

Title: Exchange Transfusion for Sickle Cell Disease	Division: Medical Management Department: Utilization Management
Approval Date: 2/9/18	LOB: Medicaid, Medicare, HIV SNP, CHP, MetroPlus Gold, Goldcare I&II, Market Plus, Essential, HARP
Effective Date: 2/9/18	Policy Number: UM-MP224
Review Date: 1/29/2021	Cross Reference Number:
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1. POLICY:

Exchange Transfusion for Sickle Cell Disease

2. RESPONSIBLE PARTIES:

Medical Management Administration, Utilization Management, Integrated Care Management, Claims Department, Provider Contracting

3. DEFINITIONS

- Sickle cell disease – Sickle cell disease (SCD) refers to a group of inherited disorders characterized by sickled red blood cells (RBCs), caused either by homozygosity for the sickle hemoglobin mutation (HbSS; sickle cell anemia) or by compound heterozygosity for the sickle mutation and a second beta globin gene mutation (e.g., sickle-beta thalassemia, HbSC disease). In either HbSS or compound heterozygotes, the majority of Hgb is sickle Hgb (HgbS; i.e., >50 percent).
- Transfusion – Simple transfusion refers to transfusion of RBCs without removal of the patient's blood.
- Exchange Transfusion – Exchange transfusion involves transfusion of RBCs together with removal of the patient's blood. Exchange transfusion can be performed manually or via apheresis (also called cytopheresis or hemapheresis) using an extracorporeal continuous flow device.

4. PROCEDURE:

- A. Exchange transfusion for sickle cell disease will be covered as an ambulatory surgery procedure when all the following criteria are met:
- i) The member has documented SCD.
 - ii) The exchange transfusion is a pre-scheduled procedure.
 - iii) The purpose of the exchange transfusion is to prevent stroke, acute chest syndrome, or recurrent painful episodes.

5. EXCEPTION:

- A. Exchange transfusion for Sickle cell Disease will be covered as an inpatient procedure when it is done immediately prior to elective surgery (within 24 hours) and the inpatient stay is greater than 48 hours.

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6. APPLICABLE PROCEDURE CODES

36455 Exchange transfusion
 36450 Exchange transfusion, neonatal
 36456 Exchange transfusion, neonatal
 36512 Therapeutic apheresis for red blood cells

7. APPLICABLE DIAGNOSIS CODES

D57.0 Hb-SS disease with crisis
 D57.00 Hb-SS disease with crisis unspecified
 D57.01 Hb-SS disease with acute chest syndrome
 D57.02 Hb-SS disease with splenic sequestration
 D57.1 Sickle-cell disease without crisis
 D57.2 Sickle-cell/Hb-C disease
 D57.20 Sickle-cell/Hb-C disease without crisis
 D57.21 Sickle-cell/Hb-C disease with crisis
 D57.211 Sickle-cell/Hb-C disease with acute chest syndrome
 D57.212 Sickle-cell/Hb-C disease with splenic sequestration
 D57.219 Sickle-cell/Hb-C disease with crisis unspecified
 D57.3 Sickle-cell trait
 D57.4 Sickle-cell thalassemia
 D57.40 Sickle-cell thalassemia without crisis
 D57.41 Sickle-cell thalassemia with crisis
 D57.411 Sickle-cell thalassemia with acute chest syndrome
 D57.412 Sickle-cell thalassemia with splenic sequestration
 D57.419 Sickle-cell thalassemia with crisis unspecified
 D57.8 Other sickle-cell disorders
 D57.80 Other sickle-cell disorders without crisis
 D57.81 Other sickle-cell disorders with crisis
 D57.811 Other sickle-cell disorders with acute chest syndrome
 D57.812 Other sickle-cell disorders with splenic sequestration

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D57.819 Other sickle-cell disorders with crisis unspecified

8. BACKGROUND

Individuals with sickle cell disease (SCD) have chronic anemia that can worsen abruptly (e.g., from splenic sequestration or transient red cell aplasia), and they are at risk of vaso-occlusive events (e.g., stroke) due to the high concentration of sickle hemoglobin (HgbS) associated with their condition. Transfusion of red blood cells (RBCs) can be life-saving in these settings.

Blood transfusion therapy in SCD can serve two roles, either for therapy (typically for life-threatening SCD related complication) or for prophylaxis, to decrease the incidence of specific SCD related complications. In both cases, blood transfusion does more than simply raise the hemoglobin (Hgb) level for oxygen delivery; transfusion also lowers the percentage of sickle Hgb (HgbS) and increases Hgb oxygen saturation, both of which decrease the propensity for vaso-occlusion. The potential benefit of transfusion therapy must be weighed against potential risks, including transfusion reactions, blood-borne viral infection, iron overload, and alloimmunization.

Exchange transfusion involves removing some of the patient's own blood and transfusing allogeneic blood, thereby lowering the concentration of HgbS through dilution. A cardinal principle in transfusing individuals with SCD who are critically ill is that exchange transfusion provides greater benefit compared with simple transfusion because only exchange transfusion can significantly lower HgbS levels (i.e., to <30 percent of total Hgb). The lessened effects on viscosity for a given Hgb level are critical in potentially reversing vaso-occlusion and improving blood flow.

Exchange transfusion therapy can involve full blood volume exchange by manual or automated apheresis. A full exchange transfusion allows for rapid lowering of the HgbS level to 30 percent or less, and correction of anemia. Partial exchange transfusion refers to a limited exchange transfusion that is less effective in lowering the HgbS level but is more easily performed. To lower the HgbS below 30 percent, repeat partial exchange transfusions may be necessary.

Randomized trials analyzing the benefit of simple versus exchange transfusion for treating specific complications in SCD have not been performed. Clinical experience coupled with several limited observational studies suggests that exchange transfusion, either automated

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apheresis or manual, is superior to simple blood transfusion in suspected stroke, respiratory failure, and multi-organ failure.

In clinical situations where the exchange may be considered as part of standard care (acute chest syndrome, multi-organ failure, or strokes) without availability of apheresis or local expertise to perform a manual exchange, the patient should be transferred to a facility to perform apheresis or manual exchange, as these decisions are often time sensitive.

9. REFERENCES:

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10. ATTACHMENTS:

	Title	Attachment
1		
2		
3		

11. REVISION LOG:

REVISIONS	DATE
Annual Review – FIDA removed from LOB	1/18/19
Annual Review	1/31/2020
Annual Review	1/29/2021

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Approved: <i>Glendon Henry</i>	Date:	Approved: 	Date: 03.17.2021
Glendon Henry, MD Clinical Medical Director	1/29/2021	Sanjiv Shah, MD Chief Medical Officer	1/29/21

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Medical Guideline Disclaimer:

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All coding and website links are accurate at time of publication.

MetroPlus Health Plan has adopted the herein policy in providing management, administrative and other services to our members, related to health benefit plans offered by our organization.