Hearing loss and Health Related Quality of Life (HRQoL) in sickle cell disease and traits

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1. Introduction

Sickle cell disease(SCD) and Sickle cell traits(SCT) are genetically inherited red blood cell disorders. The prevalence is relatively high among sub-Saharan Africans and African Americans [1,2] SCD is associated with chronic haemolytic anaemia, increased risk of infections, vasoocclusive crisis and end-organ damage if poorly managed.[2] Hearing loss and overall-poor health status have been associated with SCD but no consensus on SCT.[3] Hence, the need for a comprehensive study of the hearing loss in SCD and SCT

2. Objectives

The study objectives were to assess and characterize the hearing loss and HRQoL in SCD and SCT and compare with matched controls.

3. Methodology

This is a comparative cross-sectional study of children and adults aged 6-months to 55years to assess the association between SCD/SCT and hearing impairment. Participants were divided into two groups Cohort (test group) and Control groups. The cohort group consisted of individuals diagnosed with SCD/SCT and the control group those with normal haemoglobin gene(HbAA). It was conducted at UATH Nigeria (02/02/20222-30/06/2023). Audiometric tests and WHOADS2 [4] questionnaire were used to assess for hearing loss and the HRQol respectively. Statistical analyses were done with *Statistical Analysis System (SAS 9.4)*. The study was approved for ethics by UCT CapeTown(HREC REF 228/2022) and UATH Nigeria (UATH/HREC/PR/2020/08/007).

4. Results

A total of 212 participants were enrolled , 106 (cohort) had Sickle /trait and another 106 (control) normal genotype(AA). No significant difference between the mean age, sex and educational background of cohort and control groups (p = 0.77, 0.074 and 0.904 respectively).Overall Mean 23±2. Mean ages for SCD/SCT and HbAA are 15.6 and 15.1 years respectively. The gender distribution is shown in Figure 1.

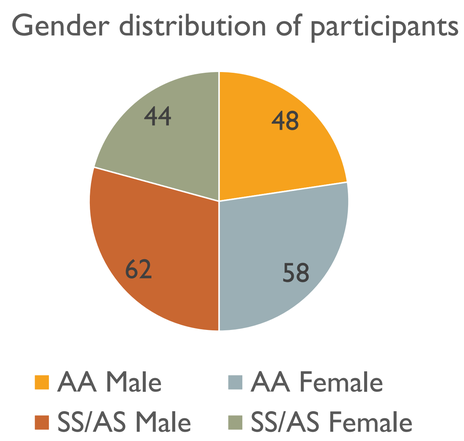

Figure 1: Features the gender distribution in the order of genotypes of the participants

The prevalence of auditory impairment assessed in the left (29.3 vs 11.3%, p=0.002), the right (31.1 vs 18.9%, p=0.056) and the two ears considered together 44.3% vs 26.4%, p=0.009) were significantly higher in case compared with control subjects SNHL, mainly moderate to severe) was significant in SCD but not SCT. The educational level of parents(P=0.016) and the duration of diagnosis of SCD genotype for (P< 0.001) were strong demographic factors that influenced the health status (See Table 1).

Results -contrds

Table 1. Auditory condition in AA patients compared with AS/SS

	AA			AS/SS			P value
Hearing impairment	N	n (%)	95%CI	N	n (%)	95%CI	
All	106	28 (26.4)	18.3-35.9	106	47 (44.3)	34.7-54.3	0.009
Left	106	12 (11.3)	6.0-18.9	106	31 (29.3)	20.8-38.9	0.002
Right	106	20 (18.9)	11.9-27.6	106	33 (31.1)	22.5-40.9	0.056
Symmetry of impairment							
Asymmetrical	28	24 (85.7)	67.3-96.0	47	30 (63.8)	48.5-77.3	0.062
Symmetrical		4 (14.3)	4.0-32.7		17 (36.2)	22.7-51.5	
Type of Hearing impairment							
Left							
Sensori-neural	6	1 (16.7)	0.4-64.1	22	2 (9.1)	1.1-29.2	0.643
Mixed		5 (83.3)	35.9-99.6		19 (86.4)	65.1-97.1	
Normal		0 (0.0)	-		1 (4.5)	0.1-22.8	
Right							
Sensori-neural	12	0 (0.0)	-	20	1 (5.0)	0.1-24.9	0.044
Mixed		9 (75.0)	42.8-94.5		19 (95.0)	75.1-99.9	
Normal		3 (25.0)	5.5-57.2		0 (0.0)	-	
Sensori-neural left/right		1 (6.3)	0.2-30.2		2 (6.7)	0.8-22.1	>0.999
Mixed left/right		12 (75.0)	47.6-92.7)		28 (93.3)	77.9-99.2	0.163
Degree of hearing impairment							
Left	106	1 (1-2)	0.70-2.20	106	1 (1-1)	1.00-1.67	0.779
Right	106	1 (1-2)	0.95-2.12	106	1 (1-3)	1.30-2.29	0.437

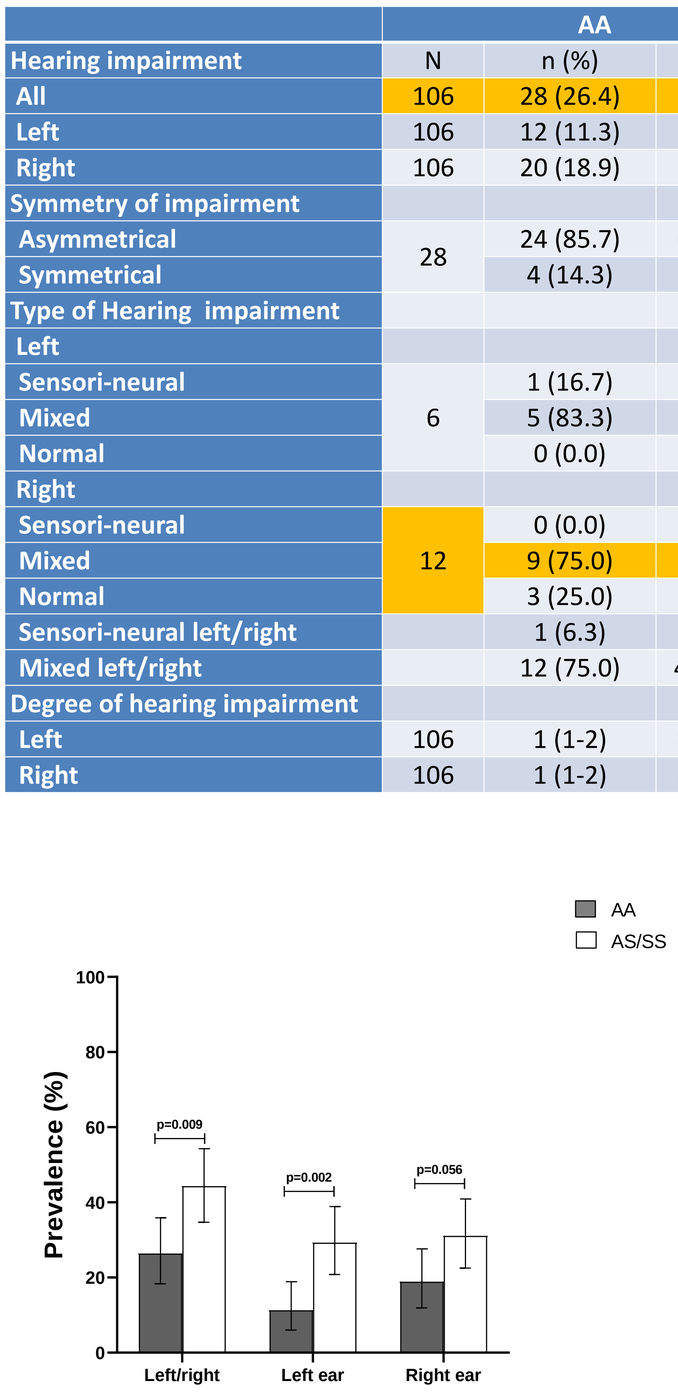

Figure 2: Auditory impairment in AA compared with AS/SS subjects The height of each horizontal bar , lower and upper cap represent the proportion and 95% lower and upper limits respectively.

The pattern of impairment in right ear only was different (p=0.044) between the two study groups with perhaps higher prevalence of mixed hearing impairment (95.0 vs 75.0%) and lower prevalence of normal impairment (0.0 vs 25.0%) in cases compared with control subjects. The Prevalence of hearing loss in cases (sickle cell disease and trait) is 47/106(44.3%). Of the six domains assessed, subjects with AS/SS genotypes had higher median (interquartile range) score in four domains including cognitive [0 (0-1) vs 0 (0-0), p=0.002], mobility [2 (0-3) vs 0 (0-2), p=0.000], work [1(0-3) vs 0 (0-2), p=0.003], socialisation [1 (0-2) vs 0 (0-0), p=0.000] and overall [4.5(2-10) vs 1 (0-14), p=0.003] in comparison to those with AA genotype. Table 3 shows summary of severity of HRQoL across domains.

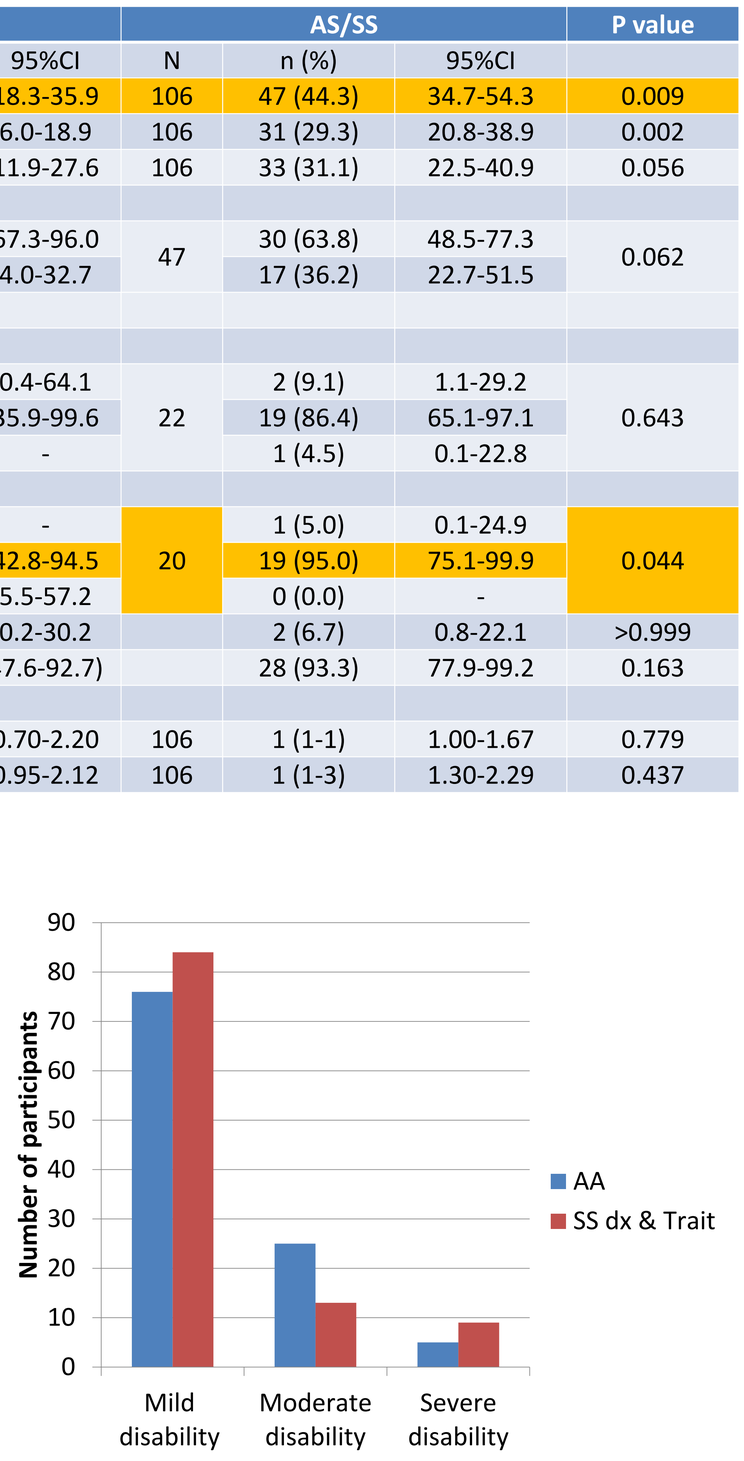

Figure 3: Health Related Quality of Life (HRQoL)

Recommendations: We recommend for longitudinal studies in this field and early commencement of regular hearing assessment in both Sickle cell disease and trait patients in children and adults, to aid early diagnosis and intervention.

Study limitations : Longitudinal study would provide a more comprehensive study period and follow-up for the hearing Impairment in the HbSS and HbAS. However we were limited with time line since this is a PhD thesis with tenured period. A follow-up longitudinal study could be done afterwards.

5.Conclusions

Hearing loss is significantly associated with Sickle cell disease, whereas the Health Related qualities of lives are depressed in both SCDs and SCTs with higher impacts on the SCDS. Awareness and early diagnosis appears to be strong positive predictive factors.

6. References

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