Walter Clement Noel—First Patient Described With Sickle Cell Disease

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Historical reviews of medical discoveries usually focus on the physician or scientist who first reported the novel finding or the scientific and intellectual climate at the time of the development. The individual study patient is rarely considered—sometimes because of privacy concerns but perhaps more often because the specific afflicted person is not considered of universal interest. However, for sickle cell disease, first described in 1910 by noted Chicago-based internist James Bryan Herrick (1861-1954), enough details are available about the first patient for a compelling story because of the determined research efforts of a medical historian and a physician (Savitt TL and Goldberg MF, *JAMA* 1989;261(2): 266-271).

Walter Clement Noel (1884-1916), the sickle cell proband, was born on a large estate in the hilly bush country at the north end of Grenada, a small Carribean island that at that time was a British colony. His family was wealthy landholders. Despite chronic health problems, Noel received a quality education and attended Harrison College in Barbados, completing his undergraduate studies in the summer of 1904. In September of that year, Noel sailed from Barbados to New York on the *SS Cearense*; he developed a leg ulcer (a common complication of sickle cell disease) during the week-long journey. After customs and immigration processing in New York, he sought medical attention and was treated with topical iodine, and the leg wound quickly healed. Noel then traveled by train to Chicago, where he had been accepted as a dental student at the Chicago College of Dental Surgery on the city's West Side, and he took a room on West Congress Street in the heart of the Chicago medical district. It was unusual in 1904 for a student of African descent such as Noel to be permitted to study in the United States outside of a traditionally "blacks-only" secondary school; however, Noel was well-to-do and was also a foreigner, and educational opportunities for black students were slowly beginning to expand.

In late November 1904, Noel developed respiratory problems, now recognized as the leading acute cause of death in patients with sickle cell disease, that persisted for more than a month. He finally sought medical attention at the Presbyterian Hospital in Chicago (a private facility) and was evaluated by an intern, Ernest Irons (1877-1959). Irons performed a peripheral blood smear, which was a relatively recent addition to the clinical testing battery, and noted that Noel's blood smear contained "many pear-shaped and elongated forms—some small." Irons discussed the case at length with Herrick, his supervising physician. A thorough search for potential causes of these oddly shaped cells was unrevealing; malaria or a parasitic infection was suspected but not substantiated. With supportive care, Noel eventually recovered and returned to school. Throughout the next 2½ years, as Noel progressed through his dental studies, he experienced several illnesses: he was hospitalized briefly for bronchitis and later confined for 2 months at the Frances Willard Hospital for "a bilious and muscular attack" (patients with sickle cell syndromes frequently have musculoskeletal painful crises and pigment gallstones). Irons also made house calls for Noel's knee pain and for bronchitis. Irons kept dutiful case notes (Noel is described as "bright and intelligent") and gave all these to Herrick at the end of his training. Irons later achieved distinction as a rheumatologist, and he was the president of the American College of Physicians and the American Medical Association in the 1940s.

Herrick presented Noel's case (without giving any credit to Irons) at a national meeting in 1910 and published a detailed report later that same year, but then he turned his attention to other matters; he is customarily given credit for being the first to describe myocardial infarction in 1912. A few months after the Noel case was published, a second, similar case was described in rural Virginia; the patient was a cook and housemaid named Ellen Anthony. The next clear instance was not published for 5 more years; however, by the early 1920s, enough experience had accumulated that Vernon Mason was able to name the illness *sickle cell anemia*. By the 1940s, the inheritance pattern and physical chemistry of hemoglobin S were well enough understood that renowned scientist Linus Pauling (1901-1994) could call sickle cell anemia "the first molecular disease."

Despite his illnesses, Noel graduated from dental school with his entering class in 1907 and then returned to Grenada to set up a private general dentistry practice in the capital city of St George's. His mother owned the building where the practice was located; Noel lived upstairs, and his offices were at street level. Few details are available

about Noel's life between the time of his return to Grenada and his final illness, but it is known that he was successful enough that he was eventually able to hire an assistant. He never married. In 1915, Noel drafted a will, suggesting the possibility that he was becoming worried about his health. In April 1916, he overexerted himself, with fatal consequences. He attended a horse race on the far side of Grenada, traveled a considerable distance home, and bathed, all on the same day. He then developed a "chill," followed by a serious respiratory infection. Noel's condition steadily worsened, and he died in May 1916 in his home, at the age of 32 years. His death certificate lists the cause of death as "asthenia from pneumonia." Noel is buried in a churchyard that overlooks the Caribbean Sea in the parish of Sauteurs (most famous as the site of a French massacre of Carib Indians in the early 1650s), next to his sister Jane, who also died in early adulthood of pulmonary disease, and his father, who died at age 36 years, ostensibly from kidney disease.

Approximately 8% of African Americans are asymptomatic heterozygotes for hemoglobin S, and about 1 in 400 has sickle cell disease. In 2004, the US Post Office released a postage stamp (Scott No. 3877) encouraging early testing for sickle cell disease. The stamp is not a semipostal; it was designed to increase social awareness rather than to raise funds. The message urges early testing for sickle cell disease, although sickle cell screening at birth is now required in almost every state in the United States because early recognition and treatment can prevent complications and death. The stamp was designed by illustrator James Gurney, better known for his popular *Dinotopia* book series for children.