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Von Willebrand Factor as a Prognostic Index in Sickle Cell Disease- Kapoona I Iwara Eteng- UCH

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Abstract

Sickle cell disease is a genetic disorder of haemoglobin with a world-wide distribution. Nigeria the most populous African Country, has the highest cohort of sickle cell anaemia patients. Von Willebrand factor (vWF) is an acute phase reactant stored exclusively in the Weibel-Palade bodies of the endothelial cells and α-granules in platelets. vWF is generally considered as a marker of injury to the endothelium and has been implicated in the process of adhesion of sickled erythrocytes to the vascular endothelium.

This study was aimed at ascertaining if vWF: Ag level is increased during bone pain crises in sickle cell anaemia patients, and if it can be used as a predictor of disease severity, when compared with other known prognostic factors such as the steady state haematocrit and white cell count.

The study was carried out at the University College Hospital, Ibadan. The study population comprised of 80 HbS patients in steady state, 15 HbS patients in bone crises and 30 HbA individuals. The mean age of the patients was 26 years and that for controls was 28 years.. There were more females (59%) with SS disease compared with males (41%). This did not affect the interpretation of the results as there is no variation in vWF:Ag with gender. vWF:Ag was assayed using a sandwich ELISA method.

There was no significant increase in the vWF:Ag in the SCA patients in steady state (93.74±16.74%) compared with the values during bone pain crises (102.4±13.98%; p=0.59). However, there was a statistically significant increase in vWF:Ag in the SCA patients both in steady state and bone pains crises compared with the HbA control group (77.50±15.43%; p=<0.01). The levels of vWF:Ag correlated with the ISC index (p<01) but not with the level of leukocytosis or steady state haematocrit. This could mean that the ISC index which is a simple morphological evaluation could be used as a measure of the chronic inflammation or disease severity in SCD patients in conjunction with vWF:Ag.

Endothelial actuation assumes a focal part in the pathophysiology of vaso-impediment in sickle cell sickness (SCD), encouraging glue cooperations with coursing platelets. Upon initiation, different cement atoms are communicated, including von Willebrand factor (VWF). Expanded VWF levels have been seen in patients with SCD during consistent state. Be that as it may, the job of VWF in the pathogenesis of SCD vaso-impediment is hazy. Destinations to longitudinally evaluate the amount and reactivity of VWF and its managing protease ADAMTS-13 during vaso-occlusive emergency (VOC). Strategies In this observational investigation, we got consecutive blood tests in grown-up SCD patients during VOC. Results VWF reactivity was altogether higher during VOC (dynamic VWF, VWF glycoprotein Ib-restricting movement, and high sub-atomic weight multimers), though platelet tally and levels of ADAMTS-13 antigen and ADAMTS-13 action were correspondingly lower than during consistent state. Levels of VWF antigen, VWF propeptide (VWF:pp) and ADAMTS-13 explicit action didn't change during VOC.

VWF reactivity connected firmly with markers of irritation and neutrophil enactment, and was conversely associated with the platelet check. In patients who created intense chest condition, levels of VWF, VWF:pp and dynamic, hyperadhesive VWF were altogether higher, though ADAMTS-13 movement was lower, than in patients without this intricacy. Ends we give the main proof that VOC in SCD is related with expanded reactivity of VWF, without an articulated ADAMTS-13 insufficiency. This hyper-reactivity might be clarified by obstruction of VWF to proteolysis, auxiliary to cycles, for example, irritation and oxidative pressure. Hyperadhesive VWF, searching platelets in the microcirculation, may along these lines intensify and support VOC in SCD.