

Patient Guide:

**How automated red blood cell
exchange can help you manage the
balancing act of sickle cell disease.**

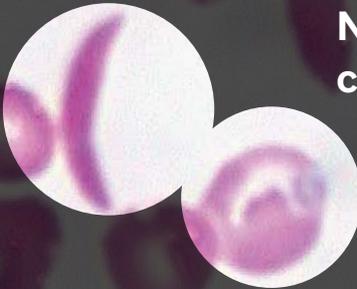


Sickle Cell Disease

In sickle cell disease, red blood cells (RBCs) contain an abnormal form of hemoglobin called hemoglobin S. The presence of hemoglobin S can cause RBCs to change shape and become sticky, making it difficult for them to pass through small blood vessels. As a result, the body's tissues and organs are deprived of the oxygen they need to function properly.

This often leads to pain and other disease-related complications.¹

**Sickled cells live
15 to 20 days.²**



**Normal red blood
cells live 120 days.²**

Three Ways You Can Manage Disease-Related Symptoms³

- ✓ Get regular checkups with your doctor
- ✓ Follow doctor-prescribed treatments
- ✓ Eat a healthy diet and stay hydrated

Transfusion Therapies

There are several types of transfusion therapy to help manage sickle cell disease and the related complications.



Automated red blood cell exchange (RBCX)

Rapidly removes your red blood cells and replaces them with healthy donor cells using an apheresis device.



Manual exchange

Removes your red blood cells and subsequently replaces them with healthy donor cells. This is performed manually and does not require a device.



Simple or “top-up” transfusion

Transfuses packed red blood cells from a healthy donor without removing any of your blood volume.

Not all blood transfusion therapies are the same.

Compare the differences among the three different transfusion therapy options.

	Automated RBCX	Manual Exchange	Simple Transfusion
Provides healthy donor cells ¹	✓	✓	✓
Increases RBC oxygen-carrying capacity ⁴	✓	✓	✓
Removes sickled cells ^{1,3,5}	✓	✓	
Automatically manages and maintains fluid levels ^{1,4}	✓*		
Manages iron levels ¹	✓	✓	
Average procedure time (minutes) ^{5,6,7}	86 to 127	120 to 245	180 to 360
Average procedure frequency ^{5,7,8,9,10,11}	Every 4 to 6 weeks	Every 3 to 4 weeks	Every 2 to 4 weeks

*Requires specialized equipment.





Finding balance. Improving quality of life.

What does an improved quality of life look like for you? One survey showed patients experienced 25% higher health-related quality of life when treated with automated RBCX versus simple transfusions. The factors that determined quality of life included¹²:

- ✓ More energy
- ✓ Increased mobility and physical functioning
- ✓ Less pain
- ✓ Greater independence
- ✓ Improved emotional well-being and mental health
- ✓ Improved social life



80% of sickle cell patients preferred automated RBCX over simple transfusion.¹²



More time for life. Less time in the hospital.

Automated RBCX can be performed less frequently than manual exchange or simple transfusion, and the procedure requires less time.^{6,7,8,9,10}

An Automated RBCX Procedure: What to Expect

Before the procedure

Single-use sterile tubing is connected to one or both of your arms with a needle. A clinician may decide that a more permanent kind of access connection is needed.

During the procedure

1. A small portion of your blood is removed.
2. It is then mixed with a fluid that prevents the blood from clotting.
3. The system spins the blood in a centrifuge to separate it into red blood cells and other blood components.

At any point during the procedure, the amount of blood in the tubing set is less than the amount of cola in a can.

4. Your red blood cells flow to a waste bag, and healthy donor cells are returned to you.

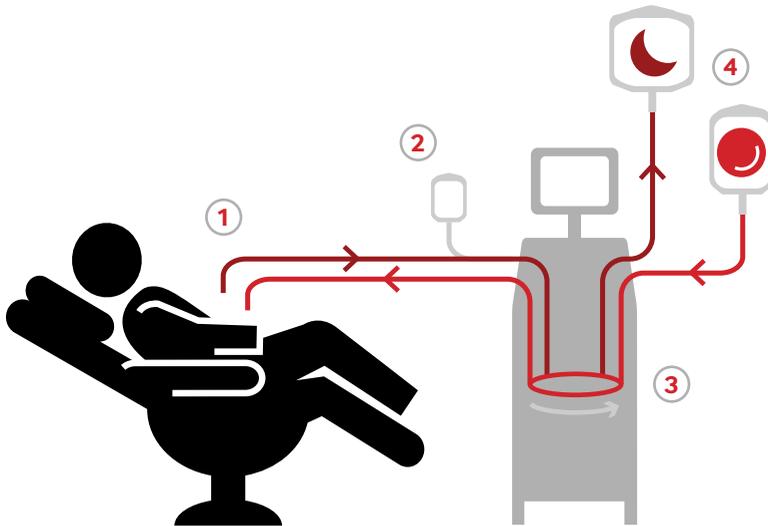
After the procedure

The tubing set and the removed red blood cells are properly discarded.

Vascular access varies per patient and device.

Common vascular access options:

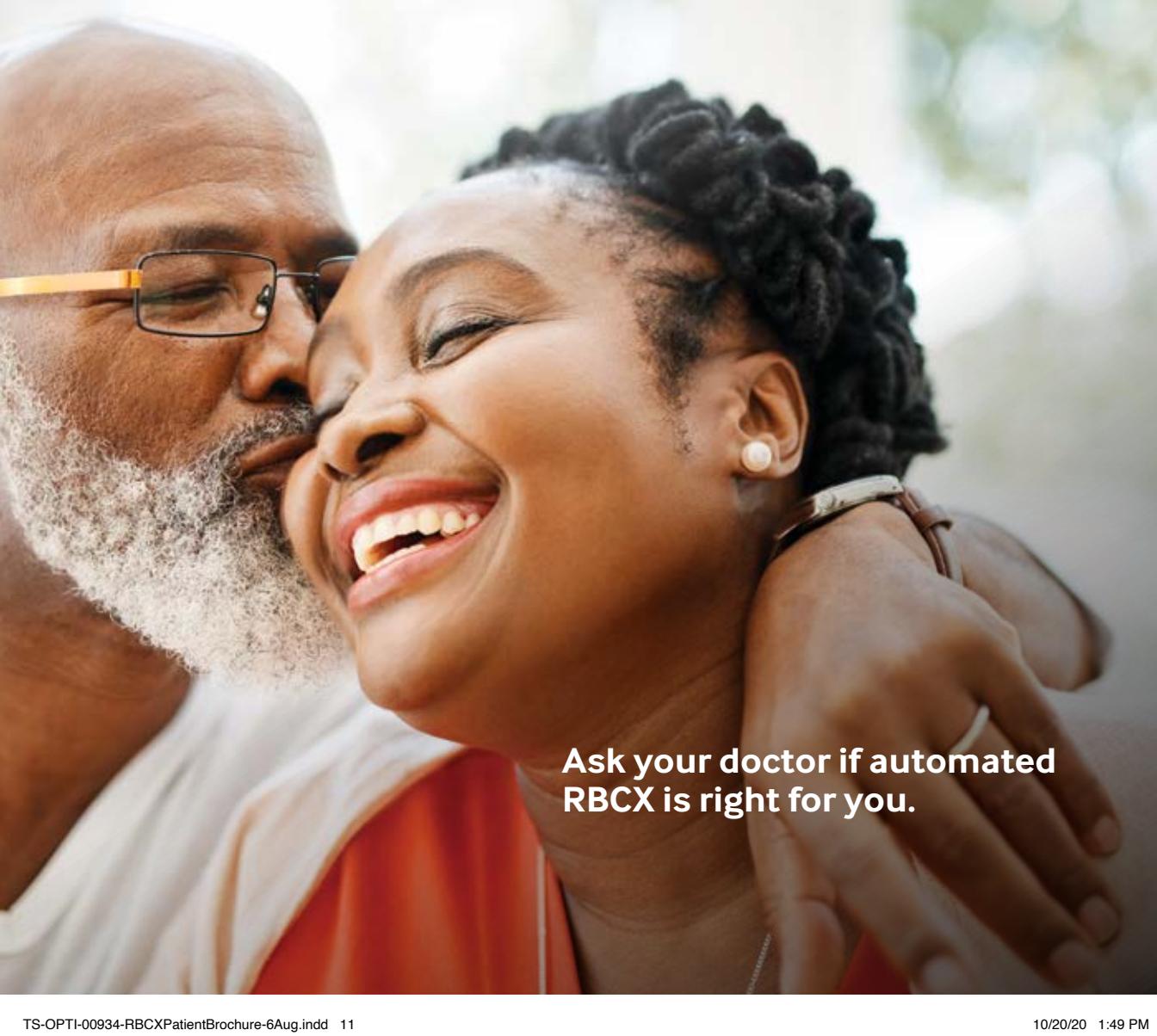
- Peripheral access
- Central venous catheter
- Arteriovenous (AV) fistula
- AV graft
- Implanted port





**I really didn't find anything
that was effective for keeping
me away from having crises,
except for automated RBCX."**

Rona Wiggins, sickle cell disease patient



**Ask your doctor if automated
RBCX is right for you.**

Side effects of automated RBCX may include:

Anxiety, headache, light-headedness, digital and/or facial paresthesia, fever, chills, hematoma, hyperventilation, nausea and vomiting, syncope (fainting), urticaria, hypotension, allergic reactions, infection, hemolysis, thrombosis in patient and device, hypocalcemia, hypokalemia, thrombocytopenia, hypoalbuminemia, anemia, coagulopathy, fatigue, hypomagnesemia, hypogammaglobulinemia, adverse tissue reaction, device failure/disposable set failure, air embolism, blood loss/anemia, electrical shock, fluid imbalance and inadequate separation of blood components.¹³

Transfusion-Related Safety Information**Reactions to blood products transfused during procedures can include:**

Hemolytic transfusion reaction, immune-mediated platelet destruction, fever, allergic reactions, anaphylaxis, transfusion-related acute lung injury (TRALI), alloimmunization, posttransfusion purpura (PTP), transfusion-associated graft-versus-host disease (TA-GVHD), circulatory overload, hypothermia, metabolic complications and transmission of infectious diseases and bacteria.^{14,15}

Restricted to prescription use only.

- Operators must be familiar with the system's operating instructions.
- Procedures must be performed by qualified medical personnel.

¹ National Institute for Health and Care Excellence. Spectra Optia for automatic red blood cell exchange in patients with sickle cell disease. Medical technologies guidance (MTG28). <https://www.nice.org.uk/guidance/mtg28>. Published March 2016. Accessed 09 April 2019.

² Houwing ME, de Pagter PJ, van Beers EJ, et al. Sickle cell disease: clinical presentation and management of a global health challenge. *Blood Rev*. 2019;37:100580.

³ National Center on Birth Defects and Developmental Disabilities, Centers for Disease Control and Prevention. Sickle cell disease. Updated 15 June, 2020. Accessed 30 June, 2020. <https://www.cdc.gov/ncbddd/sicklecell/index.html>.

⁴ Danielson CFM. The role of red blood cell exchange transfusion in the treatment and prevention of complications of sickle cell disease. *Ther Apher*. 2002;6(1):24-31.

⁵ Singer T, Quirolo K, Nishi K, Hackney-Stephens E, Evans C, Vichinsky E. Erythrocytapheresis for chronically transfused children with sickle cell disease: an effective method for maintaining a low HbS level and reducing iron overload. *J Clin Apher*. 1999;14(3):122-125.

⁶ Kuo K, Ward R, Kaya B, Howard J, Telfer P. A comparison of chronic manual and automated red blood cell exchange transfusion in sickle cell disease patients. *Br J Haematol*. 2015;170(3):425-428.

⁷ Duclos C, Merlin E, Paillard C, et al. Long-term red blood cell exchange in children with sickle cell disease: manual or automatic? *Transfus Apher Sci*. 2013;48(2):219-222.

⁸ Dedeken L, Le PQ, Rozen L, et al. Automated RBC exchange compared to manual exchange transfusion for children with sickle cell disease is cost-effective and reduces iron overload. *Transfusion*. 2018;58(6):1356-1362.

⁹ Howard J. The role of blood transfusion in sickle cell disease. *ISBT Science Series*. 2013;8:225-228.

¹⁰ Al-Salem AH. *Medical and Surgical Complications of Sickle Cell Anemia*. Springer; 2016.

¹¹ Wayne AS, Kevy SV, Nathan DG. Transfusion management of sickle cell disease. *Blood*. 1993;81(5):1109-1123.

¹² Cobianchi C, Fafoutis D, Roig J, Dierick K, Comasolivas N. Measuring health-related quality of life in individuals with sickle cell disease undergoing automated red blood cell exchange. Poster presented at: Sickle Cell Disease Association of America Annual Meeting; October 2018; Baltimore, MD.

¹³ Crookston KP. *Therapeutic Apheresis: a Physician's Handbook*. 5th ed. Bethesda, MD: AABB/ASFA; 2017.

¹⁴ AABB. Circular of Information for the Use of Human Blood and Blood Components. Bethesda, MD: AABB; 2017.

¹⁵ European Directorate for the Quality of Medicines & HealthCare (EDQM). Guide to the Preparation, Use and Quality Assurance of Blood Components. 19th Edition Strasbourg, France: EDQM Council of Europe; 2017.