Case histories
Sickle cell anaemia

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For more on Case histories see **Comment** *Lancet* 2016; **387:** 211, **Perspectives** *Lancet* 2016; **387:** 217, 737, 1265, 1711, 2082, 2495, *Lancet* 2016; **388:** 228, 649, 1148, e10, 2467, and *Lancet* 2017; **389:** 25, 591

Intersectional scholarship is all the rage, and if one wished to write an intersectional history of modern medicine there are few better case studies than sickle cell anaemia (SCA). The story of SCA is the story of an encounter between two harbingers of modernity: a potent molecular frame for disease, rooted in laboratory science, and a set of anxieties and prejudices about race and human difference.

Early 20th-century physicians were deeply ambivalent about the place of laboratory science in clinical practice. The notion of diagnosis by chemical test or by machine, in the hands of expert technicians rather than experienced clinicians, threatened their position as members of an independent and authoritative profession. Tensions over expertise and experience found one expression in the work of James B Herrick, a physician at the Presbyterian Hospital in Chicago, USA. In 1904 Herrick treated Walter Clement Noel, a Grenadian dental student with an unusual "symptom-complex". Using a microscope—still a fairly novel clinical tool—he observed "peculiar elongated and sickle-shaped corpuscles" in a sample of Noel's blood. But Herrick was a laboratory sceptic, carrying out his own chemical assays, and in his 1910 description of the case he emphasised traditional clinical observation over laboratory work.

7 years later Victor Emmel, an anatomist at the University of Washington, embraced laboratory techniques and brought a new dimension to "Herrick's disease". A simple innovation—a ring of Vaseline sealing a coverslip on a microscope slide—revealed that sickling was not present permanently, appearing only when blood oxygen concentrations fell. Emmel's work framed SCA as a counterintuitive kind of disease, one that could be diagnosed from a blood sample rather than an encounter with a patient, and which might appear in potential form even when symptoms were absent.

Through the first half of the 20th century clinical research became ever more entangled in what might, misquoting Eisenhower, be called the medical–industrial complex: a network of intellectual and financial relationships linking laboratories, medical schools, hospitals, pharmaceutical companies, governments, and military. One product of the medical–industrial complex was penicillin; another was a compelling molecular explanation for SCA. In the late 1930s, the chemist Linus Pauling had been headhunted by the Rockefeller Foundation to pursue the "physico-chemical study of life". Using the new technique of electrophoresis, Pauling found that at a particular voltage and pH haemoglobin from patients with SCA separated into two fractions. In a 1949 paper Pauling deployed his electrophoresis studies and a Mendelian analysis to show that SCA was the result of a genetic mutation, leading to the formation of unstable haemoglobin S.

Pauling's research cast new light on the molecular basis of SCA just as the meaning of the condition was growing ever more contested. The higher incidence of SCA in those with sub-Saharan African heritage, and the long and dismal history of institutionalised racism in the USA, generated a general impression that SCA was a so-called "negro disease". Opponents of racial integration played on fears that the disease might become one of many hidden consequences of miscegenation, and Pauling himself suggested that SCA carriers be tattooed on the inner arm. Civil rights activists struck back, citing evidence that SCA had persisted because it conferred a degree of protection against malaria, and so could be reclaimed as a symbol of black history and pride.

In the late 1960s and early 1970s, the Black Panther Party offered testing and treatment with opiates for SCA as part of its inner-city "survival programme"—a reproach to what they saw as the medical and political establishment's indifference to the suffering of poor African Americans. Richard Nixon's 1972 Sickle Cell Control Act provided limited funds from a frozen health-care budget, but (as its name suggests) it continued to emphasise control over treatment, and neither this nor the Black Panthers' survival programmes had a major impact on long-term survival.

In the past 30 years, a number of therapies, notably folic acid and hydroxycarbamide, have been found to be useful in managing SCA, and bone marrow transplants have provided effective cures in children. But the history and the present of SCA—not only in the wealthy west but in low-income settings like sub-Saharan Africa—also prompt reflections on the question of what the remarkable explanatory power of biomedicine has meant for patients and their communities.

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Further reading

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