# CORRESPONDENCE

## Sickle Cell Anemia in Septuagenarians

*To the Editor:*

Sickle cell anemia was once deemed a disease of childhood. Recently, there has been a vast improvement in the longevity of sickle cell anemia patients in the United States.[1] The median ages of death for males and females are now 42 and 48 years, respectively.[2] More severe clinical complications portended an earlier death, and fetal hemoglobin (Hb F) had a protective effect.[2] Most clinics caring for substantial numbers of sickle cell anemia patients have seen individuals in their sixth and seventh decade. Rare patients with sickle cell anemia have a nearly normal lifespan; we describe five of these individuals in this report. Key laboratory features noted in the eighth decade of life, at a time when their disease was stable, are presented in Table 1. A summary of the clinical features of these patients is shown in Table 2.

While the life-span of these individuals was near normal, they were not unaffected by their disease. As expected, most serious complications became apparent in their later years. Although anecdotal, some laboratory observations are curious. Hb F levels—a predictor of diminished longevity and increased pain—were lower than average.[2,3] Higher than usual numbers of dense cells, a predictor of less pain, were found. The leukocyte counts were lower than usual—a good prognostic finding in sickle cell anemia.[2] Leukocyte counts and Hb F levels decrease with age in sickle cell anemia.[1] Their levels in these patients may be an effect—not a cause—of their extended life. Low hemoglobin levels predict less pain and more leg ulcers. Two individuals had coincident $\alpha$ thalassemia. However, evidence suggests that $\alpha$ thalassemia—perhaps mediated through its effects on PCV—is a mortality risk in sickle cell anemia.[4] As predicted by their Hb F concentration, no patient had a favorable $\beta$-globin gene cluster haplotype.[5] One earlier account of a 70-year-old man with sickle cell anemia and a normal $\alpha$-globin genotype reported a PCV of 18, 6.4% Hb F, and a WBC count of $10.3 \times 10^6$/L.[6]

We do not understand why some patients with sickle cell anemia survive their peers by decades just as we have little insight into why occasional normal individuals live far beyond the average number of years. It seems unlikely that one or a few laboratory measurements will explain this observation. However, these unusual patients can serve as exemplars to others with this disease and should alert physicians that this diagnosis cannot be excluded by age alone.

**Table 2. Clinical Findings in Septuagenarians With Sickle Cell Anemia**

| Pain | CHF | Ulcer | Chest | ON | CVA | Retina | TX | Other |
|---|---|---|---|---|---|---|---|---|
| 0 | Y | Y | N | N | N | N | Y | A |
| 10 | Y | Y | Y | Y | Y | N | N | D |
| 3 | N | Y | N | Y | N | N | N | A |
| 0 | Y | Y | N | N | N | N | Y | D |
| 0 | Y | N | N | Y | N | N | Y | A |

Abbreviations: Pain, average numbers of pain crises yearly; CHF, congestive heart failure; Y, yes; N, no; Ulcer, presence or history of leg ulcers; Chest, history of acute chest syndrome; ON, osteonecrosis; CVA, history of cerebrovascular accidents; Retina, sickle retinopathy; TX, regular transfusions; A, alive; D, deceased at age given in Table 1.

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**Table 1. Laboratory Values in Septuagenarians With Sickle Cell Anemia**

| Age/Sex | Hb | PCV | DS | MCV | Retic | WBC | PLT | HbF | HAPLO | $\alpha$ Genes | Cr | ALT | Bili |
|---|---|---|---|---|---|---|---|---|---|---|---|---|---|
| 75/M | 5.6 | 19 | 26 | 80 | 13 | 6.4 | 249 | 4.0 | BEN/Atyp | 3 | 1.5 | 23 | 3.3 |
| 71/M | 7.8 | 24 | 13 | 86 | 7 | 6.8 | 170 | 5.3 | BEN/BEN | 3 | 1.3 | 27 | 2.4 |
| 71/F | 7.5 | 23 | 14 | 95 | 14 | 10.7 | 433 | 2.8 | BEN/CAR | 4 | 1.5 | 22 | 1.7 |
| 76/M | 5.5 | 15 | — | 96 | 7 | 7.6 | 162 | 0.2 | — | 4 | 4.2 | 25 | 2.9 |
| 73/M | 5.5 | 16 | — | 91 | 5 | 11.4 | 272 | <1 | — | — | 1.5 | 7 | 4.7 |

Abbreviations: Hb, hemoglobin level in grams per deciliter; PCV, packed cell volume; DS, dense cells; MCV, mean corpuscular volume (fL); Retic, reticulocyte count (%); WBC, leukocyte count ($10^6$/L); PLT, platelet count ($10^9$/L); Hb F, fetal hemoglobin (%); HAPLO, $\beta$-globin gene cluster haplotype; BEN, Benin; CAR, Central African Republic; Atyp, atypical; $\alpha$ Genes, $\alpha$-globin genotype; Cr, creatinine (milligrams per deciliter); ALT, alanine aminotransferase (units); Bili, total bilirubin (milligrams per deciliter).

## REFERENCES

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