## ADAMTS13 Activity and Inhibitor

Collected 12/16/07 06:59:00

| Results | | Units | Reference Interval |
|---|---|---|---|
| ADAMTS13 Inhibitor | Not Indicated | | <=0.4 |
| ADAMTS13 Activity | 67 | % | >=67 |

12/16/2007 6:59:00 ADAMTS13 Activity:
METHOD: ADAMTS13 activity is directly measured by a Fluorescence Resonance Energy Transfer (FRET) assay whereby a fluorescent signal is detected when a synthetic substrate (FRETS-VWF73) is cleaved by ADAMTS13 (Kokame K, et al. Br J Haematol. 2005;129:93-100). Inhibitor activity is determined by measuring the ability of heat-treated patient plasma to inhibit ADAMTS13 present in normal pooled plasma, with inhibitor units calculated by Bethesda-type method. One inhibitor unit is defined as the concentration of inhibitor able to reduce the ADAMTS13 activity of an equal volume of normal pooled plasma by half (Zheng XL, et al. Blood. 2004;103:4043-4049).
INTERPRETIVE COMMENTS: Severe deficiencies of ADAMTS13 (activity <5%) appear to be a relatively specific finding in thrombotic microangiopathy patients with a clinical diagnosis of idiopathic TTP or the congenital form of TTP (Upshaw-Schulman Syndrome). The sensitivity of a severe deficiency for the diagnosis of idiopathic TTP is controversial, with reports ranging from 30 to 100%. Severe deficiencies have also been observed with such conditions as sepsis, disseminated intravascular coagulation and metastatic malignancy. Concomitant detection of low ADAMTS13 activity and an inhibitor has increased specificity in idiopathic TTP. In one prospective study, inhibitors were observed in 44% of patients with TTP. The consistent finding of a proportion of patients with idiopathic TTP/HUS without severe ADAMTS13 deficiency suggests that alternative mechanisms that are not yet known can also lead to TTP/HUS. Examples include marrow transplant-associated thrombotic microangiopathy and childhood hemolytic uremic syndrome. Mild to moderate deficiency of ADAMTS13 activity has been observed in multiple medical conditions. While ADAMTS13 assay results may assist in diagnosis and have prognostic value regarding likelihood of relapse of TTP, the utility of serial measurement for ongoing management of patients with acute thrombotic microangiopathies remains to be defined. Recent plasma exchange therapy may raise the observed ADAMTS13 activity and reduce the observed inhibitor levels. Hemolysis with plasma free hemoglobin >2 gm/L causes inhibition of ADAMTS13, which can cause artifactually low ADAMTS13 activity and false positive inhibitor results (Lammle B, et al. Thrombotic Thrombocytopenic purpura. J Thromb Haemost. 2005;3:1663-1675).
Effective 4/20/06