

Katherine C. Silver, MD, MSCR, and Richard M. Silver, MD, MACR

Dr. Katherine C. Silver ($A\Omega A$, Medical University of South Carolina, 2008) is Assistant Professor, Department of Medicine and Department of Pediatrics, Medical University of South Carolina in Charleston.

Dr. Richard M. Silver (AΩA, Vanderbilt University, 1974) is Distinguished Professor, Department of Medicine and Department of Pediatrics, Medical University of South Carolina in Charleston.

The name of my dread disease is Lupus Erythematosus, or as we literary people prefer to call it, Red Wolf. —Flannery O'Connor¹

Flannery O'Connor, the mid-20th century Southern Gothic author, achieved notable and enduring fame despite completing only two novels and 32 short stories. Her career was cut short by systemic lupus erythematosus (lupus), a disease difficult to diagnose and even more difficult to treat in the 1950s and 1960s. Lupus is a systemic autoimmune disease characterized by exaggerated B-cell and T-cell responses and loss of immune tolerance to self-antigens.² Despite today's better understanding of pathogenesis, lupus remains a mysterious and, too often, fatal disease.

Enduring her "dread disease," O'Connor would become one of the towering figures of the American short story, comparable to Poe, her favorite writer as a child; Hawthorne, her idol; and others such as Stephen Crane, Henry James, and Ernest Hemingway.³ O'Connor's legacy is all the more amazing when one considers that the totality of her literary work was completed during her 20s and 30s while ill.

Before the wolf came knocking

Mary Flannery O'Connor was born in Savannah, Georgia, March 25, 1925, the only child of Edward Francis O'Connor and Regina Cline. The O'Connor family was an integral member of the Irish Catholic community of Savannah, a largely Protestant southern town where



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O'Connor's childhood home in Savannah, Georgia. Photographs by Richard M. Silver

Flannery would live until she was 13 years old. With the financial setbacks of the Great Depression, the O'Connors left in 1938 for Atlanta, where Edward went to work for the Federal Housing Administration, part of Franklin Roosevelt's New Deal.

One year prior to the move, Edward had been diagnosed with lupus, his illness beginning as a whitish patch of skin on his forehead, and arthritis. His initial diagnosis was rheumatoid arthritis.⁴ When Edward's health deteriorated further, the O'Connors moved to the Cline family home in Milledgeville, Georgia. Later, Flannery would move with her mother to Andalusia Farm, a nearby estate purchased in the early 1930s by her uncle, Dr. Bernard Cline, a prominent Atlanta physician.⁴ Flannery's father died just one month after his 45th birthday when Flannery was 15-years-old.

Flannery felt the loss of her father deeply and, although she rarely spoke of him, her fiction is replete with widows and orphans. As a spiritually precocious 17-year-old, during her first year at college O'Connor wrote:

The reality of death has come upon us and a consciousness of the power of God has broken our complacency like a bullet in the side. A sense of the dramatic, of the tragic, of the infinite, has descended upon us, filling us with grief, but even above grief, wonder. Our plans were so beautifully laid out, ready to be carried to action, but with magnificent certainty God laid them aside and said, "You have forgotten—mine?"⁴

In June 1945, O'Connor graduated from Georgia State College for Women (now Georgia College & State University) with a degree in social sciences. She departed Georgia the following year to attend the State University of Iowa, where she enrolled in the Writers' Workshop. There, O'Connor became acquainted with writers and critics of the day including Robert Penn Warren, John Crowe Ransom, and Andrew Lytle, the editor of the *Sewanee Review*. Her thesis comprised a collection of short stories entitled *The Geranium*, which would contain the seed for her first novel, *Wise Blood*, published in 1952.⁵ After two years, O'Connor received a Master of Fine Arts degree.

She remained in Iowa for another year before going to the Yaddo artists' colony near Saratoga Springs, New York. There, she was introduced to the poet, critic, and translator of the classics, Robert Fitzgerald. While writing *Wise Blood*, O'Connor lived with Fitzgerald and his wife, Sally, who would become her close friend and later would edit *The Habit of Being: Letters of Flannery O'Connor*, winner of a National Book Critics Circle Special Award.¹

O'Connor returned to Andalusia Farm in 1951, where she surrounded herself with birds of all sorts, most notably the peacocks of which she was so proud. Her passion for birds had begun as early as age five, documented when the newsreel producer *Pathe News* sent a photographer to Savannah to film the young O'Connor and her chicken that "had the distinction of being able to walk either forward or backward."⁶ In her 1961 essay *The King of Birds*, O'Connor estimated that she had about 40 beaks to feed, "though for some time now I have not felt it wise to take a census."⁷

The peacock feather would become her literary shorthand, as denoted on the 2015 Flannery O'Connor commemorative stamp issued by the United States Postal Service.

O'Connor would remain with her mother at Andalusia Farm, surrounded by her beloved birds, where she continued to live and write for the next 13 years until her untimely death.



Plantation house at Andalusia Farm near Milledgeville, Georgia, where O'Connor lived and wrote for much of her life. It is now on the National Register of Historic Places. Georgia College & State University



Flannery O'Connor commemorative stamp issued June 5, 2015, by the U.S. Postal Service depicting the author as a young woman, before the onset of lupus. ©Used by permission from The Mary Flannery O'Connor Charitable Trust and the U.S. Postal Service. All rights reserved

The wolf at the door

An early indication of O'Connor's health problems occurred in December 1949, when at age 24 she was traveling to Georgia for a holiday visit.⁸ Once back in Milledgeville, she fell seriously ill, and during a month-long hospitalization surgery was performed to correct a floating kidney. She wrote, "I have to go to the hospital Friday and have a kidney hung on a rib."⁴

The diagnosis of floating kidney (nephroptosis) was often proposed to explain pain in the back, abdomen, groin, or flank called Dietl's crisis, and treated surgically (nephropexy) in the early 20th century.⁸ Later, after learning of her lupus, a physician friend would speculate, "the Dietl's crisis, if that's what it really was, may actually have been the opening salvo in her battle with that cruel disease."⁴ Lupus nephritis, however, is usually not a source of pain, so whether this episode was the initial manifestation of her lupus is unclear.

The following year, while living in Connecticut and working on *Wise Blood*, O'Connor developed joint pain. Rheumatoid arthritis was suspected, but as with her father's illness, this diagnosis would later prove incorrect. Her friend Sally Fitzgerald wrote:

When, in December 1950, I had put Flannery on the train for Georgia she was smiling perhaps a little wanly but wearing her beret at a jaunty angle. She looked much as usual, except that I remember a kind of stiffness in her gait as she left me on the platform to get aboard. By the time she arrived she looked, her uncle later remarked, "like a shriveled old woman."¹

Upon arrival back in Milledgeville, O'Connor was immediately hospitalized. From Baldwin Memorial Hospital she wrote to a friend, "I am languishing on my bed of semi affliction, this time with AWTHRITUS...."¹ Arthritis is often an early manifestation of lupus, and mimics rheumatoid arthritis, as in her father's case. Recurrent bouts of lupus arthritis may give rise to what is known as Jaccoud arthropathy with characteristic hand deformities that are, unlike those of rheumatoid arthritis, usually reversible and non-erosive.⁹ Photographs of O'Connor suggest that she may have developed Jaccoud arthropathy, as evidenced by ulnar deviation of the fifth metacarpal phalangeal joint along with multiple Boutonniere deformities in her fingers.

During this hospitalization kidney disease was suspected, so her physician consulted Dr. Arthur J. Merrill (A Ω A, Emory University, 1943), Georgia's first nephrologist. Merrill, an early president of the Southern Society for Clinical Investigation,¹⁰ suspected lupus and upon his recommendation O'Connor was transferred to Emory Hospital in Atlanta.

Letters several years later suggest that O'Connor may also have had a typical (malar) facial rash early in the course of her illness.⁸ Later photographs demonstrate a facial rash with the typical malar distribution of lupus.



Arthritis was one of the earliest manifestations of O'Connor's lupus. This photograph appears to show ulnar deviation of the fifth metacarpal phalangeal joint and Boutonniere deformity suggestive of Jaccoud arthropathy. There may also be swelling over the dorsum of the wrist. "Flannery O'Connor" by bswise is licensed under CC BY-NC-ND 2.0

A diagnosis

At Emory Hospital, O'Connor was found to have a positive LE (lupus erythematosus) cell test, confirming the diagnosis of systemic lupus erythematosus (SLE).

The LE cell was discovered in bone marrow at the Mayo Clinic by Dr. Malcolm Hargraves (A Ω A, Ohio State University, 1933), and had been reported only three years before it was used to confirm O'Connor's diagnosis.¹¹

Fearing the shock of discovering she had the same disease that killed her father, her mother chose to conceal from Flannery the news that she was suffering from lupus.⁴ Familial cases of lupus are not rare. Cumulative studies have shown the tendency of SLE to cluster within families,

with first-degree relatives of patients having a greater than 10-fold relative risk compared to the general population.¹²

Following another month-long hospitalization with blood transfusions and steroid treatment, O'Connor improved and was discharged on daily ACTH (adrenocorticotropic hormone) injections. One year earlier, Kendall, Hench, and Reichstein shared a Nobel Prize for their discoveries relating to the hormones of the adrenal cortex, their structure and biological effects. In his Nobel lecture, Dr. Philip S. Hench (A Ω A, University of Pittsburgh, 1925) listed lupus as one of the rheumatic conditions responding to steroid therapy.¹³

By June 1951, O'Connor was well enough to return to Connecticut, and it was only then that she learned from Sally Fitzgerald that her diagnosis was lupus, not rheumatoid arthritis. "Flannery, you don't have arthritis," Sally related. "You have lupus."⁴ After a few moments of silence, Flannery responded, "Well, that's not good news, but I can't thank you enough for telling me..., I thought I had lupus, and I thought I was going crazy. I'd a lot rather be sick than crazy."³

The treatment

Steroid therapy with cortisone and ACTH was a great step forward in the management of lupus. O'Connor tersely assessed her father's earlier treatment saying, "at



Photograph of O'Connor taken in 1952 for a book signing party to celebrate publication of her novel *Wise Blood*. ©Courtesy of Special Collections, Russell Library, Georgia College

that time there was nothing to do for it but the undertaker."¹ Reports as early as 1956, however, noted, "although these steroids modify the acute phase of the disease, there is less evidence that they influence the subsequent course."¹⁴

While corticosteroids markedly improved O'Connor's symptoms, this was certainly not without high cost. In January 1953, O'Connor wrote, "I am doing fairly well these days, though I am practically bald-headed on top and have a watermelon face," 1 signifying alopecia and iatrogenic Cushing syndrome. Another likely steroid complication occurred by early 1954, when O'Connor developed a limp and progressive hip pain,

subsequently attributed to avascular necrosis of the femoral head. O'Connor would require the use of crutches for most of her remaining years. In a letter to a friend, she wrote:

My last x-rays were very bad, and it appears the jaw is going the same way as the hip is. I had noticed a marked change in the position of my mouth.¹

Suggesting that avascular necrosis may have also affected her jaw.

A few weeks later she wrote:

What they found out at the hospital is that my bone disintegration is being caused by the steroid drugs which I have been taking for 10 years. So they are going to try to withdraw the steroids.¹

O'Connor's steroid therapy was tapered during the first half of 1961, perhaps aided by the addition of an antimalarial drug. In May of that year, O'Connor was taking chloroquine (Aralen[®]). Ten years prior, the first report of an antimalarial (mepacrine) for lupus had been published.¹⁵ Flannery wrote to a friend who also suffered from lupus to say, "Dr. Merrill tells me that they can control the lupus skin rash (when it is just the skin-type of lupus and not systemic lupus) entirely with Aralen."¹ Today, antimalarials, e.g., hydroxychloroquine (Plaquenil[®]), are a cornerstone of the management of skin and joint manifestations of lupus.

After steroid withdrawal, her lupus appeared to be in remission, as O'Connor's correspondence mentioned no new health problems until Christmas Day of 1963, when she reported to a friend that she had fainted several days earlier and was restricted to bed. She was found to be anemic, due to bleeding from a fibroid tumor, and she was hospitalized for a hysterectomy.

The surgery appeared to be successful, however she returned to the hospital for 10 days the third week in April because "I woke up covered from head to foot with the lupus rash."⁴ Lupus often flares following stress including that induced by surgical trauma.

In late May, O'Connor was admitted to Piedmont Hospital in Atlanta with anemia, weakness, and a 20-pound weight loss. She reported that her blood pressure was "dangerously" high.⁸ She was placed on a low-protein diet,



O'Connor at Andalusia Farm with one of her peacocks. For much of her later life she required the use of crutches due to hip pain from avascular necrosis. The Habit of being / photo by Joe McTyre. Georgia, printed ca. 1979

suggesting she was azotemic, and she received transfusions because her hemoglobin was "down to 8."⁸ After almost a month, she was discharged home. "My dose of prednisone has been cut in half on Dr. Merrill's orders because the nitrogen content of the blood has increased by a third."¹

In late July, she was given another blood transfusion and wrote about receiving "a double dose of antibiotic for the kidney," also writing that "they are withdrawing the cortisone." ¹ Soon afterward, O'Connor became critically ill and was rushed to the hospital. O'Connor, at only 39 years of age, lapsed into a coma and died August 3, 1964.

Living with the red wolf

It has been said that one of the most remarkable features of O'Connor's letters is how little she says about her personal suffering.⁴ Instead, she often made self-deprecating quips about her illness and its treatment.

Regarding her painful daily injections of ACTH, O'Connor wrote to a friend, "I owe my existence and cheerful countenance to the pituitary glands of thousands of pigs butchered daily in Chicago, Illinois at the Armour packing plant. If pigs wore garments I wouldn't be worthy to kiss the hems of them."¹ On one occasion, when a viral infection led to a flare of her lupus and the dose of ACTH had to be increased, O'Connor found comfort in the fact that the cost of the ACTH not covered by insurance had been "...reduced from \$19.50 to \$7.50," which she described as "a kind of Guggenheim," referring to the coveted fellowships for gifted writers and artists.⁴

By the spring of 1954, O'Connor was beginning to limp and was noted to use a cane. To a friend she wrote, "I am doing very well these days except for a limp, which I am informed is rheumatism. Colored people call it 'the misery.' Anyway I walk like I have one foot in the gutter but it's not an inconvenience and I get out of doing a great many things I don't want to do."⁴

On her later requirement for crutches, O'Connor wrote, "They say if I keep the weight off it entirely for a year or two, it may harden up again; otherwise in my old age I will be charging people from my wheelchair...."¹

Despite limited weight bearing, the hip never improved. At one point, hip arthroplasty was discussed but later dismissed for fear the stress of surgery might flare her lupus.

O'Connor continued to deal with her requirement for crutches in her inimitable style. An elderly woman she encountered in an elevator of an Atlanta department store whispered in her ear, "Remember what they said to John at the gate, darling!" In a letter to a friend Flannery recounted this experience noting that she had learned the meaning of the elderly lady's words, i.e., "The lame shall enter first." O'Connor continued, "This may be because the lame will be able to knock everybody else aside with their crutches."⁴

In O'Connor's short story *The Lame Shall Enter First*, the main character, Rufus, is described as having a "monstrous club foot," and she describes a brace shop where "the walls were hung with every kind of crutch and brace." ¹⁶

In the spring of 1958, O'Connor, made a pilgrimage to Lourdes with her mother and a wealthy Savannah cousin who financed the trip. O'Connor wrote, "Lourdes was not as bad as I expected it to be....They passed around a thermos bottle of Lourdes water and everybody had a drink out of the top. I had a nasty cold so I figured I left more germs than I took away."¹

It was not physical health that O'Connor was after at Lourdes or anywhere else, "I prayed there for the novel I was working on, not for my bones, which I care less about," she wrote.⁴

In early 1960, soon after her second book *The Violent Bear It Away* went on sale, O'Connor realized her greatest fear of having her life and her disease exposed.⁴ In an unflattering book review for *Time*, a critic mentioned (and mischaracterized) her lupus. To a friend, O'Connor wrote, "My lupus has no business in literary considerations."¹

In her introduction to *The Habit of Being*, Sally Fitzgerald wrote of O'Connor, "…her offhand way of speaking of her physical ordeal, when she did, tells more about her gallantry than any encomium could make real."¹ One month before her death O'Connor wrote, "The wolf, I'm afraid, is inside tearing up the place."¹

Undeterred by the "Red Wolf," O'Connor will be remembered for her darkly comic and unsettling stories, and as one of the most brilliant and provocative writers of the 20th century.

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The corresponding author's E-mail address is silverr@musc.edu.



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