4191-02U

SOCIAL SECURITY ADMINISTRATION

[Docket No. SSA-2017-0007]

Social Security Ruling, SSR 17-3p;

Titles II and XVI: Evaluating Cases Involving Sickle Cell Disease (SCD)

AGENCY: Social Security Administration.

ACTION: Notice of Social Security Ruling (SSR).

SUMMARY: We are providing notice of SSR 17-3p. This SSR provides guidance on SCD and how we evaluate SCD in disability claims under titles II and XVI of the Social Security Act.

DATES: This SSR is applicable on [INSERT DATE OF PUBLICATION IN THE FEDERAL REGISTER].

FOR FURTHER INFORMATION CONTACT: Cheryl A. Williams, Office of Disability Policy, Social Security Administration, 6401 Security Boulevard, Baltimore, Maryland 21235-6401, (410) 965-1020. For information on eligibility or filing for benefits, call our national toll-free number, 1-800-772-1213 or TTY 1-800-325-0778, or visit our Internet site, Social Security Online, at *http://www.socialsecurity.gov.*

SUPPLEMENTARY INFORMATION: Although 5 U.S.C. 552(a)(1) and (a)(2) do not require us to publish this SSR, we are doing so in accordance with 20 CFR 402.35(b)(1).

Through SSRs, we make available to the public precedential decisions relating to the Federal old-age, survivors, disability, supplemental security income, and special veterans' benefits programs. We may base SSRs on determinations or decisions made at all levels of administrative adjudication, Federal court decisions, Commissioner's

decisions, opinions of the Office of the General Counsel, or other interpretations of the law and regulations.

Although SSRs do not have the same force and effect as statutes or regulations, they are binding on all components of the Social Security Administration. 20 CFR 402.35(b)(1).

This SSR will remain in effect until we publish a notice in the Federal Register that rescinds it, or until we publish a new SSR that replaces or modifies it.

(Catalog of Federal Domestic Assistance, Programs Nos. 96.001, Social Security—Disability Insurance; 96.002, Social Security—Retirement Insurance; 96.004, Social Security—Survivors Insurance; 96.006—Supplemental Security Income.)

Nancy A. Berryhill,
Acting Commissioner of Social Security.

POLICY INTERPRETATION RULING

Titles II and XVI: Evaluating Cases Involving Sickle Cell Disease (SCD)

PURPOSE: This Social Security Ruling (SSR) provides background information on SCD and how we evaluate SCD during our adjudication process. We provide this guidance to help adjudicators consistently apply our policies in disability claims involving SCD.

CITATIONS: Sections 216(i), 223(d), 223(f), 1614(a)(3) and 1614(a)(4) of the Social Security Act, as amended; Regulations No. 4, subpart P, sections 404.1502, 404.1505, 404.1509, 404.1512, 404.1513, 404.1520, 404.1520a, 404.1520b, 404.1521, 404.1522, 404.1523, 404.1525, 404.1526, , 404.1529, 404.1545, 404.1560-404.1569a, 404.1593, 404.1594, appendices 1 and 2; and Regulations No. 16, subpart I, sections 416.902, 416.905, 416.906, 416.909, 416.911, 416.912, 416.913, 416.920, 416.920a, 416.920b, 416.921, 416.922, 416.923, 416.924, 416.924a, 416.925,416.926, 416.926a, , 416.929, 416.945, 416.960-416.969a, 416.987, 416.993, 416.994, and 416.994a.

INTRODUCTION:

SCD is the most common inherited blood disease in the United States, affecting an estimated 100,000 Americans.[1] SCD is not always easy to evaluate due to its varying nature and complications. In this SSR, we provide basic information about SCD and its

[1] See Centers for Disease Control and Prevention, "Sickle Cell Disease." (https://www.cdc.gov/ncbddd/sicklecell/data.html.)

variants and clarify that sickle cell trait is not a variant of SCD. We also provide guidance for assessing SCD under the hematological disorder listings and determining how this impairment may affect the residual functional capacity finding for adults and the functional equivalence finding for children.

POLICY INTERPRETATION:

We consider all medical evidence when we evaluate a claim for disability benefits. The following information is in a question and answer format that provides guidance about SCD and how to consider evidence regarding this impairment. Questions 1 and 2 provide basic background information about SCD and its variants. Question 3 clarifies that sickle cell trait is not a variant of SCD. Question 4 discusses the complications and symptoms of SCD. Questions 5 through 7 explain how adjudicators should evaluate SCD at various points of the adjudication process, including the adult and child hematological listings we consider.

List of Questions

1. What is SCD?

2. What are the different variants of SCD?

3. Is sickle cell trait a variant of SCD?

4. What are the common complications and symptoms of SCD?

5. How do we evaluate the complications of SCD under the hematological disorder listings?

6. How do we evaluate the complications of SCD when assessing residual functional capacity (RFC) for adults?

7. How do we evaluate the complications of SCD under functional equivalence for children?

Answers

1. <u>What is SCD?</u>

SCD is a type of hemolytic anemia and an inherited hematological disorder that affects the hemoglobin within a person's red blood cells (RBC). Hemoglobin is the protein within RBC that carries oxygen. The abnormal hemoglobin makes the RBC more prone to distortion ("sickling"), which results in blocked blood vessels and a shortened RBC lifespan. Hemolytic anemia results when the abnormal RBC are destroyed faster than the body can produce them.

When hemoglobin is normal, a person's RBC are round and easily travel through

blood vessels, bringing oxygen to the body's organs and tissues. SCD causes sickle-shaped RBC that are not flexible and can stick to vessel walls, causing blockages (vaso-occlusion) that slow or stop the flow of blood and oxygen. This blockage may in turn cause pain. Persons with SCD are predisposed to pain, infection, and other complications. Because people inherit SCD, the disease is present at birth, but the age when children display symptoms varies.[2]

2. What are the different variants of SCD?

The different variants of SCD may indicate the severity of complications and the resulting functional limitations caused by SCD. Laboratory blood tests such as hemoglobin electrophoresis establish the existence and the variants of SCD. The following are the most common variants of SCD:[3]

- Hemoglobin (Hb) SS (HbSS) — a person with this form of SCD inherits one sickle cell gene from each parent. HbSS is the most common and usually most severe form of SCD.

[2] See National Heart, Lung, and Blood Institute, "What Are the Signs and Symptoms of Sickle Cell Disease?" (https://www.nhlbi.nih.gov/health/health-topics/topics/sca). Health problems usually do not appear until an infant is around 5 to 6 months of age.
[3] See Centers for Disease Control and Prevention, "Sickle Cell Disease." (https://www.cdc.gov/ncbddd/sicklecell/facts.html).

- HbSC — a person inherits one sickle cell gene from one parent, and another gene for an abnormal hemoglobin called "C" from the other parent. HbSC is usually a milder type of SCD.

- Hb S-beta (Sβ) thalassemia — a person inherits one sickle cell gene from one parent, and a gene for beta thalassemia from the other parent. There are two forms of beta thalassemia, sickle beta zero thalassemia (Hb $S\beta^0$ thalassemia) and sickle beta plus thalassemia (Hb $S\beta^+$ thalassemia). Sickle beta zero thalassemia is usually a more severe form of SCD. People with sickle beta plus thalassemia tend to have a milder form of SCD.

- HbSD, HbSE, and HbSO— people with these variants of SCD have one sickle cell gene plus another abnormal hemoglobin gene, "D," "E," or "O." These are rarer types of SCD with varying severity.

3. Is sickle cell trait a variant of SCD?

No. Sickle cell trait is not a variant of SCD. Sickle cell trait occurs when a person inherits one sickle hemoglobin gene from one parent and a normal gene from the other parent. People with sickle cell trait rarely have signs and symptoms associated with SCD and usually do not need treatment. However, in rare cases and under extreme conditions such as intense exercise, people with sickle cell trait have a higher risk of severe breakdown of muscle tissue (exertional rhabdomyolysis) that can lead to serious

complications.[4] In spite of this higher risk, recent evidence indicates that sickle cell trait is not associated with an increased probability of death.[5]

Sickle cell trait alone is not an impairment. As defined by the Social Security Act, an impairment must result from anatomical, physiological, or psychological abnormalities that can be shown by medically acceptable clinical and laboratory diagnostic techniques. To establish an impairment in this context, we require objective medical evidence (medical signs and laboratory findings) from an acceptable medical source of complications from sickle cell trait. In addition, a person's complications from sickle cell trait must meet the statutory duration requirement, i.e., be expected to result in death or last or be expected to last for a continuous period of not less than 12 months. Therefore, we cannot find a person disabled due to sickle cell trait if there are no medical signs or laboratory findings of complications from sickle cell trait and the complications from sickle cell trait do not meet the duration requirement.

4. <u>What are the common complications and symptoms of SCD?</u>

Complications of SCD may include, but are not limited to pain crises, anemia, osteomyelitis, leg ulcers, pulmonary infections or infarctions, acute chest syndrome, pulmonary hypertension, chronic heart failure, gallbladder disease, liver failure, kidney

[4] Other conditions that could be harmful for people with sickle cell trait include high altitudes, dehydration, low oxygen levels in the air, and increased pressure in the atmosphere. We evaluate impairments that result from sickle cell trait under the affected body system.
[5] See Nelson D.A., et al. Sickle Cell Trait, Rhabdomyolysis, and Mortality among U.S. Army Soldiers. New England Journal of Medicine, Aug; 375(17), 1695-6 (2016).

failure, nephritic syndrome, aplastic crisis, stroke, and mental impairments such as depression. Examples of symptoms that may stem from these complications include pain, fatigue, malaise, shortness of breath, and difficulty feeding in infants. The symptoms of SCD vary from person to person and can change over time.

A. Pain (vaso-occlusive) crisis is a common complication of SCD. Pain crises are either acute or chronic. Acute pain crises occur suddenly when sickled RBC stop blood flow and reduce oxygen delivery. This pain can be intense, stabbing, or throbbing. Pain can strike almost anywhere in the body and in more than one spot at a time. The pain often occurs in the lower back, legs, arms, abdomen, and chest.[6] Chronic pain in SCD is more than a continuation of acute pain crisis. It usually occurs when lack of oxygen to the bone due to vaso-occlusion results in the death of bone tissue (avascular necrosis) at various joints such as the hips, shoulders and ankles.[7]

B. Anemia is another complication of SCD. It occurs when sickled RBC die prematurely, which reduces the amount of oxygen-carrying hemoglobin in the blood. Symptoms from anemia can include fatigue, weakness, shortness of breath, and dizziness. Chronic deprivation of oxygen-rich blood can damage nerves and organs in the body, including the spleen, brain, eyes, joints, bones, lungs, liver, heart, kidneys, and other organs.

---

[6] See National Heart, Lung, and Blood Institute, "What Are the Signs and Symptoms of Sickle Cell Disease?" (http://www.nhlbi.nih.gov/health/health-topics/topics/sca/signs).

[7] See Okpala I, Tawil A. Management of Pain in Sickle-Cell Disease. Journal of the Royal Society of Medicine, Sep; 95(9), 456-458, 2002 (available at: http://www.ncbi.nlm.nih.gov/pmc/articles/PMC1279994/).

C. Pulmonary complications such as acute chest syndrome (ACS) and pulmonary hypertension are the leading cause of death for SCD patients.[8] ACS is a vaso-occlusion of the pulmonary vessels. Symptoms of ACS include but are not limited to chest pain, fever, tachypnea (abnormally rapid breathing), wheezing, or coughing. Pulmonary hypertension can occur when sickled RBC cause pulmonary arteries to become narrow and blocked. The result of this damage to the pulmonary arteries is high blood pressure in the lungs. Symptoms of pulmonary hypertension include shortness of breath, fatigue, and chest pain.[9]

D. Strokes and silent strokes affect people with SCD at a higher rate because sickled RBC clump along the walls of larger arteries going to the brain. Strokes can result in full or partial paralysis on one side of the body, problems with balance, or difficulty speaking or understanding. Silent strokes can occur without outward symptoms and are only detectable by brain imaging. However, silent strokes can impair intellectual ability, attention, visual-spatial skills, language, and long-term memory.[10]

E. Bacterial infections are often severe complications in people with SCD. Anemia from SCD and vaso-occlusions can damage the spleen, which ultimately

---

[8] See Gladwin MT, Miller, A. Pulmonary Complications of Sickle Cell Disease. American Journal of Respiratory and Critical Care Medicine, Jun; 185(11), 1154-1165, 2012 (available at: https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3373067/).
[9] See National Institutes of Health. MedlinePlus. "Pulmonary Hypertension." (https://medlineplus.gov/pulmonaryhypertension.html).
[10] See, The Internet Stroke Center, "Stroke as a Complication of Sickle Cell Disease." (http://www.strokecenter.org/patients/about-stroke/pediatric-stroke/stroke-as-a-complication-of-sickle-cell-disease/).

increases risk of infection and damages other organs. Infection frequently leads to hospitalization and is the primary cause of death in young children with SCD.[11]

F. Mental disorders in people with SCD are often secondary to the impact of treatment, pain, and other symptoms. For example, depression from reoccurring pain is especially common in people with SCD.[12] Other mental disorders that may occur include, but are not limited to, anxiety and cognitive disorders from stroke.[13]

5. How do we evaluate the complications of SCD under the hematological disorder listings?

We may evaluate SCD under the following hematological disorder listings:

- Listing 7.05 and 107.05, Hemolytic anemias; or

- Listing 7.17 and 107.17, Hematological disorders treated by bone marrow or stem cell transplantation; or

- Listing 7.18, Repeated complications of hematological disorders.

[11] See Booth, C., et al. Infection in Sickle Cell Disease: A Review. International Journal of Infectious Diseases, Jan; 14(1), e2-e12, 2010 (available at: http://www.sciencedirect.com/science/article/pii/S1201971209001453).
[12] See Jonassaint CR, Jones VL, Leong S, Frierson GM. A Systematic Review of the Association between Depression and Health Care Utilization in Children and Adults with Sickle Cell Disease. British Journal of Hematology, Jul; 174(1), 136-47, 2016.
[13] Becker M, Axelrod DJ. Hematologic Problems in Psychosomatic Medicine. Psychiatric Clinics of North America, Dec; 30(4), 739-759, 2007 (available at: http://www.sciencedirect.com/science/article/pii/S0193953X07000767).

Under listing 7.05 and 107.05, we assess hemolytic anemias, including sickle cell disease, thalassemia, and their variants. We evaluate pain crises caused by SCD under listings 7.05A and 107.05A. We assess complications of SCD requiring hospitalizations under listings 7.05B and 107.05B. Listings 7.05C and 107.05C describes the criteria we use to evaluate SCD that results in anemia with low hemoglobin levels.

Under listings 7.17 and 107.17, we consider people who receive bone marrow or stem cell transplantation to treat their SCD, to be disabled for at least 12 months after the date of transplant.

We evaluate adults who have repeated complications from SCD, but do not have the requisite findings for listing 7.05 or 7.17, under listing 7.18.[14] To meet listing 7.18, SCD must cause repeated complications, resulting in significant, documented symptoms or signs and a "marked" level of limitation in one of the general areas of functioning: activities of daily living, social functioning, or completing tasks because of deficiencies in concentration, persistence, or pace. We use listing 7.18 to evaluate only hematological disorders.[15]

[14] We evaluate a child's functioning under the rules for functional equivalence. See 20 CFR 416.926a.
[15] We use listing 7.18 to evaluate hematological disorders and complications caused by hematological disorders. We can only evaluate anemia under 7.18 if it results from an underlying hematological disorder. If the person's anemia results from a condition that is not a hematological disorder, we would evaluate the anemia under the listing for that impairment.

If a person's SCD does not meet a hematological listing, we will compare the specific findings in each case to any appropriate hematological listings to determine whether medical equivalence may exist. We may also find medical equivalence if the person has multiple impairments, including SCD, none of which meet or medically equal the requirements of a listing alone, but the combination of impairments is medically equivalent in severity to a listed impairment.

If the person's SCD does not meet or equal the criteria in a listing, we will consider whether he or she has an impairment that satisfies the criteria in a listing in another body system. For example, we may evaluate the effects of intracranial bleeding or stroke under 11.00 or 12.00.

6. How do we evaluate the complications of SCD when assessing residual functional capacity (RFC) for adults?

For adults, we assess RFC when the effects of a person's SCD, either alone or in combination with another impairment(s), do not meet or medically equal a listing. We base the RFC assessment on all the relevant evidence in the record, including the effects of treatment.[16] In assessing RFC, we must consider all of a person's work-related limitations, whether due to SCD, other impairment(s), or a combination of impairments. For example, adults with SCD may have pain, fatigue, and shortness of breath that may affect their ability to stand and walk. In addition, a person experiencing repeated acute

[16] See 20 CFR 404.1545 and 416.945, and SSR 96-8p.

pain crises may have difficulty maintaining concentration to complete tasks and have frequent absences from work.

7. How do we evaluate the complications of SCD under functional equivalence?[17]

Children with SCD that does not meet or medically equal a listing may nevertheless have an impairment(s) that functionally equals the listings under our rules for evaluating disability in children.[18] When we determine whether a child's impairment(s) functionally equal the listings, we use the six domains of functioning.

When we evaluate a child's functioning in these six domains, we consider how the child functions compared to children the same age who do not have impairments. We must explain any limitation in a child's ability to function appropriately for his or her age based on a medically determinable impairment(s).[19] It is important to remember that the cumulative physical effects of SCD and its treatment can vary in kind and intensity, affecting each child differently. The six domains of functioning are:

[17] Functional equivalence applies only to claims for children under title XVI. All claims for title II, even if the claimant is under age 18, are decided under the adult rules.
[18] See 20 CFR 416.926a, SSR 09-1p, 74 FR 7527 (2009) also available at https://www.ssa.gov/OP_Home/rulings/ssi/02/SSR2009-01-ssi-02.html, and SSR 09-2p, 74 FR 7525 (2009) also available at https://www.ssa.gov/OP_Home/rulings/ssi/02/SSR2009-02-ssi-02.html. For the complete titles of all SSRs cited in this footnote and those following, see the CROSS-REFERENCES section at the end of this SSR.
[19] See 20 CFR 416.924a(b) and 416.926a.

Acquiring and using information. Some children with SCD may have limitations in acquiring and using information due to stroke, including silent stroke.[20] A stroke can cause brain injury that impairs a child's ability to learn, concentrate, speak, and remember.

Attending and completing tasks. Frequent pain crises can result in limitations in attending and completing tasks at school and at home.[21] If a child does not feel well due to pain, it may be difficult for him or her to stay focused on activities long enough to complete them in an age-appropriate manner. A child with SCD who is experiencing pain may also have difficulty paying attention to details and may make mistakes on schoolwork due to an inability to concentrate.

Interacting and relating with others. SCD can also cause limitations interacting and relating with others.[22] The unpredictable nature of pain in SCD may cause anxiety and difficulty maintaining relationships. Children suffering from complications of SCD may become withdrawn, uncooperative, or unresponsive.

Moving about and manipulating objects. If SCD limits a child's ability to move and manipulate objects, we evaluate those effects in the domain of "Moving about and manipulating objects."[23] For example, sickling in the hip bones, knees, and ankles due to

[20] See 20 CFR 416.926a(g) and SSR 09-3p.
[21] See 20 CFR 416.926a(h) and SSR 09-4p.
[22] See 20 CFR 416.926a(i) and SSR 09-5p.
[23] See 20 CFR 416.926a(j) and SSR 09-6p.

SCD may cause joint pain and problems with walking, running, and climbing up and down stairs.

Caring for yourself. Caring for yourself involves a child's basic understanding of his or her body's normal functioning and the adequate emotional health for carrying out self-care tasks.[24] A child with SCD may avoid taking medication or ignore complications of the disease out of frustration with the limitations of SCD.

Health and physical well-being. The ongoing effects of SCD and its treatment may affect a child's health and physical well-being.[25] In this domain, we evaluate the effects of periodic exacerbations of pain crises due to sickle cell anemia. We consider the frequency and duration of the exacerbations as well as the extent to which they affect a child's ability to function physically.

This SSR is applicable on [INSERT DATE OF PUBLICATION IN THE FEDERAL REGISTER][26]

[24] See 20 CFR 416.926a(k) and SSR 09-7p.
[25] See 20 CFR 416.926a(l) and SSR 09-8p.
[26] We will use this SSR beginning on its applicable date. We will apply this SSR to new applications filed on or after the applicable date of the SSR and to claims that are pending on and after the applicable date. This means that we will use this ruling on and after its applicable date in any case in which we make a determination or decision. We expect that Federal courts will review our final decisions using the rules that were in effect at the time we issued the decisions. If a court reverses our final decision and remands a case for further administrative proceedings after the applicable date of this SSR, we will apply this SSR to the entire period at issue in the decision we make after the court's remand.

CROSS REFERENCES: SSR 86-8: Titles II and XVI: The Sequential Evaluation Process; SSR 96-3p: Titles II and XVI: Considering Allegations of Pain and Other Symptoms in Determining Whether a Medically Determinable Impairment is Severe; SSR 96-8p: Titles II and XVI: Assessing Residual Functional Capacity in Initial Claims; SSR 09-1p: Title XVI: Determining Childhood Disability Under the Functional Equivalence Rule – The "Whole Child" Approach; SSR 09-2p: Title XVI: Determining Childhood Disability – Documenting a Child's Impairment-Related Limitations; SSR 09-3p: Title XVI: Determining Childhood Disability – The Functional Equivalence Domain of "Acquiring and Using Information"; SSR 09-4p: Title XVI: Determining Childhood Disability – The Functional Equivalence Domain of "Attending and Completing Tasks"; SSR 09-5p: Title XVI: Determining Childhood Disability – The Functional Equivalence Domain of "Interacting and Relating with Others"; SSR 09-6p: Title XVI: Determining Childhood Disability – The Functional Equivalence Domain of "Moving About and Manipulating Objects"; SSR 09-7p: Title XVI: Determining Childhood Disability – The Functional Equivalence Domain of "Caring for Yourself"; SSR 09-8p: Title XVI: Determining Childhood Disability – The Functional Equivalence Domain of "Health and Physical Well-Being"; SSR 16-3p: Titles II and XVI: Evaluation of Symptoms in Disability Claims; and Program Operations Manual System (POMS) DI 22001.001, DI 22505.001, DI 22505.003, DI 24501.021, DI 24510.005, DI 25201.005, DI 25220.010, DI 25505.025, and DI 25505.030.

[FR Doc. 2017-19551 Filed: 9/14/2017 8:45 am; Publication Date: 9/15/2017]