Learning About Sickle Cell: The Patient in Early Sickle Cell Disease Case Reports, 1910–1933

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Case reports of sickle cell disease (SCD) from its discovery in 1910 to 1933 provide glimpses into the disease's impact on patients and families. Attending physicians, trying to understand the pathophysiology of and treatments for this newly recognized disease, reported also on the effect of SCD on patients' ability to attend school, play, and work, the kinds and severity of the pain patients endured, the late onset of puberty and slowed development of secondary sex characteristics, and the ways families dealt with loved ones who had the disease. These anonymous patients and families helped "teach" physicians about SCD in the early years after its discovery. The current study uses information gleaned from the third published article in 1915 to 1933.

Key Words: sickle cell disease ■ history ■ pain

Medical case reports are intentionally cold, analytical, scientific. Their authors' goal is to present the facts of a case in order to understand a patient's situation, illustrate a specific point, provide information to medical colleagues so they can determine if they have encountered similar cases, and/or alert physicians to be on the lookout for similar cases. Chicago doctor James B. Herrick published his report of a patient with severe anemia and odd crescent-shaped red blood cells in 1910 because he and his colleague Ernest E. Irons had never come upon a case like it before and wanted to put out the information to other practitioners.[1] His plan worked. A fourth-year medical student at the University of Virginia School of Medicine read that report and, four months later, published his own paper about a patient at the university's hospital with similar findings.[2] Those two cases formed the basis for the third and fourth case reports in 1915[3] and 1922,[4] and the steady stream of case reports and analytical articles that followed in the subsequent decade, discussing the new disease that came to be known as sickle cell anemia. Sickle cell anemia is now recognized to include several types of hemoglobinopathies that are called, in aggregate, sickle cell disease (SCD).

Historians can and have told the story of the scientific discovery of SCD[5, 6] as well as the human stories of the first two patients and their physicians.[7, 8] The current paper looks at what can be learned about the effects of SCD on patients from case reports published after the first two in 1910 and 1911 until 1933. In many ways, the descriptions in those articles of symptoms, medical findings, and the course of the disease, as well as the effects of SCD on patients and families, will be familiar to present-day health care professionals, patients and families.

By 1933, more than fifty SCD articles had appeared in print, but though the disease was well-described, it remained puzzling to clinicians and medical scientists. In addition to investigations into the nature of the disease, authors of medical journal papers on SCD during this period included clinical information about patients' chief complaints, past medical history, present situation, laboratory findings, family history, diagnosis, treatment, disease course, and outcome. They also included bits and pieces of personal patient information—facts about individuals and statements about their experience of illness. It is from these fragments of personal data that this article is constructed. What can we learn about these usually anonymous (in a few cases authors revealed a patient's full name) human beings whose medical problems and histories helped shape physicians' understanding of SCD?

The writers of these early articles were trying to paint a picture of a new disease, to characterize it. They built on each other's findings to work out what was occurring within their patients' bodies and the various ways the disorder manifested itself in individuals. Physicians were just learning to include SCD in their differential diagnoses, so they sometimes failed to make the correct judgment the first time (or second or third) a patient was seen. As new articles appeared, physicians had more opportunity to learn about SCD and incorporate it into their diagnostic thinking. Their patients were in some ways their teachers.

These patients were people who felt very sick or whose loved ones considered them very sick and in pain. They hurt and wanted relief from the acute symptoms and chronic problems that beset them, as did their parents and other family members who accompanied them to the doctor or waited at home. If they were in what came to be called (in the late 1920s) sickle cell pain "crisis," as was probably often the case when they presented at the clinic or hospital or dispensary, their physicians were seeing them at their lowest.

The physicians who wrote the case reports were all or almost all white. They were dealing with a disease that appeared almost always to affect people of color. Most practitioners saw SCD as a black disease,[9, 10] but the case reports they wrote were about individuals, human beings, not a race. The authors

did sometimes employ racial stereotypes in their analyses. For example, physicians often mentioned syphilis in their differential or initial diagnosis, syphilis being a disease many whites thought especially rampant among African Americans and which can manifest some of the findings common in SCD. Many of the patients were children. Most patients, when they were hospitalized, remained there for weeks or months, and often returned for second and third stays. This amount of contact provided willing physicians an opportunity to get to know their patients over time, though little evidence of physician-patient or physician-family relationships is revealed in the case reports.

Personal information in the case reports came from the physicians who met their patients in clinic or hospital, observed them, listened to them, and wrote down their stories. We learn about the patients' experiences with SCD through what their physicians thought was important and recorded in the case histories. Some authors wrote brief, terse, single-page case descriptions, while others provided longer and fuller discussions. The current article focuses on what patients endured as a result of their illness and the impact of SCD on their lives. It begins not with descriptions of pain and physical symptoms but with comments about the profound effect the pain and physical symptoms had on how patients lived. The cases that made it to publication were probably the severe ones, the ones that stood out. Some patients with SCD then, as now, developed less dramatic symptoms, and physicians did not report on them. Some patients dealt with their SCD pain without consulting a physician and so were not seen in hospitals and clinics where their cases would have been noticed and recorded.

Physicians could offer few treatments to relieve the various disabling symptoms of SCD. Reliable analgesics for pain relief, salves and healing creams for leg ulcers, and antibiotics for respiratory and other intercurrent infections had not yet been developed, leaving patients to suffer the natural consequences of their illness. They almost certainly tried various home remedies and perhaps patent medicines, but no published case histories of the time recorded such information.

Not counting the first two from 1910 and 1911, 40 articles published on SCD between 1915 and 1933 contained case histories from which patient information beyond the strictly medical was gleaned. (See Table 1) From these 40 articles 54 patient histories were used. (See Table 2) The patients ranged in age from six months to 38 years. Twenty were aged 10 or lower, 15 were between 11 and 19, 14 were in their twenties, four in their thirties, and one was an adult of unstated age. Twenty were female, 34 male. Almost all lived east of the Mississippi River. (The two or three case reports from outside the United States during this period are not included.) Because scientists did not have the means to identify variations in patients' hemoglobin until much later, some of these early patients may not have been S-S, the traditional hemoglobin configuration for sickle cell disease. Some may have had combinations of hemoglobins or thalassemia or another hemoglobinopathy.

During physician-patient encounters doctors heard about and saw the impact of SCD on people's lives. In these early days, of course, the vast majority of patients and families did not know of the existence of sickle cell disease and so thought in terms of and treated symptoms, as often did their physicians.[11] H.S., a "colored maid of eighteen" reported that she had "always been sickly and easily fatigued . . . [and] troubled with 'shaking chills' and night sweats for many years."[12 p51] The mother of L.P., "a colored lad of eight," told her son's doctor that "he has been sickly since infancy." [12 p51] Similarly, I.H., a "tall, poorly developed and poorly nourished" 22-year-old male, had been told by his mother that "he was a normal healthy baby until he was 9 months of age, but that since then he had never been in robust health."[13 p636] Sickle cell was a chronic disease.

As a result of feeling so poorly, patients were unable to participate in the usual childhood or adult activities. As a youngster, Tom Anderson (identified by name in the published case history), a 24-year old Louisiana moss picker, told his physician, he was considered "sickly" and was "never able to play much with the other children," because he "would tire so easily." All through his life he had never been able to "pick up any strength" and so "has never been able to do heavy work, or any kind of work when hurried. When attempting moderately heavy work, or on being hurried," he explained, he "starts to 'tremble all over,' and has to lie down until it passes off."[14 p1192] Another young man told a Johns Hopkins physician a similar story: "As far back as he [the patient] could remember," his doctor reported, "he had been weak and sickly and unable to work or play as hard as other persons of his age." [4 p1318] For individuals such as this, day-to-day life was unpredictable, making regular attendance at school impossible and holding a regular job difficult. R.B., for example, a 20-year-old living in California, attended school very irregularly as a youngster because of repeated illness.[15 pp768-769] Similarly, the physician caring for P.R., "a young mulatto girl" seen at (New Orleans) Charity Hospital's outpatient department, reported that she was "forced to remain away from school" when her symptomatic "periods of distress" became too severe.[16 p1175] N.T., an unmarried 20-year-old in Maryland, had "never been able to work hard or attend school regularly, and, as an adult "is always weak and tired, and is able to do only the lightest kind of work."[17 p338] Thirty-eight-year-old W.T., married with five children, "at times . . . 'had to stop work for a week or two and rest, because of weakness;' at other times does fairly hard work."[17 p338] Staying home was often the best option. A 25-year-old woman admitted to Georgetown University Hospital had, eight days earlier, in the midst of doing housework, developed "severe pain in both legs . . . which forced her to bed where the "pain continued unremittingly."[18 p1672]

Of all the complaints that doctors reported in these case histories, severe pain stands out—joint and muscle pain,

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