Who we are
Majority owned by the NHS, but with the commercial freedom to invest in innovation, Viapath are on a mission to transform pathology services in the UK. We provide pathology services to the NHS, private hospitals and other organisations both across the country and internationally.

What we do
All our laboratories are either accredited or working towards accreditation by UKAS to ISO15189. To view our laboratory accreditation status please follow this link:
http://www.viapath.co.uk/about-viapath/quality-and-governance/accreditations

TEST OVERVIEW

Description
The ADAMTS-13 activity assay is a chromogenic ELISA employing a recombinant fragment of a region of the A2 domain of VWF, which contains the cleavage site of ADAMTS-13, the VWF73 region. The process of manufacturing the recombinant VWF73 region involves tagging the N-terminal with glutathione S-transferase (GST), a property that is used in the design of the assay. The ELISA plate is coated with anti-GST antibody and the GST-VWF73 added to act as a substrate. The resultant antibody:antigen complex is then incubated with test samples whereby the ADAMTS-13 cleaves the immobilised recombinant VWF73, exposing the cleavage site. The plate is washed to remove unbound protein and then incubated with a horse radish peroxidise (HRP) conjugated monoclonal antibody to N10, the C-terminal edge residue of the VWF-A2 domain generated after ADAMTS-13 cleavage. The HRP is provided with a substrate, the product of which is coloured, and the degree of colour formation is directly proportional to the ADAMTS-13 activity that cleaved VWF73 and exposed N10.

Clinical details
Vascular endothelial cells and platelets contain ultra-large VWF multimers that are highly adhesive. However, they are only transiently detectable in plasma as they are cleaved by the circulating protease ADAMTS-13 (A zinc and calcium dependent Disintegrin and Metalloprotease with ThromboSpondin type 1 motifs, member 13), also known as VWF-cleaving protease. ADAMTS-13 degrades the ultra-large multimers into smaller forms ranging in size from 500 to ~20 000kD. Deficiency of ADAMTS13 leads to a condition called thrombotic thrombocytopenic purpura (TTP), which can be either congenital or acquired. In congenital TTP, also known as Upshaw-Schulman syndrome, mutations cause deficiency of ADAMTS13 which generally affects synthesis or secretion of the enzyme rather than causing production of dysfunctional molecules. In acquired TTP, an IgG inhibitory antibody prevents the normal action of ADAMTS13. Each mechanism results in an excess of ultra-large VWF multimers being produced from the Weibel-Palade bodies which become anchored to the endothelium, where they can bind platelets via GpIbα on the platelet membrane. This can cause aggregation of platelets which results in microvascular thrombosis and haemolytic anaemia. Coagulation screening tests are usually normal or only slightly disturbed, which helps to distinguish TTP from disseminated intravascular coagulation, though thrombocytopenia and a microangiopathic haemolytic anaemia will be evident from a full blood count and blood film. Haemoglobin is usually

Related condition or disease
Thrombotic thrombocytopenic purpura (TTP)

Reference range
66.4 – 107.9

Synonyms or keywords
Thrombotic thrombocytopenic purpura (TTP), ADAMTS13 activity, adams13, adam

Units
%

Department
Haemostasis and Thrombosis Department

Laboratory
Diagnostic Haemostasis and Thrombosis Laboratory at St Thomas’

Location
Viapath at St Thomas’ Hospital
below 10.5 g/dL and MCV normal unless there is marked red cell fragmentation, which will be evident from the blood film. The red cell fragments arise from being sheared as they travel past and through the micro-thrombi, which is the cause of the intravascular haemolysis. Reticulocytes are increased, which can increase the MCV, and nucleated red cells can be seen in the peripheral blood, both indicators of a bone marrow response to the haemolysis. Serum lactate dehydrogenase is also elevated. Neurological signs, such as coma, stroke, seizures and even personality change, are a presenting feature due to formation of thrombi in the cerebral circulation.

ORDERING INFORMATION

Sample type and Volume required
External requests: Citrated platelet poor plasma 500µl x 1 aliquot
Internal requests: please refer to EPR label

Turnaround time
5 - 7 working days.

Contacts
Diagnostic Haemostasis and Thrombosis Department
020 7188 2797
St Thomas’ Hospital
North Wing - 4th and 5th Floors
Westminster Bridge Road
London SE1 7EH

Laboratory opening times
24/7

How can we help?

We have a number of partnering options to suit your needs, whether you require this specific test or a range of services, we are here to help. Contact one of our friendly Business Development Managers for more information, or visit our website.