## Haematology audit template

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| Date of completion  | (To be inserted when completed) |
| Name of lead author/participants | (To be inserted) |
| Specialty | Haematology |
| Title | An audit of compliance with the British Society for Haematology guideline for the diagnosis and management of thrombotic thrombocytopenic purpura and other thrombotic microangiopathies |
| Background | The British Society for Haematology (BSH) has published revised guidance on the diagnosis and management of thrombotic thrombocytopenic purpura (TPP) and other thrombotic microangiopathies (TMAs).1 This audit will review compliance with some of the level 1 recommendations made. |
| Aim and objectives | This audit template is a tool to review whether:investigations are performed appropriately in the diagnosis of TTPpatients with TTP and related TMAs are being managed in an appropriate way. |
| Standards and criteria | **Criteria range:** 100%, or if not achieved, there is documentation in the case notes that explains the variance.**Diagnosis of TTP**1. Pretreatment samples should be obtained to measure ADAMTS13 activity levels and to detect anti-ADAMTS13 antibodies.
2. Serological tests for human immunodeficiency virus (HIV), hepatitis B virus (HBV) and hepatitis C virus (HCV), autoantibody screen and, when appropriate, a pregnancy test should be performed at presentation.

**Initial management of acute TTP**1. Platelet transfusion should be avoided.

**Therapies and evidence for use in TTP**1. Caplacizumab should be initiated on confirmation of acute immune-mediated TPP (iTTP).
2. Intravenous daily methylprednisolone (e.g. 1 g/day for 3 consecutive days [adult dose]) or high-dose oral prednisolone (e.g. 1 mg/kg/day) should be considered, with tapering when there is a sustained increase in ADAMTS13 activity levels.
3. Plasma exchange (PEX), with OctaplasLG should be started with 1.5 PV exchanges and reassessed daily, reducing to 1.0 V when the clinical picture and laboratory tests are stabilising.
4. Monoclonal anti-CD20 therapy should be initiated within 3 days of acute immune-mediated TTP (iTTP) admission.
5. All hospitalised/immobilised patients should receive thromboprophylaxis once platelet counts are ≥50×109/L, even when treated with caplacizumab.

**Follow-up after an TTP episode**1. Pre-emptive therapy with rituximab should be given when ADAMTS13 activity <20 IU/dL or higher levels associated with clinical symptoms.

**Management of congenital TTP (cTTP)**1. Diagnosis of cTTP is confirmed by: persisting low ADAMTS13 activity <20 IU/dL; no anti-ADAMTS13 antibody and confirmation of homozygous or compound heterozygous variants in the *ADAMTS13* gene.
2. ADAMTS13 prophylaxis should be considered for all patients with cTTP, with an individualised approach to dose and frequency according to symptoms, whether overt or non-overt.

**Management of pregnancy-associated TTP**1. Patients presenting for the first time with TTP in pregnancy should initially be treated as per iTTP with PEX and steroids.
2. Women presenting with TTP in pregnancy should have investigations to determine whether they have iTTP or a first presentation of cTTP.
3. For pregnant women with cTTP, regular solvent/detergent fresh frozen plasma (SD-FFP) replacement therapy/ADAMSTS13 replacement therapy should be given prophylactically to prevent clinical TTP relapse in subsequent pregnancies if not on regular prophylaxis.

**HIV-associated TTP**1. HIV-associated iTTP should be treated with highly active antiretroviral therapy (HAART) and PEX/steroids/caplacizumab.
2. In patients with low/undetectable viral load, ADAMTS13 relapse or clinical relapse should be treated as standard iTTP.

**Haemolytic uraemic syndrome (HUS)**1. In TMAs associated with renal impairment, ADAMTS13 activity should be checked to exclude TTP.
2. Complement-mediated HUS (CM HUS) is a clinical diagnosis (that can sometimes be confirmed by detection of a pathogenic complement gene variant or relevant autoantibody) for which prompt complement inhibitor therapy should be initiated.
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| Method | **Sample selection:** (To be completed by the author)All patients diagnosed with TPP in the preceding 12 months up or 10 consecutive patients.**Data to be collected on proforma (see below).** |
| Results | (To be completed by the author)The results of this audit show the following compliance with the standards.

| Investigation | No. audited | No. compliant | % compliance |
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| Diagnosis of TTP |
| Pretreatment samples were obtained to measure ADAMTS13 activity levels and to detect anti-ADAMTS13 antibodies |  |  |  |
| Serological tests for HIV, HBV and HCV, autoantibody screen and when appropriate, a pregnancy test, were performed at presentation |  |  |  |
| Initial management of acute TPP |
| Platelet transfusion was avoided |  |  |  |
| Therapies and evidence for use in TTP |
| Caplacizumab was initiated on confirmation of acute iTTP  |  |  |  |
| Intravenous daily methylprednisolone (e.g. 1 g/day for 3 consecutive days [adult dose]) or high-dose oral prednisolone (e.g. 1 mg/kg/day) was considered, with tapering when there was sustained increase in ADAMTS13 activity levels |  |  |  |
| PEX, with OctaplasLG was started with 1.5 PV exchanges, and reassessed daily, reducing to 1.0 V when the clinical picture and laboratory tests were stabilising |  |  |  |
| Monoclonal anti-CD20 therapy was initiated within 3 days of acute iTTP admission |  |  |  |
| All hospitalised/immobilised patients received thromboprophylaxis once platelet counts were ≥50×109/L, even when treated with caplacizumab |  |  |  |
| Follow-up after an TTP episode |
| Pre-emptive therapy with Rituximab was given when ADAMTS13 activity <20 IU/dL or higher levels associated with clinical symptoms |  |  |  |
| Management of cTTP |
| Diagnosis of cTTP was confirmed by: persisting low ADAMTS13 activity <20 IU/dL; no anti-ADAMTS13 antibody and confirmation of homozygous or compound heterozygous variants in the *ADAMTS13* gene |  |  |  |
| ADAMTS13 prophylaxis was considered for all patients with cTTP, with an individualised approach to dose and frequency according to symptoms, whether overt or non-overt |  |  |  |
| Management of pregnancy-associated TTP |
| Patients presenting for the first time with TTP in pregnancy were initially treated as per iTTP with PEX and steroids |  |  |  |
| Women presenting with TTP in pregnancy underwent investigations to determine whether they had iTTP or a first presentation of cTTP |  |  |  |
| For pregnant women with cTTP, regular SD-FFP replacement therapy was given prophylactically to prevent clinical TTP relapse in subsequent pregnancies if not on regular prophylaxis |  |  |  |
| Management of HIV-associated iTTP |
| HIV-associated iTTP was treated with HAART and PEX/steroids/caplacizumab |  |  |  |
| In patients with low/undetectable viral load, ADAMTS13 relapse or clinical relapse was treated as standard iTTP |  |  |  |
| Management of HUS |
| In TMAs associated with renal impairment, ADAMTS13 activity was checked to exclude TTP |  |  |  |
| CM HUS is a clinical diagnosis (that can sometimes be confirmed by detection of a pathogenic complement gene variant or relevant autoantibody) for which prompt complement inhibitor therapy was initiated |  |  |  |

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| Conclusion | (To be completed by the author) |
| Recommend-ations for improvement | Present the result with recommendations, actions and responsibilities for action and a timescale for implementation. Assign a person(s) responsible to do the work within a timeframe. |
| Action plan | (To be completed by the author – see attached action plan proforma) |
| Re-audit date | (To be completed by the author) |
| References | 1. Scully M, Rayment R, Clark A, Westwood JP, Cranfield T, Gooding R *et al*. A British Society for Haematology Guideline: Diagnosis and management of thrombotic thrombocytopenic purpura and thrombotic microangiopathies. *Br J Haematol* 2023;203:546–563.
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## Data collection proforma for patients (Diagnosis and management of thrombotic thrombocytopenic purpura and other thrombotic microangiopathies)

## Audit reviewing practice

Unit number(s)

Date of transfusion:

(Note: a separate form should be completed for each transfusion episode.)

**Given to:**

Patient name:

Hospital number:

Date of birth:

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|  | **1**Yes  | **2**No | **3** If shaded box not ticked, was there documentation to explain the variance?**Yes/No** plus free-text comment | **4** Compliant with guideline if shaded box ticked or an appropriate explanation from column 3. **Yes/No**(Record if standard not applicable) |
| **Diagnosis of TTP** |
| **1** Pretreatment samples were obtained to measure ADAMTS13 activity levels and to detect anti-ADAMTS13 antibodies |  |  |  |  |
| **2** Serological tests for HIV, HBV and HCV, autoantibody screen and when appropriate, a pregnancy test, were performed at presentation |  |  |  |  |
| **Initial management of acute TTP** |
| **1** Platelet transfusion should be avoided |  |  |  |  |
| **Therapies and evidence for use in TTP** |
| **1** Caplacizumab was initiated on confirmation of acute iTTP |  |  |  |  |
| **2** Intravenous daily methylprednisolone (e.g. 1 g/day for 3 consecutive days [adult dose]) or high-dose oral prednisolone (e.g. 1 mg/kg/day) was considered, with tapering when there was sustained increase in ADAMTS13 activity levels |  |  |  |  |
| **3** PEX, with OctaplasLG was started with 1.5 PV exchanges, and reassessed daily, reducing to 1.0 V when the clinical picture and laboratory tests were stabilising |  |  |  |  |
| **4** Monoclonal anti-CD20 therapy was initiated within 3 days of acute iTTP admission |  |  |  |  |
| **5** All hospitalised/immobilised patients received thromboprophylaxis once platelet counts were ≥50×109/L, even when treated with caplacizumab |  |  |  |  |
| **Follow-up after an TTP episode** |
| **1** Pre-emptive therapy with Rituximab was given when ADAMTS13 activity <20 IU/dL or higher levels associated with clinical symptoms |  |  |  |  |
| **Management of cTTP** |
| **1** Diagnosis of cTTP was confirmed by: persisting low ADAMTS13 activity <20 IU/dL; no anti-ADAMTS13 antibody and confirmation of homozygous or compound heterozygous variants in the *ADAMTS13* gene |  |  |  |  |
| **2** ADAMTS13 prophylaxis was considered for all patients with cTTP, with an individualised approach to dose and frequency according to symptoms, whether overt or non-overt |  |  |  |  |
| **Management of pregnancy-associated TTP** |
| **1** Patients presenting for the first time with TTP in pregnancy were initially treated as per iTTP with PEX and steroids |  |  |  |  |
| **2** Women presenting with TTP in pregnancy underwent investigations to determine whether they had iTTP or a first presentation of cTTP |  |  |  |  |
| **3** For pregnant women with cTTP, regular SD-FFP replacement therapy was given prophylactically to prevent clinical TTP relapse in subsequent pregnancies if not on regular prophylaxis |  |  |  |  |
| **Management of HIV-associated iTTP** |
| **1** HIV-associated iTTP was treated with HAART and PEX/steroids/caplacizumab |  |  |  |  |
| **2** In patients with low/undetectable viral load, ADAMTS13 relapse or clinical relapse was treated as standard iTTP |  |  |  |  |
| **Management of HUS** |
| **1** In TMAs associated with renal impairment, ADAMTS13 activity was checked to exclude TTP |  |  |  |  |
| **2** CM HUS is a clinical diagnosis (that can sometimes be confirmed by detection of a pathogenic complement gene variant or relevant autoantibody) for which prompt complement inhibitor therapy was initiated |  |  |  |  |

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| **Audit action plan** An audit of compliance with the BSH guideline (Diagnosis and management of thrombotic thrombocytopenic purpura and other thrombotic microangiopathies) |
| Audit recommendation | Objective | Action | Timescale | Barriers and constraints | Outcome | Monitoring |
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