

## ADAMTS-13 ACTIVITY

### Description:

ADAMTS-13 (a disintegrin-like and metalloproteinase with thrombospondin type 1 motif 13) is an enzyme (vWF-cleaving protease or vWF-CP) that specifically cleaves unusually large von Willebrand factor (vWF) multimers, which induce platelet thrombus formation under high shear stress. If the activity of ADAMTS13 is lowered for some reason, however, unusually large vWF multimers may accumulate within blood causing thrombosis due to platelet aggregation, which in turn may lead to TTP (thrombotic thrombocytopenic purpura)

The TECHNOZYM® ADAMTS-13 Activity ELISA is a chromogenic test for the determination of ADAMTS-13 activity in human plasma.

### Indication:

Investigation of microangiopathic haemolytic anaemias to diagnose thrombotic thrombocytopenic purpura (TTP). Monitoring of patients with known TTP.

### Reference Ranges (Adult normal ranges)

Assay	Range	Units
ADAMTS-13 Activity	40-130	%

### Concurrent Tests:

ADAMTS-13 assay usually follows examination of a blood film in a patient with normal coagulation profile and microangiopathy. If indicated or requested by a Consultant, an ADAMTS-13 Inhibitor assay can be performed.

### Interpretation:

Unexpected or unknown results <20 need to urgent referral to a Haematology clinician as TTP is a considered a clinical emergency.

### Collection Conditions:

A minimum of 3ml citrated plasma is required. Samples should be collected before transfusion of blood and/or blood components. Samples must be correctly filled as the ratio of anticoagulant to blood is crucial for accurate test results. Samples will be rejected by the laboratory if they are under or over filled. Samples containing EDTA cannot be used because EDTA is a strong inhibitor of ADAMTS13 function.

Samples from other laboratories should arrive separated and frozen unless delivery to laboratory within 4 hours.

### Frequency of Testing:

The Royal Liverpool and Broadgreen University Hospital Trust (RLBUHT) is a regional specialist referral centre for TTP with samples arriving into the laboratory from all over the North West area.

ADAMTS-13 Activity ELISA is run weekly to coincide with outpatient clinics (Thursday am) but urgent requests can be arranged. Urgent results available next working day by request. Please contact Specialist Coagulation Laboratory for more information.

**Clinical Advice Contact:** Haematology registrar/ TTP Nurse Specialist