

Original Research Article

KNOWLEDGE OF HAEMOGLOBIN TYPES AT UNION AMONG COUPLES AND IMPACT ON OFFSPRING; A CROSS-SECTIONAL STUDY IN SOUTHERN NIGERIA

ABSTRACT

Aims: This study aimed at assessing haemoglobin (Hb) types of couples, their knowledge of Hb types at the time of union and outcome on offspring.

Study design: Cross-sectional descriptive study.

Place and Duration of Study: This study was carried out in Abi local government area of Cross River State between March and July, 2018.

Methodology: Two hundred consenting couples and their 445 offspring were enrolled in this study. A pre-tested structured questionnaire was administered by two trained interviewers to capture the bio-data and other pertinent information in relation to couple's knowledge of Hb types. Blood sample was collected from each participant and Hb type was determined by electrophoresis in alkaline pH using cellulose acetate method.

Results: Thirty-eight percentage (38%) of the females and 30% of the males who participated in this study lacked knowledge of their haemoglobin type at the time of producing offspring. Haemoglobin type AA predominated (70% of females and 72% of males), while 30% females and 28% males accounted for the presence of AS. The Hb types of their offspring were 79% AA, 15% AS and 6% SS. Among AS versus AS couples, 67% had no knowledge of their Hb type prior to having children and contributed to 85% of children with Hb SS.

Conclusion: Ignorance of haemoglobin types prior to having children contributed to the high prevalence of Hb SS observed in the present study.

Keywords: Haemoglobin types, Sickle cell disease, Offspring.

1. INTRODUCTION

Sickle cell disease has been observed as a significant contributor to global child mortality. This is even more pronounced in Africa where healthcare is generally inadequate [1, 2]. Over the years, efforts to reduce the mortality of sickle cell disease and consequently the associated health burden have been largely successful in developed countries compared to the developing regions. While the mortality rate of sickle cell disease among children and teenagers has been observed to be reducing in advanced societies, findings from Africa and South America reveal higher rates [1, 3-5]. It appears that the persistence of sickle cell disease and its impact on health particularly that of children and teenagers has been underappreciated. This may have conferred some resilience to this global scourge.

Ideally, newborn and prenatal screening for sickle cell disease undoubtedly aid in early intervention for affected persons. An important gain of this approach is the prospect of early intervention and better outcomes [6-11]. It is rather unfortunate that in the very regions ravaged by sickle cell disease, adequate

screening is very much lacking [12-15]. A situation that is not unlinked to the deplorable state of primary healthcare found in resource-poor settings.

A common approach to combating sickle cell disease in Nigeria is that of promoting premarital counselling and testing, together with campaigns to raise awareness among the populace. The idea is to encourage intending couples to avoid marital unions with the potential of producing affected offspring [16,17]. This strategy has proved unreliable majorly due to poor knowledge and perception of the importance of one's haemoglobin type as well as lack of adequate healthcare provision in most rural areas in Nigeria. Thus, intending couples may not even be aware of their haemoglobin types prior to having children [12,14]. It is not also uncommon to find cohabiting unions often fostered by unplanned pregnancy. In this manner, there is increased risk of sustaining sickle cell disease within such communities.

The commonest sickle cell disease in Nigeria remains sickle cell anaemia (SCA) and the initial efforts towards its control brought about massive campaigns on the awareness of sickle cell anaemia among members of the public [18-20]. Popularly targeted during these campaigns are schools and churches with recommendations for the participants to visit the hospital for testing, while sometimes, the services are rendered on site. Inclusion of haemoglobin type testing as part of general medical examination required from students at all levels by admitting schools, from intending couples by their religious institutions and from prospective employees by the various employer bodies has helped to sustain some level of awareness. In spite of all this progress, SCA persists in the Nigerian population and more often than not ignorance is implicated among the parents of affected subjects. This situation puts to test the population reach of our campaigns and the availability of supportive health facilities to our people. Cross River State in Southern Nigeria has been a tourism destination with reasonable migration activities in both urban and rural communities. This part of the country therefore represents an important setting for investigating genetic distribution patterns.

2. MATERIALS AND METHODS

This study was carried out in Abi local government area of Cross River State between March and July, 2018. It was a cross-sectional descriptive study that enrolled 200 consenting couples and their 445 offspring. All the participants were indigenes of the study area. Ethical approval was granted by the

departmental ethical board, while informed consent was granted by each participating family. A pre-tested structured questionnaire was administered by two trained interviewers to capture the bio-data of enrolled subjects and other pertinent information in relation to couple's knowledge of Hb types. Blood sample was collected from each participant and Hb type was determined by electrophoresis in alkaline pH using cellulose acetate method. Data analysis was carried out using SPSS 20.0 to extract frequency distributions.

3. RESULTS AND DISCUSSION

3.1 Results

This study observed that among the 200 couples who participated, 124 (62%) females and 140 (70%) males had knowledge of their Hb types before having children, while 76 (38%) females and 60 (30%) males lacked knowledge at the time of producing offspring. Haemoglobin type AA predominated (140 (70%) of females and 144 (72%) of males), while 60 (30%) females and 56 (28%) males accounted for the presence of AS. Among couples, AA and AA was 108 (54%), AA and AS was 68 (34%), while AS and AS stood at 24 (12%). The Hb types of their offspring were 352 (79%) AA, 67 (15%) AS and 26 (6%) SS (Table 1). Among AS versus AS couples, 16 (67%) had no knowledge of their Hb type at the time of producing offspring and contributed to 23 (85%) of children with Hb SS (Table 2).

Table1. Couple's knowledge of haemoglobin type at time of producing offspring and haemoglobin types of subjects

Variables	Distribution of participants n (%)
Couples who had Knowledge of Hb Type at Time of producing offspring	
Females	124 (62%)
Males	140 (70%)
Couples who lacked Knowledge of Hb Type at Time of producing offspring	
Females	76 (38%)
Males	60 (30%)
Hb Type of Couples	
Females with Hb AA	140 (70%)
Females with Hb AS	60 (30%)
Males with Hb AA	144 (72%)
Males with Hb AS	56 (28%)

Combination of Hb Types

Among couples

Hb AA and AA	108 (54%)
Hb AA and AS	68 (34%)
Hb AS and AS	24 (12%)

Hb Types among Offspring

Hb AA	352 (79%)
Hb AS	67 (15%)
Hb SS	26 (6%)

Table 2. Influence of couple's knowledge of haemoglobin type on choice of spouse and outcome of offspring among Double heterozygous unions

Variables	Distribution of participants n (%)
Double heterozygous unions (AS vs AS)	
Couples who had Knowledge	8 (33%)
Couples who had no Knowledge	16 (67%)
Hb SS Offspring	
Couples who had Knowledge	4 (15%)
Couples who had no Knowledge	22 (85%)

3.2 Discussion

Continuous appearance of the sickling haemoglobin among children implies sustenance of the disorder through generations and Nigeria is among the nations in need of interventions [21]. A common practice aimed at circumventing future occurrence of sickle cell disease is that of premarital counselling and testing for haemoglobin types of intending couples. This is typically the case in urban areas and is often encouraged by most religious institutions where marriage is celebrated. In urban areas also, schools and organized employers of labour demand for medical examination including testing for haemoglobin type as a prerequisite for admission and employment among other requirements. On the whole, these various avenues serve to keep the urban populace fairly knowledgeable about the subject of sickle cell anaemia. This is however, not the case in the rural areas. Those living in the rural areas may not have been sufficiently sensitized as observed in this study where 38% of females and 30% of males who participated lacked knowledge of their Hb types at union. Nigeria is known to have a haemoglobin type distribution in

which Hb AA predominates [18-20]. It was therefore not surprising that the current study recorded for Hb AA 70% of females and 72% of males, while 30% females and 28% males accounted for the presence of AS. What is rather worrisome in a situation like this is that in a population with sickle cell anaemia trait as high as 30% and 28% across females and males, some members of the same population lacked knowledge of their Hb types prior to having children as earlier mentioned. Far from being a mere finding, this has revealed an important evidenced-based explanation to the persistence of SCA in Nigeria, particularly from the rural areas.

The unions with double heterozygosity turned out to be 12%, while 6% Hb SS was recorded among all the children from the participating couples. Furthermore, 67% of those double heterozygosity couples had no knowledge of their Hb type at union and contributed to 85% of children with Hb SS. Ignorance is obviously a challenge to possible eradication of SCA within the studied population . When ignorance is not completely driven out in the presence of a threat, avoidable casualties are bound to occur. The unfortunate thing about our current campaign achievement is that the neglected part of the population has become a fueling place for the persistence of the disorder. This has to be urgently addressed in order to effectively control an avoidable situation that continues to harass our population. The emergence of other threatening diseases such as human immunodeficiency virus infection/ acquired immune deficiency syndrome could have taken some attention off the awareness of sickle cell disease in our society [12,22]. Unfortunately, the pressure of natural selection allegedly operating in malaria endemic regions continue to ensure the retention of the Hb S gene [23,24]. Therefore, except where precautions have been intentionally taken, producing children with sickle cell disease would remain a viable probability. Strategies to combat sickle cell anaemia in Nigeria need to be reinvented and extended to reach the rural communities where urgent attention is needed.

4. CONCLUSION

Ignorance of haemoglobin types prior to having children contributed to the high prevalence of Hb SS observed in the present study. Rural areas with inadequate enlightenment on haemoglobin types and health consequences contribute to the persistence of Hb SS in Nigeria.

REFERENCES

1. Grosse SD, Odame I, Atrash HK, Amendah DD, Piel FB, Williams TN. Sickle cell disease in Africa: a neglected cause of early childhood mortality. *Am J Prev Med.* 2011;41(6 Suppl. 4): S398–S405.
2. McGann PT. Sickle cell anemia: an underappreciated and unaddressed contributor to global childhood mortality. *J Pediatr.* 2014;165(1):18–22.
3. Hamideh D, Alvarez O. Sickle cell disease related mortality in the United States (1999–2009) *Pediatr Blood Cancer.* 2013;60(9):1482–6.
4. Lobo CL, do Nascimento EM, de Jesus LJ, de Freitas TG, Lugon JR., Ballas SK. Mortality in children, adolescents and adults with sickle cell anemia in Rio de Janeiro, Brazil. *Hematol., Transfus. Cell Ther.* 2018, 40(1): 37-42.
5. Wastnedge E, Waters D, Patel S, Morrison K, Goh MY, Adeloye D. et al. The global burden of sickle cell disease in children under five years of age: a systematic review and meta-analysis. *Journal of Global Health.* 2018; 8: 021103.
6. Rahimy MC, Gangbo A, Ahouignan G, Adjou R, Deguenon C, Goussanou S. et al. Effect of a comprehensive clinical care program on disease course in severely ill children with sickle cell anaemia in sub-Saharan Africa setting. *Blood.* 2003; 102 (3): 834-8.
7. Ohene-Frempong K, Oduro J, Tetteh H, Nkrumah F. Screening newborns for sickle cell disease in Ghana. *Pediatrics.* 2008; 121 (Issue Supplement 2).
8. Tshilolo L, Kafando E, Sawadogo M, Cotton F, Vertongen F, Ferster A. et al. Neonatal screening and clinical care programmes for sickle cell disorders in sub-Saharan Africa: Lessons from pilot studies. *Public Health.* 2008; 122 (9): 933-41.
9. Sadarangani M, Makani J, Agbo T, Newton CR, Marsh K, Williams TN. An observational study of children with sickle cell disease in Kalifi Kenya. *British Journal of Haematology.* 2009; 146 (6): 675-82.
10. Rwezaula S, Cox S, Lowe B, Magesa, P, Massawe A, Newton C et al. Neonatal screening for haemoglobinopathies at Muhimbili National Hospital, Dar es Salaam. A pilot study. *British Journal of Haematology.* 2011; 153: 27.
11. Williams H, Silva S, Simmons LA, Tanabe P. A telephonic mindfulness-based intervention for persons with sickle cell disease: study protocol for a randomized controlled trial. *Trials.* 2017; 18(1): 218.
12. Abioye-Kuteyi EA, Oyegbade O, Bello I, Osakwe C. Sickle cell knowledge, premarital screening and marital decisions among local government workers in Ile-Ife, Nigeria. *Afr J Prim Health Care Fam Med.* 2009;1 (1): 022.
13. Makani J, Ofori-Acquah SF, Nnodu O, Wonkam A, Ohene-Frempong K. Sickle Cell Disease: New Opportunities and Challenges in Africa. *The Scientific World Journal,* Volume 2013, Article ID 193252.
14. Akodu SO, Disu EA, Njokanma OF. Pattern and factors associated with hemoglobin genotype testing among children attending a University Teaching Hospital in Lagos, Nigeria. *The Nigerian Journal of General Practice.* 2015; 13(1): 16-20.

15. Mulumba LL, Wilson L. sickle cell disease among children in Africa: An integrative literature review and global recommendations. *International Journal of Africa Nursing Sciences*. 2015; 3: 56-64.
16. Odunvbun ME, Okolo AA, Rahimy CM. Newborn screening for sickle cell disease in a Nigerian hospital. *Public Health*. 2008; 122 (10): 1111-6.
17. Aneke JC, Okocha CE. Sickle cell disease genetic counselling and testing: A review. *Archives of Medicine & Health Sciences*. 2016; 4: 50-7.
18. Akhigbe R.E, Ige SF, Afolabi AO, Azeez OM, Adegunlola GJ, Bamidele JO. Prevalence of Haemoglobin Variants, ABO and Rhesus Blood Groups in Ladoke Akintola University of Technology, Ogbomoso, Nigeria. *Trends in Medical Research* 2009; 4(2): 24-9.
19. Umoh AV, Abah GM, Ekanem TI, Essien EM. Haemoglobin Genotypes: A prevalence study and implications for reproductive health in Uyo, Nigeria. *Nigerian Journal of Medicine*. 2010; 19 (1): 36-41.
20. Akwiwu EC, Usanga EA, Akpotuzor JO. Comparative study of haemoglobin types in coastal (Yenogoa) and hinterland (Owerri) communities in South-Eastern Nigeria. *Journal of Medical Laboratory Science*. 2011; 20 (2): 16-20.
21. Piel FB, Patil AP, Howes RE, Nyangiri OA, Gething PW, Dewi M, et al. (2013). Global epidemiology of sickle haemoglobin in neonates: a contemporary geostatistical model-based map and population estimates. *Lancet*. 2013; 381: 142-51.
22. Ware RE. Is Sickle Cell Anemia a Neglected Tropical Disease? *PLoS Neglected Tropical Diseases*. 2013; 7: e2120.
23. Luzatto, L. Sickle cell anaemia and malaria. *Mediterranean Journal of Hematology and Infectious Diseases*. 2012; 4: e2012065.
24. Bunn, H. F. The triumph of good over evil: protection by the sickle gene against malaria. *Blood*. 2013; 121(1): 20-5.