfer and TTP is listed as an agreed condition for transfer even if the patient is not being transferred between ICU wards. It operates 24/7 and referrals are made by calling **0333 016 9859 (option 1)** to request a transfer team. When the referral phone call to ACCTS is placed, the team will direct the referrer to an online form to complete on their website (<https://www.transfer-eastofengland.nhs.uk/>).

The referring hospital haematology consultant or delegate should call the service. This is to ensure the correct clinical details and level of urgency is conveyed for prioritisation of transfer, and because they have the patient information at hand to make the booking.

[2] **Second line option.** Only in the event that the Adult Critical Care Transfer Service to do not have capacity to facilitate transfer, then the process is to phone the Ambulance Service.

The referring hospital haematology consultant or delegate should call the service. This is to ensure the correct clinical details and level of urgency is conveyed for prioritisation of transfer, and because they have the patient information at hand to make the booking.

Healthcare professional contact phone numbers for the ambulance service for each county:

- Cambridgeshire 01603 888 060
- Bedfordshire & Luton 01234 716 120
- Essex 01245 443 241
- Hertfordshire 01234 716 120
- Norfolk 01603 888 060
- Suffolk 01603 888 060

During the call, you will be asked clinical questions about the patient's condition.

In a life-threatening situation or an emergency request, it is the responsibility of the attending clinician to make the request to the ambulance trust. Where delegation is unavoidable, the individual making the request should be able to answer triage questions about the patient's condition. When you book transport, a clinician from the Ambulance Operations Centre may get back in touch with you for further assessment to ensure the right response is sent to the patient.

Emergency ambulance transport must not be booked for repatriations or step-down transfers/discharges to non-hospital facilities and outpatient appointments.

TTP would be categorised as an 'inter-facility transfer (IFT) category 2 (IFT2)' and the level of ambulance response is 'emergency'. This level of response is based on the clinical condition of the patient and the need, or a high likelihood of the need, for further treatment and management at the destination facility rather than the patient's diagnosis. Immediate life, limb or sight (globe trauma) threatening (ILT) situations that require immediate management in another healthcare facility should receive this level of response. Other examples include patients going directly to theatre for immediate neurosurgery, primary percutaneous coronary intervention, stroke thrombolysis, mechanical thrombectomy, surgery for ruptured aortic aneurysm,

emergency surgery for ectopic pregnancy, limb or sight saving surgery or mental health patients being actively restrained (National Framework for Inter-Facility Transfer).

10 Consideration for giving steroids and fresh frozen plasma at the referring hospital

Methylprednisolone 1 gram intravenously and Fresh Frozen Plasma (FFP) infusion (preferably solvent detergent plasma (OctaplasLG) (10-15ml/kg)) should be given (at a rate of 10ml/kg/hr or as clinically tolerated) while awaiting transfer if there are any delays in cases of suspected TTP. This should not delay the transfer.

11 Consideration for giving caplacizumab at Norfolk and Norwich University Hospital

A single dose of caplacizumab 10mg will be stocked at the Norfolk and Norwich University Hospital. If there are concerns over the clinical state of the patient (e.g. severe neurological or cardiac involvement) or delays in ambulance transfer then it can be given intravenously to the patient prior to departure from NNUH after discussion between the Norfolk and Norwich University Hospital haematology team and CUH non-malignant haematology consultant. **This should not delay the transfer.**

Caplacizumab requires a Blueteq form being completed for funding, so the approving CUH consultant will need to complete this on behalf of the Norfolk and Norwich for them to be reimbursed.

For CUH staff: See the appendix of the [CUH TTP guideline](#) for details of supplying caplacizumab to CUH inpatients or on discharge.

12 Paediatric patients

Referrals will be via direct liaison between the hospital and paediatric haematology-oncology consultant. These patients should be transferred urgently via the paediatric ambulance service.

13 Operational policy where there is no bed available at CUH

For all Trusts in the East of England network then contact UCLH TTP consultant to make the referral, (UCLH switchboard is on 020 3456 7890; ask for the on-call TMA consultant).

It is the responsibility of the CUH non-malignant consultant to phone UCLH and start the referral process and liaison. This should not be delegated to the referring hospital.

14 Monitoring compliance with and the effectiveness of this document

Periodic audit of referrals to CUH TTP centre. The outcome of interest is time from referral (as recorded in the patient record) to plasma exchange. A target of <4-8 hours is the standard.

15 References

National Framework for Inter-Facility Transfer (2019). Accessible at: [NHS England » National Framework for Inter-Facility Transfers](#), last accessed 24.8.2022.

Scully M, Hunt BJ, Benjamin S, Liesner R, Rose P, Peyvandi F *et al.*, (2012). Guidelines on the diagnosis and management of thrombotic thrombocytopenia purpura and other thrombotic microangiopathies. *BJH*. 158(3); 323-35.

<https://www.england.nhs.uk/wp-content/uploads/2022/08/1668-Service-Specification-2022.pdf>

last accessed 29/05/2025

16 Associated documents

- [Acute treatment of thrombotic thrombocytopenic purpura \(TTP\)](#).

Equality and diversity statement

This document complies with the Cambridge University Hospitals NHS Foundation Trust service equality and diversity statement.

Disclaimer

It is **your** responsibility to check against the electronic library that this printed out copy is the most recent issue of this document.

Document management

Approved by:	E Gowers		
Approval date:	19 June 2025		
JDTC approval date:	12/05/2025		
Owning department:	Medical haematology		
Author(s):	J. Griffin		
Pharmacist:	A Saoulidis		
Version number:	4		
Local reference:		Document ID:	102706

Appendix 1: Specimen request form for ADAMTS13 testing (first-time or urgent samples):

ADAMTS13 Request Form

This request form should be utilised for all initial presentations and any urgent requests, such as suspected relapse. For non-urgent requests in known TTP patients, please continue to use your standard request forms. Please complete all details in shaded boxes or request will not be processed.

Surname		NHS Number	
First name(s)		Date of Birth	
Sex			
Requesting Hospital		Requesting Clinician	
Phone number of requesting clinician		CUH Consultant Discussed with	
Urgency			

Please note: **All** sample requests must be discussed with the CUH non-malignant consultant on call (contact via CUH switchboard 01223 805000) and will not be processed as urgent without prior agreement.

Clinical Details:

Plasmic Score:		
Platelet Count <30x10 ⁹ /L	No = 0, Yes = +1	
Haemolysis: Reticulocyte count >2.5%, haptoglobin undetectable, or indirect bilirubin >2.0 mg/dL (34.2 µmol/L)	No = 0, Yes = +1	
Active cancer Treated for cancer within the past year	No = +1, Yes = 0	
History of solid-organ or stem-cell transplant	No = +1, Yes = 0	
MCV <9.0 x 10-14 L (<90 fL)	No = 0, Yes = +1	
INR <1.5	No = 0, Yes = +1	
Creatinine <2.0 mg/dL (176.8 µmol/L)	No = 0, Yes = +1	
Plasmic Score:		

PLASMIC Score	Risk group
0-4	Low
5	Intermediate
6-7	High

Sample Collection	
Collection Date:	
Collection Time:	