2128: Aplastic Anemia Post-HCT

The Aplastic Anemia Post-HSCT Data Form is one of the Comprehensive Report Forms. This form captures aplastic anemia-specific post-HSCT disease assessment data for the reporting period.

This form must be completed for all recipients whose primary disease, as reported on the Pre-TED Disease Classification Form (Form 2402) as severe aplastic anemia, paroxysmal nocturnal hemoglobinuria, or one of the following inherited abnormalities of erythrocyte differentiation or function: Shwachman-Diamond syndrome, Diamond-Blackfan anemia (pure red cell aplasia), or other constitutional anemia. Fanconi Anemia and Sickle Cell Anemia each have their own forms to complete (Forms 2129 and 2130, respectively).

The Aplastic Anemia Post-HSCT Data (Form 2128) must be completed in conjunction with each Post-HSCT follow-up form completed (Forms 2100, 2200, and 2300). Form 2128 is designed to capture specific data occurring within the timeframe of each reporting period (e.g., between day 0 and day 100 for Form 2100, between day 100 and the six-month date of contact for Form 2200 Six-Month follow-up, between the date of contact for the six-month follow-up Form 2200 and the date of contact for the one-year follow up Form 2200, etc).

Q1-6: Disease Assessment at the Time of Assessment for This Reporting Period

Manual Updates:
Sections of the Forms Instruction Manual are frequently updated. The most recent updates to the manual can be found below. For additional information, select the manual section and review the updated text.

Date	Manual Section	Add/ Remove/ Modify	Description
4/7/ 2020	2128: Aplastic Anemia Post-HCT	Modify	Updated red blood cell independence instructions (question 1) to make determinations based on the date of contact rather than the date of last report. Also added guidance that **if the recipient was transfusion independent for part of the reporting period, but then is dependent again at the end of the reporting period, select "no" and continue to question 2.**
3/11/ 2020	2128: Aplastic Anemia Post-HCT	Modify	Updated the platelet transfusion independence instructions by removing (strike through) and adding (red) text as indicated below: *Indicate if the recipient was platelet transfusion independent ~~since the date of the last report~~ on the date of contact for the current reporting period. A general guideline for platelet transfusion independence is that platelet transfusions have not been required for seven or more days.* *If the recipient was platelet transfusion independent ~~since the date of the last report~~ on the date of contact, select "yes" and continue with question 5.* *If the recipient was not platelet transfusion independent ~~since the date of the last report~~ on the date of contact, select "no" and continue with question 4.*

			If the recipient is transfusion independent for a portion of the reporting period, but then is dependent again at the end of the reporting period, select "no" and continue with question 4. *If it is unknown if the recipient was platelet transfusion independent ~~since the date of the last report~~ on the date of contact, select "unknown" and continue with question 5.*
2/24/2017	Comprehensive Disease-Specific Manuals	Modify	Updated explanations of triggers for disease inserts to refer to the primary disease reported on the Pre-TED Disease Classification Form (Form 2402) instead of the Pre-TED Form (Form 2400)
9/14/2015	2028/2128: Aplastic Anemia	Modify	Updated questions number is 2028 and 2128 Aplastic Anemia Pre- and Post-HCT

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Q1-6: Disease Assessment at the Time of Assessment for This Reporting Period

Question 1: Was the recipient red blood cell (RBC) transfusion independent since the date of the last report?

Indicate if the recipient was RBC transfusion independent on the contact date of for the current reporting period. A general guideline for RBC transfusion independence is that RBC transfusions have not been required for four or more weeks.

Some discretion may be required if the recipient received a transfusion for a surgical procedure or other reason. If a recipient received an RBC transfusion for a procedure and would otherwise be transfusion independent, the recipient may still be reported as being transfusion independent.

If the recipient was RBC transfusion independent on the date of contact, select “yes” and continue with question 3.

If the recipient was not RBC transfusion independent on the date of contact, select “no” and continue with question 2.

If the recipient is transfusion independent for a portion of the reporting period, but the is dependent again at the end of the reporting period, select “no” and continue with question 2.

If it is not known if the recipient was RBC transfusion independent on the date of contact, select “unknown” and continue with question 3.

Question 2: Date of the most recent RBC transfusion:

Indicate the date of the most recent RBC transfusion.

If the recipient was RBC transfusion independent for ≥ one month, but subsequently experienced a decline in RBCs and required transfusions, record the date of the last RBC transfusion before the date of decline.

If the date reported on question 2 is more than one month prior to the date of contact, evaluate if the recipient is RBC transfusion independent.

Question 3: Was the recipient platelet transfusion independent since the date of the last report?

Indicate if the recipient was platelet transfusion independent on the date of contact for the current reporting period. A general guideline for platelet transfusion independence is that platelet transfusions have not been required for seven or more days.

If the recipient was platelet transfusion independent on the date of contact, select “yes” and continue with

question 5.

If the recipient was not platelet transfusion independent on the date of contact, select “no” and continue with question 4.

If the recipient is transfusion independent for a portion of the reporting period, but is dependent again at the end of the reporting period, select “no” and continue with question 4.

If it is unknown if the recipient was platelet transfusion independent on the date of contact, select “unknown” and continue with question 5.

If the recipient was never dependent on platelet transfusions or if this question is not applicable, select “not applicable/never dependent” and continue with question 5.

Question 4: Date of the most recent platelet transfusion:

Indicate the date of the most recent platelet transfusion.

If the recipient was platelet transfusion independent for ≥ 14 days but subsequently experienced a decline in platelets and required transfusions, record the date of the last platelet transfusion before the date of decline.

If the date reported on question 4 is more than seven days prior to the date of contact, evaluate if the recipient is platelet transfusion independent.

Questions 5-6: Specify reticulocyte level (uncorrected):

Indicate whether the uncorrected reticulocyte count in the blood is “known” or “not known/transfused” since the date of last report. If the reticulocyte count was assessed multiple times during the reporting period, report the results of the latest reticulocyte count. If “known,” report the value documented on the laboratory report.

If the recipient had an RBC transfusion within 30 days prior to the latest reticulocyte count, select “not known/transfused” and do not report a value.

Report the absolute value of reticulocytes in __ x 10^9/L. Do not report a percentage, the corrected reticulocyte count, or the reticulocyte production index.

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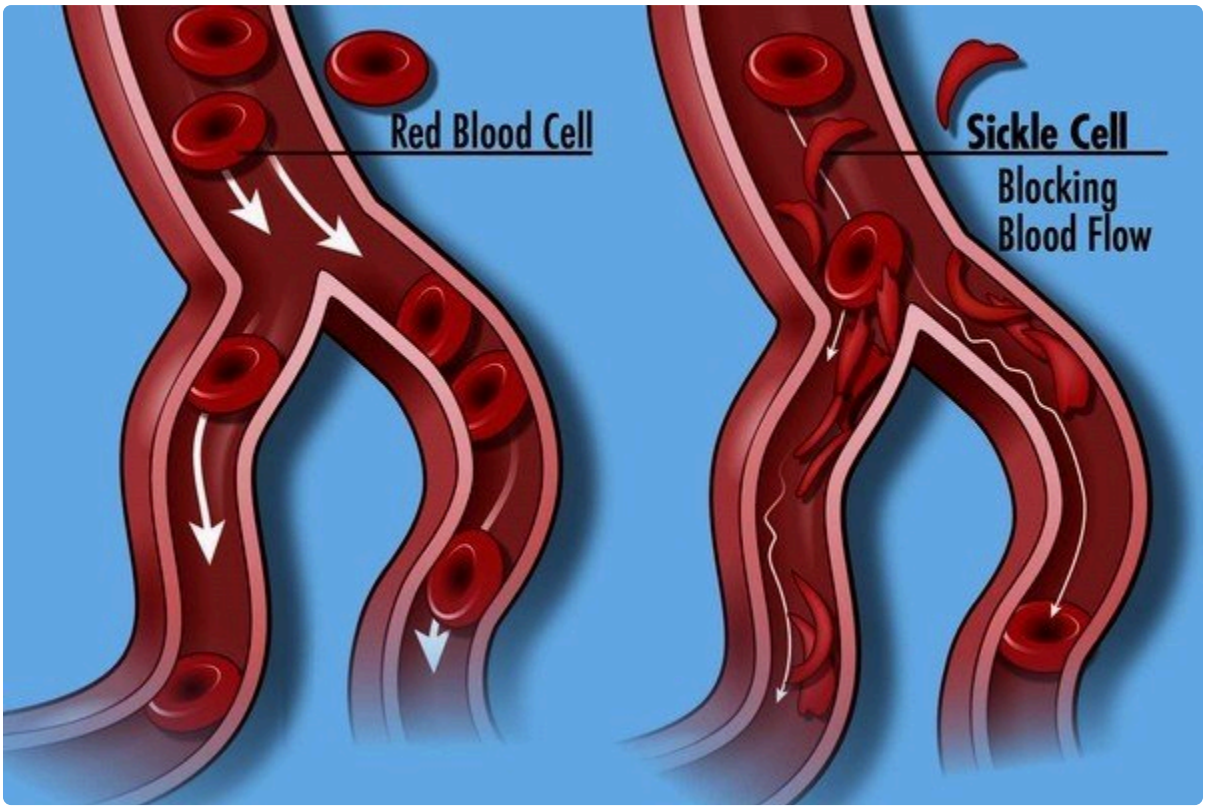
2030/2130: Sickle Cell Disease (SCD)

Sickle Cell Disease (SCD) is a group of disorders that adversely affect the body's production of hemoglobin, the component in red blood cells that delivers oxygen throughout the body.[1] Individuals with these disorders possess atypical hemoglobin molecules, called hemoglobin "S," which can distort the red blood cell morphology into a sickle, or crescent, shape. Multiple variants of the hemoglobin S molecule exist, and each genetic sub-type is characterized by different prognostic indications.

Graphic 1. Depiction of a Sickle Cell



Sickle cell disease (SCD) is often diagnosed at birth following newborn screening or following severe and / or persistent infections. Symptoms of sickle cell disorders include anemia (low red blood cell counts) as a result of the sickle cells dying prematurely, repeat infections, and periodic episodes of pain (i.e., pain crisis). The severity of symptoms varies from person to person ranging from mild to severe. Refer to *Table 1. Sickle Cell Disease Symptoms* for additional symptoms associated with SCD. Hematopoietic cell transplant (HCT) is currently the major curative treatment for Sickle Cell Disease (SCD).

Table 1. Sickle Cell Disease Symptoms[2]

Common SCD Symptoms
100% of people have these symptoms:
Chronic hemolytic anemia

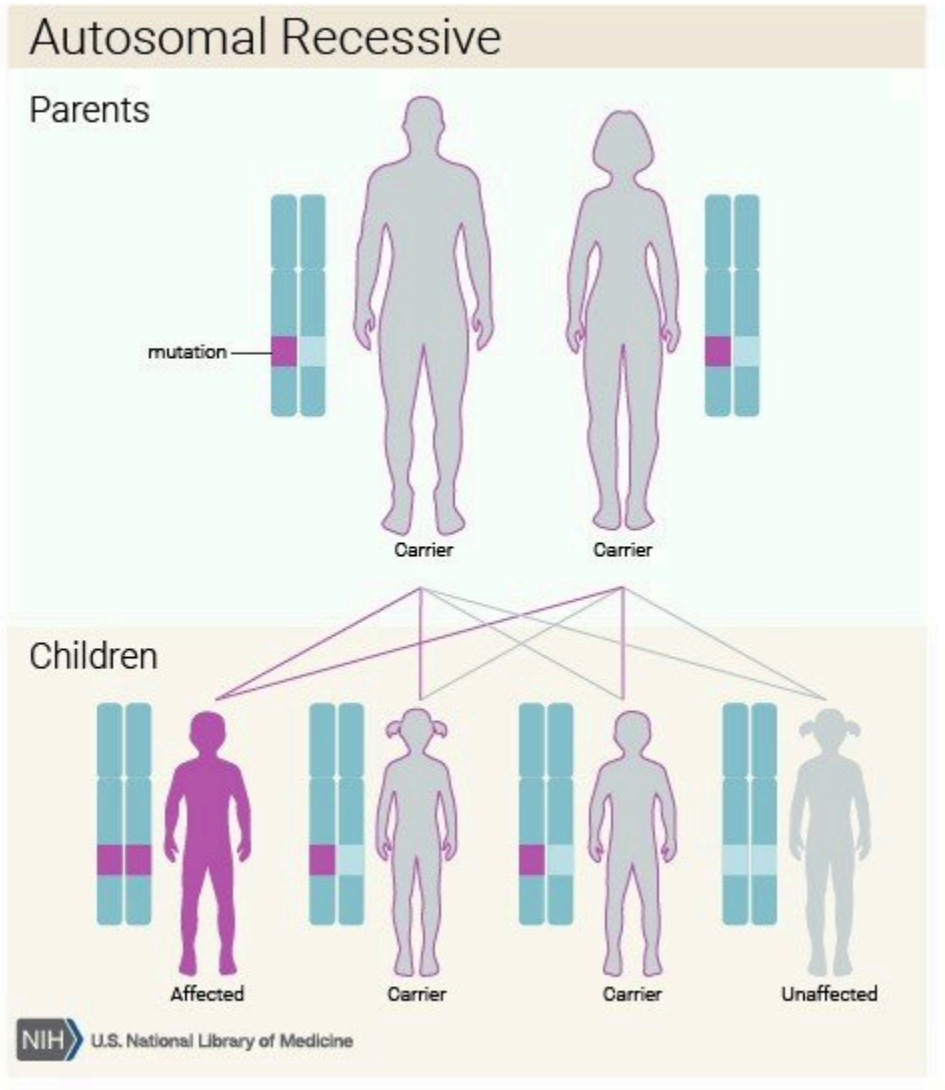
80 – 99% of people have these symptoms:
Recurrent infections
30 – 79% of people have these symptoms:
Abnormality of the spleen
Avascular necrosis
Chest pain
Iron deficiency
Leukocytosis
Osteomyelitis
Osteoporosis
Pigment gallstones
Reticulocytosis
Thrombocytosis
5 – 29% of people have these symptoms:
Abnormality of the nervous system
Cholestasis
Elevated serum creatinine
Hypoxemia
Increased LDH activity
Persistence of hemoglobin F
Unconjugated hyperbilirubinemia
1 – 4% of people have these symptoms:
Increased mean corpuscular volume
Microcytic anemia
Additional symptoms that recipients may experience:
Abdominal pain
Cardiomegaly
Hematuria
Hepatomegaly
Hypertension
Jaundice
Priapism
Renal insufficiency

Retinopathy
Splenomegaly
Stroke

The table above contains information collected from the Human Phenotype Ontology (HPO) database.

Sickle cell disease is inherited in an autosomal, recessive pattern. A person who carries one copy of the mutated gene is said to be a carrier of the sickle cell trait but will not exhibit symptoms of sickle cell disease. If two people are both carriers for the sickle cell trait and have a child, there is a 25% chance that this child will have SCD, a 50% chance of being a carrier, and a 25% chance that the child will not be a carrier nor have SCD. Reference *Graphic 2. Inheritance of Sickle Cell Disease* below for a depiction of this description:

Graphic 2. Inheritance of Sickle Cell Disease



- 2030: Sickle Cell Disease (SCD) Pre-HCT
- 2130: Sickle Cell Disease (SCD) Post-HCT

[1] Genetics Home Reference: Your Guide to Understanding Genetic Conditions. 2020. *Sickle Cell Disease*. [online] Available at: < https://ghr.nlm.nih.gov/condition/sickle-cell-disease > .

[2] Genetics Home Reference: Your Guide to Understanding Genetic Conditions. 2020. *Sickle Cell Disease*. [online] Available at: < https://ghr.nlm.nih.gov/condition/sickle-cell-disease >.

Last modified: Jul 26, 2020