Quiz Jan 2025 TTP Diagnosis Results guide

JANUARY 14

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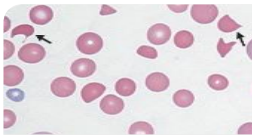
Q1. . What is your profession?

Responses.

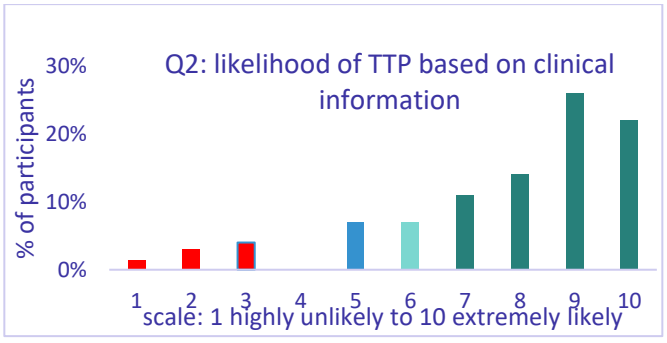
47% answered as Biomedical Scientist for this question. The remaining was mixture of the below:

Q2. How likely is a diagnosis of TTP with patient presenting with a TMA and the following clinical details: Recent travel to India. Unwell from for a week- intermittent dark urine, had electrolyte drink, abdominal pain. Flagged bloods results: (1 point)

1. WBC: 13.0
2. PLTS: 15
3. HB: 67
4. LDH: 1136
5. BIL: 137
6. Cr: 236
7. Blood film showed clear fragments



Responses.



Expected answer is highly likely.
7 and above was the requirements.

I.e. based on the clinical featured there is a ~70% chance of TTP based on these finding.
Platelet counts <30, quite visible fragment on blood and the extremely high LDH film is highly suggestive of TTP.

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This patient was initially diagnosed with UTI by his GP and then HUS on admission, the dark urine and creatinine through off the initial examiners.
The abdominal pain & dark urine is also seen in TTP. This level of thrombocytopenia, and LDH and prominent red cells fragments is key that should be flagging the most to clinical team to suspect TTP.
Patient Diagnosed with ITTP: ADAMTS13 levels <5%, Antibody levels: 53%; successfully treated.

Q3. What to do with ADAMT13 activity of 4% from the Acustar method low clinical suspicion of TTP? (1 point)

Responses.

82% of participants answered this question correctly. 👏

A previous quiz on the same question performed in March 2024; only 35% of participants got this question correct.

Repeat on Acustar using another sample	X
treat as TTP	X
run ADAMTS IgG antibodies to confirm before making diagnosis	X
Repeat on FRET/ELISA method before making diagnosis	✓

Expected answer
The low clinical suspicion should prompt a repeat to confirm result to exclude spurious results

Current guidelines are set as <10% as the cut-off for TTP. However this is not platform specific. Acustar results for acute ADAMTS13 iTTP is shown to run much lower at presentation and discrepant results for non-TTP are seen yielding results between 2% to 10%. See proposed guidelines for CLIA method ADAMTS13 activity levels.

If **low clinical suspicion** then await for repeat ADAMTS13 on FRET method.

- Carefully note patient deterioration, noting both bleeding and thrombotic symptoms, raising LDH levels, platelet counts continuing to decease. If any of the above is observed then treat as TTP and do not wait for ADAMTS13 result**.**

Results classification of Acustar method

- **<1.5% = mostly certainly iTTP**
- **1.6- 3.0% = possible iTTP or cTTP***
- **>3.1% = unlikely to be iTTP – consider other TMA or cTTP**
- **cTTP* can have results ranging from 0-20%**

Q4. What to do with ADAMTS13 activity of 2.1 % from Acustar method with a high clinical suspicion of TTP? (1 point)

Responses.

60% of participants answered this question correctly. 27% stated repeat on another method which is partially correct in this scenario.

Treat as TTP	✓
Consider other TMA	X
Repeat on Acustar but using another sample before making diagnosis	X
Repeat on another method before making diagnosis	X

If High clinical suspicion of TTP then treat as TTP
Sending repeat sample to test on another method to confirm result is not incorrect though this is not the first course of

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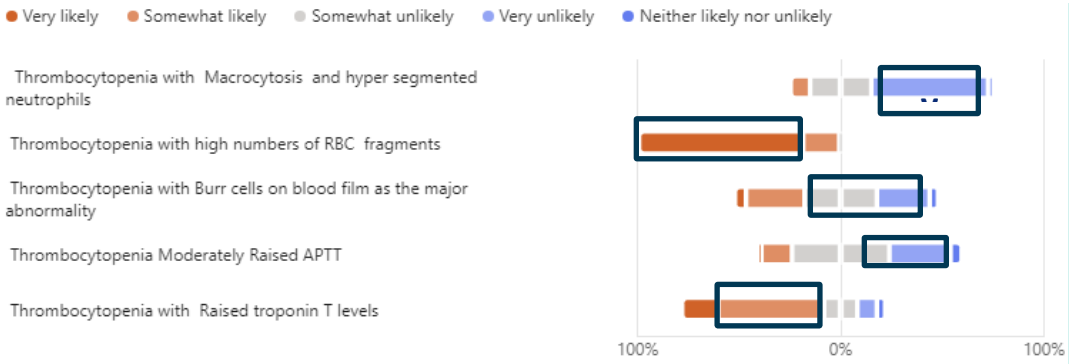
Q5. How likely is a diagnosis of TTP with patients presenting with significantly low platelets (<60x10 9/L) and following lab results (listing only next major lab abnormality). (5 points)

Responses.

Very mixed responses from the receipts, which is expected. Reviewer will examine all the key parameters before making judgment. However all these cases are referral cases made to UCLH and the below is all the information provided which illustrates the need to provide as much clinical details as possible if referring for TMA differential diagnosis.



Correct answers.

In order top to bottom
1. Very unlikely
2. Very likely
3. Somewhat unlikely
4. Very unlikely
5. Somewhat likely

Box: accepted area for correctness

Cases

1. >60% of participants got this correct. Case with thrombocytopenia of 55x $10^{/9}$/L and very occasional RBC fragments, Junior SPR orders urgent ADAMTS13 on this basis, completely overlooking the signs of B12 deficiency. Some mitigation as the patient showed signs of neurological impairment. However, on review, this was due to underlying Schizophrenia and alcohol related problems not noted at time of presentation.

2. >90% of participants got this correct. Case with genuine RBC fragments note in each field of 40x magnification view with platelet count of 29x $10^{/9}$/L.

3. Mixed responses with only 35% participants getting this correct. Burr cells and no signs of genuine RBC fragments. Patient had renal failure.

4. Prolonged APTT in isolation with thrombocytopenia warrants a close examination though TTP here is unlikely. Patient had CAPS.

5. Patients present for cardiac impairment, had raised Trop T and Thrombocytopenia. This combination of results prompted an ADAMTS13 check by a SRP (luckily) and it turns out patient had cTTP.

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Notes:

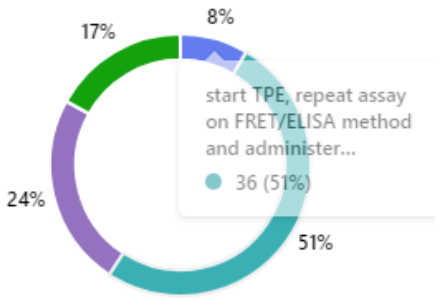
a. Genuine TTP shows a normochromic normocytic blood picture with RBC fragments and thrombocytopenia.
b. Hyper segmented are indicative of B12 deficiency, neurological disorders are common with this disorder, which can clinically resemble a TTP.
c. High number of burr cells (misleading to inexperienced blood reader) are not fragments, hence not a genuine TMA. Burr cells are associated with severe renal disorders, which are more common in HUS.
d. Raised APTT is associated with secondary Haemostasis defects. TTP is a primary hemostatic defect hence coagulation screen are usually normal.
e. Unexplained young strokes or cardiac events with thrombocytopenia is suspicious and TTP must be in consideration here.

Q6. Patient presents with TMA with suspicions of STEC UTI infection with bloody urine. ADAMTS13 in Acustar is <0.2IU/ml. How do you proceed with consideration particularly to use of caplacizumab therapy? (1 point)

Responses.

Mixed responses form participants. 51 % suggests to start plasma exchange but only administrate Caplacizumab after ADAMTS13 result confirmation.

Treat as TTP: commence TPE and administer caplacizumab urgently	6
Order repeat sample on acustar method before administering of caplacizumab	0
start TPE, repeat assay on FRET/ELISA method and administer Caplacizumab after confirmation result.	36
Treatment of UTI as priority, and retest ADAMTS13 after IV antibiotics	17
Do not treat as TTP. Repeat on another method before administering caplacizumab?	12



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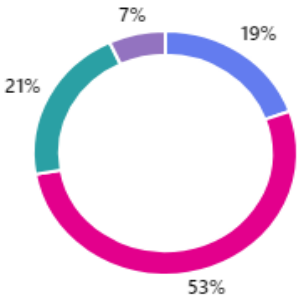
Q7. What is the expected ADAMTS13 activity results at the 1month time point after standard of care treatment of iTTP? (1 point)

Responses.

Mixed responses from participants. Consensus (53 %) think that results for ADAMTS13 activity at the 1-month post treatment time point should be >25%.

5-14%	14
>25%	38
15-24%	15
Levels not required at this time point	5



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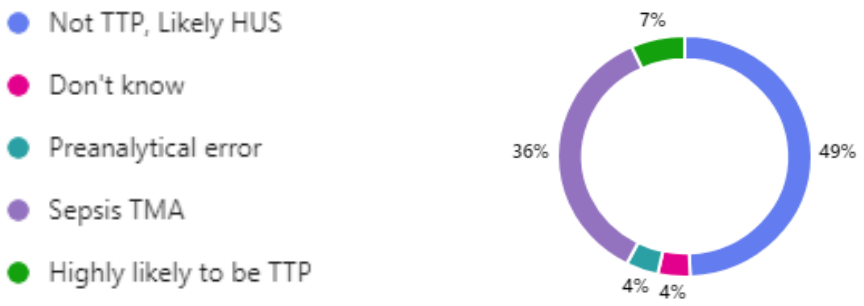
Q8. Child presents with pneumonia and TMA. FBC show platelets counts of 65 x 10^9/L, & few RBC fragments. Renal function, CRP, liver enzymes are elevated. Citrate sample analysis shows Prolonged APTT and PT mildly reduce fibrinogen and Acustar ADAMTS13 <0.2IU/ml. Results are suggestive off?

Responses.

Mixed responses form participants. 49% say likely HUS and 36% thinks it sepsis.



The correct answer is pre-analytical error.
Based on the clinical details it's not incorrect to be presumptive of HUS or Sepsis though the ADAMTS13 result is less <1% which is contrary to literature.
Patient had VITTS post adenovirus infection.
Initial citrate sample was contaminated with EDTA.

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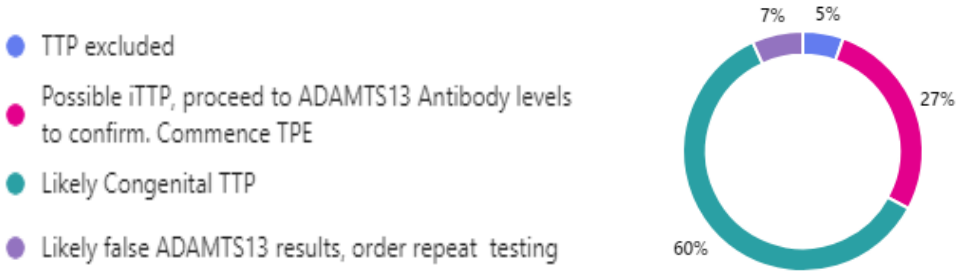
Q9. Patient with recent TIA, family history (father passed away with young stroke). Patient presents in A&E and now presents with a TMA, platelet counts is 50x 10^9/L. Acustar ADAMTS13 is 11.6%

Responses.

60% got this correct with likely to be congenital



The correct answer is likely to be congenital.

Young stroke with thrombocytopenia is suspicious of an underlying condition of which TTP is one of them

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AcuStar Presentation (1).ppt