PD018

Paediatric Cardiology & Congenital Heart Disease • Poster Presentation

LEFT VENTRICULAR DIASTOLIC DYSFUNCTION IN YOUTH WITH SICKLE CELL DISEASE: EVALUATING GENOTYPIC DIFFERENCE AND EFFECTS OF HYDROXYUREA

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Stand: POSTER #PD018 | **Venue:** Poster Exhibition Hall & Gallery - Eko Convention Centre |**Online Ref:** https://nigeriancardiacsociety.com/abstracts/PD018

Background: Sickle cell disease (SCD) encompasses a spectrum of genotypes, most commonly Hemoglobin (Hgb)SS and HgbSC. Diastolic dysfunction (DD) is common in SCD and impacts morbidity and mortality. We evaluated if rates of DD differed by SCD genotype and by hydroxyurea (HU) use among youths. Methods: This was a retrospective study of patients with HgbSS and HgbSC who received an echocardiogram, stratifying genotype and HU use. DD was assessed using mitral valve (MV) E/A ratio <0.8 or >2, septal E' < 7 cm/s, lateral E' <10 cm/s, E/e' Average > 10; and LA:Aortic root (LA:Ao) ratio >1.3:1. DD was defined as a diastolic aggregate score ?2 of 5. Multivariable regression models adjusted for age were used to estimate the associations of DD with HU use by genotype. Results: 39% of youth with SCD who had an echocardiogram had DD. DD rates were lower in those on HU (n=102, 36%) vs those not on HU (n=94, 41%). Age adjusted, youths with HgbSS on HU had lower odds of DD compared to those not on HU, OR 0.22 [95% CI (0.07, 0.59), p=0.004]. Conversely, youth with HgbSC on HU had slightly increased odds of DD compared to those not on HU, OR 1.36 [95% CI (0.35, 4.98) p=0.6]. Conclusions: Our study showed DD was less common among patients with HgbSS on HU. Larger, multi-center, prospective, studies are needed to assess longitudinal improvement of DD from baseline for SCD patients on HU.

Keywords: Sickle cell disease, hydroxyurea, left ventricular diastolic dysfunction, left ventricular systolic dysfunction, hemoglobin SS (HgbSS), hemoglobin SC (HgbSC).