2130: SCD Post-Infusion

The Sickle Cell Disease Post-HCT Data 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 Pre-TED Disease Classification (2402) Form, is “Sickle Cell Disease (SCD).” The Post-HCT Sickle Cell Disease (2130) Form must be completed in conjunction with each Post-HCT 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
Q1 – 5: Physical Assessments
Q6 – 8: Transfusion Therapy
Q9 – 13: Pulmonary Assessments
Q14 – 30: Cardiovascular Assessments
Q31 – 37: Renal Assessments
Q38 – 42: Splenic Assessments
Q43: Acute Chest Syndrome
Q44 – 46: Pain
Q47 – 49: Avascular Necrosis
Q50 – 58: Central Nervous System
Q59 – 66: Other Symptoms
Q67 – 73: Disease Modifying Therapies
Q74 – 100: Other Laboratory Studies
Q101: Disease Status

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
5/5/ 2021	2130: SCD Post-Infusion	Add	The Bedside Schwartz equation was added to questions 36 -37 to calculate the GFR for pediatric recipients.

4/28/2021	2130: SCD Post-Infusion	Modify	Instructions on when to report not applicable for questions 74 – 75 were updated: *If RBC transfusion(s) were given within four weeks prior of the hemoglobin electrophoresis, report Not applicable and go to question 89.*
11/23/2020	2130: SCD Post-Infusion	Add	Instructions added to question 101 to explain how to report the current disease status when the Hb S ≤ 50% but clinical symptoms are present: *If the Hb S is ≤ 50 % but clinical symptoms are present, leave the data field blank, override the FormsNet3 error with "unable to answer," and explain the Hb S is below ≤ 50 %; however, clinical symptoms are present in the comment section.*
7/24/2020	2130: SCD Post-Infusion	Add	Version 1 of the 2130: Sickle Cell Disease (SCD) Post-Infusion Data section of the Forms Instruction Manual released. Version 1 corresponds to revision 3 of the Form 2130.

Last modified: May 05, 2021

Q1-5: Physical Assessments

NOTE: 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 questions 2 – 5 below.

Questions 2 – 3: Abdominal girth

Indicate if the recipient’s abdominal girth was measured on the date of evaluation specified in question 1. 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** and go to question 4.

Questions 4 – 5: Blood pressure (BP)

Indicate if the recipient’s blood pressure was assessed on the date of evaluation specified in question 1. 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** and go to question 6.

Section Updates:

Question Number	Date of Change	Add/Remove/Modify	Description	Reasoning (If applicable)
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Last modified: Dec 22, 2020

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**.

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)
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Last modified: Dec 22, 2020

Q9-13: Pulmonary Assessments

Questions 9 – 10: 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 11: 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 12 – 13: 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 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)
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Last modified: Dec 22, 2020

Q14-30: Cardiovascular Assessments

Question 14: 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 15 – 19: 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 20: 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 and go to question 27.

Questions 21 – 22: 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 23 – 25: 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.

Question 26: Is a copy of the echocardiogram report attached?

Indicate whether an echocardiogram report is attached to this form. For further instructions on how to attach documents in FormsNet3 SM, refer to the Training Guide.

Questions 27 – 28: 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 and go to question 29.

Questions 29 – 30: Is there a new onset of pulmonary hypertension? (developed since last report)

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 HCT 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**.

Section Updates:

Question Number	Date of Change	Add/Remove/Modify	Description	Reasoning (If applicable)
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Last modified: Dec 22, 2020

Q31-37: Renal Assessments

Question 31: Were renal assessments conducted?

Indicate if urine albumin, serum creatinine, or GFR assessments were conducted at the time of evaluation for this reporting period. If these renal assessments were performed, report **Yes**. If these renal assessments were not performed or no information is available to determine if they were performed since the date of last report, select **No** or **Unknown**.

Questions 32 – 33: Urine albumin (if multiple, report the most recent tested)

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).

Questions 34 – 35: Serum creatinine (if multiple, report the most recent tested):

Indicate whether the serum creatinine 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 serum creatinine was measured multiple times in the reporting period, report the most recent results. If serum creatinine was not measured or if no information is available to determine if serum creatinine was measured in the current reporting period, report **Unknown**.

Questions 36 – 37: Glomerular filtration rate (GFR) (if multiple, report the most recent tested):

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.”

Section Updates:

Question Number	Date of Change	Add/ Remove/ Modify	Description	Reasoning (If applicable)
Q36 – 37	5/5/2021	Add	The Beside Schwartz Equation was added to questions 36 – 37	Equation added to calculate GFR for pediatric recipients

Last modified: May 05, 2021

Q38-42: Splenic Assessments

NOTE: Splenic Assessments
Questions 38 – 42 will only be applicable for the 100-day, 6-month, 1 year, and 2-year follow-up periods. Questions 38 – 42 will be disabled for annual reporting > 2 years.

Question 38: 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 39 – 42: 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) in question 40.

If a pitted RBC score was performed, specify the percentage in question 41.

If a splenic scan was performed, specify the results as either **Normal (radionuclide uptake)** or **Abnormal (no radionuclide uptake)** in question 42.

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)
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Last modified: Dec 22, 2020

Q43: Acute Chest Syndrome

Question 43: 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**.

Section Updates:

Question Number	Date of Change	Add/Remove/Modify	Description	Reasoning (If applicable)
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Last modified: Dec 22, 2020

Q44-46: Pain

Question 44: 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 45: 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 46: 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)
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Last modified: Dec 22, 2020

Q47-49: Avascular Necrosis

Questions 47 – 49: New onset of 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.

Question 48: 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:
p(. 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:
Question 47: Report “Yes” to capture the new development of avascular necrosis of the left shoulder.
Question 48: Select “Shoulder” and continue with the form. “Hip” will not be selected as this was diagnosed pre-transplant.

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:
Question 47: Report “Yes” to indicate the diagnosis of avascular necrosis of the right knee.
Question 48: Select “Knee.”

Six Month Post-Infusion Data Form:
Question 47: Report “No” as there was not a new onset of avascular necrosis during the reporting period.

One Year Post-Infusion Data Form:
Question 47: Report “Yes” to capture the onset of avascular necrosis of the left knee.
Question 48: Select “Knee” again.

Section Updates:

Question Number	Date of Change	Add/Remove/Modify	Description	Reasoning (If applicable)
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Last modified: Dec 22, 2020

Q50-58: Central Nervous System

Question 50: Have any new central nervous system (CNS) events occurred?

A CNS event is the development of different neurologic signs and symptoms that occur in recipients with sickle cell disease. See question 51 for a list of CNS events.

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

Report **No** in the following scenarios:

- A new CNS event did not occur in the current reporting period.
- The recipient experienced a CNS event 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 event occurred since the date of last report, report **Unknown**.

*Note: CNS Events – Reporting Multiple Events *
FormsNet3 SM application: Complete questions 51 – 58 for each new CNS event the recipient experienced by adding an additional instance in the FormsNet3 SM application. Only report the initial event and applicable information at the time of onset. Do **not** report follow-up scans.
Paper form submission: Copy questions 51 – 58 and complete for each new CNS event the recipient experienced since the date of last report. Only report the initial event and applicable information at the time of onset. Do not report follow-up scans.

Question 51: Specify type of CNS event:

Indicate the type of CNS event(s) that occurred. If multiple CNS events occurred, report each CNS event as a separate instance. If the same CNS event 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.
- **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 52 – 53: Date of onset:

Indicate if the date of onset of the new CNS event reported in question 51 is **Known** or **Unknown**. If **Known**, report the date of onset. If the date of onset is not known, select **Unknown**.

Questions 54 – 55: 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.

Questions 56 – 58: Was transcranial doppler velocity assessed at any time for this reported CNS event?

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 trans-cranially.

Indicate if transcranial doppler velocity was assessed for the reported CNS event at any time in the current reporting period, after the CNS event 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 for the reported CNS event, report **No** or **Unknown**.

Section Updates:

Question Number	Date of Change	Add/Remove/Modify	Description	Reasoning (If applicable)
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Last modified: Dec 22, 2020

Q59-66: Other Symptoms

Priapism:
Question 59 will only be applicable to biologically male recipients.

Question 59: 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.

Question 60: 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 61: 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 62: 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 63: 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 64: 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 65: 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 66: 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.

Section Updates:

Question Number	Date of Change	Add/Remove/Modify	Description	Reasoning (If applicable)
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Last modified: Dec 22, 2020

Q67-73: Disease Modifying Therapies

Question 67: Were disease modifying therapies given or stopped since the date of last report?

Indicate if the recipient received any disease modifying therapies (see question 68 for a list of common disease modifying therapies) or if the recipient discontinued a disease modifying therapy, since the date of last report. 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.

> **Note: Disease Modifying Therapies**
> **FormsNet3 SM application**: Complete questions 68 – 73 for each disease modifying therapy the recipient received by adding an additional instance in the FormsNet3SM application.
> **Paper form submission**: Copy questions 68 – 73 and complete for each disease modifying therapy the recipient received.

> **Note: Same Therapy Restarted**
> If the same therapy was started and stopped multiple times during the current reporting period, only one instance needs to be reported.

Questions 68 – 69: 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 in question 69. 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 70 – 71: 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**.

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

Questions 72 – 73: 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)
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Last modified: Dec 22, 2020

Q74-100: Other Laboratory Studies

Questions 74 – 75: Was hemoglobin electrophoresis performed?

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 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** and go to question 89.

If RBC transfusion(s) were given within four weeks prior of the hemoglobin electrophoresis, report **Not applicable** and go to question 89.

> **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-HCT Follow-Up (2100) Form. For centers that performed both hemoglobin electrophoresis and chimerism studies, please report the results of both on the appropriate forms.

Questions 76 – 88: Specify the hemoglobin allele types based on the sample tested in question 75

Specify the hemoglobin allele types identified in the reported hemoglobin electrophoresis study (reported above in question 75). 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* in question 86, specify the other hemoglobin type, and provide the percentage.

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

Questions 89 – 91: Were red blood cell counts tested?

Red blood cell counts are part of a complete blood count panel. Indicate if red blood cell counts were measured since the date of last report. If red blood cell counts were assessed during the current reporting period, report **Yes**, provide the RBC cell count in cells / µL and specify the date the sample was collected for examination. If RBC counts were measured multiple times in the reporting period, report the most recent results.

If RBC counts were not measured in the current reporting period or if no information is available to determine if RBC counts were assessed, select **No** or **Unknown**, respectively.

If the sample collection date is partially known (i.e., the recipient's RBC counts were measured in mid-July

2019), use the process described in reporting partial or unknown dates in General Instructions, Guidelines for Completing Forms.

Questions 92 – 94: Were reticulocyte counts tested?

Indicate if reticulocyte counts were assessed since the date of last report. If reticulocyte counts were measured in the current reporting period, 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 since the date of last report 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 95 – 97: 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 98 – 100: 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)
Q74 – 75	4/28/ 2021	Modify	The following update was made: *If RBC transfusion(s) were given within four weeks prior of the hemoglobin electrophoresis, report **Not applicable** and go to question 89.*	Not applicable

Last modified: Apr 28, 2021

Q101: Disease Status

Question 101: 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:

- **Disease cured: 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 in question 85), the clinical symptoms are absent, and the recipient is not receiving disease modifying agents.
- **Disease recurred: 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 in question 85), the clinical symptoms are absent, and the recipient is not receiving disease modifying agents.
- **Disease recurred: 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 in question 85), the clinical symptoms are present, and the recipient is not receiving disease modifying agents.
- **Disease recurred: on 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 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 **Disease recurred: Hb S > 50% and clinical symptoms described are absent** or **Disease recurred: 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 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)
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Last modified: Dec 22, 2020