MetroPlus Health

Policy and Procedure

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, UltraCare
Effective Date: 2/9/18	Policy Number: UM-MP224
Review Date: 1/30/2024	Cross Reference Number:
Retired Date:	Page 1 of 7

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 (Hb SS; sickle cell anemia) or by compound heterozygosity for the sickle mutation and a second beta globin gene mutation (e.g., sickle-beta thalassemia, Hb SC disease). In either Hb SS or compound heterozygotes, the majority of Hb is sickle Hb (Hb S; 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 cytapheresis or hemapheresis) using an extracorporeal continuous flow device.

4. PROCEDURE:

A. Exchange transfusion for sickle cell disease will be covered as an ambulatory 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, recurrent painful episodes, or to reduce the incidence of fetomaternal complications during pregnancy.

5. EXCEPTION:

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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.

B. For young children requiring chronic transfusion therapy, a simple transfusion combined with iron chelation is preferred to avoid the need for central venous catheter (CVC) placement. Once they reach the age of 10 years or the weight of 30 kg, a CVC or peripheral venous access for red cell exchange may be considered.

6. APPLICABLE PROCEDURE CODES

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

7. APPLICABLE DIAGNOSIS CODES

CODE:	DESCRIPTION
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
D57.819	Other sickle-cell disorders with crisis unspecified

8. BACKGROUND

Individuals with sickle cell disease (SCD) have chronic anemia that can worsen abruptly due to several reasons including (e.g., from splenic sequestration or transient red cell aplasia). These individuals are also 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 lifesaving in these settings.

Blood transfusion therapy in SCD can serve two roles, either for therapy (typically for life-threatening, SCD related complications) or for prophylaxis, to decrease the incidence of specific SCD related complications. In both cases, blood transfusion does more than simply raise the hemoglobin (Hb) level for oxygen delivery; transfusion also lowers the percentage of sickle Hgb (HbS) and increases Hb 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 Hb S 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 Hb S levels (i.e., to <30 percent of total Hgb). The lessened effects on viscosity for a given Hb 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 Hb S 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 Hb S level but is more easily performed. To lower the Hb S 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 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:

1. Up-To-Date, Literature review current through: December 2023, Last updated: May 30, 2023 https://www.uptodate.com/contents/red-blood-cell-transfusion-in-sickle-cell-disease-indications-and-transfusion-techniques?search=Exchange%20Transfusion%20for%20Sickle%20Cell%20Disease&source=search_result&selectedTitle=1~150&usage_type=default&display_rank=1

2. Journal of Clinical Apheresis, Role of prophylactic and therapeutic red blood cell exchange in pregnancy with sickle cell disease: Maternal and perinatal outcomes, August 14, 2020 https://onlinelibrary.wiley.com/doi/10.1002/jca.21819

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
Annual Review	1/28/2022
Annual Review	1/31/2023
Annual Review	1/30/2024

Approved: Sanjiv Shah MD Digitally signed by Sanjiv Shah MD Date: 2024.02.05 15:43:41 -05'00'	Date:
Sanjiv Shah, MD **Chief Medical Officer**	

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Medical Guideline Disclaimer:

 The treating physician or primary care provider must submit MetroPlus Health Plan clinical evidence that the patient meets the criteria for the treatment or surgical procedure. Without this documentation and information, Metroplus Health Plan will not be able to properly review the request for prior authorization. The clinical review criteria expressed in this policy reflects how MetroPlus Health Plan determines whether certain services or supplies are medically necessary. MetroPlus Health Plan established the clinical review criteria based upon a review of currently available clinical information(including clinical outcome studies in the peer-reviewed published medical literature, regulatory status of the technology, evidence-based guidelines of public health and health research agencies, evidence-based guidelines and positions of leading national health professional organizations, views of physicians practicing in relevant clinical areas, and other relevant factors). MetroPlus Health Plan expressly reserves the right to revise these conclusions as clinical information changes and welcomes further relevant information. Each benefit program defines which services are covered. The conclusion that a particular service or supply is medically necessary does not constitute a representation or warranty that this service or supply is covered and or paid for by MetroPlus Health Plan, as some programs exclude coverage for services or supplies that MetroPlus Health Plan considers medically necessary. If there is a discrepancy between this guidelines and a member's benefits program, the benefits program will govern. In addition, coverage may be mandated by applicable legal requirements of a state, the Federal Government, or the Centers for Medicare & Medicaid Services (CMS) for Medicare and Medicaid members. 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.