2130: SCD Post-Infusion

The Sickle Cell Disease Post-Infusion (2130) Form is one of the Comprehensive Report Forms. This form captures Sickle Cell Disease (SCD) post-HCT data for the reporting period.

This form must be completed for all recipients whose primary disease, as reported on the Disease Classification (2402) Form, is **Sickle Cell Disease (SCD)**. The Sickle Cell Disease Post-Infusion (2130) Form must be completed in conjunction with each Post-Infusion Follow-up (2100) Form. This form is designed to capture specific data occurring within the timeframe of each reporting period (i.e., between day 0 and day 100; between day 100 and the six-month date of contact for six-month follow-up; and between the date of contact for the six-month follow-up and the date of contact for the one-year follow-up, etc.).

Links to Sections of the Form

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.

If you need to reference the historical Manual Change History for this form, please click here or reference the retired manual section on the Retired Forms Manuals webpage.

Date	Manual Section	Add/ Remove/ Modify	Description
1/24/	2130	Modify	Abdominal Girth red warning box above Q2 updated: p(banner important).

2025	SCD Post-Infusion		***Abdominal girth ~~and Blood Pressure~~**: Abdominal girth ~~and blood pressure are~~ is currently disabled and cannot be answered at this time.*
10/25/2024	2130 SCD Post-Infusion	Add	Version 2 of the 2130: Sickle Cell Disease (SCD) Post-Infusion Data section of the Forms Instruction Manual released. Version 2 corresponds to revision 4 of the Form 2130.

Last modified: Jan 27, 2025

Q1 – 5: Physical Assessments

Date of Last Report Definition
If completing this form for the 100-day reporting period, "date of last report" should be interpreted as "day 0 (i.e., infusion)."

Question 1: Date of evaluation

Report the date of the most recent physical evaluation where both the abdominal girth and blood pressure were assessed in the reporting period. This evaluation will be used to answer *Abdominal girth* and *Blood pressure*.

Abdominal girth
Abdominal girth is currently disabled and cannot be answered at this time.

Questions 2 – 3: Abdominal girth

Indicate if the recipient's abdominal girth was measured on the date of evaluation specified in the question above. If **Known**, specify the recipient's abdominal measurement in centimeters. If the recipient's abdominal girth was not measured or if no information is available, select **Unknown**.

Questions 4 – 5: Blood pressure

Indicate if the recipient's blood pressure was assessed on the date of evaluation specified above. If assessed, report **Known** and provide the recipient's blood pressure. If the recipient's blood pressure was not measured or if no information is available to determine if the recipient's blood pressure was measured, select **Unknown**.

Section Updates:

Question Number	Date of Change	Add/ Remove/ Modify	Description	Reasoning (If applicable)
Q2	1/24/ 2025	Modify	Abdominal Girth red warning box above Q2 updated: p(banner important). ***Abdominal girth ~~and Blood Pressure~~***: *Abdominal girth ~~and blood pressure are~~ is currently disabled and cannot be answered at this time.*	Due to change in form validation

Last modified: Jan 28, 2025

Q6 – 8: Transfusion Therapy

Questions 6 – 7: Were red blood cell transfusions administered since the date of last report?

Red blood cell (RBC) transfusions are often given as supportive care to lower hemoglobin S for recipients with Sickle Cell Disease (SCD).

Indicate if the recipient received RBC transfusion(s) since the date of last report. If RBCs were transfused, select **Yes** and report the date of the most recent transfusion administered in the reporting period. If the recipient did not receive RBC transfusions since the date of last report, select **No**.

If the last administration date is partially known (i.e., the recipient received their last RBC transfusion in mid-July 2019), use the process described for reporting partial or unknown dates in General Instructions, Guidelines for Completing Forms.

Question 8: Was the transfusion for a sickle cell related event?

Red blood cell transfusions may be given to prevent sickle cell-related events.

Indicate **Yes** or **No** if any of the RBC transfusion(s) administered during the reporting period were given for a sickle cell related event.

Section Updates:

Question Number	Date of Change	Add/Remove/Modify	Description	Reasoning (If applicable)
.	.	.	.	.

Last modified: Oct 27, 2024

Q9 – 22: Therapy for Iron Overload

Questions 9 – 10: Serum ferritin

Ferritin is a protein that stores, transports, and release iron. Iron is toxic to cells, so it is stored within the ferritin protein for use. Ferritin that is too low might be indicative of iron deficiency related anemia. Ferritin that is too high might be indicative of iron overload.

Indicate if the serum ferritin is known. If **Known**, report the value in ng / mL (µg / L). If the serum ferritin was assessed multiple times, report the most recent results.

Question 11: Was iron chelation therapy given?

Iron chelation therapy is used to prevent or reduce iron overload. Examples include Deferoxamine (Desferal) and Deferasirox (Jadenu, Exjade).

Indicate if the recipient was on iron chelation therapy in the current reporting period. If the recipient did not receive iron chelation therapy in the reporting period or it is not known, report **No** or **Unknown**, respectively.

Question 12: Was iron chelation previously reported?

Specify if the iron chelation start date was previously reported. If iron chelation was started in prior reporting period and continued into the current, report **Yes** and continue with *Is iron chelation ongoing?*.

The **Yes** option is not applicable for the Day 100 reporting period.

Questions 13 – 14: Date started

Indicate if the iron chelation start date is known. If Known, report the date when this therapy began.

If the exact date is not known report an estimated date and check the **Date estimated** box. Refer to General Instructions, Guidelines for Completing Forms for information about reporting estimated dates.

Questions 15 – 16: Is iron chelation ongoing?

Indicate if the recipient is still receiving iron chelation therapy on the contact date. If **Yes**, continue with *Was phlebotomy performed?*.

If **No**, report the date when the recipient received the last dose of iron chelation in the reporting period.

If the exact date is not known report an estimated date and check the **Date estimated** box. Refer to General Instructions, Guidelines for Completing Forms for information about reporting estimated dates.

Question 17: Was phlebotomy performed?

Phlebotomy is a procedure in which blood is removed from the body with the goal of reducing iron overload.

Phlebotomy therapy is typically a series of these procedures.

Indicate if phlebotomy was performed in the current reporting period. If phlebotomy was not performed in the reporting period or it is not known if performed, select **No** and continue with *Were pulmonary function tests (PFTs) performed?*.

Question 18: Was phlebotomy previously reported?

Specify if the phlebotomy start date was previously reported (i.e., the first phlebotomy of the series was started in a prior reporting period). If **Yes**, continue with Is phlebotomy ongoing.

The **Yes** option is not applicable for the Day 100 reporting period.

Questions 19 – 20: Date started

Indicate if the phlebotomy start date is known. If **Known**, report the date when this therapy began.

If the exact date is not known report an estimated date and check the **Date estimated** box. Refer to General Instructions, Guidelines for Completing Forms for information about reporting estimated dates.

Questions 21 – 22: Is phlebotomy ongoing?

Indicate if phlebotomy is ongoing at the time of the contact date for the current reporting period. If **Yes**, continue with Splenic Assessments.

If **No**, report the date when phlebotomy ended in the reporting period (i.e., the last phlebotomy of the series).

If the exact date is not known report an estimated date and check the **Date estimated** box. Refer to General Instructions, Guidelines for Completing Forms for information about reporting estimated dates.

Section Updates:

Question Number	Date of Change	Add/Remove/Modify	Description	Reasoning (If applicable)
.	.	.	.	.

Last modified: Oct 27, 2024

Q23 – 27: Pulmonary Assessments

Questions 23 – 24: Were pulmonary function tests (PFTs) performed? (if PFTs tests were performed, attach the most recent report)

Indicate if pulmonary function tests (PFTs) were performed since the date of last report. If pulmonary function tests were performed in the current reporting period, report **Yes** and indicate if a PFT report is attached. If multiple PFTs were performed, attach the most recent report.

If pulmonary function tests were not performed since the date of last report, select **No**.

For further instructions on how to attach documents in FormsNet3^{SM}, refer to the Training Guide.

Question 25: For children unable to perform a PFT, was oxygen saturation on room air > 95%? *(only required to answer if children are 5 years of age or less) (if multiple, report the most recent)*

Indicate if oxygen saturation on room air was > 95% since the date of last report. If oxygen saturation is > 95% in the current reporting period, report **Yes**. If the oxygen saturation of room air was ≤ 95%, report **No**. If the oxygen saturation was assessed multiple times in the reporting period, report the most recent assessment.

If oxygen saturation was not tested, report **Not tested**.

This question is only enabled for recipients ≤ 5 years of age.

Questions 26 – 27: Was a 6-minute walk test performed?

A 6-minute walk test is used to assess total distance walked within 6 minutes to determine aerobic capacity and endurance. Indicate if a 6-minute walk test was performed since the date of last report. If **Yes**, report the total distance walked and specify the unit of measure. If multiple walk tests were performed during the reporting period, report the results of the most recent assessment.

If a 6-minute walk test was not performed, recipient was unable to walk, or cannot perform the 6-minute walk test due to their current clinical status, report **No**.

Section Updates:

Question Number	Date of Change	Add/Remove/Modify	Description	Reasoning (If applicable)
.	.	.	.	.

Last modified: Oct 27, 2024

Q28 – 48: Cardiovascular Assessments

Question 28: Were lipid profiles assessed?

A lipid profile is a blood test that measures the amount of cholesterol and triglycerides within the blood. Indicate if lipid profiles were assessed since the date of last report. If lipid profiles were assessed in the current reporting period, report **Yes**. If lipid profiles were not assessed or if no information is available to determine if lipid profiles were evaluated during the current reporting period, report **No** or **Unknown**, respectively.

Questions 29 – 33: Specify which lipids were assessed (check all that apply)

Indicate which lipids were assessed and report the value in mg / dL. Select all that apply. If multiple lipid profile assessments were performed, report the results of the most recent assessment.

Question 34: Was an echocardiogram performed?

Indicate if an echocardiogram was performed since the date of last report. If an echocardiogram was performed during the current reporting period, report **Yes**. If an echocardiogram was not performed or if no information is available to determine if an echocardiogram was performed, select **No** or **Unknown**, respectively.

Questions 35 – 36: Was tricuspid regurgitant jet velocity (TRJV) measured?

Tricuspid regurgitant jet velocity (TRJV) measurements are used in determining the pulmonary artery pressure for recipients with sickle cell and other hemolytic disorders. An elevated TRJV is an indication of pulmonary hypertension, a condition common in adults with hemolytic diseases. TRJV is typically documented in the echocardiogram report.

Report **Yes** if TRJV was measured in the current reporting period and provide the TRJV value as documented on the echo report. If the TRJV was measured multiple times in the reporting period, report the most recent value. Report **No** if TRJV was not assessed or is not documented on the echo report.

Questions 37 – 39: Was left ventricular ejection fraction (LVEF) or left ventricular shortening fraction reported?

The left ventricular ejection fraction (LVEF) is a percentage that represents the volume of blood pumped from the left ventricle into the aorta (also known as stroke volume) compared to the volume of blood in the ventricle just prior to the heart contraction (also known as end diastolic volume). The left ventricular shortening fraction is the percentage change in cavity dimensions of the left ventricle with systolic contraction.

Report **Yes** if *either* the LVEF or left ventricular shortening fraction were assessed in the current reporting period and provide the percentage(s). If the LVEF or left ventricular shortening fraction were assessed multiple times in the reporting period, report the most recent value(s). Report **No** if the LVEF and left ventricular shortening fraction were not assessed in the current reporting period.

Questions 40 – 41: Is there a new onset of pulmonary hypertension?

Pulmonary hypertension (PH) refers to elevated pulmonary arterial pressure and is diagnosed either by an echocardiogram or right heart catherization. PH can be due to a primary elevation of pressure in the pulmonary arterial system alone (pulmonary arterial hypertension) or secondary to elevations of pressure in the pulmonary venous and pulmonary capillary systems (pulmonary venous hypertension; post-capillary PH).

Indicate **Yes** if there was a new onset of pulmonary hypertension (has never been previously diagnosed) since the date of last report and specify if either an echocardiogram or right heart catherization was used to diagnose PH.

Report **No** in in the following scenarios:

- There was not a new onset of PH in the current reporting period.
- PH was diagnosed prior to infusion or in a prior reporting period, resolved, and developed in the current reporting period.
- PH was diagnosed in a prior reporting period and persisted into the current reporting period.

If documentation is not clear or there is not enough information available to determine if PH was present, report **Unknown**.

Question 42: Is a copy of the echocardiogram report attached?

Indicate whether an echocardiogram report for either LVEF or pulmonary hypertension is attached to this form. For further instructions on how to attach documents in FormsNet3^{SM}, refer to the Training Guide.

Questions 43 – 44: Was cardiac MRI performed?

Cardiac MRI is a noninvasive test that uses a magnetic field and radiofrequency waves to create detailed pictures of your heart and arteries. Specify if a cardiac MRI was completed in the current reporting period. If **Yes**, indicate if abnormal iron deposition was found based on the MRI of the heart.

Questions 45 – 46: Cardiac iron T2 imaging?

Indicate if cardiac iron T2* imaging is known during the current reporting period. If **Known**, specify the value and units of measurement. If the cardiac iron T2* imaging was done multiple times, report the most recent assessment in the reporting period.

Questions 47 – 48: Was brain natriuretic peptide (BNP) assessed?

Brain natriuretic peptide (BNP) is a hormone secreted by cardiac ventricle cells in response to increased ventricular blood volume. BNP is typically measured using various immunoassay techniques.

Indicate if the BNP was assessed during the current reporting period. If **Yes**, report the value as documented on the laboratory report (in pg / mL). If BNP was assessed multiple times, report the results of

the most recent test. If BNP was not assessed or if no information is available to determine if BNP was tested, report **No** or **Unknown**, respectively.

Section Updates:

Question Number	Date of Change	Add/Remove/Modify	Description	Reasoning (If applicable)
.	.	.	.	.

Last modified: Oct 27, 2024

Q49 – 58: Hepatic Assessments

Questions 49 – 51: Was liver iron content (LIC) tested?

Transfusions for hemolytic diseases may lead to iron build up or accumulation in the liver and other target organs. Liver iron content (LIC) is commonly used to measure total iron storage. Methods of assessment include, but are not limited to, liver biopsy, T2 MRI, FerriScan, and SQUID biomagnetometer.

Indicate if the LIC was assessed during the current reporting period. If **Yes**, report the value, units of measurement, and specify the method of assessment. If the LIC was assessed multiple times, report the most recent results.

Questions 52 – 58: Laboratory studies performed (check all that apply)

Specify if any of the laboratory values listed below were assessed in the reporting period. If the assessment was completed multiple times, report the most recent value.

If none of the laboratory values were assessed in the reporting period, leave the data fields blank and override the FormsNet3^{SM} error.

- **Total serum bilirubin**: Bilirubin is a pigment that is formed from the breakdown of hemoglobin in red blood cells. Serum bilirubin is a test of liver function that reflects the ability of the liver to take up, process, and secrete bilirubin. Total bilirubin includes the *direct* (conjugated) and *indirect* (unconjugated) bilirubin values. If your laboratory reports direct and indirect separately, add the two together to report the total serum bilirubin. Specify the most recent value since the date of the last report, and the upper limit of normal for your institution.
- **Direct bilirubin**: also known as conjugated bilirubin. Specify the most recent value since the date of the last report, and the upper limit of normal for your institution.
- **Lactate dehydrogenase (LDH)**: an enzyme found in the cytoplasm of almost all tissues, which converts L-lactate into pyruvate, or pyruvate into L-lactate depending on the oxygen level. Specify the most recent value since the date of the last report, and the upper limit of normal for your institution.

Section Updates:

Question Number	Date of Change	Add/Remove/Modify	Description	Reasoning (If applicable)
.	.	.	.	.

Last modified: Oct 27, 2024

Q59 – 65: Renal Assessments

Questions 59 – 60: Urine albumin

Indicate whether urine albumin was measured in the current reporting period. If measured, select **Known** and report the laboratory value and unit of measure documented on the laboratory report. If urine albumin was assessed multiple times within the reporting period, report the most recent results. If urine albumin was not measured or if no information is available to determine if urine albumin was measured in the current reporting period, select **Unknown**.

If the urine albumin is < 30 mg / g, report the urine albumin value as “29 mg / g” (29,000 mg / kg).

Question 61: Serum creatinine *(if multiple, report the most recent tested)*

Report the laboratory value and unit of measurement as documented on the laboratory report in the current reporting period. If serum creatinine was measured multiple times in the reporting period, report the most recent results.

If serum creatinine was not measured in the current reporting period, leave the data field blank and override the FormsNet3^{SM} error,

Questions 62 – 63: Glomerular filtration rate (GFR)

The glomerular filtration rate (GFR) estimates how much blood passes through the glomeruli each minute and is used to check how well the kidneys are working.

Indicate whether the GFR was measured in the current reporting period reporting period. If measured, select **Known** and report the laboratory value and unit of measure documented on the laboratory report. If the GFR was measured multiple times in the reporting period, report the most recent results. If the GFR was not measured or if no information is available to determine if the GFR was assessed in the current reporting period, select **Unknown**.

GFR may be reported to the CIBMTR as “actual” or “calculated.” If your center’s laboratory does not calculate the actual GFR value, the following equations may be used to determine the calculated value.

Cockcroft-Gault Equation
GFR = [(140 – age) x Wt] / (72 x Cr)

- GFR_cg = Glomerular Filtration Rate (Cockcroft) (mL / min)
- Age = Patient Age (years)
- Sex = Gender (Male)
    - If female, multiply result by 0.85
- Wt = Body Weight (kg)
- Cr = Creatinine (S, mg / dL)

An online calculator for this equation can be found here: https://www.kidney.org/professionals/kdoqi/gfr_calculatorCoc

Bedside Schwartz Equation
eGFR = K x H / Cr

- H = Height (cm)
- Cr = Creatinine (S, mg / dL)
- K =
    - 0.33 for preemies
    - 0.45 for infants to 1 year
    - 0.55 for 1 year to 18 years for females
    - 0.55 for 1 year to 13 years for males
    - 0.70 for adolescent to 18 years for males

An online calculator for this equation can be found here: https://www.kidney.org/professionals/KDOQI/gfr_calculatorPed

If the laboratory report indicates the GFR as a range, report the average. Example, the laboratory report indicates GFR is 80 – 120, report “100.”

If the GFR value is expressed as “> X,” report the value as “X+1.” Example, the laboratory report indicates the GFR is > 120, report “121.”

If the GFR value is reported as “< X”, report “X-1.” Example, the laboratory report indicates the GFR is < 80, report “79.”

Question 64 – 65: Cystatin-C

Cystatin-C is a protein in the blood that can be used to evaluate kidney function. If measured, select **Known** and report the laboratory value . If testing was performed multiple times, report the most recent laboratory value obtained. If the cystatin-C was not measured or if no information is available to determine if the cystatin-C was assessed, report **Unknown**.

Section Updates:

Question Number	Date of Change	Add/Remove/Modify	Description	Reasoning (If applicable)
.	.	.	.	.

Last modified: Oct 27, 2024

Q66 – 70: Splenic Assessments

Splenic Assessments
The following section will only be applicable for the 100-day, 6-month, 1 year, and 2-year follow-up periods. The section will be disabled for annual reporting > 2 years.

Question 66: Was splenic function assessed?

Evaluation of splenic tissue may be necessary to determine the extent of the recipient's disease. Splenic assessments include the following:

- **Complete RBC**: Also called an erythrocyte count, this assessment is used to determine how many red blood cells are present. One of the several tests included in a CBC.
- **Pitted RBC score**: As red blood cells (RBC) age, membrane vacuoles ("pits") occur. Pitted RBCs may be listed on laboratory reports as a "PIT count."
- **Splenic scan**: Radionuclide spleen scans (liver-spleen scans) are nuclear scan utilizing a radioactive tracer substance that is administered intravenously. The tracer collects in large amounts and shows as bright spots on imaging results. One of the most common methods used is single photon emission computed tomography (SPECT / CT).

Indicate **Yes** or **No** if splenic function was assessed by a complete RBC, pitted RBC score, or splenic scan at any time during the current reporting period.

If no information is available to determine if splenic function was assessed during the current reporting period, select **Unknown**. This option should be used sparingly.

Report **Not applicable** if the recipient had a prior splenectomy or was born without a spleen (congenital asplenia).

Questions 67 – 70: Select which splenic test was completed

Indicate which splenic test (completed RBC, pitted RBC score, or splenic scan) was completed.

If a complete RBC was performed, specify the value (in cells / µL).

If a pitted RBC score was performed, specify the percentage.

If a splenic scan was performed, specify the results as either **Normal (radionuclide uptake)** or **Abnormal (no radionuclide uptake)**.

If any of the assessments were performed multiple times in the reporting period, report the most recent results.

Section Updates:

Question Number	Date of Change	Add/Remove/Modify	Description	Reasoning (If applicable)
.	.	.	.	.

Last modified: Oct 27, 2024

Q71 – 74: Acute Chest Syndrome

Question 71: New onset of acute chest syndrome

Acute chest syndrome (ACS) is a term used to identify symptoms of chest pain, cough, fever, decreased oxygen (hypoxia), and lung infiltrates. Due to the sickling nature of red blood cells as a result of sickle cell disease, ACS may result in pulmonary infarction / emboli or viral / bacterial pneumonia. Diagnoses should be made based on clinical judgement.

Report **Yes** in the following scenarios:

- There was a new onset of ACS since the date of last report.
- ACS was diagnosed prior to HCT or in a prior reporting period, resolved, and developed in the current reporting period.

Report **No** in the following scenarios:

- There was not a new onset of ACS since the date of last report.
- ACS was diagnosed in a prior reporting period and persisted into the current reporting period.

If documentation is not clear or is unavailable to determine if ACS was present, report **Unknown**.

Question 72: Number of events

Specify the number of events that occurred in this reporting period. An ‘event’ is defined as having clinical symptoms requiring intervention (i.e., antibiotics, steroids, etc.) followed by resolution of symptoms and discontinuation of intervention. This should not be interpreted as the number of days the recipient was receiving treatment or had active symptoms. An event may also be referred to as an ‘episode.’

Question 73: Date of last event

Report the date of the last acute chest syndrome event requiring hospitalization or treatment in the current reporting period.

Question 74: Did any events require hospitalization?

Indicate if *any* of the acute chest syndrome events that occurred in this reporting period required hospitalization. Acute chest syndrome must be the primary indication for hospitalization.

Section Updates:

Question Number	Date of Change	Add/Remove/Modify	Description	Reasoning (If applicable)
.	.	.	.	.

Last modified: Oct 27, 2024

Q75 – 80: Pain

Question 75: Has vaso-occlusive pain occurred requiring hospitalization or treatment? *(treatment that is in a hospital or clinic setting since the date of last report)*

Vaso-occlusive pain, sometimes called a pain crisis, is a common painful complication of sickle cell disease in adolescents and adults. Recurrent episodes may cause irreversible organ damage.

Report **Yes** in the following scenarios:

- The recipient experienced vaso-occlusive pain requiring hospitalization or treatment (i.e., ER admission, day hospital, inpatient admission, etc.) in the current reporting period.
- The recipient experienced vaso-occlusive pain requiring hospitalization or treatment prior to HCT or in a prior reporting period, resolved, and developed in the current reporting period.

Report **No** in any of the following scenarios:

- The recipient did not experience vaso-occlusive pain the current reporting period
- Vaso-occlusive pain occurred in the reporting period, but hospitalization or treatment was not required
- The recipient was hospitalized or received treatment in a prior reporting period for vaso-occlusive pain and continued hospitalization or treatment into the current reporting period

If no information is available to determine if the recipient required hospitalization or treatment for vaso-occlusive pain since the date of last report, select **Unknown**.

Question 76: Number of events

Report the number of vaso-occlusive pain events requiring hospitalization or treatment within the past two years. Report the date of the last event and if any of the events required hospitalization. In this context, an ‘event’ is defined as being admitted to a hospital setting (i.e., ER admission, day hospital, inpatient admission, etc.) followed by a discharge. This should not be interpreted as the number of days for which the recipient was hospitalized.

Question 77: Date of last event

Report the date of the last vaso-occlusive pain event requiring hospitalization or treatment in the current reporting period.

Question 78: Did any events require hospitalization?

Specify if any of the vaso-occlusive pain events occurring in the current reporting period required hospitalization.

Question 79: Has there been any new onset of chronic pain?

Indicate **Yes** or **No** if the recipient experienced a new onset of chronic pain in the current reporting. Chronic

pain is defined as pain that is present a majority of days per month, lasting for ≥ 6 months with one more of the following:

- Without contributory SCD complications (acute vaso-occlusive pain, acute chest syndrome, leg ulcers, avascular necrosis; or,
- Mixed pain type in which chronic pain is occurring at site(s) (arms, back, chest, or abdominal pain) but unrelated to any sites associated with SCD complications

The progress notes should clearly document if chronic pain is present or absent. If is not clear, based on the documentation available, whether chronic pain is present or absent, report **Unknown**.

Chronic Pain Prior to Infusion
For recipients with chronic pain prior to infusion, chronic pain may be reported in the Day 100 reporting period. However, for recipients who did not have chronic pain prior to infusion, the earliest reporting period in which chronic pain should be reported is the 6-month reporting period.

Question 80: Were opioids prescribed for the treatment of pain?

Opioids are typically listed in the medical administration record (MAR) and/or in the medication list documented on the progress note within the medical record. Examples of opioid treatment may include Hydrocodone, Methadone, Hydromorphone (Dilaudid), etc.

Indicate if the recipient was prescribed opioids for treatment of either vaso-occlusive or chronic pain during this reporting period. If opioids were prescribed for treatment of pain, report **Yes**. If opioids were not prescribed for treatment of pain or if no information is available to determine if opioids were prescribed for treatment of pain, report **No**.

Section Updates:

Question Number	Date of Change	Add/Remove/Modify	Description	Reasoning (If applicable)
.	.	.	.	.

Last modified: Oct 27, 2024

Q81 – 83: Avascular Necrosis

Question 81: Is there a new area affected by avascular necrosis?

Avascular necrosis is the death of bone tissue due to a lack of adequate blood supply. It is sometimes called osteonecrosis and can lead to minute fractures in the bone followed by eventual collapse. Avascular necrosis can spread from joint to joint.

Indicate if there was a new onset of avascular necrosis in the current reporting period.

Report **Yes** in the following scenarios:

- A new onset of avascular necrosis occurred since the date of last report
- There is a prior diagnosis of avascular necrosis of a joint and a new joint(s) was diagnosed with avascular necrosis since the date of last report (see example below)

Report **No** in the following scenarios:

- If a new onset of avascular necrosis did not occur in the current reporting period
- Avascular necrosis of a specific joint occurred in a prior reporting period and persisted into the current reporting period

If no information is available to determine if a new onset of avascular necrosis was experienced since the date of last report, **Unknown**.

See examples of reporting scenarios listed below.

Questions 82 – 83: Specify joint(s) affected (check all that apply)

Specify the joint(s) affected by the new onset of avascular necrosis; check all that apply. If there was avascular necrosis of a joint not listed, report **Other** and specify the joint.

Avascular Necrosis Reporting Scenarios:
Example A. A recipient is diagnosed with avascular necrosis of the hip pre-transplant. Following infusion, the recipient develops avascular necrosis of the left shoulder.

- 100 Day Post-Infusion Data Form
  - *Is there a new area affected by avascular necrosis?*: Report **Yes** to capture the new development of avascular necrosis of the left shoulder.
  - *Specify joint(s) affected*: Select **Shoulder** and continue with the form. “Hip” will not be selected as this was diagnosed pre-transplant.

Example B. A recipient with diagnosed with avascular necrosis of the right knee and resolves in the Day 100 reporting period. In the six-month reporting period, the recipient does not experience avascular necrosis but in the one-year reporting period, the recipient is diagnosed with avascular necrosis of both the right and left

knees.

- Day 100 Post-Infusion Data Form
  - *Is there a new area affected by avascular necrosis?*: Report **Yes** to indicate the diagnosis of avascular necrosis of the right knee.
  - *Specify joint(s) affected*: Select **Knee**.

- Six Month Post-Infusion Data Form
  - *Is there a new area affected by avascular necrosis?*: Report **No** as there was not a new onset of avascular necrosis during the reporting period.

- One Year Post-Infusion Data Form
  - *Is there a new area affected by avascular necrosis?*: Report **Yes** to capture the onset of avascular necrosis of the left knee.
  - *Specify joint(s) affected*: Select **Knee** again.

Section Updates:

Question Number	Date of Change	Add/Remove/Modify	Description	Reasoning (If applicable)
.	.	.	.	.

Last modified: Oct 27, 2024

Q84 – 92: Central Nervous System

Questions 84 – 86: Was transcranial doppler velocity assessed at any time?

Transcranial doppler and transcranial color doppler are types of ultrasonography that measure the velocity of blood flow through the brain's blood vessels by measuring the echoes of ultrasound waves moving transcranially.

Indicate if transcranial doppler velocity was assessed at any time in the current reporting period, independent of any CNS event that occurred. If **Yes**, report the date of the most recent assessment and the transcranial doppler velocity value in cm / sec. If transcranial doppler velocity was not assessed or if no information is available to determine if transcranial doppler velocity was assessed, report No or **Unknown**.

Question 87: Have any new central nervous system (CNS) complications occurred?

A CNS complication is the development of different neurologic signs and symptoms that occur in recipients with sickle cell disease. See *Specify type of CNS complication* for a list of CNS events.

Report **Yes** if the recipient experienced a new manifestation of a CNS complication(s) since the date of last report.

Report **No** in the following scenarios:

- A new CNS complication did not occur in the current reporting period.
- The recipient experienced a CNS complication in a prior reporting period and follow-up scans are performed in the current reporting period.

If no information is available to determine if a new CNS complication occurred since the date of last report, report **Unknown**.

> *Multiple CNS Complications *
> For each CNS complication the recipient experienced in the reporting period, complete questions *Specify type of CNS complication*, *Year of onset*, and *Was an MRI / MRA of the brain performed for the diagnosis of this reported CNS complication?* for each complication in the reporting period by adding an additional instance in the FormsNet3^{SM} application.

Question 88: Specify type of CNS complication

Indicate the type of CNS complication(s) that occurred. If multiple CNS complications occurred, report each CNS complication as a separate instance. If the same CNS complication occurred multiple times throughout the reporting period, only one instance needs to be reported.

- **Cerebral venous thrombosis:** A blood clot in the cerebral vein in the brain.
- **Hemorrhagic stroke:** Blood vessels break and bleeds in the brain.

- **Ischemic stroke:** The most common type of stroke. Occurs when a blood vessel in the brain is blocked or narrowed, causing reduced blood flow.
- **Moyamoya:** A rare condition where the carotid artery is blocked or narrowed which reduces the blood flow to the brain.
- **Overt stroke:** A focal neurologic deficit lasting more than 24 hours. If the type of CNS event is not documented and only noted as a "stroke," select this option.
- **Posterior reversible encephalopathy syndrome (PRES)**: An acute neurotoxic syndrome that is characterized by a spectrum neurological and radiological feature from various risk factors. Common neurological symptoms includes headache, impairment in level of consciousness, seizures, visual disturbances, and focal neurological deficits
- **Seizure:** Uncontrolled electrical activity in the brain, which may produce a physical convulsion, minors physical sings, thought disturbances or a combination of symptoms.
- **Silent stroke:** Asymptomatic stroke.
- **Transient ischemic stroke:** A temporary period of mild stroke symptoms that lasts only a few minutes and does not result in permanent damage. This is also known as transient ischemic attack or a ministroke.

Questions 80 – 90: Date of onset

Indicate if the date of onset of the new CNS event reported in *Specify type of CNS complication* is **Known** or **Unknown**. If **Known**, report the date of onset. If the date of onset is not known, select **Unknown**.

If the exact date is not known report an estimated date and check the **Date estimated** box. Refer to General Instructions, Guidelines for Completing Forms for information about reporting estimated dates.

Questions 91 – 92: Was an MRI / MRA of the brain performed for the diagnosis of this reported CNS event?

Magnetic resonance imaging (MRI) is an imaging technique used to form pictures of the anatomy and the physiological processes of the body. Magnetic resonance angiography (MRA) is similar to MRI but is used to specifically examine blood vessels.

If an MRI or MRA was performed to diagnose the reported CNS event, indicate **Yes** and attach a copy of the MRI / MRA report. Only attach the MRI / MRA report performed "at diagnosis" of the CNS event. The diagnostic MRI / MRA report may not be the most recent scan performed prior to the start of the preparative regimen. Do not attach MRI / MRA reports performed at any other time-point.

If an MRI / MRA was not performed to diagnose the reported CNS event or if no information is available to determine if an MRI / MRA was performed at diagnosis of the CNS event, report **No** or **Unknown**, respectively.

For instructions on how to attach documents in FormsNet3^{SM}, refer to the Training Guide.

Section Updates:

Question Number	Date of Change	Add/Remove/Modify	Description	Reasoning (If applicable)
.	.	.	.	.

Last modified: Oct 27, 2024

Q93 – 104: Other Symptoms

Priapism:
Did one or more episodes of priapism occur? Is applicable to only biologically male recipients.

Questions 93 – 95: Did one or more episodes of priapism occur? (to be answered for males only)

Priapism is defined as prolonged erection of the penis, usually without sexual arousal.

Report **Yes** if the recipient experienced one or more episodes of priapism in the current reporting period. If the recipient did not experience any episodes of priapism in the current reporting period or if no information is available to determine if priapism occurred, report **No** or **Unknown**, respectively.

If **Yes**, specify the number of episodes occurring in the current reporting period and the date of the last episode.

If the exact date is not known report an estimated date. Refer to General Instructions, Guidelines for Completing Forms for information about reporting estimated dates.

Question 96: Has a new onset of sickle cell retinopathy developed?

Sickle cell retinopathy is an ocular manifestation of sickle cell disorders characterized by ocular damage due to trapping of sickle-shaped cells in the small blood vessels in various structures of the eye. Diagnosis of sickle cell retinopathy should be made by an ophthalmologist and is typically documented within the recipient's medical record. Seek physician clarification if it is unclear if there was a new onset of sickle cell retinopathy in the current reporting period.

Indicate **Yes** if the recipient developed a new onset of sickle cell retinopathy since the date of last report.

Report **No** in in the following scenarios:

- There was not a new onset of sickle cell retinopathy since the date of last report.
- Sickle cell retinopathy was diagnosed in a prior to HCT or in a prior reporting period, resolved, and developed again in the current reporting period.
- Sickle cell retinopathy was diagnosed in a prior reporting period and persisted into the current reporting period.

If documentation is not clear or is not available to determine if sickle retinopathy occurred in the current reporting period, report **Unknown**.

Question 97: Have chronic leg ulcers developed?

Chronic leg ulcers are defined as a defect of the skin below the level of the knee persisting for more than six

weeks with no tendency to heal after three or more months.

Indicate whether the recipient developed chronic leg ulcers since the date of last report. If chronic leg ulcers were diagnosed in the current reporting period (first onset), report **Yes**.

Report **No** in the following scenarios:

- Chronic leg ulcers did not develop in the current reporting period.
- Chronic leg ulcers developed prior to HCT or in a prior reporting period and persisted into the current reporting period.
- Chronic leg ulcers developed in a prior reporting period, resolved, and developed again in the current reporting period.

If no information is available to determine if chronic leg ulcers developed in the current reporting period, select **Unknown**.

Question 98: Is there a new diagnosis of asthma or a reactive airway disease?

Asthma is a condition where the airways narrow, swell, and produce extra mucus which results in breathing difficulty, coughing, wheezing, and shortness of breath. Reactive airway disease (RAD) is a general term used to describe coughing, wheezing or shortness of breath when a specific diagnosis has not been made (i.e., asthma, chronic obstructive pulmonary disease, etc.).

Indicate if there was a new diagnosis of asthma or a reactive airway disease since the date of last report.

If the recipient was clinically diagnosed with asthma or reactive airway disease in the current reporting period, select **Yes**.

Report **No** in the following scenarios:

- There was not a diagnosis of asthma or reactive airway disease in the current reporting period.
- Asthma and reactive airway disease were diagnosed in a prior reporting period.

If there is no information available to determine if there was a diagnosis of asthma or reactive airway disease in the current reporting period, select **Unknown**.

Question 99: Has a venous thrombosis embolism developed?

Venous thrombosis embolism, also called a venous thromboembolism, is a condition characterized by formation of blood clots in deep veins of the body. These clots typically manifest in the lower leg, thigh, or pelvis, but have been noted to occur in the arm.

Indicate if the recipient developed venous thrombosis embolism since the date of last report. If the recipient developed a venous thrombosis embolism, report **Yes**.

Report **No** in any of the following scenarios:

- A venous thrombosis did not develop in the current reporting period.
- A venous thrombosis developed in a prior reporting period and persisted into the current reporting period.

If no information is available to determine if a venous thrombosis embolism developed in the current reporting period, select **Unknown**.

Question 100: Was it associated with an indwelling (central line) catheter?

There are several types of long term indwelling central line catheters used to access veins. Examples include Hickman, Broviac, PICC, etc. which carry a risk of developing a blood clot.

Indicate **Yes** or **No** if the venous thrombosis embolism was associated with the recipient's indwelling catheter.

Question 101: Has a pulmonary embolism developed?

Pulmonary embolism is a medical condition where a blood clot gets lodged in an artery of the lung, blocking blood flow to that area.

Indicate if the recipient developed a pulmonary embolism since the date of last report. If the recipient developed a pulmonary embolism in the current reporting period, select **Yes**.

Report **No** in any of the following scenarios:

- A pulmonary embolism did not develop in the current reporting period.
- A pulmonary embolism developed in a prior reporting period and persisted into the current reporting period.

If information is available to determine if a pulmonary embolism developed in the current reporting period, select **Unknown**.

Question 102: Was it associated with an indwelling (central line) catheter?

There are several types of long term indwelling central line catheters used to access veins. Examples include Hickman, Broviac, PICC, etc. which carry a risk of developing a blood clot.

Indicate **Yes** or **No** if the pulmonary embolism was associated with the recipient's indwelling catheter.

Question 103: Number of splenic sequestration events

The diagnosis of a splenic sequestration crisis is usually clinical using patient history, physical exam, and accompanying lab values. This typically requires seeking medical care for urgent diagnosis and management. An event is defined as the documentation of this diagnosis after seeking medical care either in clinic, emergency department or hospitalization

Question 104: Date of last event

Report the date of the last splenic sequestration event requiring hospitalization or treatment in the current reporting period. This information is typically documented within the progress notes.

Section Updates:

Question Number	Date of Change	Add/Remove/Modify	Description	Reasoning (If applicable)
.	.	.	.	.

Last modified: Oct 27, 2024

Q105 – 116: Existing Organ Impairments

Multiple Organ Impairments
For each Organ Impairment the recipient experienced in the reporting period, complete questions *Specify co-existing diseases or organ impairments developed*, *Date of diagnosis*, *Method used to assess osteopathies*, *DEXA scan vertebral*, *DEXA scan hip*, *Quantitative CT vertebral* and *Quantitative CT vertebral* for each organ impairment in the reporting period by adding an additional instance in the FormsNet3^{SM} application.

Question 105: Specify co-existing diseases or organ impairments developed

Indicate if the recipient developed any of the co-existing disease or organ impairments listed below during the current reporting period. Select all that apply.

- **Amenorrhea**: Absence of menstruation
- **Cardiomyopathy**: A disease of the heart muscle that makes it more difficult for the heart to pump blood to the rest of the body
- **Cholelithiasis**: Presence of one or more gallstones in the gallbladder
- **Growth hormone deficiency / short stature**: A condition in which the body does not produce enough growth hormone / a reduced overall rate of growth.
- **Hypersplenism**: Overactive spleen. Diagnosis is typically based on a physician's exam (checking for splenomegaly), a CBC to assess the concentration of red and white blood cells, and / or an ultrasound, measuring the size of the spleen.
- **Hypogonadism / gonadal dysfunction**: A condition where there is a hyperfunction of the gonads. It can manifest as precocious puberty, and is caused by abnormally high levels of testosterone or estrogen, crucial hormones for sexual development.
- **Hypothyroidism requiring replacement therapy**: Decreased activity of the thyroid gland. Diagnosis of hypothyroidism includes high levels of thyroid-stimulating hormone (TSH). Symptoms of hypothyroidism include fatigue, depression, weakness, weight gain, musculoskeletal pain, decreased taste, hoarseness, and / or puffy face.
- **Osteonecrosis**: Flow to part of a bone is disrupted. This results in death of bone tissue, and the bone can eventually break down and the joint will collapse.
- **Osteopathies (porosis, penia)**: Includes osteoporosis or osteopenia. Osteopathies should be reported if osteopenia or osteoporosis is documented within the medical record by the physician or based on the Z or T-score. Osteopenia is defined as a Z or T-score between -1.0 and -2.0 by a DEXA or quantitative CT scan. Osteoporosis is defined as a Z or T-score less than -2.0 by a DEXA or quantitative CT scan.
- **Retinal changes**: Changes include but are not limited to macular degeneration, floaters, diabetic eye disease, retinal detachment, and retinitis pigmentosa.
- **Thrombosis**: Blood clot within the vein or artery.

The co-existing disease or organ impairment should be reported in any of the following scenarios:

- The co-existing disease or organ impairment was first diagnosed in the current reporting period.
- The co-existing disease or organ impairment was diagnosed prior to infusion or in a prior reporting period and persisted into the current reporting period.
- The co-existing disease or organ impairment was diagnosed prior to infusion or in a prior reporting period, resolved, and then recurred in the current reporting period.

Report **None** in any of the following scenarios:

- No co-existing disease or organ impairments developed during the current reporting period.
- A co-existing disease or organ impairment developed prior to start of the preparative regimen / infusion or in a prior reporting period but resolved before the current reporting period.

Question 106: Was this organ impairment previously reported?

Specify if the co-existing disease or organ impairment diagnosis date was previously reported. If **Yes**, continue with the next question.

If the co-existing disease or organ impairment was diagnosed prior to infusion or in a prior reporting period, resolved, and then recurred in the current reporting period, report No.

Question 107: Date of diagnosis

Report the diagnosis date of the co-existing disease or organ impairment.

If the co-existing disease or organ impairment was diagnosed prior to infusion or in a prior reporting period, resolved, and then recurred in the current reporting period, report the diagnosis date as the date of the most recent episode.

If the exact date is not known report an estimated date and check the **Date estimated** box. Refer to General Instructions, Guidelines for Completing Forms for information about reporting estimated dates.

Question 108: Method used to assess osteopathies *(report the most recent Z or T-score available. Z-scores are used in patients younger than or equal to 20 and T-scores in patients older than 20) (check all that apply)*

Specify the method used to assess osteopathies during the current reporting period. Select all that apply.

If the osteopathy was not assessed during the current reporting period or is not known if assessed, select **Unknown** and continue with Disease Modifying Therapies.

Questions 109 – 110: DEXA scan vertebral

Indicate if the vertebral Z-score by DEXA is known. If **Known**, report the Z or T-score. Select Negative value if the Z or T-score is a negative (i.e., Z-score is -1.0).

If multiple DEXA scans were performed during the current reporting period, report the most recent vertebral Z or T-score.

Questions 111 – 112: DEXA scan hip

Indicate if the hip Z-score by DEXA is known. If **Known**, report the Z or T-score. Select Negative value if the Z or T-score is a negative (i.e., Z-score is -1.0).
If multiple DEXA scans were performed during the current reporting period, report the most recent hip Z or T-score

Questions 113 – 114: Quantitative CT vertebral

Indicate if the vertebral Z-score by quantitative CT is known. If **Known**, report the Z or T-score. Select Negative value if the Z or T-score is a negative (i.e., Z-score is -1.0).
If multiple quantitative CT scans were performed during the current reporting period, report the most recent vertebral Z or T-score.

Questions 115 – 116: Quantitative CT hip

Indicate if the hip Z-score by quantitative CT is known. If **Known**, report the Z or T-score. Select Negative value if the Z or T-score is a negative (i.e., Z-score is -1.0).
If multiple quantitative CT scans were performed during the current reporting period, report the most recent hip Z or T-score.

Section Updates:

Question Number	Date of Change	Add/Remove/Modify	Description	Reasoning (If applicable)
.	.	.	.	.

Last modified: Oct 27, 2024

Q117 – 123: Disease Modifying Therapies

Question 117: Were disease modifying therapies given or stopped?

Indicate if the recipient received any disease modifying therapies (see below for a list of common disease modifying therapies) or if the recipient discontinued a disease modifying therapy in the current reporting period. If the recipient received disease modifying therapy(ies) or discontinued a disease modifying therapy in the current reporting period, select **Yes**.

If the recipient did not receive disease modifying therapy(ies) in the current reporting period, report **No**.

If no information is available to determine if disease modifying therapy was given in the current reporting period, select **Unknown**.

Report **Not applicable** if the recipient started a disease modifying therapy in a prior reporting period and is still receiving therapy on the date of contact for the current reporting period.

Disease Modifying Therapies
For each disease modifying therapy the recipient received in the reporting period, complete questions *Specify the disease modifying therapy*, *Date therapy started*, *Date therapy stopped* for each disease modifying therapy received in the reporting period by adding an additional instance in the FormsNet3^{SM} application.

Same Therapy Restarted
If the same therapy was started and stopped multiple times prior to the start of the preparative regimen, only one instance needs to be reported. In this case, report the therapy start date as the date when therapy first began.

Questions 118 – 119: Specify the disease modifying therapy

Indicate which disease modifying therapy the recipient received. If the administered agent is not listed, report **Other** and specify the agent. Report the generic name of the agent, not the brand name.

- **Crizanlizumab (Adakveo)**: A monoclonal antibody given to reduce the frequency of vaso-occlusive crises.
- **Hydroxyurea**: A type of chemotherapy. Common brand names include Droxia and Hydrea.
- **L-Glutamine**: An amino acid in the form of an oral powder given to reduce the complications of sickle cell disease. Also known as Endari.
- **Voxelotor (Oxbryta)**: An oral medication given to inhibit sickle hemoglobin polymerization.

Questions 120 – 121: Date therapy started:

Indicate whether the start date is **Known** or **Unknown**. If the therapy start date is **Known**, report the date

the recipient began this line of therapy. If the start date is partially known (i.e., the recipient started treatment in mid-June 2010), use the process described for reporting partial or unknown dates in General Instructions, Guidelines for Completing Forms.

If therapy was started in a prior reporting period and continued to the current reporting period, select **Previously reported**. This option is not applicable for the Day 100 reporting period.

If the therapy start date is not known, select **Unknown**.

Questions 122 – 123: Date therapy stopped:

Indicate if the therapy stop date is **Known** or **Unknown**. If the therapy stop date is **Known**, use the following instructions to report the end date:

- If the therapy is administered in cycles, report the date the recipient started the last cycle for this line of therapy.
- If the therapy is administered on a daily basis, report the last date the recipient received the line of therapy being reported.

If the stop date is partially known, use the process for reporting partial or unknown dates as described in the General Instructions, Guidelines for Completing Forms.

If the disease modifying therapy stop date is not known, select **Unknown**.

Report **Not applicable** if the recipient is still receiving therapy on the contact date for the current reporting period.

Section Updates:

Question Number	Date of Change	Add/Remove/Modify	Description	Reasoning (If applicable)
.	.	.	.	.

Last modified: Oct 27, 2024

Q124 – 147: Other Laboratory Studies

Questions 124 – 125: Was hemoglobin electrophoresis performed? *(do not include results if an RBC transfusion occurred within 4 weeks of the electrophoresis study)*

Indicate if hemoglobin electrophoresis studies were performed since the date of last report. If hemoglobin electrophoresis studies were performed, report **Yes** and provide the date of the most recent hemoglobin electrophoresis study performed in the current reporting period.

If the exact date is not known report an estimated date and check the **Date estimated** box. Refer to General Instructions, Guidelines for Completing Forms for information about reporting estimated dates.

If hemoglobin studies were not performed or if no information is available to determine if hemoglobin electrophoresis studies were conducted in the current reporting period, select **No** or **Unknown**.

If a hemoglobin electrophoresis study was performed but RBC transfusion(s) were given within four weeks prior to the study, select **Not applicable**.

Hemoglobin Electrophoresis:
Some centers do not routinely perform hemoglobin electrophoresis studies and may perform chimerism studies instead. The chimerism results are reported in the Chimerism Studies section of the Post-Infusion Follow-Up (2100) Form. For centers that performed both hemoglobin electrophoresis and chimerism studies, report the results of both on the appropriate forms.

Hemoglobin Types: Hb A and Hb A1
Hb A and Hb A1 are the same hemoglobin types. If Hb A1 is assessed, report these results under **Hb A**.

Questions 126 – 138: : Specify the hemoglobin allele types based on the reported sample

Specify the hemoglobin allele types identified in the reported hemoglobin electrophoresis study (reported above in *Was hemoglobin electrophoresis performed?*). If the hemoglobin allele type was assessed, report **Yes** and specify the percentage. If additional sickle related hemoglobin allele types are identified and not listed as options on the form, report **Yes** for *Other sickle related hemoglobin allele type*, specify the other hemoglobin type, and provide the percentage.

Report **No** if the specified hemoglobin allele type was not assessed.

Questions 139 – 141: Were reticulocyte counts tested?

Indicate if reticulocyte counts were assessed in the current reporting period. If reticulocyte counts were measured, report **Yes**, provide the reticulocyte cell count, and specify the date the sample was collected for

examination. If reticulocyte counts were measured multiple times in the current reporting period, report the most recent results.

If reticulocyte counts were not measured or if no information is available to determine if reticulocyte counts were assessed, select **No** or **Unknown**, respectively.

If the sample collection date is partially known (i.e., the recipient's reticulocyte counts were measured in mid-July 2019), use the process described for reporting partial or unknown dates in General Instructions, Guidelines for Completing Forms.

Questions 142 – 144: Were soluble transferrin receptors (sTfR) tested?

Soluble transferring receptors (sTfR) are proteins found in the blood and are used as a measure of functional iron status. These levels are typically elevated in individuals with an iron deficiency (i.e., iron deficiency anemia).

Indicate if sTfR was tested since the date of last report. If sTfR counts were assessed in the current reporting period, report **Yes**, provide the sTfR value in mg / L, and specify the date the sample was collected for. If sTfR was measured multiple times in the current reporting period, report the most recent results.

If sTfR counts were not measured since the date of last report or if no information is available to determine if sTfR counts were measured, select **No** or **Unknown**, respectively.

If the sample collection date is partially known (i.e., the recipient's sTfR counts were measured in mid-July 2019), use the process described for reporting partial or unknown dates in General Instructions, Guidelines for Completing Forms.

Questions 145 – 147: Was an erythropoietin (EPO) level obtained?

Erythropoietin (EPO) is a hormone predominantly produced in the kidneys which plays a critical role in the production of red blood cells.

Indicate if EPO was tested since the date of last report. If EPO levels were assessed in the current reporting period, report **Yes**, specify the EPO level in IU / L, and report the date the sample was collected for examination. If EPO was measured multiple dates in the reporting period, report the results of the most recent assessment.

If EPO levels were not measured or if no information is available to determine if EPO levels were assessed since the date of last report, select **No** or **Unknown**, respectively.

If the sample collection date is partially known (i.e., the recipients sTfR counts were measured in mid-2019), use the process described for reporting partial or unknown dates in General Instructions, Guidelines for Completing Forms.

Section Updates:

Question Number	Date of Change	Add/Remove/Modify	Description	Reasoning (If applicable)
.	.	.	.	.

Last modified: Oct 27, 2024

Q148: Disease Status

Question 148: What is the status of sickle cell disease at the time of this report, or at the time of death?

Report the status of the recipient's sickle cell disease based on the Hb S and clinical symptoms at the time of evaluation, or death, for this reporting period. The term "clinical symptoms" refers to the clinical complications captured above (i.e., vaso-occlusive pain, acute chest syndrome, avascular necrosis, etc.). See below for additional details regarding the available disease status options:

- **Hb electrophoresis (Hb S) ≤ 50% and clinical symptoms described are absent:** The Hb S value at the time of evaluation for the current reporting period is ≤ 50% (reported above), the clinical symptoms are absent, and the recipient is not receiving disease modifying agents.
- **Hb S > 50% and clinical symptoms described are absent:** The Hb S value at the time of evaluation for the current reporting period is > 50% (reported above), the clinical symptoms are absent, and the recipient is not receiving disease modifying agents.
- **Hb S > 50% and clinical symptoms described are present:** The Hb S value at the time of evaluation for the current reporting period is > 50% (reported above), the clinical symptoms are present, and the recipient is not receiving disease modifying agents.
- **Recipient is receiving disease modifying therapy:** Select this disease response option if the recipient is still receiving disease modifying agents on the date of contact for the current reporting period, regardless of if improvement was observed in Hb S values.

If therapy was discontinued prior to the contact date and Hb S was not re-assessed, report the disease status as either **Hb S > 50% and clinical symptoms described are absent** or **Hb S > 50% and clinical symptoms described are present** based on the absence or presence of clinical symptoms.

If the Hb S is ≤ 50% but clinical symptoms are present, leave the data field blank, override the FormsNet3^{SM} error with "unable to answer," and explain the Hb S is ≤ 50%; however, clinical symptoms are present in the comment section.

If it is unclear whether clinical symptoms are still present at the time of evaluation for the current reporting period, confirm with the attending physician.

Section Updates:

Question Number	Date of Change	Add/Remove/Modify	Description	Reasoning (If applicable)
.	.	.	.	.

Last modified: Oct 27, 2024

Q149: Marrow Evaluation

Marrow Evaluation
Complete *Was a marrow aspirate and / or biopsy performed?* for gene therapy infusions only

Question 149: Was marrow aspirate and /or biopsy performed?

Indicate **Yes** or **No** if a marrow aspirate and or biopsy was performed in this reporting period. If **Yes**, also complete the Laboratory Studies (3502) Form and Marrow Surveillance (3506) Form. The intent is to screen for and/or identify changes in the marrow such as dysplasia, MDS, or new hematologic malignancy

Report **Unknown** if not documented.

Section Updates:

Question Number	Date of Change	Add/Remove/Modify	Description	Reasoning (If applicable)
.	.	.	.	.

Last modified: Oct 27, 2024