



Emergence of sickle cell gene through migration and intermarriage: A case study

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Abstract

Sickle cell disease (SCD) is an inheritable disorder which is very common among persons of equatorial regions including Africa. The evolution of SCD is linked to malaria endemicity in which sickle cell traits are selected for against malaria and therefore perpetuate the gene to the next generations. In this study we present the pedigree of a family from Ibanda (then sub-county of Mbarara district) which represents the intermarriage between Baganda migrants and Banyankole 'natives'. The possibility of the emergence of SC anemia in this family through migration and gene admixture is therefore explored.

Introduction

In earlier studies, sickle cell disease (SCD) was thought to be a family disease until it was later discovered that it was an autosomal recessive inheritable disorder(1). SCD is therefore transmitted from one generation to another (2,3) and is very common among persons in the equatorial regions especially in Africa (4,5,6). The evolution of SCD in these regions has been linked to malaria endemicity. The sickle cell traits are selected for against malaria and they therefore perpetuate the gene to the next generations(7,8,9). Indeed, its emergence in other regions has been associated with migration and intermarriages (10). According to a previous study by Lehman and Raper, sickle cell trait was found to be very rare (1-4%) among Banyankole (11). In the current study one case of SS (a 12 year old girl) was detected in one of the families in Ibanda district which was then county in Ankole during Lehman and Rapers' study (11) and this raised the interest of the possible origin of sickle cell gene. It was against the above background that this study was done to explore whether migration and gene admixture had any role to play in the emergence of SC anemia in this family.

The Baganda are believed to have migrated to Ibanda district, then a sub county of Mbarara

district, in the mid 18th century. They were fleeing persecution by King Semakokiro after the death of king Junju (12,13,14,15,16). During the migration, the Baganda who could not walk further settled in Ibanda in a place called Kitagwenda which was coined from a word Batagwenda meaning "*those who can not go further*". Others continued to Bushenyi and settled in a place called Bunyaruguru meaning "*a place with a leg that takes them a step ahead*". This is similar to Iteso and Karamojong migration whereby people who could not continue to 'walk further' stayed in Karamoja derived from "*Ekaraki Emojong*" meaning "*the old failed to walk further*", while those who continued were looked at as persons who were going to die from wilderness. These group settled in Teso a place name derived from '*Atesia*' meaning graves) (17).

Methodology

A Case study was done on one of the descendants of Baganda who settled in Ibanda. It's pedigree were as shown in Fig. 1. For the purpose of confidentiality, the names which have been given in this pedigree are not true names of the persons in these families.

The photograph taken of family were as shown in Fig. 2. Milly, is a Muganda by tribe and she is 70 years old. She is a descendant of the Baganda who

Fig. 1. Pedigree of one of the descendants

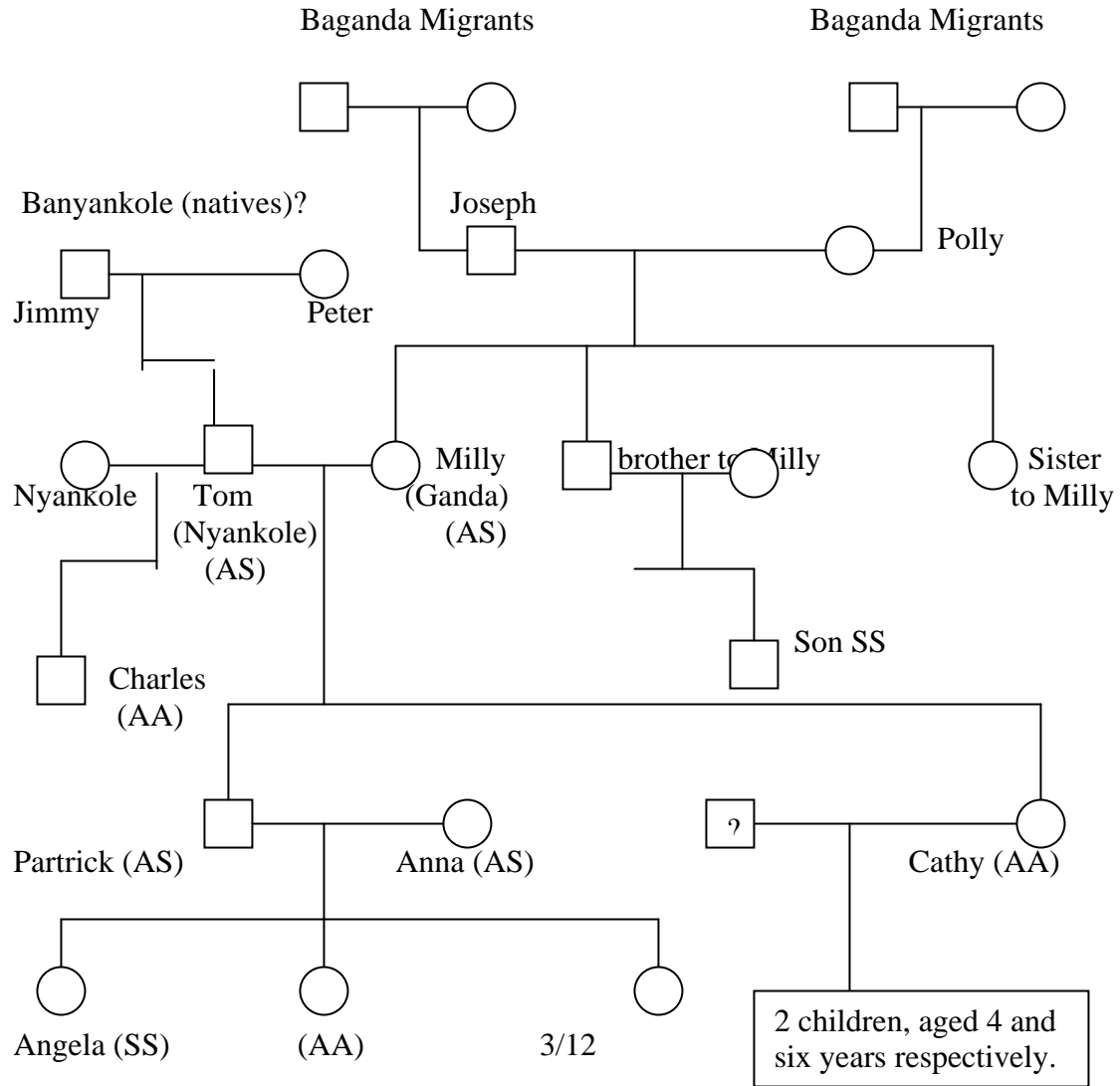
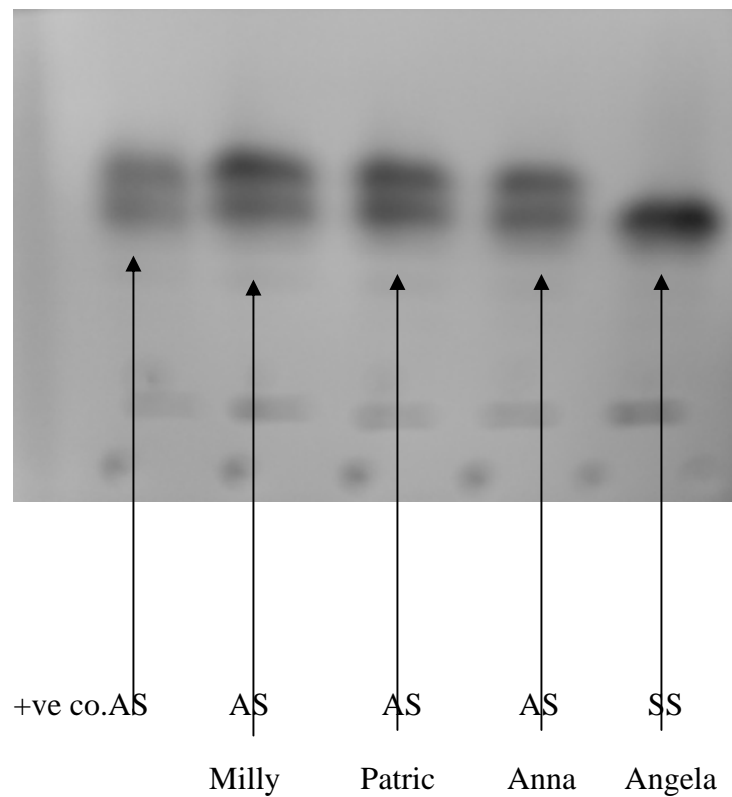


Fig. 2. Photograph of the family



Fig. 3. Some of the Hb electrophoresis results from Toms family



migrated to Ibanda. Her father was called Joseph and the mother was called Polly. She has a brother and a sister who live in Masaka. She said her brother has a child with sickle cell anemia. Milly is married to a munyankole man called Tom (70 years old) whose parents were Grace and Peter (all Banyankole by tribe). They produced five children unfortunately three died from unclear circumstances when they were less than three year old. The living children are: Patrick (38 years old) and Cathy (36 years old). Besides, Tom has a son called Charles with another wife who is a Munyankole. Patrick married a Munyankole woman called Anna (picture 2), she is 32 years old and all her parents are Banyankole. They had six children unfortunately three died of unknown cause at the age below three years. The remaining children are all girls namely; Dorothy (14 years old), Angela (picture 4) 12 years old, was diagnosed with sickle cell disease (SS) in Mbarara hospital and lastly a baby girl (picture 1) who was by then three months old. Cathy (daughter of Milly) has two children aged four and six years old respectively. However, she neither disclosed the father(s) nor childrens whereabouts.

The blood samples were taken from these people for Hb electrophoresis except from a three months old baby because Hb SS levels are low and is therefore not easily detectable by Hb electrophoresis in this age group (18). According to the results, both Milly and Joseph were found to be carriers of sickle cell disease (AS). Patrick and his wife Anna both tested positive for haemoglobin AS. Their daughter Dorothy was detected with AA. However, one of the daughters (Angela) was confirmed as SS. Cathy and Charles were both detected with haemoglobin AA. Some of the Hb electrophoresis results were as shown in Fig. 2.

Discussion

Several hypotheses may be drawn from this case study. First, the presence sickle cell anemia in this family could have been as a result of the migration and intermarriage between the native Munyankole family (natives) and the Muganda family (descendant of Baganda migrants). Secondly since both Milly and Joseph tested positive for AS and their son and his wife also tested positive for AS, it is possible that their children who died at an early age, could have died from sickle cell anemia.

Lastly it was also possible that either one or both parents of Milly and Joseph had AS and so were their great grand parents, thus perpetuating the gene into the next generations. Although we had interests in exploring further the pedigree of both Milly and Joseph in order to elucidate the role of migration and intermarriage in sickle cell involvement in these families, we were not able to do it because of limited financial resources. It was therefore not possible, at this stage, to infer from this pedigree the route of emergence of sickle cell gene in this family.

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