

Stigma in Sickle Cell Disease: What do we know & how can we address this?

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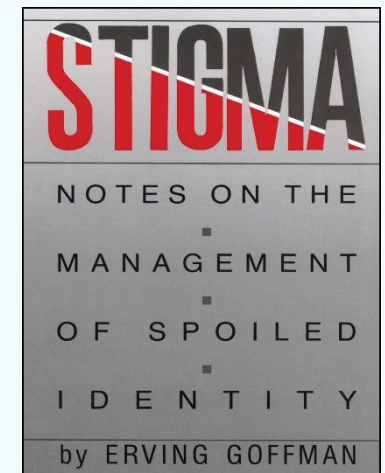
What is Stigma?

- * Shame & disgrace that result from prejudice associated with something regarded as socially unacceptable
- * A multi-dimensional concept

Social Stigma - I

- * Ostracized or treated badly because of some characteristic
 - * individual has little or no control
 - * varies across different groups & in different time periods

- * Dates back to Greeks
 - * cut or burned marks into skin of criminals, slaves & traitors to identify them as tainted or immoral people that should be avoided



Social Stigma - II

- * Physical Features
 - * undesirable visible disabilities within a culture
- * Mental Health & Intellectual Ability
 - * eg. schizophrenia, depression, learning difficulties
- * Behaviour
 - * failing to adhere to specific norms of society or cultural group
- * Group Exclusion
 - * membership in a race, caste, tribe, religion or other group devalued in society

Stigma

- * Guilt
- * Humiliation
- * Disgrace
- * Prejudice
- * Shunned
- * Embarrassed
- * Isolated
- * Ashamed
- * Lonely
- * Disapproved
- * Different
- * Devalued
- * Worry
- * Sadness

Stigma & SCD: Genetic History

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SCIENCE

543

* LINUS PAULING - Nobel Laureate

Sickle Cell Anemia, a Molecular Disease¹

Linus Pauling, Harvey A. Itano,² S. J. Singer,² and Ibert C. Wells³

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“I have suggested that the time might come in the future when information about heterozygosity in such serious genes as the sickle cell anemia gene would be tattooed on the forehead of the carriers, so that young men and women would at once be warned not to fall in love with each other.”

[Linus Pauling, August 15 1966]

Bad Genes and Marriage

By BARBARA YUNCKER

Persons who carry the recessive genes of severe inheritable disease should probably not have children, two famed scientists advised today in order not to increase the bad-gene load of the race.

The two are Nobel laureates Sir Peter Medawar of London and Dr. Linus Pauling, who holds the prize both for chemistry and for peace efforts. They were in town to participate in the dedication of the new Mount Sinai School of Medicine and inauguration of its president and dean, Dr. George James.

Sir Peter, whose prize honored pioneer work in the understanding of immunology and tissue rejection, said two carriers of the same defective gene—for Cooley's anemia or the retardation deficiency PKU, for example—should be discouraged from marrying each other because half their children will be carriers and a quarter of their children will be victims.

“It is humbug to say that such a policy violates an elementary right of human beings,” Medawar said. “No one has conferred upon human beings the right knowingly to bring maimed or biochemically crippled children into the world.”

Genetic Inflation
Even with such limitations, he said, “The frequency of the malignant gene will steadily rise. . . . We are dealing here with a genetic equivalent of inflationary economics; we seem to be getting on all right, but the currency is deteriorating.” Countries to this, he said, could come through medical advances such as direct genetic

repair which he called “not likely, but not inconceivable . . . or, more likely, very early embryonic diagnosis of gross derangements of the chromosomal apparatus—presumably with therapeutic abortion.”

Pauling, speaking later at the “Future of Medicine” session at City College, said: “I agree we should keep these carriers from marrying one another. I have advised, not entirely joking, that individuals should have tattooed on their foreheads symbols for the defective genes they carry”

“Because of certain objections which might be raised, a ribald friend suggests it would be better to tattoo the symbols in Braille on their abdomens.”

Carriers who marry normals, he said, can produce carriers, so they have an obligation to produce a decreased number of children, at least.”

Dr. Francis H. C. Crick of Cambridge, who won a Nobel for his share in deciphering the structure of DNA, the basic genetic material, predicted the new area of ferment in medicine would be in study of the nervous system.

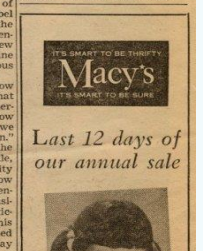
“If all the things we know about man, compared to what we would like to know, the nervous system is the one we know least. . . . It is complex and we are complex for that reason.”

The fourth Nobelist on the panel, Dr. George W. Beadle, now president of the University of Chicago, who elucidated how genes direct production of enzymes, stressed social responsibility of medical schools, particularly in urban centers, as his own and Sinai are. He warned that “black separatism may

counteract and even reverse the trend . . . to [help] disperse, dilute and cure the concentrated social and medical ills” of urban slum areas in cooperative programs with their residents.

James in his inaugural address stressed the need to “redefine the very concept of medical care. The tenfold higher tuberculosis rates in certain areas of our city do not mean that the tuberculosis organism is more virulent there or that our drugs are any less effective in such areas. It must be the poverty and all that accompanies this which is responsible.”

Gov. Rockefeller, in closing the day-long celebration of the new school affiliated with the City University of New York, praised it as proof that “private citizens and private initiative and private enterprise can do a large job on a large scale.”



THE SMARTER TO BE SMARTER
Macy's
IT'S SMARTER TO BE SURE
Last 12 days of
our annual sale

Stigma & SCD: USA History

- * Early 1970's African Americans stigmatised by carrier status
 - * denied health & life insurance
 - * prevented employment opportunities eg. US Air Force Academy
- * Sickle cell screening & discrimination
 - * African Americans anger at being further discriminated against for simply being a carrier of sickle cell trait



Impact of Stigma in SCD: Public Attitudes

 **GiveBlood NHS** ❤️ @GiveBloodNHS · Jun 19
Black people with sickle cell disease urgently need black donors.



Be proud to say #ImThere. Register to #Give blood
blood.co.uk

109 2.7K 1.9K

Braunbär 
@ImGrunenWalde [Follow](#)

Replying to @GiveBloodNHS

If we deport all blacks, this will stop being an issue.

"OR.. we could just deport you."

Impact of Stigma in SCD: Society & Culture

- * Society's attitudes to SCD & those affected
 - * myths, misconceptions
- * Cultural & religious factors
 - * beliefs & practices
 - * divine retribution & intervention
- * Family genetics
 - * blame on mothers i.e. childbearing



Impact of Stigma in SCD: Psychosocial

- * Newborn screening
 - * home visits for newborns & return of results
 - * community nurses are bearers of bad news
- * Visible complications
 - * delayed puberty & growth - small stature
 - * jaundice, leg ulcers
- * Morbidity
 - * 'Sickler', always sick

Impact of Stigma in SCD: Education & Employment

- * Sickle cell pain & fatigue
 - * frequent pain episodes & extreme fatigue can lead to absenteeism from both school & work
 - * false impression of being lazy
- * Insufficient support
 - * special educational needs not addressed
 - * individual requirements of employment not met eg. working from home

Impact of Stigma in SCD: Healthcare - I

- * Sickle cell pain
 - * frequent pain & hospitalisations
 - * mistrust of patients about pain severity
 - * pain levels underestimated
 - * seen as opioid dependent
- * Stigma associated with increased depressive symptoms & hospitalisations for SCD pain^{1,2}



Impact of Stigma in SCD: Health Services - II

- * Emergency Department
 - * Poor experience in A&E due to staff not understanding SCD¹
- * Use of term ‘Sickler’ by Emergency Physicians²
 - * associated with negative attitudes
 - * ‘not seeing SCD patients as people’
 - * inevitably deliver low quality of care




Impact of Stigma in SCD: COVID-19

* Feeling different

* “The Covid-19 Pandemic has really opened my eyes onto how different I am from my able-bodied friends....

It has brought to life all our differences, all our disparities, and all our most vulnerabilities, whether in health, race, or class.”



**Coronavirus
Shielding**

Some people are **Extremely Vulnerable** and coronavirus could kill them. The NHS sent letters to these people about **Shielding**.

If you are shielding

- Stay home for 12 weeks on your own.
- Talk to carers, family or friends. You will need support.
- If you do not have support phone 0800 028 8327

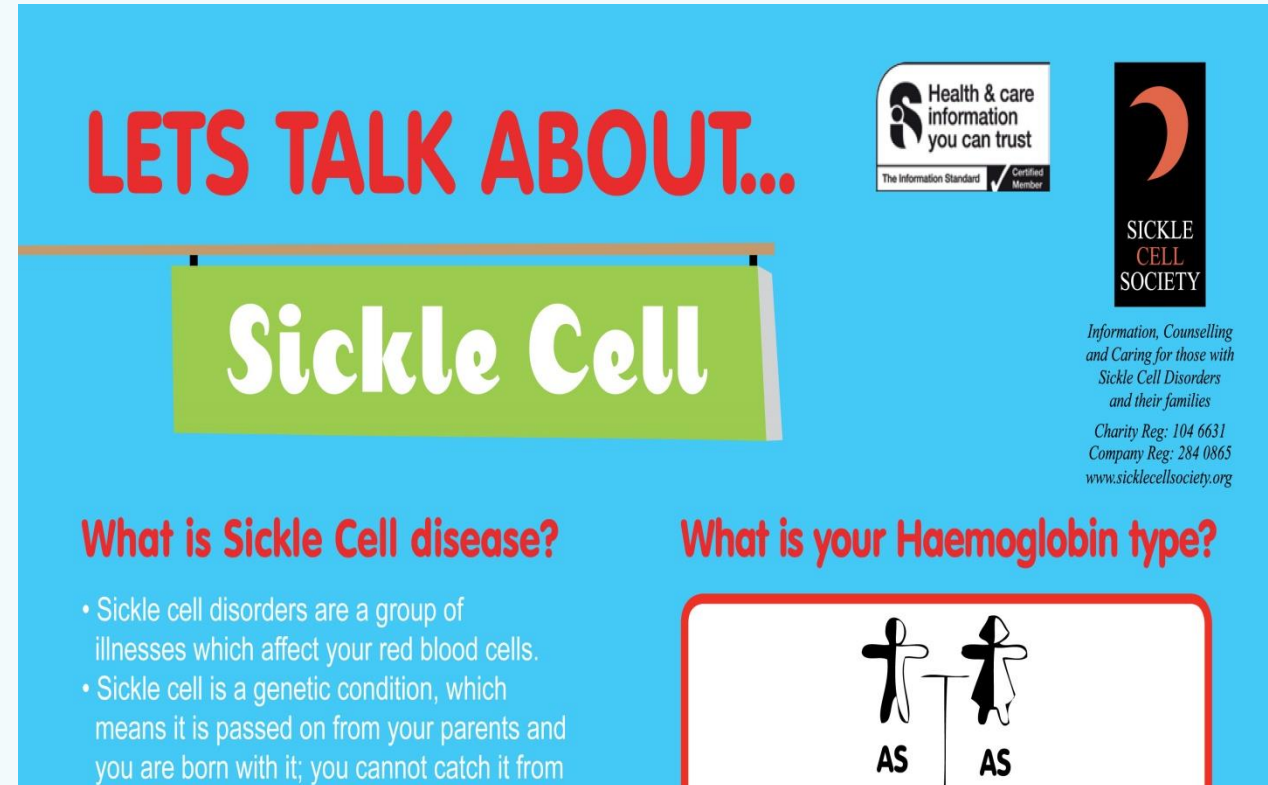
111
111.nhs.uk

**If you feel ill stay home
Get advice from NHS 111
DO NOT go to your GP
pharmacy or hospital**

Updated April 2020 - nhs.uk

Impact of Stigma in SCD: How can we address this?

- * General public & society
 - * more public awareness
 - * improve knowledge & understanding
 - * dispelling myths
- * Health professionals
 - * education & training
 - * all disciplines & levels



LET'S TALK ABOUT...

Sickle Cell

What is Sickle Cell disease?

- Sickle cell disorders are a group of illnesses which affect your red blood cells.
- Sickle cell is a genetic condition, which means it is passed on from your parents and you are born with it; you cannot catch it from

What is your Haemoglobin type?

AS AS

Health & care information you can trust
The Information Standard Certified Member

SICKLE CELL SOCIETY
Information, Counselling and Caring for those with Sickle Cell Disorders and their families
Charity Reg: 104 6631
Company Reg: 284 0865
www.sicklecellsociety.org

Impact of Stigma in SCD: How can we address this?

* Patients & parents

- * empowerment
- * advocacy
- * support groups
- * self-help
- * social media



Impact of Stigma in SCD: How can we address this?

- * Health professionals
 - * patient & parent education
 - * counselling
 - * psychological therapy
- * Other professionals
 - * education
 - * employment
 - * social services, welfare

Summary

- * Stigma in sickle cell disease continues to pose difficulties for patients & families
- * Needs to be addressed with multifaceted strategies involving general public, health professionals, educational institutions, employers, social services, etc.