Background: Thrombotic microangiopathy (TMA) with non-deficient ADAMTS-13 (a disintegrin-like and metalloprotease with thrombospondin type 1 motif 13) outcome is unknown hence the survival analysis correlating with ADAMTS-13 activity is conducted in Malaysia.

Methods: This was a retrospective epidemiological study involving all cases of TMA from year 2012-2016.

Results: The study evaluated 243 patients with median age of 34.2 years and 57.6% of female. Majority of the patients were malay (62.5%), followed by chinese (23.5%), indian (8.6%) and others (5.4%). There was 20.9% of thrombotic thrombocytopenic purpura (TTP) in which 72.2% were acquired while 27.8% were congenital. Patients with ADAMTS-13 activity ≥5 % had the odds of death of 4 times higher than those with ADAMTS-13 activity <5% (OR: 4.133, p=0.0425). There was 22.6% of mortality (n=55). The most common etiology was secondary causes (42.5%), followed by acquired TTP (16.6%), atypical hemolytic uremic syndrome (HUS) or HUS (12.8%) and congenital TTP (6.4%). The TMA with secondary causes had an inferior overall survival (OS) (p=0.0387), which consisted of systemic lupus erythematosus (30%), infection (29%), pregnancy (10%), transplant (8%), malignancy (6%) and drug (3%). Transplant related TMA had the worst OS (p=0.0016) among the secondary causes. Plasma exchange, methylprednisolone and intravenous immunoglobulin were recorded in 162 patients as first line treatment while rituximab, bortezomib, vincristine, azathioprine, cyclophosphamide, cyclosporin and tacrolimus were described in 78 patients as second line treatment.

Conclusion: This study illustrates that non-deficient ADAMTS-13 TMA has a tendency of inferior outcome compared to severely deficient ADAMTS-13 TMA albeit the statistical insignificance.