The double-edged sword of bloodlines - Classification of colour, race and ethnicity in sickle cell research and screening

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Sickle cell disease (SCD) – do we need to worry?

Almost one in thirty deaths of children under-5 worldwide are linked to SCD. SCD can be life threatening or complications such as stroke, organ failure and acute chest syndrome. The UN General Assembly has declared SCD to be a major public health concern

Who is at risk?

SCD is caused by the inheritance of a sickle haemoglobin gene from both biological parents. The gene is thought to have emerged as an protective adaptation to malaria over 7,500 years ago. The map below shows the populations where the sickle gene is most common today but the gene could be found anywhere. An estimated 4.4 million people have sickle cell disease and 300 million people have sickle cell trait (SCT) globally. Newborn screening programmes are rare globally and first symptoms may appear in infancy but may be fatal so early identification is vital

'African American'

'In parts of Asia'

'Latinos'

'Brazilian'

'Africa'

'Black'

'African descent'

'Sub Saharan Africa'

'Madhya Pradesh – Vidarbha region, Chaudry, Gamit, Rohit, Vasava tribal groups'

'Black/African American'

'Caribbean'

'Mediterranean'

'Malaria hyperendemic country'

'Arab countries'

'Ethnic African'

'India'

'America'

Methods

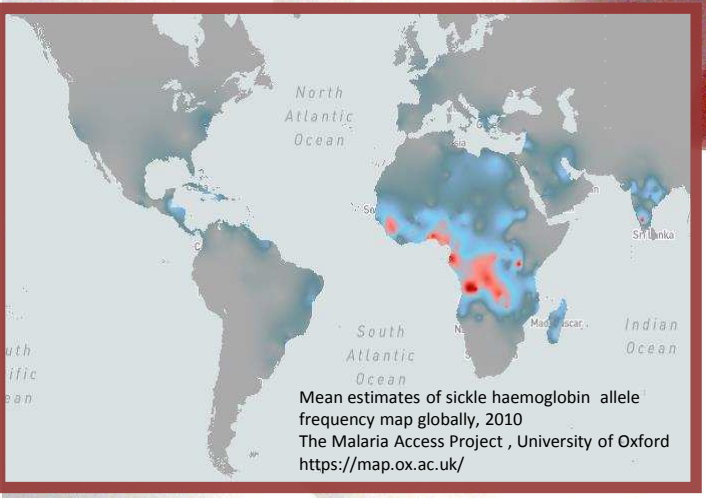
A pubmed search of 'sickle cell screening', free full text past 10 years, yielded 1193 articles of which 100 were randomly selected. Policies for screening searched on google for English speaking countries Terms for at risk groups were extracted by 2 reviewers

Results

At risk groups are described in research and screening policies by ethnic group, region, sub-region, continent, country, race, skin colour or as someone originating from current or past endemic malarial areas. 'Origin' could be defined as recent as biological parents or as ancient as being a descendent of someone of endemic malarial areas of .. Neolithic Africa or countries .. within these latitudes.

The terms found are given between the red blood cells on this poster

The descriptions of risk were usually provided as a list of 2 or more of these terms, never all. The majority of SCD/SCT research is generated in the U.S.A. hence 'African American' was an extremely common description of risk. There is no consensus of how to describe an individual who may be at risk



Mean estimates of sickle haemoglobin allele frequency map globally, 2010
The Malaria Access Project , University of Oxford
https://map.ox.ac.uk/

'French Islands'

'Mijikenda tribe'

'West African'

'Past or present history of malaria endemnicity'

'Black Africans'

'Gulf countries'

'Mixed race'

'In parts of Eastern Mediterranean'

'Historic incidence of death from Malaria'

'Certain African tribes'

'Hispanic'

'Pacific Islander'

'Afro-Carrbbean'

'India – Surat, South Gujarat'

'India – tribal belts of Central zones comprised of Maharashtra, Madhya Pradesh, Vasava tribal groups'

Pitfalls of the terms used

The at risk groups are too numerous, complex and widespread to describe in a simplistic, concise fashion. The list of countries are too numerous, naming regions such as 'Sub Saharan Africa' includes low risk countries such as South Africa and Botswana and skin colour is not an indicator as high risk groups include all skin colours. Almost all of the terms used refer to recent ethnic background whereas the gene could literally be carried by anyone

'Asian'

'Middle East'

'Nigerian'

'Lakh tribal populations'

'Hawaiian'

'Indian Sub-continent'

'Hispanic descent'

'Arabian peninsula'

'Tropics and sub-tropics

'Sukuma tribe'

Conclusion

Highlighting its prevalence in certain communities may lead to better targeting of resources but may also stereotype people being at risk. Thus there is a double edge sword of stigmatisation of the groups considered to be high risk and exclusion of other groups from research, screening and treatment. It seems impossible to cover all groups so is universal screening for all age groups more appropriate given the severity of the disease versus the relatively low cost screening?

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