HEMOGLOBIN SC DISEASE IN CHRONIC KIDNEY DISEASE STAGE III, IV AND RARE ESRD REQUIRE CLOSE MONITORING OF HEMOGLOBIN AND FREQUENT TITRATION OF ERYTHROPOIETIN TO AVOID CRISIS AND ANGINA.

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Background: Hemoglobin(Hb) SC disease is the second most common of Sickle Cell Syndromes with inheritance of sickle cell gene in compound heterozygosity with other mutant β-globin genes causing more sickling. Complications are crises: vasoocclusive (painful), aplastic, sequestration, hemolysis, autosplenectomy and chronic kidney disease (CKD).

Purpose: Prevalence of sickle cell disease plus trait in ESRD is 0.1% and Hb SC disease is 0.06%. Multifactorial etiology of anemia in Hb SC plus CKD makes maintaining Hb goal challenging to avoid crisis and angina.

Case: A 78 yo AAF with history of Hb SC Disease, Hypertension, Diabetes Mellitus, Coronary Artery Disease had CKD stage III. Hb SC was confirmed by Hb electrophoresis: Hb A1= 0%, Hb A2<0.1%, Hb S= 54% and Hb C= 46% on two separate occasions. Peripheral Blood smear: target cells, sickle cells, Howell jolly bodies, marked poikilocytosis and anisocytosis, nucleated RBCs and mild polychromasia. Hemoccult: negative. Patient had bone crisis if Hb > 10 and angina with Hb < 8. Anemia was initially treated with up to 11 units of packed RBC/year. As the kidney disease progressed, Erythropoietin (EPO) replacement was started. Over 4 years, CKD stage 4 & 5 progressed to ESRD requiring hemodialysis (HD). ESRD worsened the anemia with bloodloss during HD, secondary hyperparathyroidism and functional iron deficiency. Hb, iron and hemoccult were monitored frequently with EPO dosage adjustments. EPO dose: Stage IV & V CKD - 10,000 U/wk; ESRD 9,000 to 21,000 U/wk.

Conclusion: Close monitoring and frequent dosing of Erythropoietin Stimulating Agents (ESA) is required in Hb SC Disease with ESRD, to avoid crisis and angina. Maintenance of Hemoglobin at a lower goal i.e. 9 to 10 mg/dL is desirable in this delicate population